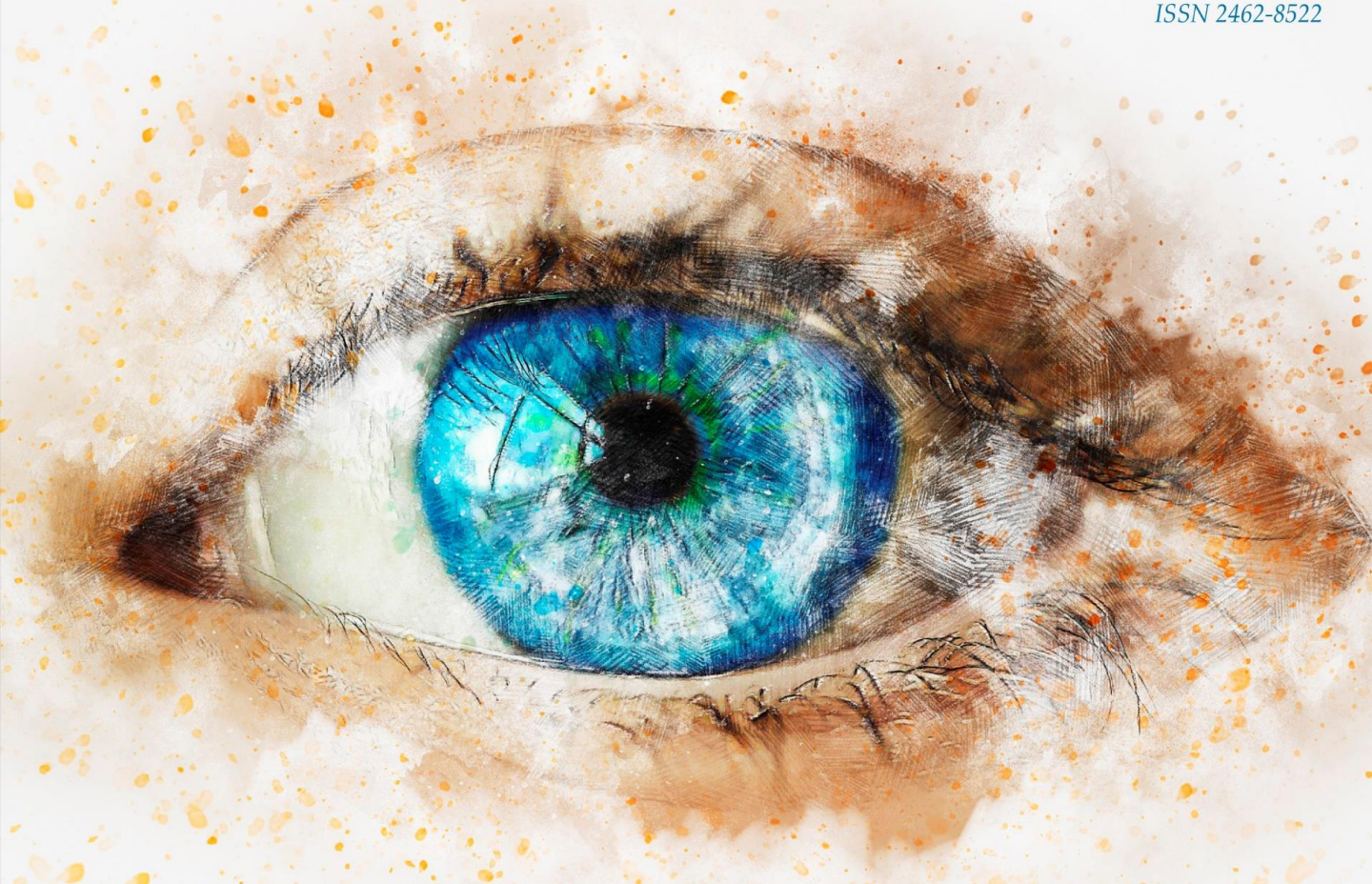
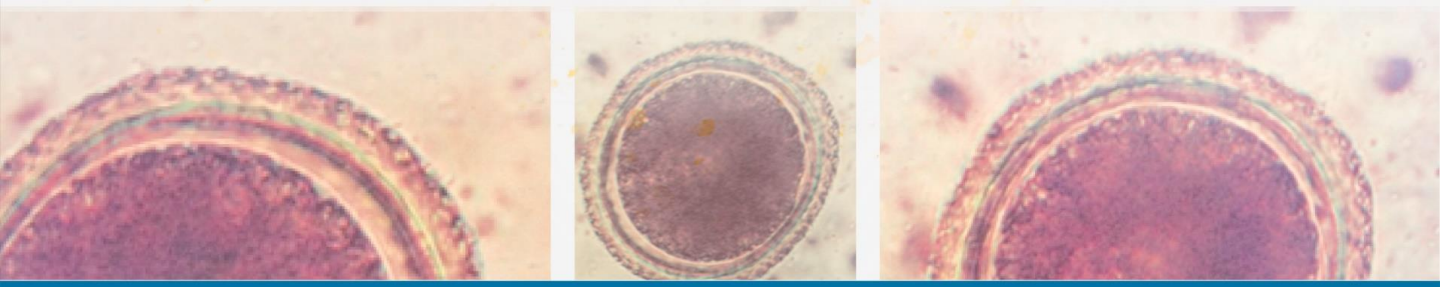
TOXOCARIASIS OCULAR EN MUJER ADULTA

CASE REPORTS
REPORTES DE CASO
ISSN 2462-8522



CASE REPORTS
2018 Año 4, Vol.4 No. 1



Editores: Edith Ángel Müller
Bibiana Jeannette Escobar
David Rincón Valenzuela

Fotografías: Cortesía Laboratorio Parasitología Veterinaria, FMVZ-UN

Centro Editorial
Facultad de Medicina
Sede Bogotá

UNIVERSIDAD NACIONAL DE COLOMBIA

Case Reports
Reportes de Caso

e-ISSN 2462-8522

The Case reports Journal is an official publication of the Faculty of Medicine of Universidad Nacional de olombia, in digital versions, specialized in case reports, images in medicine, case series, systematic review of cases studies and case studies of adverse effects, that are relevant to the advance in Medicine and other Health sciences. Is indexed in: REDIB, DRJI, OJS (Open Journal Systems), the Universidad Nacional de Colombia OJS website and Google Scholar. The journal is published biannual: an annual volume consisting of two issues (January-June, July-December). The purpose of the journal is to be a leader and model in the field of national and international case report.

CONTENTS

Editorial:

COLOMBIA AS A MEGADIVERSE COUNTRY: CHALLENGES OF MIXED EPIDEMIOLOGICAL BEHAVIOR

Ligia I. Moncada-Álvarez
Universidad Nacional de Colombia
- Bogotá Campus - Faculty of Medicine -
Department of Public Health - Bogotá D.C. -
Colombia.

Corresponding author
Ligia I. Moncada-Álvarez.
Department of Public Health, Faculty of Medicine,
Universidad Nacional de Colombia. Bogotá D.C.
Colombia. Email: limoncadaa@unal.edu.co

https://doi.org/10.15446/cr.v4n1.71846

The Colombian epidemiological profile has transcended to chronic diseases since life expectancy has increased substantially, especially in large cities. (1,2) Nevertheless, infectious diseases caused by pathogens, viruses, bacteria, fungi and parasites persist in some areas of the country.

According to the Ministry of Social Protection, the improvement of the living conditions of the population has allowed for great achievements in terms of health, resulting in the increase of life expectancy and the decrease of mortality caused by communicable diseases. (3) Many of these diseases are recognized by the World Health Organization (WHO) or by the Pan American Health Organization (PAHO) as "neglected diseases", because they occur in populations that suffer from poverty and marginality. (4)

Colombia is a megadiverse country, which means that there are many and varied species of flora and fauna that, on the one hand, provide a suitable scenario for research on new molecules with therapeutic effects and, on the other, are involved in the cycles of the pathogens or produce diseases. Several of these diseases are "neglected", which is evident in scarce or nonexistent research, as well as in the poor development on the issue in related academic programs. Some of these diseases include poisoning by toxins from animals such as jellyfish (known in Colombia as "aguamala"), ophidic accidents or poisoning by other animals, and pathologies originated by the contact with larvae of *Lepidoptera* of some genera such as *Loxoceles* or *Tunga penetrans*, etiological agent of tungiasis, which may lead to mutilation of some parts of the body, among others.

This issue of the Case Reports Journal includes reports of diseases caused by helminths and of neurocysticercosis, which is caused by the larval stage of *Taenia solium*. These diseases are considered as neglected by the WHO and the PAHO. (4)

Parasitic diseases etiologically originated by helminths are still prevalent in many regions of the country. All these infections are preventable, if patients comply with some recommendations, and through community support, good epidemiological surveillance and implementation of intervention measures that go from the improvement of the living conditions of the population, which requires improving existing disparity conditions, until access to the health care system.

Neurocysticercosis is well known for its biological cycle and the entity involved in its development; in other words, knowledge about the tools to control or eliminate it is adequate, as in the case of developed countries. Thus, the question is: what should be done in Colombia to achieve its control or elimination? To this end, it is necessary to extrapolate the existing knowledge to the domestic reality in order to encourage surveillance by the corresponding institutions interested in the parasite's biological cycle, that is, not only the health sector but also the agriculture sector and all those that may be involved. The role that the education sector may play in this regard is also relevant. As exposed by several authors, this transdisciplinary and comprehensive work should be done by territorial entities and the population, while the management of these diseases should involve the community itself as established by the WHO. (5)

On the other hand, the prevalence of some parasitisms may increase as the number of pets found in cities increases; this is the case of toxocariasis, whose larva penetrates the human body as an accidental host since this parasite is acquired by soil contaminated with dog feces.

According to a study conducted in the Faculty of Veterinary Medicine and Zootechnics of the Universidad Nacional de Colombia (6), larvae eggs of *Toxocara spp* and *Ancylostoma spp* were found in parks of the Suba locality in Bogotá D.C., which can produce visceral larva migrans and cutaneous larva migrans syndromes. This implies that adequate conditions for the development of larvae are found in many cities and, therefore, there is a risk of infection with these parasites. The Ministry of Health of Bogotá addressed this issue when it defined the concept of environmental health (7) and some of its activities highlight the protection against diseases transmitted by animals.

This convergence has a very special meaning from a public health perspective since, first, zoonotic diseases are still a major issue in our context —not only in rural areas with low technification of food production, both animal and vegetable—, which are controllable with measures involving different entities and, second, the fact of being a megadiverse country also generates risks of transmission zoonotic diseases or of those originated by animal hosts, as in the case of American trypanosomiasis or accidents caused by poisonous animals, which are frequent in Colombia.

The current situation of the country requires following the initiatives established by other countries to address their own pathologies labeled as "neglected diseases", for which more human and financial resources are needed to facilitate research and management. Within the lines of research supported by Colombian funding agencies, research institutes and the academia, with a transdisciplinary and collaborative approach, there should be a special line that addresses these issues. This approach should consider the beneficial use of Colombian biodiversity to produce new therapeutic molecules or to strengthen the most adequate measures for disease control, and also ensure a place for learning about these diseases in the academic programs of human and animal health care students. In conclusion, it is necessary to address the epidemiological and biological reality in a transdisciplinary way.

REFERENCES

1. Ministerio de Salud y Protección Social. Análisis de la situación de salud, Colombia, 2016. Bogotá D.C.: MinSalud; 2016 [cited 2018 Apr 20]. Available from: https://goo.gl/e5bwds.
2. Departamento Administrativo Nacional de Estadística (DANE). Atlas Estadístico de Colombia. Bogotá D.C.: DANE; 2012 [cited 2018 Mar 23]. Available from: https://goo.gl/Locdtc.
3. World Health Organization. Global Health: today's challenges. In: WHO. The world health report 2003 - shaping the future. Geneva: WHO; 2013 [cited 2018 Abr 23]. Available from: https://goo.gl/bR7P5k.
4. World Health Organization. Neglected zoonotic diseases. New York: WHO [cited 2018 Mar 24]. Available from: https://goo.gl/Xg2a7y..
5. World Health Organization. 10 facts about zoonotic neglected tropical diseases. New York: WHO; 2015 [Cited 2018 May 15]. Available from: https://goo.gl/sb2PhS.
6. **Polo-Terán LJ, Cortés-Vecino JA, Villamil-Jiménez LC, Prieto E.** Contaminación de los parques públicos de la localidad de Suba, Bogotá con nematodos zoonóticos. *Rev. salud pública.* 2007;9(4):550-7.
7. **Henández-Florez LJ, Sarmiento R, Osorio-García SD.** Enfoques conceptuales de la relación ambiente y salud. In: Eslava-Castañeda JC, Sarmiento R, Osorio-García SD, Mesa-Cuadros G, Rojas-Roa N, Ágreda JA, *et al.* Reflexiones acerca de la relación ambiente salud. Bogotá D.C.: Editorial Universidad Nacional de Colombia. p. 15-36.

https://doi.org/10.15446/cr.v4n1.65310

OCULAR TOXOCARIASIS CAUSED BY *TOXOCARA CANIS* IN AN ADULT WOMAN. CASE REPORT

Keywords: Larva Migrans; *Toxocara canis*; Neglected Diseases; Diagnosis; Colombia.
Palabras clave: Larva Migrans; *Toxocara canis*; Enfermedades desatendidas; Diagnóstico; Colombia.

Germán Santiago Barahona-Botache
Program of Medicine - Faculty of Medicine -
Universidad Nacional de Colombia – Bogotá
Campus –Bogotá D.C. - Colombia.

Mario Javier Olivera MSc
Parasitology Group
- Instituto Nacional de Salud -
Bogotá D.C. - Colombia.

Corresponding author:
Mario Javier Olivera, Instituto Nacional de Salud
- Parasitology Group - Bogotá D.C. - Colombia.
E-mail: mjoliverar@gmail.com.

Received: 28/05/2017 **Accepted:** 22/01/2018

ABSTRACT

Introduction: Toxocariasis is a zoonosis caused by the *Toxocara canis* and *Toxocara cati* nematodes larvae. These are intestinal parasites found in canids and felids, respectively.

Case presentation: This paper presents the case of a 22-year-old woman from Caquetá, with a three-month history of vision loss in her left eye, eye pain, diplopia, photophobia and bilateral red eye. Retinal detachment in the left eye was diagnosed by ultrasound. The ophthalmology service made a differential diagnosis of retinoblastoma and pars planitis. Symptomatic management was initiated with oral and topical corticosteroids, obtaining symptom improvement, although loss of visual acuity persisted.

Discussion: Coexistence and cohabitation of the patient with pets (dogs and cats) was an important factor to consider ocular toxocariasis. Complete blood count revealed mild leukocytosis and lymphocytosis without eosinophilia. Antibodies against *Toxoplasma gondii* and *Taenia solium* cysticercus were negative, as well as the VDRL. The ELISA test for *Toxocara canis* was positive, with IgG titers of 1:64 (positive ≥1:32, specificity >90%). Management with ophthalmic and systemic corticosteroids, albendazole and vitrectomy was initiated. The vitreous band was released by means of surgery, making the diplopia disappear, although residual peripheral granuloma persisted and will be operated by ophthalmology.

Conclusions: Knowledge on ocular toxocariasis is important for physicians because it might be mistaken with retinoblastoma, a malignant neoplasm that may require enucleation of the eye.

RESUMEN

Introducción. La toxocariasis es una zoonosis producida por las larvas de los nematodos *Toxocara canis* y *Toxocara cati,* parásitos intestinales de los cánidos y los félidos, respectivamente.

Presentación del caso. Paciente femenino de 22 años de edad, procedente de Caquetá, Colombia, con un cuadro de 3 meses de evolución con pérdida de visión en ojo izquierdo, acompañada de dolor ocular, diplopía, fotofobia y ojo rojo bilateral, quien presenta desprendimiento de retina en ojo izquierdo diagnosticado por ecografía. La mujer asiste a oftalmología, donde se hace diagnóstico diferencial con retinoblastoma y pars planitis. Se inicia manejo sintomático con corticoides orales y tópicos, con mejoría de la sintomatología, pero persistencia de pérdida de la agudeza visual.

Discusión. Al revisar historial de antecedentes socioeconómicos, la paciente convive con perros y gatos, por lo que se piensa en toxocariasis ocular. En el cuadro hemático se observa ligera leucocitosis y linfocitosis, sin eosinofilia. La determinación de anticuerpos anti-*Toxoplasma gondii* y anti-cisticerco de *Taenia solium* resultaron negativas, al igual que el VDRL. El test de ELISA para T. canis fue positivo, con unos títulos IgG de 1:64 (positivo ≥1:32, especificidad >90%). Se inicia manejo con corticoide oftálmico, sistémico, albendazol y vitrectomía. Con la intervención quirúrgica se libera banda vítrea, logrando la desaparición de la diplopía, pero persiste granuloma periférico residual, el cual será intervenido por oftalmología.

Conclusiones. Dada la posibilidad de ser confundida con un retinoblastoma —una neoplasia maligna que puede requerir la enu-

cleación del ojo—, el conocimiento de esta forma de presentación de la toxocariasis es de suma importancia para los clínicos.

INTRODUCTION

Toxocariasis is a neglected parasitic disease that affects mostly poor and isolated communities in low-income countries. For this reason, little attention has been paid to this condition in terms of surveillance, prevention and control. (1-3) Toxocariasis is distributed worldwide and the seroprevalence of *Toxocara* infection varies from 2.4% to 76.6%. (4-5) This zoonosis is caused by *Toxocara* nematodes, particularly *Toxocara canis*, a dog parasite and main etiological agent, and *Toxocara cati*, found in the intestine of cats. (1,3,6)

In general, humans are infected through ingestion of embryonated eggs in contaminated soil and, therefore, contaminated hands. In consequence, children of preschool and school age are the most affected, although adults may also develop the disease. (1,6) Ocular toxocariasis occurs when larvae migrate to the eye and cause inflammation and scarring that can lead to vision loss. (1)

The purpose of this work is to report a case of ocular toxocariasis in an adult patient, highlighting the limitations found when establishing the correct diagnosis in adults, even though this is one of the most common zoonotic infections in the world. The relevance of this clinical case is that it provides specific clinical signs of ocular toxocariasis that help to achieve a diagnosis using complementary serological methods that provide evidence on *Toxocara* infection to minimize anatomical and functional sequelae.

CASE PRESENTATION

22-year-old female university student, living in Bogotá D.C.-Colombia, without a significant pathological history, who referred vision loss in the left eye of 3 months of evolution, accentuated in the last 2 weeks, accompanied by eye pain, diplopia, photophobia and bilateral red eye. The patient presented retinal detachment in the left eye diagnosed by ultrasound. She consulted with ophthalmology, where a differential diagnosis of retinoblastoma and pars planitis was made. Symptomatic management was initiated with oral and topical corticosteroids. She was referred to the Instituto Nacional de Salud (National Health Institute) due to suspicion of ocular toxocariasis.

Upon reviewing her socioeconomic background, coexistence with dogs and cats during childhood in rural Caquetá was observed. At the time of consultation, she had been living for 6 months with a dog and two kittens that had not been vaccinated nor dewormed. The woman said that she occasionally took her pets to the park, where they were in contact with soil and feces and reported feeling better with the drugs prescribed by ophthalmology, although her visual acuity continued to be affected.

Upon physical examination, her vital signs were within the normal range and without fever. Ophthalmological examination showed visual acuity in the right eye sc: 1.00 and in the left eye sc: 0.5. Hyperemic tarsal papillae were also observed in both eyes. Ophthalmoscopy in the right eye was normal, while a peripheral granuloma and a fibrous band pulling the macula were observed in the left eye. The rest of the physical examination did not show any alterations.

When analyzing the complete blood count made on March 9, 2016, slight leukocytosis and lymphocytosis without eosinophilia were observed. However, the values returned to normal after the symptomatic treatment ended on June 14, 2016 (Table 1). The results of the coprological and partial urine tests were normal. In addition, anti-*Toxoplasma gondii*

and anti-*Taenia solium* cysticerco antibodies were negative, as well as the VDRL. The ELISA test for *T. canis* was positive, with IgG titres of 1:64 (positive ≥1: 32, specificity >90%).

Table 1. Results of blood counts at two different times

Date Day/Month/ Year	LEU (CEL/mm^3)	NEU (%)	LYMP (%)	MON (%)	EOS (%)	BAS (%)	HEM (g/ dL)	HCT (%)	PLAT (CEL/mm^3)
09/03/2016	12.100	47.2	50.1	1.4	0.8	0.5	15.6	47	269.000
14/06/2016	9.800	61.6	30.8	5.7	1.4	0.5	15.2	46	272.000

LEU: leukocytes; NEU: neutrophils; LYMP: lymphocytes; MON: monocytes; EOS: eosinophils; BAS: basophiles; HEM: hemoglobin; HCT: hematocrit; PLAT: platelets; CEL: cells.

Source: Own elaboration based on the data obtained in the study.

Management was initiated with two doses of oral prednisone 2 mg/kg/day, topical prednisolone 1 drop/2hr and albendazole 400 mg/day for 5 days. The patient required surgical management with vitrectomy in the left eye to release the vitreous band, making the diplopia disappear. However, residual peripheral granuloma persisted and will be operated by ophthalmology. Hygiene recommendations were provided to the patient and her relatives, the consumption of well-cooked meats was suggested and control was scheduled at 1 month.

DISCUSSION

Knowing about this clinical case is relevant to the medical and scientific community for three reasons: first, because this is a case of ocular toxocariasis, a parasitic disease neglected worldwide that is of special interest for Latin American countries such as Colombia, where it is considered endemic (7); second, because current medical literature mentions that toxocariasis is predominant in children, but some cases are associated with the adult population (8-11), and third, because it is related to the importance of differentiating this ocular pathology from others that require differential diagnosis such as retinoblastoma, toxoplasmosis and syphilis. (8)

Toxocariasis has been described more frequently in children of preschool and school age due to the permanent presence of risk factors in this particular age group. (1) Nevertheless, this disease should not be ignored in the adult population, in which cases have also been reported, since a less common way of acquiring the infection is through the intake of raw or undercooked foods contaminated with the larvae of the parasite. (3,7,12) In addition, it is worth mentioning that the type of syndrome that appears due to infection with *Toxocara* seems to be related to age, as visceral larva migrans appears mostly during childhood, whereas ocular toxocariasis is seen in advanced ages, but still, there is controversy around this particular issue, since many authors report that it is predominant during childhood. (7,13)

Definitive diagnosis of ocular toxocariasis is obtained by demonstrating the presence of migrating larvae in the biopsies of the compromised tissues, which is rather exceptional since it is an invasive procedure. Coproparasitological examination is not useful because the parasite is unable to mature inside a human host. (3,13,14) Therefore, current diagnosis

is made based on typical ophthalmological signs, which, in general, are unilateral and supported by immunological tests. (3)

The indirect ELISA test, used as an aid to obtain a diagnosis, uses excretion/secretion antigens of the *T. canis* larvae to detect anti-*Toxocara spp*. in serum or other body fluids such as vitreous humor, which is used especially in ophthalmic cases. The sensitivity of this test is 80-100%, with specificity of 90-95%, but these figures may vary according to the geographical region where it is applied and the quality of the antigen obtained. (13) According to other sources, an ELISA test with serum titers ≥1:32 has a sensitivity of 73% and a specificity >90% (2), although a titre of 1:8 in serum is sufficient to support the diagnosis if the patient has clinical manifestations compatible with this zoonosis. (3) Due to potential cross-reactivity with the ELISA test, some authors suggest confirming the result with a Western Blot test. (13)

On the other hand, although eosinophilia is an important marker of systemic toxocariasis, it is not usually observed in ocular form (1,3), but its presence may indicate the coexistence of both forms of toxocariasis in the same patient. (3) Needless to say, for accurate diagnosis, it is necessary to know the complete clinical history of the patients, as well as their signs and symptoms and socioeconomic situation, to identify predisposing factors such as cohabitation with dogs or cats and geophagy. (14)

There are four clinical presentations of ocular toxocariasis, but only two are more common: posterior pole granuloma and peripheral granuloma. (3) They usually present with unilateral vision loss, sometimes with strabismus and sometimes with leukocoria, although granulomatous inflammation can cause diverse manifestations such as keratitis, iridocyclitis, chronic endophthalmitis, retinal detachment and optic neuritis. (7) For this reason, differential diagnosis should be made with other ocular granulomatous diseases such as ocular toxoplasmosis, sarcoidosis, tuberculosis and fungal infections. (3,15) Since leukocoria and strabismus are the two most common signs of retinoblastoma, it is also necessary to perform a differential diagnosis with this neoplasm. (7,13,16) The patient described in this case did not present any of those ocular alterations.

CONCLUSIONS

Diagnosis of ocular toxocariasis is based on the identification of particular signs and symptoms; it should be supported by a complete clinical history that provides a detailed report of socioeconomic background and should be complemented by an indirect ELISA test performed on a serum sample or vitreous humor. This process is carried out in order to avoid erroneous diagnoses such as retinoblastoma and other granulomatous diseases of the eye.

Since ocular toxocariasis is a neglected and global disease, it is important to implement prevention and control measures to reduce the prevalence of this parasitic disease in the population. For this purpose, awareness must be generated in the community about this type of preventable diseases through the promotion of good hygienic practices such as hand washing, identification of transmission sources and reduction of exposure to etiological agents. Furthermore, it is necessary to develop programs for deworming pets, both dogs and cats, in order to control the transmission of this disease.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Woodhall D, Jones JL, Cantey PT, Wilkins PP, Montgomery SP.** Neglected parasitic infections: what every family physician needs to know. *Am Fam Physician.* 2014 [cited 2018 Jan 30];89(10):803-11. Available from: https://goo.gl/6mK8F9.
2. **Woodhall DM, Eberhard ML, Parise ME.** Neglected Parasitic Infections in the United States: Toxocariasis. *Am J Trop Med Hyg.* 2014;90(5):810-3. http://doi.org/f52s2z.
3. **Ahn SJ, Ryoo NK, Woo SJ.** Ocular toxocariasis: clinical features, diagnosis, treatment, and prevention. *Asia Pac Allergy.* 2014;4(3):134-41. http://doi.org/gb93fx.
4. **Stensvold CR, Skov J, Moller LN, Jensen PM, Kapel CM, Petersen E, *et al.*** Seroprevalence of human toxocariasis in Denmark. *Clin Vaccine Immunol.* 2009;16(9):1372-3. http://doi.org/brhdpx.
5. **Fan CK, Hung CC, Du WY, Liao CW, Su KE.** Seroepidemiology of Toxocara canis infection among mountain aboriginal schoolchildren living in contaminated districts in eastern Taiwan. *Trop Med Int Health.* 2004;9(12):1312-8. http://doi.org/cft97p.
6. **Botero D, Restrepo M.** Parasitosis Humanas. 5th ed. Medellín: Corporación para Investigaciones Biológicas; 2012.
7. **Delgado O, Rodríguez-Morales AJ.** Aspectos clínico-epidemiológicos de la toxocariasis: una enfermedad desatendida en Venezuela y América Latina. *Bol Mal Salud Amb.* 2009 [cited 2018 Jan 30];49(1):1-33. Available from: https://goo.gl/qy6R6X.
8. **Azira NM, Zeehaida M.** A case report of ocular toxocariasis. *Asian Pac J Trop Biomed.* 2011;1(2):164-5. http://doi.org/cjz4.
9. **Good B, Holland CV, Taylor MR, Larragy J, Moriarty P, O'Regan M.** Ocular Toxocariasis in Schoolchildren. *Clin Infect Dis.* 2004;39(2):173-8. http://doi.org/bwhbkx.
10. **Gómez L, Rueda T, Pulido C, Sánchez-Román J.** Toxocariasis Ocular. A propósito de un caso. *Arch Soc Esp Oftalmol.* 2007;83(1):49-52. http://doi.org/bbk2mz.
11. **Rey A.** Nematode Endophthalmitis due to Toxocara. *Br J Ophthalmol.* 1962;46(10):616-8. http://doi.org/brb8rn.
12. **Pak KY, Park SW, Byon IS, Lee JE.** Ocular toxocariasis presenting as bilateral scleritis with suspectretinal granuloma in the nerve fiber layer: a case report. *BMC Infect Dis.* 2016;16(1):426. http://doi.org/cjz6.
13. **Roldán WH, Espinoza YA, Huapaya PE, Jiménez S.** Diagnóstico de la Toxocarosis Humana. *Rev Perú Med Exp Salud Pública. 2010;27(4):613-20. http://doi.org/dj55sv.*
14. **Rojas-Salamanca AC, León-Bustamante MC, Bustamante-Saavedra OR.** Toxocara canis: una zoonosis frecuente a nivel mundial. *Rev Cien Agri.* 2016;13(1):19-27. http://doi.org/cjz8.
15. **Bae KW, Ahn SJ, Park KH, Woo SJ.** Diagnostic Value of the Serum Anti-Toxocara IgG Titer for Ocular Toxocariasis in Patients with Uveitis at a Tertiary Hospital in Korea. *Korean J Ophthalmol.* 2016;30(4):258-64. http://doi.org/cjz9.
16. **Zepeda-Gómez EM, Vázquez-Tsuji O, Campos-Rivera T.** Larva Migrans Ocular vs Retinoblastoma en Pediatría. Presentación de un Caso. *Rev Enfer Infec Pediatr.* 2013 [cited 2018 Jan 30];26(104):307-13. Available from: https://goo.gl/jy9S9T.

https://doi.org/10.15446/cr.v4n1.65326

GASTROSCHISIS. CASE REPORT AND MANAGEMENT IN PRIMARY CARE SERVICES

Keywords: Gastroschisis; Congenital Abnormalities; Primary Health Care.
Palabras clave: Gastrosquisis; Anomalía congénita; Atención primaria de salud.

Ricardo Ibarra-Calderón, MD
Oscar Octalivar Gutiérrez-Montufar, MD
Department of Pediatrics
- Faculty of Health Sciences -
Universidad del Cauca
- Popayán - Colombia.
General Medicine Service
- Hospital Universitario San José -
Popayán - Colombia.

Jhan Sebastián Saavedra-Torres
Luisa Fernanda Zúñiga Cerón
Faculty of Health Sciences
- Grupo de Investigación en Salud (GIS) -
Universidad del Cauca - Popayán - Colombia.

Corresponding author:
Ricardo Andrés Ibarra-Calderón. Departamento de Pediatría, Facultad de Ciencias de la Salud, Universidad del Cauca. Popayán. Colombia.
Email: ribarra@unicauca.edu.co.

Received: 29/05/2017 **Accepted:** 19/02/2018

ABSTRACT

Introduction: Gastroschisis is a low-prevalence disease with a very good prognosis, if initial management is adequate. This paper attempts to describe the disease and highlight the importance of correct treatment at the primary care level.

Case presentation: Newborn child diagnosed with gastroschisis in a primary care center, referred to the Neonatology Service of a tertiary care institution. He received interdisciplinary management and underwent gradual surgical closure, with favorable outcome after a three-month hospitalization.

Discussion: There is no clarity about the exact cause of gastroschisis, since it is a multifactorial disease. It can be diagnosed during the prenatal stage by means of ultrasonography, which has high sensitivity and specificity for its detection.

Conclusion: Gastroschisis is a disease that requires adequate knowledge from both specialized and primary care personnel, as it ensures a correct initial management and avoids future complications.

RESUMEN

Introducción. La gastrosquisis es una enfermedad de baja prevalencia, pero de muy buen pronóstico si se realiza un adecuado manejo inicial. El presente escrito tiene como objetivo realizar una descripción de esta patología, destacando la importancia de su correcto manejo en el primer nivel.

Presentación del caso. Neonato a término con hallazgo de gastrosquisis en primer nivel quien fue remitido al servicio de neonatología de una institución de tercer nivel. El infante recibió manejo interdisciplinario y cierre quirúrgico gradual y tuvo evolución favorable tras 3 meses de hospitalización.

Discusión. No existe claridad sobre la causa exacta de la gastrosquisis, ya que es una enfermedad multifactorial. Su diagnóstico puede realizarse desde la etapa prenatal mediante la ultrasonografía, un método que posee alta sensibilidad y especificidad para su detección.

Conclusión. La gastrosquisis es una enfermedad que para su diagnóstico y tratamiento requiere de personal especializado en primer nivel, lo que garantiza un correcto manejo inicial y evita futuras complicaciones.

INTRODUCTION

Gastroschisis can be defined as a congenital defect of the anterior abdominal wall, characterized by evisceration of the abdominal organs through an opening in the absence of membranous coverage; this defect is usually observed to the right of the navel (1-3), involving, in all cases, the small intestine (3) and sometimes the stomach, colon or gonads. (1,3) As they are not covered by membranes, the eviscerated structures are exposed to amniotic fluid and external substances after birth, which increases the risk of infection and injuries. (1,3,4) This is a low-prevalence disease (5-7) of great importance due to the excellent prognosis and survival of patients (8-10) when providing adequate management. (11,12)

CASE PRESENTATION

This paper reports the case of a full-term male infant born at 37 weeks, who was transferred from Florencia, Colombia to the Neonatology Service. The child was born by vaginal delivery at a primary care center, with an incidental finding of protruding, violaceous and wet intestinal loops, associated with respiratory distress.

The patient received oxygen therapy through cannula and nasogastric tube. Ophthalmic prophylaxis was performed and then, he was referred to a secondary care institution, where gastric lavage was performed, a polyethylene bag was placed, and antibiotic treatment with ampicillin-gentamicin was initiated. Ringer's lactate solution and dextrose 10% in distilled water at 100 cm^3/kg/day were administered with a metabolic flow of 6.7 mg/kg/min, and inotropic management was initiated due to hemodynamic instability. The child was referred to a tertiary care institution for management by Pediatric Surgery.

The child was the firstborn of a teenager (17 years old) with O+ blood type, who underwent eight prenatal care checkups, serology and protocol blood tests with negative results. Obstetric ultrasounds at weeks 19 and 29 of pregnancy did not report alterations and fetal movements were positive since month two. The infant was a vaginal delivery product with cephalic presentation and without premature rupture of ovular membranes; Apgar: 6/8/10. The child was fully vaccinated. No pathological, infectious, pharmacological or transfusion history were observed other than maternal poisoning during the first trimester of pregnancy with insecticide, since the mother lives in an area where constant fumigations are performed.

On physical examination, the patient presented with stable vital signs and normal anthropometric measurements (abdominal perimeter was not assessed due to the protrusion of intestinal loops). The thorax showed a slight intercostal retraction and the abdomen, a protrusion of intestinal loops covered with a viaflex container, pink, well perfused and with a foul odor; the skin was pale and poorly perfused.

Table 1. Synthesis of the evolution of the patient.

Days	Event
Day 1	Birth and primary care
Day 3	Admission to tertiary care institution First surgery. Abdomen in viaflex container
Day 6-8	Three plications of viaflex container
Day 14	Total closure of the wall
Since day 15	Favorable evolution
Day 109	Discharged with interdisciplinary follow-up recommendations

Source: Own elaboration based on the data obtained in the study

Based on clinical findings, gastroschisis, respiratory distress syndrome and early neonatal sepsis were diagnosed. Renal ultrasound and echocardiogram were requested to rule out associated congenital malformations; the results were normal. Clinical genetics determined a chemical teratogenic disruptive process during the first trimester of pregnancy as probable etiology. Taking into account his history, a k-band karyotype was requested, which was not authorized by the health service provider, so it was not possible to use it as a diagnostic tool to establish management. This case report does not address the importance of the denied examination.

The patient required mechanical ventilation and inotropic support. The Pediatric Surgery Service proposed closing the abdominal wall gradually and adding metronidazole to antibiotic management. During surgery, severe gastro-

schisis was found with exposure of stomach, small and large intestines, intestinal malrotation with thickened meso, and leaky and thickened intestine due to intrauterine exposure.

The umbilical border was cleared, the umbilical and vesical arteries were ligated and a Bogota bag was attached to the skin covered with gauze impregnated with Furacin®.

The procedure was well tolerated at first, but a deterioration of the clinical condition was observed subsequently with hemodynamic instability, which required inotropic support with dopamine and dobutamine; mechanical ventilation with high parameters; sedation with fentanyl and morphine; relaxation with rocuronium, and follow-up with antibiotic therapy with ampicillin-gentamicin and metronidazole.

The Pediatric Surgery Service decided to perform plications of the viaflex container. The first was done 3 days after the first surgery and the second and third were performed at intervals of 24 hours after the first plication.

A second surgery was planned 24-48 hours after the last plication. However, chest x-ray findings were interpreted as possible acute disseminated candidiasis, so the procedure was postponed. Pediatric Pneumology ruled out said infection, so the second surgery was performed 4 days after the last plication (Figure 1). During the procedure, gastroschisis was corrected with myocutaneous and fasciocutaneous flap. After removing the viaflex container, a thickened, dysmorphic and malrotated intestine was observed.

If gastroschisis is a small defect (only a part of the intestines protrudes from the abdomen), it is usually treated with surgery soon after birth (Figure 1). However, if gastroschisis is a large defect (many organs protrude from the abdomen), repair could be done slowly, in stages, covering the exposed organs with a special material and placing them slowly in the abdomen. After the organs have been arranged inside the abdomen, the opening is closed.

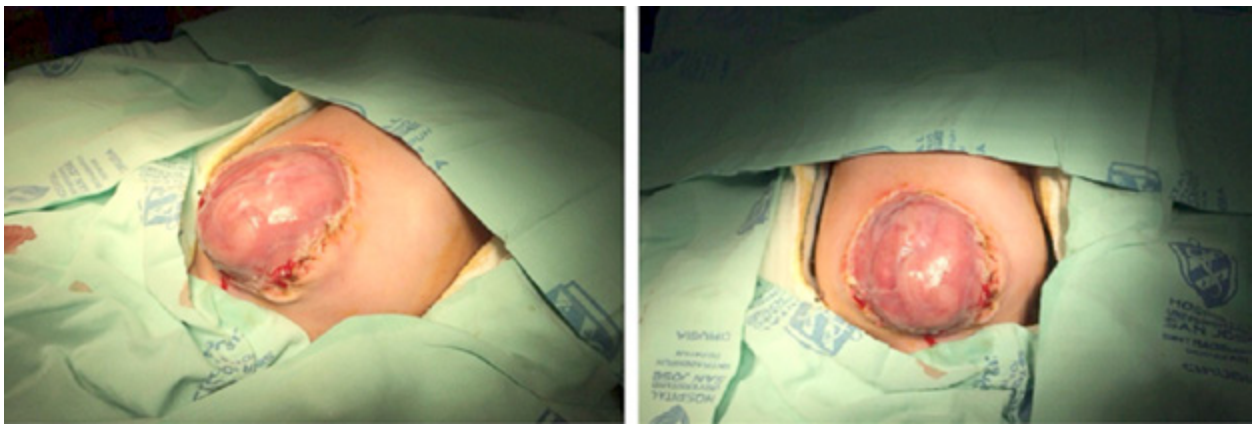


Figure 1. Secong surgery.
Source: Own elaboration based on the data obtained in the study.

In the postoperative period, the patient remained hemodynamically stable, achieving inotropic and vasoactive weaning. Pharmacological relaxation and morphine were discontinued and fentanyl was administered only at analgesic doses. Mechanical ventilation was continued in a controlled assisted manner with minimal parameters and intra-abdominal pressure between 6-11 mmHg. Trophic stimulation with dextrose at 5% was initiated.

The patient remained hospitalized for 109 days; his evolution was satisfactory and the food was well tolerated with normal stools and adequate weight gain (reaching 3 875 grams). The patient was discharged with breastfeeding on demand, supplemented extensively with hydrolyzed milk formula. Currently, the child continues to be monitored by High-risk Pediatrics, Clinical Genetics, Pediatric Cardiology and Nutrition.

DISCUSSION

The first gastroschisis report was published in 1773. (4,6) Around 1894, Taruffi coined this term to group diseases of different etiology (omphalocele, bladder exstrophy, amniotic hernia, gastroschisis). (4) The distinction between gastroschisis and other abdominal wall defects, especially omphalocele, was successfully achieved at the beginning of the 21st century with the implementation of the International Classification of Diseases, tenth edition (ICD-10). (4) Table 2 summarizes the differences between gastroschisis and omphalocele. (1-3,5,6)

Table 2. Differences between gastroschisis and omphalocele.

Gastroschisis	Omphalocele
Location: right side	Location: center
Content not covered by membranes	Presence of peritoneum-amniotic membrane
No umbilical cord	Umbilical cord inserted in caudal area of the hernial sac
Embryopathy	Fetopathy
Content: intestine (100%), colon, bladder, gonads (occasionally)	Content: intestine, liver (in most cases); spleen, colon, bladder (occasionally)
Rarely associated with other congenital anomalies (15%)	Frequently associated with other congenital anomalies (40-80%)

Source: Own elaboration based on (1,3,5,6)

EPIDEMIOLOGY

In recent decades, the incidence of gastroschisis has increased worldwide (1,5,10) and is observed in 1/3 000 to 1/10 000 births. A review of the 1991-2001period in the Clinical Hospital of the University of Chile showed that the figure was 2.1/10 000 births. (1-4) In Colombia, an incidence of 7.8/10 000 births was reported, more frequently seen in 37-week-old male newborns. (13,14) The risk factors associated with the disease are prematurity; small for gestational age newborns; maternal age <20 years; being born to a primigravida mother; Caucasian race; Hispanic mothers; maternal malnutrition; exposure to nitrosamines; teratogens and agrochemicals; consumption of nonsteroidal anti-inflammatory drugs and acetaminophen in the first trimester; cigarette, alcohol and illicit drugs consumption; low socioeconomic class; absence of prenatal checkups, and short cohabitation with the father of the child. (1,2,5,7-11,13) The average age of mothers with affected children is 21.1 years; women aged 14 to 19 have a 7.2 times higher risk of having a child with gastroschisis compared to 25 to 29-year-old mothers. (15)

It should be noted that the mother of the studied patient was 17 years old, primigravida, exposed to a toxic substance (insecticide) in the first trimester of pregnancy and of low socioeconomic status.

ETIOPATHOGENESIS

There is no certainty about the exact cause of gastroschisis, since it is a multifactorial disease. Embryologically, the abdominal wall originates from the lateral mesoderm and by the fusion of four folds (cephalic, caudal and two lateral foldings), which grow towards the midline, con-

verging in the umbilical ring that is completed around the fourth week. (2,12)

Current accepted causal theories affirm that gastroschisis is caused by vascular disruptions, either by intrauterine occlusion of the omphalomesenteric artery or by early atrophy (<28 days) of the right umbilical vein, which causes wall infarction with rupture of the umbilical ring and eventration of the intestine. (2,16-18)

A new theory proposes that there is a defect in the inclusion of the yolk sac in the fetal body stem, with the consequent formation of an additional opening through which the intestine is eventracted, instead of doing it through the umbilical cord. (16-18) The final outcome is the eventration of abdominal contents in utero that, regardless of the size and quantity of viscera exposed, is associated with a mortality of 5% and 3-15% after birth. (5,7,8,12) However, these deaths may be associated with complications and significant morbidity, prolonged hospital stays, need for mechanical ventilation, prolonged parenteral nutrition, multiple surgical interventions and diseases such as intestinal atresia, short bowel syndrome, neonatal sepsis and necrotising enterocolitis. (7-9,12)

DIAGNOSIS

A gastroschisis diagnosis can be achieved in the prenatal stage by means of an ultrasonography, which has high sensitivity and specificity for its detection. Detecting the disease is possible since week 12 (19) with rates of up to 90%, depending on the quality of the equipment used, the institution were the examination is performed and the experience of the staff (19-22), which would explain why, in this case, the pathology was not detected prenatally, despite the adequate number of controls and two ultrasound scans taken after week 12.

Prenatal detection of this disease is important because it allows timely genetic counseling, since performing a karyotype is not recommended in these patients given the limited association of this defect with other genetic syndromes. (9) Furthermore, diagnosis facilitates a better monitoring of pregnancy, which avoids complications. (19,23)

There are useful ultrasound predictors to estimate the possibility of neonatal complications, such as intestinal atresia. (7,23) Some predictors are intra-abdominal dilation of the bowel (7,8), intrauterine growth restriction (8), thickness of the abdominal wall (8) and liver herniation. (8)

MANAGEMENT OF GASTROSCHISIS AT PRIMARY CARE

Once the prenatal diagnosis is made, a multidisciplinary approach (obstetrician, neonatologist, pediatric surgeon) and bi-monthly sonographic controls are required to monitor markers to predict complications. (23) Although some studies postulate that there are no differences in the outcome of neonates in relation to prenatal diagnosis (20), these reports come from developed countries, where care delivery is done in centers of medium or high complexity, while in Colombia undetected cases may be handled in centers that do not have technological resources and adequate personnel, as in this case.

Several studies suggest that early caesarean section (36-37 weeks) decreases morbidity with respect to vaginal delivery due to the supposed risk of infection or perforation of the viscera exposed during the latter, while other authors do not find significant differences. (20,23,24)

The scheme presented below should be followed after the birth of a child without a prenatal diagnosis, which is similar to what was presented in this clinical case.

First, adequate resuscitation and oxygen support should be initiated, for which up to 170 mL/kg of dextrose solutions may be required within the first 24 hours, since the metabolic demands of these patients are greater due to the exposure of intestinal loops, with consequent loss of fluids and hypothermia. (23-25) Then, gastric decompression is performed using a probe, washing the intestinal loops with 0.9% saline and covering them with plastic bags or sterile viaflex (23,24), thus reducing the risk of infection. (23-25) The patient is placed in the right lateral decubitus position to reduce the risk of intestinal ischemia (24) and empirical antibiotic management is initiated, preferably with ampicillin-gentamicin, adding vasoactive support if required. These basic measures help to decrease mortality in patients —a study in Africa found that 25% of neonatal deaths with gastroschisis were related to some deficiency in initial management in primary care. (26) Once stabilized, the patient should be referred to a more complex level with Pediatric Surgery and Neonatology Services, maximum 4-7 hours later, since this is the preferred time to perform the surgical closure. (23)

There are two types of closures: primary and gradual. Some of the factors associated with the success of primary closure include patients classified as low risk and born intrainstitutionally and in reference centers. (27) On the other hand, gradual closure has some advantages over the primary closure, such as lower incidence of compartment syndrome and intra-abdominal hypertension, lower requirement of mechanical ventilation and vasoactive support. (28) Regarding the surgical technique, in recent years the sutureless closure technique has been implemented using flaps with autologous tissue, mainly umbilical cord. Several studies have found that this technique has an effectiveness profile similar to conventional closure, and that, in fact, in low-risk patients, it is associated with a lower requirement of mechanical ventilation and a decrease in the incidence of surgical wound infections.

In addition, the closing without sutures technique, using flaps with autologous tissue, can be performed outside the operating room, decreasing anesthesia requirements and costs for health institutions. (15,29,30)

ETHICAL CONSIDERATIONS

According to bioethical parameters, the efforts during any procedure should be directed to achieve the optimal resolution of the beneficence, nonmaleficence, autonomy, justice and equity principles, which guarantee adequate interdisciplinary management. (31)

When analyzing the conflict of principles, the lack of a timely prenatal diagnosis was evident (20), thus preventing adequate follow-up at an appropriate level of complexity and the choice of early cesarean section, which has shown effects on mortality. (23,24) This leads to an initial transgression of the justice and equity principles, since no adequate equipment or human resources were available for prenatal diagnosis in ultrasound after week 12. Regarding the management of this case, it is worth highlighting the optimal initial treatment, timely referral from the primary care institution, adequate information to relatives and the successful interhospital communication, which demonstrate full support to the beneficence and autonomy principles.

This research was authorized by the legal guardian of the minor and respected the confidentiality of the patient and his relatives.

CONCLUSION

Gastroschisis is a congenital defect that, despite its low frequency, requires adequate knowledge not only from specialized personnel, but also from primary care physicians, taking into account that they are obliged to ensure an appropriate and timely referral of the patient to a higher complexity level to avoid complications.

CONFLICT OF INTERESTS

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Frolov P, Alali J, Klein MD.** Clinical risk factors for gastroschisis and omphalocele in humans: a review of the literature. *Pediatr Surg Int.* 2010;26(12):1135-48. http://doi.org/cqfrgz.
2. **Ionescu S, Mocanu M, Andrei B, Bunea B, Carstoveanu C, Gurita A, *et al.*** Differential diagnosis of abdominal wall defects - omphalocele versus gastroschisis. *Chirurgia.* 2014;109(1):7-14
3. **Van Dorp DR, Malleis JM, Sullivan BP, Klein MD.** Teratogens inducing congenital abdominal Wall deffects in animal models. *Pediatr Surg Int.* 2010;26(2):127-39. http://doi.org/b8qb36.
4. **Svetliza J.** Reseña histórica de la gastrosquisis. Bahia Blanca: Sítio de Gastrosquisis en Español; 2010 [cited 2015 Mar 2]. Avilable from: https://goo.gl/KodZk9.
5. **Poddar R, Hartley L.** Exomphalos and gastroschisis. *Continuing Education in Anaesthesia, Critical Care and Pain.* 2009; 9(2);48-51. http://doi.org/ck2qp5.
6. **Romay-Bello AB, Soler-Ruiz P, de la Fuente EA, Costales-Badillo C, Montalvo-Montes J.** Defectos de cierre de la pared abdominal: Gastrosquisis. *Prog Obstet Ginecol.* 2011;54(12):612-17. http://doi.org/fbp23s.
7. **Tassin M, Benachi A.** Diagnosis of abdominal wall defects in the first trimester. *Curr Opin Obstet Gynecol.* 2014;26(2):104-9. http://doi.org/ck68.
8. **Page R, Ferraro ZM, Moretti F, Kee-Fung KF.** Gastroschisis: Antenatal sonographic predictors of adverse neonatal outcome. *Journal of Pregnancy.* 2014;(2014):13. http://doi.org/gb6x5n.
9. **Fernández-Ibieta M, Aranda-García MJ, Cabrejos-Perotti C, Reyes-Ríos P, Martínez-Castaño I, Sánchez-Morote JM, *et. al.*** Resultados iniciales de un protocolo de manejo terapéutico de la gastrosquisis. *Cir Pediatr.* 2013;26:30-6.
10. **Waller SA, Paul K, Peterson SE, Hitti JE.** Agricultural-related chemical exposures, season of conception, and risk of gastroschisis in Washington State. *Am J Obstet Gynecol.* 2010;202(3):241.e1-6. http://doi.org/b6tpz3.
11. **Agopian AJ, Langlois PH, Cai Y, Canfield MA, Lupo PJ.** Maternal residential atrazine exposure and gastroschisis by maternal age. *Matern Child Health J.* 2013;17(10):1768-75. http://doi.org/f5h2fg.
12. **Torres US, Portela-Oliveira E, Braga Fdel C, Wemer H, Daltro PA, Souza AS.** When clossure fails: What the radiologist needs to know about the embryology, anatomy, and prenatal imaging of ventral body wall defects. *Semin Ultrasound CT MRI.* 2015;36(6):522-36. http://doi.org/ck69.
13. **Soares H, Silva A, Rocha G, Pissarra S, Correia-Pinto J, Guimaraes H.** Gastroschisis: Preterm or term delivery?. *Clinics.* 2010;65(2):139-42. http://doi.org/d3577d.
14. **Pachajoa H, Saldarriaga W, Isaza C.** Gastrosquisis en un hospital de tercer nivel de la ciudad de Cali, Colombia durante el período marzo 2004 a febrero 2006. *Colomb Med.* 2008;39(Suppl 2):35-40.

15. **Rasmussen SA, Frías JL.** Non-genetic risk factors for gastroschisis. *Am J Med Genet C Semin Med Genet.* 2008;148C(3):199-212. http://doi.org/cn9r6q.
16. **López-Valdéz JA, Castro-Cóyotl DM, Venegas-Vega CA.** Nuevas hipótesis embriológicas, genética y epidemiología de la gastrosquisis. *Bol Med Hosp. Infant. Mex.* 2011;68(3):245-52.
17. **Jones KL, Benirschke K, Chambers CD.** Gastroschisis: etiology and developmental pathogenesis. *Clin Genet.* 2009;75(4):322-5. http://doi.org/cc6hvv.
18. **Stevenson RE, Rogers RC, Chandler JC, Gauderer MW, Hunter AG.** Escape of the yolk sac: a hypothesis to explain the embryogenesis of gastroschisis. *Clin Genet.* 2009;75(4):326-33. http://doi.org/bmqxv3.
19. **Radulescu M, Coriolan-Ulmeanu E, Nedelea M, Comsa A.** Diagnosticul ecografic prenatal al gastroschizisului. *Revista Romana de Pediatrie.* 2012;61(4):344-8.
20. **Murphy FL, Mazlan TA, Tarheen F, Corbally MT, Puri P.** Gastrosquisis and exomphalos in Ireland 1998-2004. Does antenatal diagnosis impact on outcome? *Pediatr Surg Int.* 2007;23:1059-63. http://doi.org/c884ks.
21. **Barisic I, Clementi M, Häusler M, Gjergja R, Kern J, Stoll C.** Evaluation of prenatal ultrasound diagnosis of fetal abdominal wall defects by 19 European registries. *Ultrasound Obstet Gynecol.* 2001;18(4):309-16. http://doi.org/b48b4x.
22. **Walkinshaw SA, Renwick M, Hebisch G, Hey EN.** How good is ultrasound in the detection and evaluation of anterior abdominal wall deffects? *Br J Radiol.* 1992;65(772): 298-301. http://doi.org/c6rwkt,
23. **Gamba P, Midrio P.** Abdominal wall defects: prenatal diagnosis, newborn management, and long-term outcomes. *Semin Pediatr Surg.* 2014;23(5):283-90. http://doi.org/f6q8b7.
24. **Christison-Lagay ER, Kelleher CM, Langer JC.** Neonatal abdominal wall defects. *Semin Fetal Neonatal Med.* 2011;16(3):164-72. http://doi.org/c6vfsh.
25. **Eltwab-Hashish AA, Elhalaby E.** Evolution of management of gastroschisis. *Annals of Pediatric Surgery.* 2011;7(1):10-5. http://doi.org/cdvhzf.
26. **Sekabira J, Hadley GP.** Gastroschisis: a third world perspective. *Pediatr Surg Int.* 2009;25(4):327-9. http://doi.org/d6ktj6.
27. **Stanger J, Mohajerani N, Skarsgard ED.** Practice variation in gastroschisis: Factors influencing closure technique. *J Pediatr Surg.* 2014;49(5):720-3. http://doi.org/f55hj6.
28. **Kunz SN, Tieder JS, Whitlock K, Jackson JC, Avansino JR.** Primary fascial closure versus staged closure with silo in patients with gastroschisis: a meta-analysis. *J Pediatr Surg.* 2013;48(4):845-57. http://doi.org/ck7c.
29. **Emami CN, Youssef F, Baird RJ, Laberge JM, Skarsgard ED, Puligandla PS.** A risk-stratified comparison of fascial versus flap closure techniques on the early outcomes of infants with gastroschisis. *J Pediatr Surg.* 2015;50(1):102-6. http://doi.org/f6wfxc.
30. **Chesley PM, Ledbetter DJ, Meehan JJ, Oron AP, Javid PJ.** Contemporary trends in the use of primary repair for gastroschisis in surgical infants. *Am J Surg.* 2015;209(5):901-5. http://doi.org/f7bj56.
31. **Maldonado JA, García-García E, García-Rodríguez E, Garrido-Ocaña AI.** Caso clínico neonatal. Cuestiones de justicia y no maleficencia. *Debática*. 2012;1(1).

https://doi.org/10.15446/cr.v4n1.61288

FEBRILE INFECTION-RELATED EPILEPSY SYNDROME (FIRES), A POSSIBLE CAUSE OF SUPER-REFRACTORY STATUS EPILEPTICUS. CASE REPORT.

Keywords: Epilepsy; Refractory Epilepsy; Child, Polytherapy; Fever.+.
Palabras clave: Epilepsia; Epilepsia refractaria; Niño; Polifarmacia; Estado epiléptico; Fiebre.

Arbey Aponte-Puerto • Juan David Rozo-Osorio
Jennifer J Guzmán-Porras • Álvaro Daniel Patiño-Moncayo
Medicine Program – Faculty of Medicine –
Universidad Nacional de Colombia
Bogotá – Colombia
Neuropedunal Research Group - Faculty of Medicine -
Universidad Nacional de Colombia -
Bogotá D.C - Colombia.

Jorge A Amórtegui-Beltrán
Medicine Program – Faculty of Medicine –
Universidad Nacional de Colombia
Bogotá – Colombia

Angélica M. Uscátegui-Daccarett
Department of Pediatrics - Faculty of Medicine -
Universidad Nacional de Colombia - Bogotá D.C. - Colombia.
Neuropedunal Research Group - Faculty of Medicine -
Universidad Nacional de Colombia -
Bogotá D.C - Colombia.

Corresponding author:
Arbey Aponte-Puerto.
Faculty of Medicine - Universidad Nacional
de Colombia - Bogotá D.C. - Colombia
Email: araponteпu@unal.edu.co

Received: 27/11/2016 **Accepted:** 28/05/2017

ABSTRACT

Introduction: Super-refractory status epilepticus (SRSE) is a pathology that affects the neuronal environment depending on the types of seizure and their duration.

Case presentation. This paper presents the case of a 7-year old child presenting with super-refractory status epilepticus and multifocal seizures. Metabolic, structural, infectious, toxicological and autoimmune causes were discarded, while different anticonvulsive agents were administered without any clinical improvement; seizures were controlled 6 weeks after admission to ICU. A 12-year follow-up was performed, during which time the patient presented recurrent status epilepticus with autonomic seizures and progressive cognitive decline.

Discussion: This type of status epilepticus is part of the syndrome known as Febrile Infection-Related Epilepsy Syndrome (FIRES), a possibly autoimmune form of epileptic encephalopathy that is refractory to acute and chronic management. There is no report in the literature that includes long term follow-up, therefore, there is no actual consensus about the appropriate management of the chronic phase of the disease.

Conclusion: FIRES must be considered as one of the possible etiologies of super-refractory status epilepticus, so early management strategies (like ketogenic diet) can be used in order to achieve control of the critically ill patient, control long term seizures and improve cognitive outcomes, having as the final result a positive impact on the quality of life of the patient.

RESUMEN

Introducción: El estado epiléptico superrefractario (EES) es una patología con importante morbimortalidad que afecta el ambiente neuronal según el tipo y duración de las crisis.

Presentación del caso: Se presenta el caso de un escolar con estado epiléptico superrefractario y crisis multifocales. Se descartaron causas metabólicas, estructurales, infecciosas, toxicológicas y autoinmunes y se utilizaron diferentes manejos anticonvulsivantes sin respuesta, lográndose control de las crisis 6 semanas después del ingreso a UCI. Se realizó un seguimiento de 12 años, periodo en el que el paciente presentó múltiples recaídas del estado epiléptico asociadas a la presencia de epilepsia refractaria con múltiples tipos de crisis, en su mayoría vegetativas; además se dio involución cognitiva.

Discusión: Esta forma de estado epiléptico corresponde al síndrome de estado epiléptico facilitado por fiebre (FIRES), entidad de posible origen inmunológico conocida por ser refractaria al tratamiento agudo y al manejo crónico de la epilepsia y que se presenta como secuela. Su evolución no se ha descrito a largo plazo y por tanto no hay consenso sobre el manejo en la fase crónica.

Conclusión: Es importante considerar esta etiología en estado epiléptico superrefractario para utilizar de forma temprana diferentes estrategias terapéuticas, como la dieta cetogénica, que permitan, por un lado, controlar su condición crítica y las crisis epilépticas a largo plazo y, por el otro, mejorar el pronóstico cognitivo, logrando así un impacto en la calidad de vida.

INTRODUCTION

Super-refractory status epilepticus (SRSE) is a pathology, with significant morbidity and mortality rates (1), that affects the neuronal environment depending on the types of seizure and their duration (2). Generally, its causes are divided into genetic and symptomatic status epilepticus (SE); the latter, in turn, is divided into acute and remote and may be caused by structural alterations or metabolic or immunological disorders. The last category includes Febrile Infection-Related Epilepsy Syndrome (FIRES), an encephalopathy characterized by multifocal and refractory SE that occurs in previously healthy children or adolescents.

The onset of SRSE is acute and, despite a usual history of febrile symptoms, the cerebrospinal fluid (CSF) does not present alterations or infectious focus documented frequently (3). Initially, isolated seizures, which quickly evolve to SE, may be observed (4). This paper presents a FIRES case that describes the diagnostic and therapeutic challenges posed by this condition, as well as its management difficulties in the long-term and its consequences on quality of life.

CASE PRESENTATION

7-year-old male patient from Bogotá, Colombia, without relevant family, psychosocial, prenatal or perinatal or pathological history and normal neurodevelopment. The child consulted due to severe headache of 8 days of evolution after a febrile episode managed with cephalexin, considering suspicion of unconfirmed sinusitis. Subsequently, seizures appeared in the following 48 hours, characterized by cephalic and labial version to the left and clonic generalization lasting 1-2 minutes. The seizures became more frequent until consciousness deteriorated and motor status epilepticus appeared. In consequence, management with barbiturate coma was initiated in the ICU, where multifocal clonic and myoclonic seizures continued. Different anticonvulsant treatments were used for 6 weeks: thiopental sodium, lidocaine, propofol, midazolam, valproic acid, topiramate, carbamazepine, phenytoin and clonazepam; the last 6 antiepileptic drugs were used simultaneously and at maximum recommended doses, achieving a poor initial response, but eventually leading to total crisis control.

Possible infectious, inflammatory, metabolic, autoimmune and toxicological etiologies were discarded, while initial neuroimages did not show any alteration (Table 1).

After the initial symptoms resolved, the patient developed a focal drug-resistant epilepsy, with recurrent status epilepticus. Currently, after a 12-year follow-up and treatment with oxcarbazepine, phenobarbital, valproic acid, topiramate, levetiracetam and clobazam in different associations and at maximum tolerated doses, the patient presented with focal ictal frequency of 2 to 3 events per week and altered consciousness. Many of these episodes were associated with bradycardia, decline in oxygen saturation and a monthly event with tonic-clonic generalization, as well as moderate cognitive deficit.

At the time of the case analysis, lacosamide was added to the topiramate, clobazam and levetiracetam scheme in association with immunoglobulin, which was administered for 6 months, and a low glycemic ketogenic diet (KD). This scheme did not help to decrease ictal frequency, but his relatives reported cognitive improvement with the use of KD.

Cerebral nuclear magnetic resonance (NMR) showed generalized cortical atrophy and bilateral hippocampal atrophy (Figure 1), while electroencephalogram revealed independent bilateral temporal ictal activity, frequently associated with bradycardia and hypoxemia (Figure 2).

Table 1. Paraclinical tests of the patient

Infectious	IgM negative for measles and rubella. Negative VDRL test on CSF, negative CSF ADA. Negative CMV IgM and IgG. HIV negative. Negative Toxoplasma-specific IgM and IgG. CMV by PCR+ in CSF. (Considered as a polyclonal reaction due to autoimmune disease since previous CMV results were negative)
Autoimmune	Anti TPO 8.16 (Normal). ANAs 1/800. Normal protein electrophoresis. Somatomedin C 301.2. Anti-Ro 3.3; Anti-La 4; Anti Sm 2.7; Anti-RNP 7.4 (Normal). Negative AntiDNA, ANAs, RF.
Metabolic	Aminoacids in urine. Negative dinitrophenylhydrazine. Negative nitrosonaphthol. Negative sodium nitroprusside. Negative ferric chloride. Lactic acid 0.870 mmol/L (0.5-2.2 mmol/L). Pyruvic acid 0.050 mmol/L (0.034-0.102 mmol/L). Lactate-to-pyruvate ratio 17.40 mmol/L (0-25 mmol/L) Porphobilinogen 0.97 mg/24 h (0-2 mg/24 h). Vanilmandelic acid 6.5 mcg/mg creat (0-10 mcg/mg creat). Creatinuria 75.4 mg/dL. Delta-aminolevulinic acid 3.1 mg/day (1-7 mg/day) Lysine-Histidine-Ornithine band (Observed in dibasic aminoaciduria, problems of the urea cycle, or patients who have been administered primidone, phenobarbital, or L-Dopa metabolites). Citrulline 5.1 (<20)
Toxic	Heavy metals (negative for mercury, lead and thallium). Negative TCAs and barbiturates in urine.

CMV: cytomegalovirus; ADA: adenosine deaminase; anti-TPO: anti thyroid peroxidase antibody; Anti-Sm: anti-Smith antibody; anti-RNP: anti-ribonucleoprotein antibody; ANAs: antinuclear antibodies; RF: rheumatoid factor; TCA: tricyclic antidepressants.

Source: Own elaboration based on the data obtained in the study.

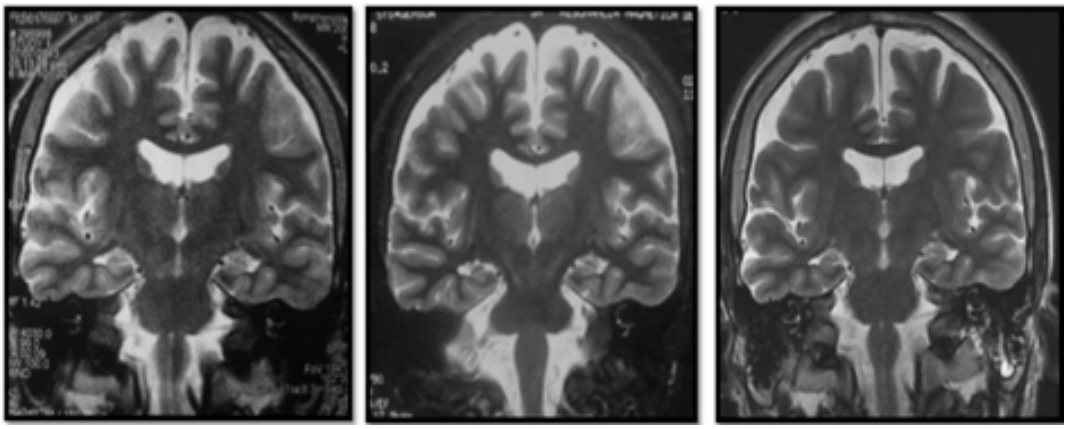


Figure 1. Simple cerebral magnetic resonance, T2 sequence. From left to right: 2008 (11 years old), 2011 (14 years old), 2016 (19 years old).

Source: Own elaboration based on the data obtained in the study.

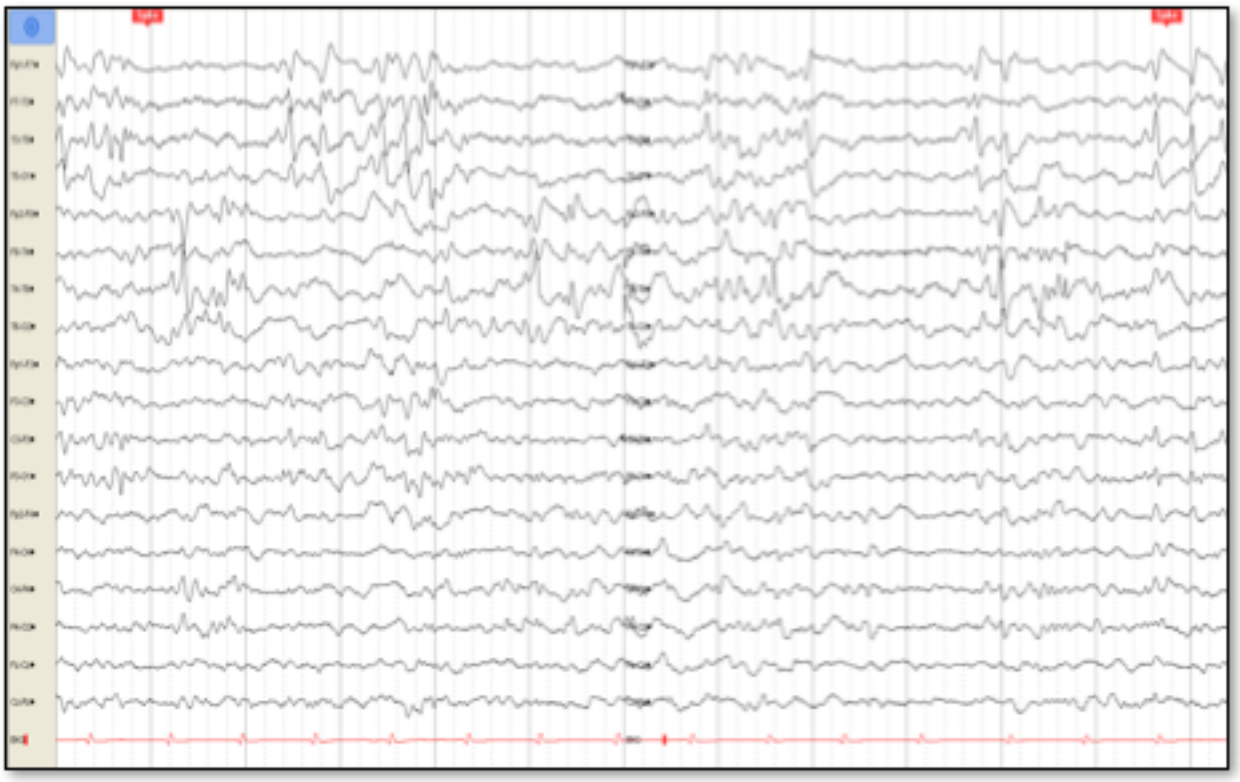


Figure 2. Electroencephalogram that reflects temporary, bilateral and independent epileptiform activity in the interictal sleep records. November 2015.

Source: Patient's studies. Image edited by the authors.

DISCUSSION

Etiology

The temporary correlation between fever and the onset of SE has led to consider an autoimmune origin for this syndrome, although current evidence does not confirm this suspicion.

In autoimmune epilepsies, epileptic seizures relate to the production of autoantibodies against central nervous system (CNS) molecules, which is confirmed when some patients respond to immunomodulatory therapy (5,6) and when some autoimmune diseases have a higher crisis incidence (7).

Innate and adaptive immunity has been observed in patients with autoimmune epilepsies, which increases inflammatory mediators, macrophages and neutrophils, activating glial cells and stimulating neuronal death. Mediators seem to act as excitatory agonists, generating a postictal state (4,8-11). Autoantibodies have been proposed as the etiology of FIRES, without finding a direct causal association in all cases or being clear whether autoimmune inflammatory processes arise first or if inflammatory activity is the result of the epileptic event (3,11). However, evidence does not support this hypothesis completely given the variability of antineuronal antibody findings in FIRES patients and inconstant response to immunotherapy (12).

Likewise, FIRES is associated with monogenic mutations in PCDH19, SNC1A and POLG1, which are in turn associated with epi-

leptic encephalopathies of similar characteristics: temporary association to infection, rapid onset of refractoriness, absence of encephalitis, and cognitive impairment markers in previously healthy patients (13-15). The presence of prolonged SE can be related to mitochondrial disorders etiologies and affect the reserve of energy and, therefore, neuronal function (11,16).

Autoimmunity plays an important role in epileptic disorders unrelated to infection or neoplasms (10,17) and in refractory epileptic encephalopathies with inconsistent findings of antibodies against neural surface antigens (antibodies anti-NMDA, anti-VGKC and anti-GAD) (3,4,10,11,18,19). The presence of antibodies and an excessive inflammatory response could justify the use of immunotherapy as a treatment for FIRES (3,16).

VKKL-ab, anti-GAD and GluR3 autoantibodies and oligoclonal bands in CSF are observed in a third of the patients (4). However, the reason for their presence, as well as their consequences and correlation with the phenotypic variants of FIRES, is unknown.

Manifestations and clinical course

FIRES usually develops in children under 15 years of age, who have fever between 2 and 14 days before the onset of symptoms, with a male-female ratio of 4:3. The febrile syndrome is attributed, mostly, to infections of the upper respiratory tract (more than 50%) and, in a smaller proportion, to gastroenteritis (4,20). Once epileptic seizures appear, a SE is established and then an SRSE-like behavior is observed; this SE usually lasts from 1 to 12 weeks (4). The seizures type at the beginning of the disease are mainly focal-motor, with clonic manifestations or segmental myoclonus that can compromise the face and other body segments (21,22), sometimes reaching up to 100 daily crises (23) (Figure 3).

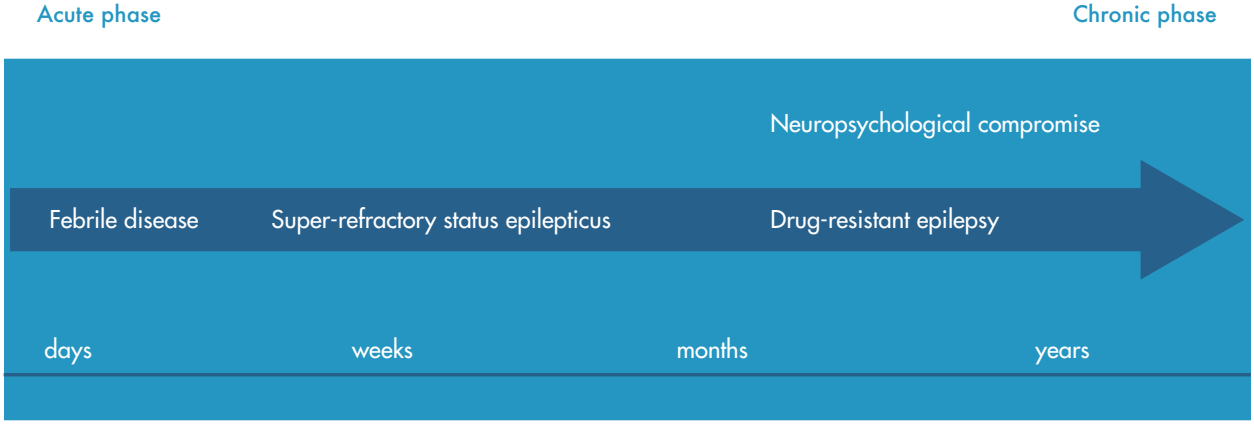


Figure 3. Natural history of the febrile infection-related epilepsy syndrome.
Source: Own elaboration based on Baalen *et al.* (24).

Except for the presence of burst suppression in barbiturate coma, EEG shows multifocal activity with greater involvement of the temporal and frontal lobes (4).

SRSE control in this patient may be related to the natural course of the disease and not to the pharmacological interventions.

With respect to neuroimaging, the variations observed in the MRI range between normal findings and different degrees of atrophy and hyperintensities in the temporal lobe, mainly in the hippocampal region (12,25,26), as well as in the insula and basal ganglia (11), perhaps secondary to prolonged neuronal activation (4).

Treatment of FIRES

Most of the medications indicated for this emergency, and even other non-usual medications such as carbamazepine or topiramate, have not shown a good response in the acute phase during status epilepticus (4,22,27,28). During this phase, barbiturates can be useful at high doses, achieving up to 50% of crisis control transiently; however, there are SE recurrences (27,29).

Other series (19,30) show that levetiracetam at regular doses for status epilepticus management could have a good response to control seizures in about half of FIRES patients. However, data are contradicted in another series of 12 patients (27), where control was not achieved with this medication. This treatment is effective in the long term to reduce seizures in more than 75% of patients at doses between 750 mg/d and 1500 mg/d, although adverse effects such as aggressiveness or impulsivity are frequent (31).

Immunomodulation

Plasmapheresis tests have been performed without obtaining a response; adrenocorticotropic hormone (ACTH), intravenous immunoglobulin (IVIG), and other steroids have shown responses in less than 5% of patients. There is no standardized scheme for the use of immunoglobulin in status epilepticus, so different treatment schemes have been used in these patients. In some cases 1.2 g IVIG were administered every 10 and 14 days, in others it was administered every 21 days for 4, 6 and 8 months or was used at a dose of 30 mg/Kg/day over 5 days, followed by 1 mg/kg/day; however, the dose used in many of the reports is not clear (4,22,26,27,28).

Ketogenic diet

Different series of FIRES patients have described the use of KD, finding a good response in terms of crisis control, thus turning this option into a promising therapy. A smaller proportion of patients did not respond and others presented recurrence when suspending it (21,26,27,32-34).

Mikaeloff *et al.* (21) used KD in two FIRES patients; one of them had more than 100 seizures per day and achieved cessation of epileptic activity two days after initiating the treatment (21).

The use of KD in a study of 9 patients showed that 8 of them achieved ketonuria 2 to 4 days after initiating the diet, while seizures stopped in 7 of them 2 to 4 days after achieving ketonuria. Patients who responded to the treatment regained consciousness 24-58 hours after the cessation of seizures and motor functions in the following weeks. In one patient, KD was interrupted abruptly, which made the status recurrent in a short period of time and caused death 10 days later (33).

Vaccarezza *et al.* (34) reported the use of KD in three patients with possible FIRES who presented SE of 52, 30 and 18 days of evolution, in whom SE stopped after 24 hours in the first two cases, and after 3 days

in the last. After establishing KD, the first patient remained free of crisis, the second had ten partial seizures per month and the third had a crisis per month, evidencing a good response to this therapeutic measure.

During the chronic phase, one study assessed six patients treated with KD for a period between 6 months and 2 years, with a marked decrease in the frequency of seizures and recurring once or twice a week (33).

Other therapies

During the acute phase, no response has been obtained with medications such as lacosamide, lidocaine, ketamine, verapamil, magnesium, vitamin B6, folic acid, biotin, paraldehyde or dextromethorphan (4,35). However, hypothermia has reported an acceptable response and fewer sequelae (36). Electroconvulsive therapy (ECT), as described by Mirás-Veiga *et al.* (37) in one patient, also showed a decreased epileptiform activity.

IVIG has not shown good results during the chronic phase of FIRES, so other treatments such as surgical management have been included, with 50% reduction of seizures (27).

In the Howell series, six patients who overcame the acute phase of FIRES presented with drug-resistant epilepsy. The vagus nerve stimulator was used in two patients, obtaining a reduction of 30-40% of seizures; one was treated with immunotherapy, without obtaining a good response (18).

PROGNOSIS

A third of patients die, usually, 4-8 months after the acute phase or suffer from refractory epilepsy and intellectual disability (3,4,21,31); only a few survive the episode without any neurological sequelae (21,31,32,38,39). Of 68 patients followed in the series published by Kramer *et al.* (4), 12 (18%) developed attention deficit or learning problems, 11 (16%) borderline cognition, 26 (38%) mild-moderate intellectual disability and 11 (16%) were in a vegetative state; some presented secondary peripheral neuropathy and ataxia.

Only KD, even used tardily, has shown efficacy in at least 50% of patients. The crises decrease during the following weeks or months, but patients remain with a cognitive deficit.

Regarding cognitive evolution, a follow-up report of two patients with FIRES treated with multiple medications showed emotional lability and significant alteration of memory, including working memory, evocation of words and auditory verbal narrations, as well as tangentiality during conversations and phonetic and verbal fluency alterations (12).

The use of barbiturates during the acute phase is significantly related to cognitive sequelae in memory, expressive language or executive function, as well as motor disability (3,31,36). About 15% of patients have a good evolution in the medium term even though FIRES is considered a “catastrophic syndrome” (4).

Early suspicion of this etiology, considering the absence of efficient diagnostic tests, may favor the timely establishment of KD, which shows the best results. Perhaps, the rapid control of crisis and the lower frequency of complications in the acute phase favor a better prognosis for the patient in the long term.

CONFLICT OF INTERESTS

None declared by the authors.

FUNDING

None declared by the authors.

REFERENCES

1. **Shorvon S.** Super-refractory status epilepticus: An approach to therapy in this difficult clinical situation. *Epilepsia.* 2011;52(Suppl 8):53-6. http://doi.org/dskhtk.
2. **Trinka E, Cock H, Hesdorffer D, Rossetti AO, Scheffer IE, Shinnar S, *et al.*** A definition and classification of status epilepticus - Report of the ILAE Task Force on Classification of Status Epilepticus. *Epilepsia.* 2015;56(10):1515-23. http://doi.org/f7t5nx.
3. **Nabbout R.** FIRES and IHHE : Delineation of the syndromes. *Epilepsia.* 2013;54(Suppl 6):54-6. http://doi.org/f49p4z.
4. **Kramer U, Chi CS, Lin KL, Specchio N, Sahin M, Olson H, et al.** Febrile infection – related epilepsy syndrome (FIRES): Pathogenesis, treatment, and outcome: a multicenter study on 77 children. *Epilepsia.* 2011;52(11):1956-65. http://doi.org/c658fq.
5. **Nabbout R.** Autoimmune and inflammatory epilepsies. *Epilepsia.* 2012;53(Suppl 4):58-62. http://doi.org/cdht.
6. **Britton J.** Autoimmune epilepsy. *Handb Clin Neurol.* 2016;133:219-45. http://doi.org/cdhv.
7. **Greco A, Rizzo MI, De Virgilio A, Conte M, Gallo A, Attanasio G, et al.** Autoimmune epilepsy. *Autoimmun Rev.* 2016;15(3):221-5. http://doi.org/cdhw.
8. **Vezzani A, Rüegg S.** Introduction. *Epilepsia.* 2011;52(Suppl 3):1-4. http://doi.org/fsfjs8.
9. **Nabbout R, Vezzani A, Dulac O, Chiron C.** Acute encephalopathy with inflammation-mediated status epilepticus. *Neurology.* 2011;10(1):99-108. http://doi.org/cmck7r.
10. **Illingworth MA, Hanrahan D, Anderson CE, O'Kane K, Anderson J, Casey M, *et al.*** Elevated VGKC-complex antibodies in a boy with fever-induced refractory epileptic encephalopathy in school-age children (FIRES). *Dev Med Child Neurol.* 2011;53(11):1053-7. http://doi.org/d7w3bj.
11. **Van Baalen A, Häusler M, Plecko-Startining B, Strautmanis J, Vlaho S, Gebhardt B, *et al.*** Febrile infection-related epilepsy syndrome without detectable autoantibodies and response to immunotherapy: a case series and discussion of epileptogenesis in FIRES. *Neuropediatrics.* 2012;43(4):209-16. http://doi.org/f36dt5.
12. **Singh RK, Joshi SM, Potter DM, Leber SM, Carlson MD, Shellhaas RA.** Cognitive Outcomes in Febrile Infection-Related Epilepsy Syndrome Treated With the Ketogenic Diet. *Pediatrics.* 2014;134(5):1431-5. http://doi.org/f6ndfd.
13. **Specchio N, Fusco L, Vigevano F.** Acute-onset epilepsy triggered by fever mimicking FIRES (febrile infection-related epilepsy syndrome): The role of protocadherin 19 (PCDH19) gene mutation. *Epilepsia.* 2011;52(11):172-5. http://doi.org/fjvff5.
14. **Appenzeller S, Helbig I, Stephani U, Häusler M, Kluger G, Bungeroth M, *et al.*** Febrile infection-related epilepsy syndrome (FIRES) is not caused by SCN1A, POLG, PCDH19 mutations or rare copy number variations. *Dev Med Child Neurol.* 2012;54(12):1144-8. http://doi.org/cdhx.
15. **Carranza-Rojo D, Simon-Harvey A, Iona X, Dibbens LM, Damiano JA, Arsov T, *et al.*** Febrile infection-related epilepsy syndrome is not caused by SCN1A mutations. *Epilepsy Res.* 2012;100(1-2):194-8.
16. **Pardo CA, Nabbout R, Galanopoulou AS.** Mechanisms of Epileptogenesis in Pediatric Epileptic Syndromes: Rasmussen Encephalitis, Infantile Spasms, and Febrile Infection-related Epilepsy Syndrome (FIRES). *Neurotherapeutics.* 2014;11(2):297-310. http://doi.org/cdhz.
17. **Kröll-Seger J, Bien CG, Huppertz HJ.** Non-paraneoplastic limbic encephalitis associated with antibodies to potassium channels leading to bilateral hippocampal sclerosis in a pre-pubertal girl. *Epileptic Disord.* 2009;11(1):54-9. http://doi.org/cdh2.
18. **Howell KB, Katanyuwong K, Mackay MT, Bailey CA, Scheffer IE, Freeman JL, *et al.***

Long-term follow-up of febrile infection – related epilepsy syndrome. *Epilepsia.* 2012;53(1):101-10. http://doi.org/fxs83r.

19. **Varadkar S, Cross H.** Rasmussen Syndrome and Other Inflammatory Epilepsies. *Semin Neurol.* 2015;35(3):259-68. http://doi.org/f7fvgt.
20. **van Baalen A, Häusler M, Boor R, Rohr A, Sperner J, Kurlemann G, et al.** Febrile infection - related epilepsy syndrome (FIRES): A nonencephalitic encephalopathy in childhood. *Epilepsia.* 2010;51(7):1323-8. http://doi.org/cb9x8g.
21. **Mikaeloff Y, Jambaqué I, Hertz-Pannier L, Zamfirescu A, Adamsbaum C, Plouin P, et al.** Devastating epileptic encephalopathy in school-aged children (DESC): A pseudo encephalitis. *Epilepsy Research.* 2006;69(1):67-79. http://doi.org/d9q7kc.
22. **Sakuma H, Awaya Y, Shiomi M, Yamanouchi H, Takahashi Y, Saito Y, *et al.*** Acute encephalitis with refractory, repetitive partial seizures (AERRPS): a peculiar form of childhood encephalitis. *Acta Neurol Scand.* 2010;121(4):251-6. http://doi.org/fsngn7.
23. **Sharma VK, Hegde AU, Nabi F.** Refractory status epilepticus: Febrile Illness Related Epileptic Syndrome (FIRES). *Int J Epilepsy.* 2015;2(2):90-3. http://doi.org/cdj8.
24. **van Baalen A, Vezzani A, Häusler M, Kluger G.** Febrile Infection – Related Epilepsy Syndrome: Clinical Review and Hypotheses of Epileptogenesis. *Neuropediatrics.* 2017;48(1):5-18. http://doi.org/f9mn9f.
25. **Suleiman J, Brilot F, Lang B, Vincent A, Dale RC.** Autoimmune epilepsy in children: Case series and proposed guidelines for identification. *Epilepsia.* 2013;54(6):1036-45. http://doi.org/f42gmq.
26. **Rivas-Coppola MS, Shah N, Choudhri AF, Morgan R, Wheless JW.** Chronological evolution of Magnetic Resonance Imaging (MRI) Findings in children with Febrile Infection-Related Epilepsy Syndrome. *Pediatr Neurol.* 2015;55:22-9. http://doi.org/cdj9.
27. **Caraballo RH, Reyes G, Avaria MF, Buompadre MC, González M, Fortini S, et al.** Febrile infection-related epilepsy syndrome : A study of 12 patients. *Seizure.* 2013;22(7):553-9. http://doi.org/f5cp4v.
28. **Specchio N, Fusco L, Claps D, Vigevano F.** Epileptic encephalopathy in children possibly related to immune-mediated pathogenesis. *Brain Dev.* 2010;32(1):51-6. http://doi.org/fjs6cz.
29. **Cherian PJ, Nickels KC.** Febrile infection-related epilepsy syndromes and their treatment. *Journal of Pediatric Epilepsy.* 2014;3(2014):67-74. http://doi.org/cdkb.
30. **Ueda R, Saito Y, Ohno K, Maruta K, Matsunami K, Saiki Y.** Effect of levetiracetam in acute encephalitis with refractory, repetitive partial seizures during acute and chronic phase. *Brain Dev.* 2015;37(5):471-7. http://doi.org/f6742j.
31. **Kramer U, Chi CS, Lin KL, Specchio N, Sahin M, Olson H, *et al.*** Febrile infection-related epilepsy syndrome (FIRES): Does duration of anesthesia affect outcome? *Epilepsia.* 2011;52(Suppl 8):28-30. http://doi.org/dm2hmr.
32. **Sort R, Born AP, Pedersen KN, Fonsmark L, Uldall P.** Ketogenic diet in 3 cases of childhood refractory status epilepticus. *Eur J Paediatr Neurol.* 2013;17(6):531-6. http://doi.org/f5m2zz.
33. **Nabbout R, Mazzuca M, Hubert P, Peudennier S, Allaire C, Flurin V, *et al.*** Efficacy of ketogenic diet in severe refractory status epilepticus initiating fever induced refractory epileptic encephalopathy in school age children (FIRES). *Epilepsia.* 2010;51(10):2033-7. http://doi.org/c4964k.
34. **Vaccarezza M, Silva W, Maxit C, Agosta G.** Estado de mal epiléptico superrefractario: tratamiento con dieta cetogénica en pediatría. *Rev Neurol.* 2012;55(1):20-4.
35. **Tan WW, Chan DWS, Lee JH, Thomas T, Menon AP, Chan YH.** Use of Magnesium Sulfate Infusion for the Management of Febrile Illness-Related Epilepsy Syndrome : A Case Series. *Child Neu-*

rology Open. 2015;2(1): 2329048X14550067. http://doi.org/cdkc.

36. **Lin JJ, Lin KL, Hsia SH, Wang HS.** Therapeutic Hypothermia for Febrile Infection-Related Epilepsy Syndrome in Two Patients. *Pediatric Neurology.* 2012;47(6):448-50. http://doi.org/f4d57n.
37. **Mirás-Veiga A, Moreno DC, Menéndez AI, Siscart IM, Fernández MD, Sánchez EG, *et al.*** Effectiveness of Electroconvulsive Therapy for Refractory Status Epilepticus in Febrile Infection-Related Epilepsy Syndrome. *Neuropediatrics.* 2017;48(1):45-8. http://doi.org/f9njw2.
38. **Kramer U, Shorer Z, Ben-zeev B, Lerman-sagie T, Goldberg-Stern H, Lahat E.** Severe Refractory Status Epilepticus Owing to presumed encephalitis. *J Child Neurol.* 2005;20(3):184-7. http://doi.org/cdkd.
39. **Covanis A.** Epileptic encephalopathies (including severe epilepsy syndromes). *Epilepsia.* 2012;53(Suppl 4):114-26. http://doi.org/f37p2r.

https://doi.org/10.15446/cr.v4n1.65333

PURULENT PERICARDITIS AS A COMPLICATION OF PNEUMONIA IN AN INFANT. CLINICAL CASE REPORT

Keywords: Pericarditis; Pericardiostomy; Purulent pericarditis; Bacterial pericarditis; Pediatrics.
Palabras clave: Pericarditis; Pericardiostomía; Pericarditis purulenta; Pericarditis bacteriana; Pediatría.

Diego Alejandro Lozano Espinosa
Olga Yamile Castiblanco Rubio
Department of Pediatrics - Universidad Nacional de Colombia - Faculty of Medicine - Bogotá D.C. - Colombia.

Juan Carlos Bustos Acosta
Department of Pediatrics - Universidad Nacional de Colombia - Faculty of Medicine - Bogotá D.C. - Colombia.
Fundación Hospital Pediátrico La Misericordia - Bogotá D.C. - Colombia.

Germán Camacho Moreno
Department of Pediatrics - Universidad Nacional de Colombia - Faculty of Medicine - Bogotá D.C. - Colombia.
Infectious Disease Service - Fundación Hospital Pediátrico La Misericordia - Bogotá D.C. – Colombia.

Edgar Hernando Rojas Soto
Department of Pediatrics - Universidad Nacional de Colombia - Faculty of Medicine - Bogotá D.C. - Colombia.

Roy Sanguino Lobo
Pediatric Cardiology Unit. Fundación Hospital Pediátrico La Misericordia - Bogotá D.C. - Colombia.

Received: 29/05/2017 **Accepted:** 17/01/2018

ABSTRACT

Introduction: Purulent pericarditis is an inflammatory process in the pericardium caused by bacterial infection. If experienced during childhood and with untimely diagnosis, it has a high mortality rate.

Case presentation: A 10-month-old infant was admitted to a high complexity pediatric hospital in the city of Bogotá D.C, Colombia, due to clinical symptoms including cough, respiratory distress and fever. A chest x-ray was taken showing cardiomegaly and multilobar pulmonary involvement. The echocardiogram showed global pericardial effusion managed with pericardiotomy, in which 50 mL of turbid fluid with whitish membranes was obtained. Cytochemical test revealed 2 600 mm3 leukocytes with 90% PMN and protein elevation. Purulent pericarditis was diagnosed based on imaging and laboratory findings. Treatment was initiated with ceftriaxone and clindamycin for four weeks, obtaining effective clinical and echocardiographic resolution.

Discussion: The clinical presentation and imaging, paraclinical and electrocardiographic findings suggested purulent pericarditis as the first possibility. This diagnosis was confirmed considering the characteristics of the pericardial fluid, which was compatible with an exudate. Clinical resolution supported by antibiotic management corroborated the diagnosis, even though microbiological isolation was not obtained in cultures.

Conclusion: Purulent pericarditis is a rare disease in pediatrics and has a high mortality rate. Making a timely diagnosis and administering early treatment are related to a better prognosis of this pathology.

RESUMEN

Introducción. La pericarditis purulenta es un proceso inflamatorio del pericardio producto de una infección bacteriana. De no lograrse un diagnóstico oportuno, se convierte en una patología con alta mortalidad en la infancia.

Presentación del caso. Lactante de 10 meses de edad que ingresó a un hospital pediátrico de alta complejidad en Bogotá D.C., Colombia, por un cuadro clínico dado por tos, dificultad respiratoria y fiebre. Se tomó una radiografía de tórax donde se observó cardiomegalia y compromiso neumónico multilobar. El ecocardiograma mostró un derrame pericárdico global que requirió pericardiotomía, en la cual se obtuvo 50 mL de líquido turbio con membranas blanquecinas. En la prueba citoquímica se encontraron 2 600mm3 leucocitos, polimorfonucleares del 90% y elevación de proteínas. Con los hallazgos de imagenología y laboratorio se hizo el diagnóstico de pericarditis purulenta, por lo que se inició tratamiento con ceftriaxona y clindamicina por 4 semanas, obteniendo una resolución clínica y ecocardiográfica efectiva.

Discusión. La presentación clínica y los hallazgos imagenológicos, paraclínicos y electrocardiográficos sugirieron como primera posibilidad pericarditis purulenta, lo cual se confirmó por las características de líquido pericárdico, que era compatible con un exudado. La resolución clínica, apoyada por el manejo antibiótico y a pesar de no obtener aislamiento microbiológico en los cultivos, corroboró el diagnóstico.

Conclusiones. La pericarditis purulenta es una enfermedad poco frecuente en pediatría pero con alta mortalidad. Realizar un diag-

nóstico oportuno sumado a un tratamiento tempano se relaciona con un mejor pronóstico de esta patología.

INTRODUCTION

Pericarditis is rarely seen in pediatrics, and few reports on this condition in infants are found. (1) Mortality rates of bacterial pericarditis are high, which may be associated with secondary dissemination of other infectious foci such as pneumonia, meningitis, osteomyelitis, septicemia or subdiaphragmatic abscesses. (2-4)

In general, the symptoms and signs of bacterial pericarditis are non-specific, thus making timely diagnosis difficult, which is associated with poor prognosis. The diagnostic suspicion, in this case, led to early diagnosis and favorable clinical outcome.

CASE PRESENTATION

A 10-month old male patient from Bogotá-Colombia (urban area), weighing 8 kg and mestizo was admitted to the emergency department with fever of two days (38.5°C) and respiratory distress. No history of importance and complete vaccination for his age were reported. The child was hospitalized in another institution two months before hospital admission due to bacterial pneumonia managed with crystalline penicillin. Since then, he was an ambulatory oxygen user.

On admission, the patient presented: heart rate of 186 beats per minute; no hypotension (blood pressure at 106/65 mmHg); respiratory frequency of 50 breaths per minute; fever (38.3°C); SaO_2 in 74% with oxygen at one liter per minute; mucocutaneous pallor; somnolence; no jugular vein engorgement; audible, rhythmic and tachycardic heart sounds; no pericardial rub, rhonchi and crepitus in both lungs; subcostal and suprasternal retractions; no hepatomegaly or edema; peripheral pulses and capillary refill without alterations, which required increased oxygen delivery (FIO_2 0.5). His hemogram showed leukocytosis of

30 230mm3 with polymorphonuclear leukocytes of 24 360mm^3, thrombocytosis (platelets of 1 006 000mm^3); 5.8g/d serum proteins, and increase in acute phase reactants: C reactive protein of 96mg/L and procalcitonin of 5.24ng/mL. Initial chest X-ray showed cardiomegaly, bibasilar and retrocardiac parenchymal opacities, without signs of pulmonary congestion or pleural effusion (Figure 1).

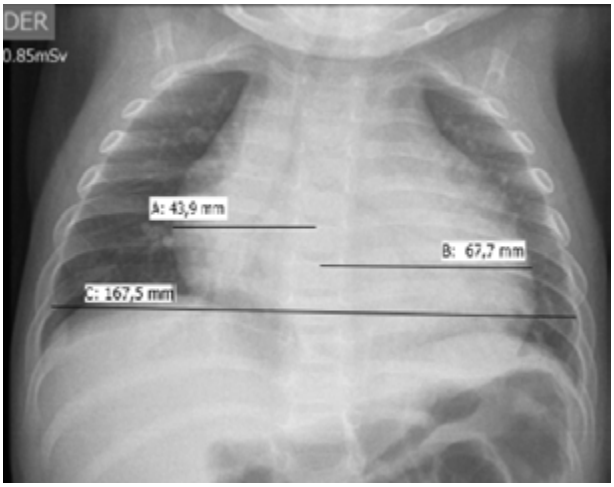


Figure 1. Initial chest x-ray.
Source: Own elaboration based on the data obtained in the study.

Pericardial effusion, myocarditis and dilated cardiomyopathy were suspected considering septic status and cardiomegaly, since they are differential diagnoses associated with multilobar pneumonia. However, no signs of pulmonary hyperflow, cardiac failure or tamponade or pulmonary or systemic congestion were observed which, added to electrocardiographic changes (Figure 2) that showed repolarization and supra-ST elevation disorder in V2 to V6,

DI and DII (evident changes in the early stages of pericarditis), led to suspect pericarditis associated with pericardial effusion, without cardiac tamponade, in the first place.

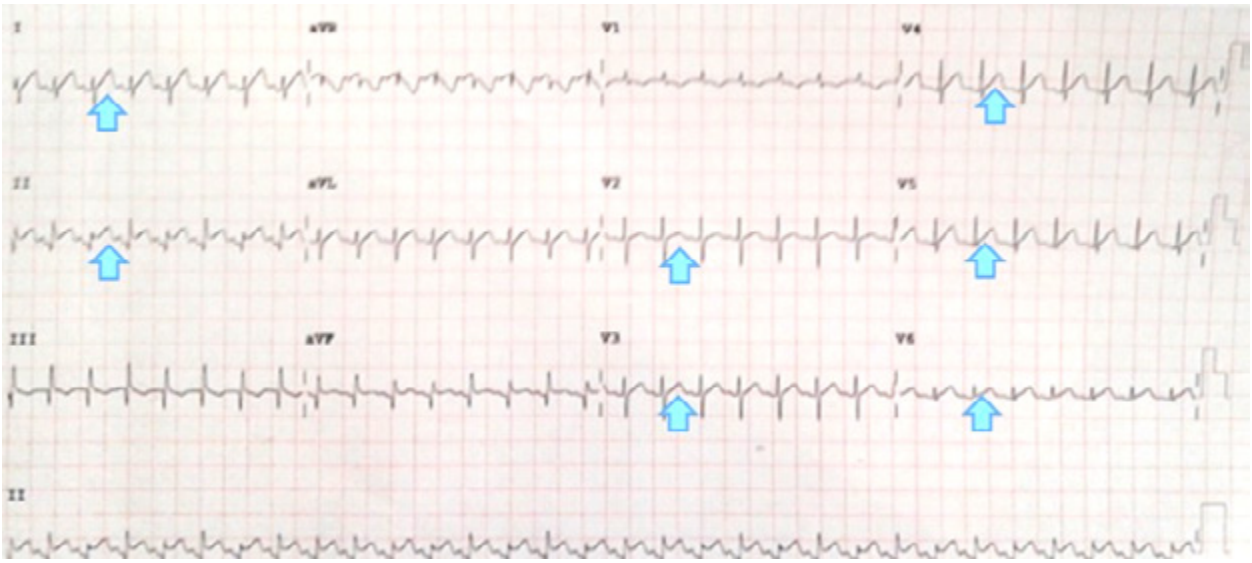


Figure 2. Evidence of electrocardiographic changes in the patient.
Source: Own elaboration based on the data obtained in the study.

In addition, an echocardiogram showed global pericardial effusion and the presence of thick membranes between the visceral and parietal pericardium, without signs of cardiac tamponade, adequate ventricular function, ventricular ejection fraction of 77% and central venous pressure of 8-10mmHg, without signs of pulmonary hypertension (Figure 3).

Respiratory distress, fever, tachycardia and altered state of consciousness without hypotension or alteration in distal perfusion were interpreted as a septic shock of pulmonary origin in hyperdynamic phase. The septic pattern and the echocardiographic findings increased the diagnostic probability of purulent pericarditis, so management with ceftriaxone (400 mg IV every 12 hours) and vancomycin (360 mg IV a day) was initiated. Then, the pediatric surgery service performed a pericardiotomy in the operating room and obtained about 50mL of turbid fluid with whitish membranes. Pericardial fluid analysis showed 2 600mm^3 of polymorphonuclear leukocytes at 90%, monocytes at 10%, glucose of 0.2 mg/dL, lactate dehydrogenase (LDH) of 591 IU/L, fresh red blood cells of 1 500, crenated red blood cells at 2 080 and total protein of 5.23 g/dL (pericardial fluid/serum proteins ratio: 0.9), which suggested a bacterial etiology.

Following the surgical procedure, a right jugular central venous catheter was passed (central venous pressure was not measured) and coupled to mechanical ventilation (CPAP PS), without requiring vasoactive support. The patient was admitted to the pediatric intensive care unit in normotensive condition, with tachycardia, adequate distal perfusion and under the effects of sedation and anesthesia (midazolam and fentanyl). Transfusion of 120cm^3 of red blood cells was carried out when arterial blood gas analysis yielded a figure of 7.1gr/dL.

Extubation was achieved within a few hours with adequate response, while fever was observed until the day after antibiotic management was initiated. The control blood count at that moment showed decreased white blood cell count, polymorphonuclear cells and platelets in relation to the previous exam (leukocytes: 24 600mm^3, polymorphonuclear leukocytes: 13 530mm^3, Hb: 11.4 g/dL, HCTO: 35%, platelets: 820 000mm^3) as well as decreased procalcitonin levels (5.21 ng/mL). The pericardiostomy tube was maintained for 4 days and blood cultures, as well as Ziehl-Neelsen, KOH and pericardial fluid culture tests, were negative.

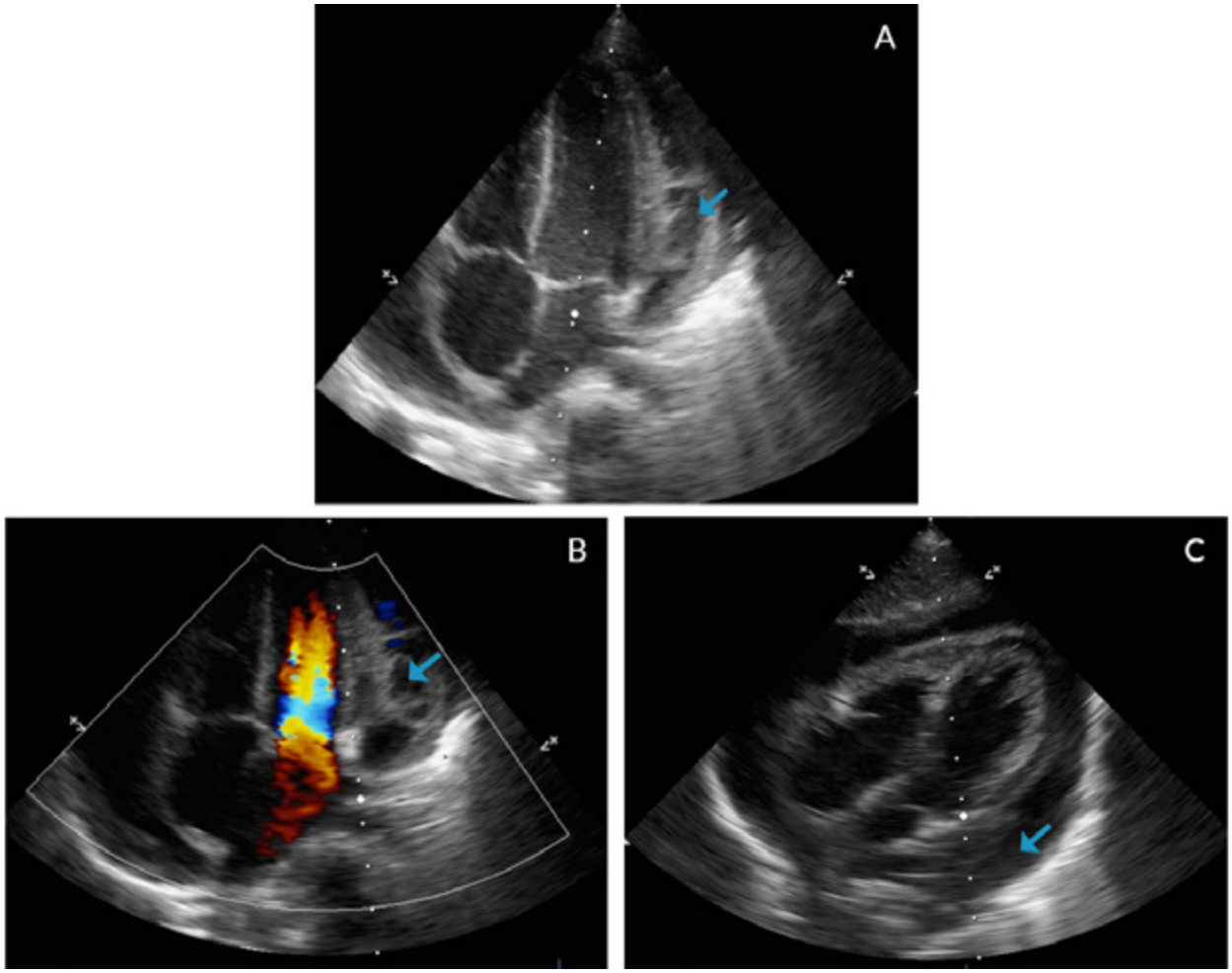


Figure 3. Echocardiogram with apical four chamber view (A, B) and coronal axial plane (C).
Source: Own elaboration based on the data obtained in the study.

To reduce the risk of toxicity and before clinical improvement, management with vancomycin was adjusted to clindamycin (80 mg IV every 6 hours) to complete 4 weeks of treatment. Studies to rule out immunological involvement were within normal limits (Table 1). Given the cyclic and echocardiographic improvement, discharge was authorized (Figure 4).

Table 1. Interpretation of laboratory results.

Exam	Interpretation
Maternal HIV	Negative
Immunoglobulins	IgE 3.45 UI/mL, IgA 84 mg/dL, IgG 641 mg/dL, IgM 161 mg/dL, within normal limits
Lymphocytes	CD3+: 74.3% (5 235.78), CD3+CD8+:27.17% (1 914.84), CD3+CD4: 43.79% (3 085.57), CD45+ (7 046.43), CD16+CD56+:4.33% (296.96): within normal limits
Antibodies against hepatitis B surface antigen	Reactive (422.7 U/L)
Thyroid function	TSH: 0.551 uU/ml, free T4: 1.58 ng/dL, T3: 0.87 ng/dL, within normal limits
Complement	Initial: C3 (73 mg/dL), C4 (8 mg/dL), low levels, Control: C3: 96 mg/dL, C4: 17 mg/dL, within normal limits
Culture of pericardial fluid	Culture for bacteria and M. tuberculosis: Negative
Dehidrorodamine test	Negative
Blood cultures	Negative

Source: Own elaboration based on the data obtained in the study.

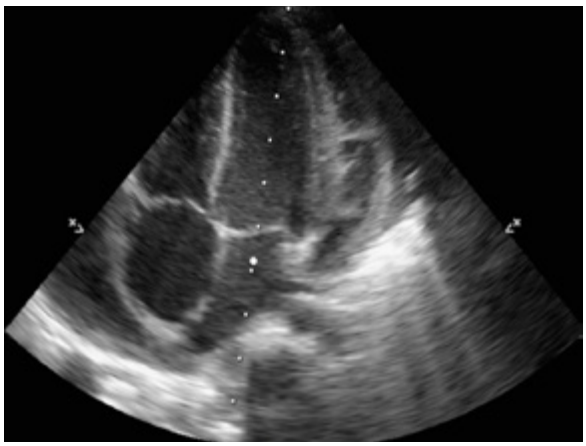


Figure 4. Echocardiogram taken before discharge.
Source: Own elaboration based on the data obtained in the study.

DISCUSSION

Purulent pericarditis is rarely observed in pediatrics and its incidence has not been properly established in this population. Most studies are documented in case reports and reports in infants are scarce. (5-10)

A study conducted in Uruguay (1) followed 19 pediatric patients with pericarditis, finding that 9 of them had a purulent origin and only 2 were younger than 12 months. This research showed that bacterial etiology is <50% and its occurrence in infants is unusual.

Considering a history of previous infections such as pneumonia, meningitis, osteomyelitis or subdiaphragmatic abscesses is important in children with purulent pericarditis. (2) In this case the infant presented bacterial pneumonia, which required hospitalization 2 months prior to this event.

Upon admission, the patient presented with respiratory distress, fever, tachycardia and increased oxygen requirements, which was explained by a septic pattern of pulmonary origin and pericarditis. Chest pain, despite being one of the most common symptoms in adults and older children, is difficult to interpret in infants. Clinically, there were no signs of cardiac tamponade (hypotension, jugular vein or veiled cardiac sounds), a complication that increases mortality and is explained by volume, accumulation rate, and the nature and etiology of the exudate between the pericardial layers that compress the cardiac chambers. (11)

The initial chest X-ray showed bibasilar and retrocardiac consolidations. The cardiothoracic index was 0.66 (>0.55) and was interpreted as cardiomegaly considering the patient's age. There was no evidence of pulmonary hyperflow or pulmonary venous congestion. The latter, together with electrocardiographic alterations (repolarization

disorders compatible with acute pericarditis) and absence of signs of heart failure (no signs of systemic or pulmonary congestion or low cardiac output), led to rule out pericarditis and associated pericardial effusion, without cardiac tamponade at first. (3,12)

The echocardiogram is the diagnostic method of choice to clarify radiological findings. (2,3) In this case, it showed a global pericardial effusion associated with the presence of thick membranes between the visceral and parietal pericardium, without cardiac tamponade, systolic-diastolic function involvement or alterations in the myocardium. Computed tomography (CT) is indicated when emergency echocardiogram is not available and when inconclusive echocardiographic results, poor response to treatment, atypical presentation, penetrating lesion, suspicion of neoplasms and pulmonary infections or mediastinitis are observed. (13) This patient did not require chest CT because of his rapid improvement, which reduced the possibility of complications.

Cytological and biochemical evaluation of pericardial effusion can help to obtain a diagnosis. The increase of LDH, the protein ratio in the pericardial fluid and serum >0.5 are characteristics that can lead to conclude that the fluid is compatible with the exudate. (14)

The protein ratio of the pericardial fluid and the serum proteins was 0.9 (>0.5), which suggested exudate characteristics. In the culture of this liquid, no bacterial isolation was obtained, despite the presence of purulent pericardial fluid. Some possible explanations include that these cultures are based on the detection of bacteria in the planktonic form (individual bacterial cells), but, over time, it has been demonstrated and accepted that the bacteria responsible for infections (including *Staphylococcus aureus* and *Streptococcus pneumoniae*) are associated in the form of biofilms, in such a way that they guarantee their survival by changing their properties and enhancing their virulence factors.

Associated bacteria in biofilms yield a negative result in culture tests directed at individual bacteria. (15) For this reason, it is necessary to use different diagnostic tests that can provide a fast identification of microorganisms, including PCR (polymerase chain reaction) (16) and ELISPOT (enzyme-linked immunospot) (17); the latter detects T cells specific for microbacteria and other microorganisms.

The treatment of purulent pericarditis includes pericardiocentesis, which is indicated for diagnosis and treatment, especially in the presence of significant effusion. (2-4) In this case, the septic status and the presence of pericardial effusion together with adherent membranes suggested purulent pericarditis. Empirical antibiotic treatment initiates with vancomycin and ceftriaxone and is maintained for 4 to 6 weeks, in order to eliminate the main microorganisms involved, namely, *S. aureus*, *Haemophilus influenzae* and *S. pneumoniae*. (2,3) In addition, bed rest and management with NSAIDs are recommended (2-4). In this case, antibiotic management with vancomycin was adjusted to clindamycin to reduce the risk of toxicity and support clinical and imaging improvement.

Other condition that can be considered for a pediatric patient presenting with this pathology is immunodeficiency, either primary or acquired and sometimes associated with malnutrition. (18) The main immunological etiologies were discarded for this patient and no degree of malnutrition was documented during the assessment.

The limitations of this study include the lack of availability of the clinical records of the patient prior to admission and non-ambulatory

follow-up of the patient. On the other hand, its strengths include comprehensive and interdisciplinary management, as well as permanent imaging support throughout the recovery process.

CONCLUSIONS

The relevance of this case lies in the presentation of the disease at a rare age. Signs and symptoms are non-specific, so high clinical suspicion is required based on the symptomatology and clinical and paraclinical findings that include radiography, electrocardiogram and echocardiography. (2-4) Timely diagnosis and early treatment decrease the probability of complications and favor better results.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Cabo E, Rios M, Assandri E, Vázquez M, Pujadas M, Chiesa P, *et al.*** Pericarditis aguda en pediatría. Incidencia y características. *Arch Pediatr Urug. 2015 [cited 2016 Nov 16];86(4):284-93. Available from: https://goo.gl/hZ93xw.*
2. **Johnson WH, Moller JH.** Pediatric cardiology. The essential pocket guide. *3rd ed. Minneapolis: Wiley Blackwell; 2014 [cited 2016 Oct 30]. Available from: https://goo.gl/PXehbz.*
3. **Park MK.** Park's the pediatric cardiology handbook. 5th ed. *Philadelphia: Elsevier; 2016.*
4. **Bergmann KR, Kharbanda A, Haveman L.** Myocarditis And Pericarditis In The Pediatric Patient: Validated Management Strategies. *Pediatr Emerg Med Pract. 2015 [cited 2016 Dec 28];12(7):1-22. Available from: https://goo.gl/Tt9p4A.*
5. **Flores-González JC, Rubio-Quiñones F, Hernández-González A, Rodríguez-González M, Blanca-García JA, Lechuga-Sancho AM, *et al.*** Pneumonia and Purulent Pericarditis Caused by Streptococcus pneumoniae, An Uncommon Association in the Antibiotic Era. *Pediatr Emerg Care. 2014;30(8):552-4. http://doi.org/f6f6wz.*
6. **Feinstein Y, Falup-Pecurariu O, Mitrica M, Berezin EN, Sini R, Krimko H,** *et al.* Acute pericarditis caused by *Streptococcus pneumoniae* in young infants and children : Three case reports and a literature review. *Int J Infect Dis. 2010;14(2):175-8. http://doi.org/d2q79t.*
7. **Donoso A, Díaz F, Bertrán K, Cruces P.** Pericarditis purulenta primaria por *Streptococcus pneumoniae*, a propósito de un caso clínico. *Rev Chil Pediatr. 2008 [cited 2016 Dec 4];79(6):623-8. Available from: https://goo.gl/YBbbVW.*
8. **Presnell L, Maeda K, Griffin M, Axelrod D.** A child with purulent pericarditis and Streptococcus intermedius in the presence of a pericardial teratoma: An unusual presentation. *J Thorac Cardiovasc Surg. 2013;147(3):e23-4. http://doi.org/cjxs.*
9. **Mirón L, Neyro S, Cheistwer A, Muracciole B, Ortellao G, Martinez-Iriart E.** Pericarditis como presentación inicial de enfermedad meningocócica diseminada. *Arch Argent Pediatr. 2013;111(6):144-7. http://doi.org/cjxt.*
10. **Cakir O, Gurkan F, Balci AE, Eren N, Dikici B.** Purulent pericarditis in childhood: Ten years of experience. *J Pediatr Surg. 2002;37(10):1404-8. http://doi.org/cqx8tk.*
11. **Douchin S. Bost-Bru C, Pagnier A.** Pericarditis aguda del niño. *EMC - Pediatría. 2017 [cited 2018 Jan 10];52(4):1-9. Available from: https://goo.gl/kNUJ4i.*
12. **Bruch C, Schmermund A, Dagres N, Bartel T, Caspari G, Sack S, *et al.*** Changes in QRS Voltage in Cardiac Tamponade and Pericardial

Effusion: Reversibility After Pericardiocentesis and After Anti-Inflammatory Drug Treatment. *J Am Coll Cardiol. 2001;38(1):219-26. http://doi.org/fshckq.*

13. **Klein AL, Abbara S, Agler DA, Appleton CP, Asher CR, Hoit B, *et al.*** American Society of Echocardiography Clinical Recommendations for Multimodality Cardiovascular Imaging of Patients with Pericardial Disease. Endorsed by the Society for Cardiovascular Magnetic Resonance and Society of Cardiovascular Computed Tomography. *J Am Soc Echocardiogr. 2013;26(9):965-1012. http://doi.org/cjx4.*
14. **Ben-Horin S, Bank I, Schinfel A, Kachel E, Guerra V, Livneth A.** Diagnostic Value of the Biochemical Composition of Pericardial Effusions in Patients Undergoing Pericardiocentesis. *Am J Cardiol. 2007;99(9):1294-7. http://doi.org/b42hkd.*
15. **Chao Y, Marks LR, Pettigrew MM, Hakansson AP.** Streptococcus pneumoniae biofilm formation and dispersion during colonization and disease. *Front Cell Infect Microbiol. 2015;4:194. http://doi.org/f3nktt.*
16. **Levy PY, Habib G, Collart F, Lepidi H, Raoult D.** Etiological diagnosis of pericardial effusion. *Future Microbiol.* 2006;1(2):229-39. http://doi.org/dr3zpn.
17. **Ewer K, Deeks J, Alvarez L, Bryant G, Waller S, Anderson P, *et al.*** Comparison of T-cell-based assay with tuberculin skin test for diagnosis of Mycobacterium tuberculosis infection in a school tuberculosis outbreak. *Lancet. 2003;361(9364):1168-73. http://doi.org/dv8csw*
18. **Onyeama CO, Okomo U, Garba D, Njai PC, Tapgun M, Corrah T.** Staphylococcal purulent pericarditis in a malnourished Gambian child: A case report. *Int J Cardiol. 2007;119(3):392-4. http://doi.org/fbprw2.*

https://doi.org/10.15446/cr.v4n1.65771

BOUVERET SYNDROME, A RARE FORM OF GALLSTONE ILEUS. CASE REPORT

Keywords: Cholelithiasis; Biliary fistula; Duodenal obstruction; Gallstones.
Palabras clave: Colelitiasis; Fistula biliar; Obstrucción duodenal; Cálculos biliares.

Lúver Macías MD
Adriano Rocha MD
Marcos Alvarado MD
José Chung MD
David Yépez MD
Department of General Surgery
- Hospital Luis Vernaza - Guayaquil - Ecuador.

Corresponding author:
Lúver Macías. Department of General Surgery
- Hospital Luis Vernaza - Guayaquil - Ecuador.
luver16mj@hotmail.com

Received: 20/06/2017 **Accepted:** 15/01/2018

RESUMEN

Introducción. El síndrome de Bouveret es una entidad poco frecuente de íleo biliar que provoca obstrucción gástrica debido a la presencia de un lito biliar a nivel de píloro o duodeno y secundaria a una fistula biliodigestiva. Esta enfermedad es de difícil diagnóstico y una de sus complicaciones es la hemorragia digestiva alta; su manejo es quirúrgico y se recomienda en dos tiempos, aunque se puede manejar por endoscopia alta en aquellos cálculos <2.5cm. El pronóstico es bueno, con un post-operatorio sin novedades.

Presentación del caso. Paciente de 63 años con cuadro clínico de 15 días de evolución de dolor en epigástrico y vómitos biliosos incontables. Se realizaron imágenes diagnósticas donde se evidenció masa calcificada en duodeno II, por lo que la paciente fue intervenida quirúrgicamente: se progresó el cálculo a yeyuno, se realizó enterotomía y se extrajo el lito. Al quinto día fue dada de alta sin novedades.

Conclusión. El síndrome de Bouveret es una entidad rara que requiere de una adecuada valoración médica y de métodos auxiliares de imágenes para un diagnóstico oportuno.

ABSTRACT

Introduction: Bouveret syndrome is a rare form of gallstone ileus that causes gastric obstruction due to the presence of a gallstone in the pylorus or duodenum, secondary to biliodigestive fistula. This condition is difficult to diagnose and one of its main complications is hemorrhage in the digestive tract. Two-stage surgical management is recommended, although it can also be managed through upper endoscopy in gallstones smaller than 2.5cm. Prognosis and postoperative period are good.

Clinical case: 63-year-old patient with a clinical picture of 15 days of pain in the epigastrium and bilious vomiting. Imaging showed a calcified mass in the second part of the duodenum, which was surgically treated by entering the jejunum, performing an enterotomy and extracting the gallstone. The patient was discharged on the fifth day without any symptoms.

Conclusion: Bouveret syndrome is a rare entity that requires better medical assessment and ancillary imaging techniques to achieve a timely diagnosis.

INTRODUCTION

Gallstone ileus (GI) refers to a mechanical obstruction of the gastrointestinal tract caused by impaction of one or more gallstones in the bowel lumen. This condition accounts for 1-4% of all intestinal occlusions in adults. In most cases, obstruction occurs in the terminal ileum (60%), followed by the proximal ileum (25%), and less frequently by the jejunum (9%), the sigmoid colon (4%) and the duodenum (2%). (1)

Bouveret syndrome (BS) is a very rare form of gallstone ileus caused by the passage and impaction of a gallstone through a cholecystoduodenal fistula into the duodenum, which results in gastric outlet obstruction; this condition was first described by Leon Bouveret in 1896. (2,3)

Six risk factors are involved in BS presentation: 1) history of cholelithiasis; 2) repeated episodes of cholecystitis; 3) female gender; 4) age >60 years; 5) comorbidity, and 6) gall-

stones >2.5cm in diameter. (4) This disease usually occurs in elderly women, and in people with an average age of 70-75 years due to the presence of a large gallstone (only one in more than 90% of cases) within the pyloric or duodenal canal. (5)

The clinical picture of BS is usually non-specific, and may present with clinical symptoms of intestinal obstruction characterized by abdominal pain, nausea, vomiting, food intolerance and absence of flatus and stools. (6) The objectives of this work are to report a case of this rare pathology, to comment on its particular characteristics and to present a brief literature review.

CASE PRESENTATION

A 63-year-old male, mestizo patient, who works as a farmer, from the rural area of Milagro-Ecuador, with a history of cholelithiasis 3 years before consultation, visited the emergency service of the Luis Vernaza Hospital due to a clinical picture of 15 days of evolution characterized by moderate epigastric pain that exacerbated after the excessive intake of food. Symptoms included nausea, countless bilious vomiting and malaise.

The patient reported that he had been hospitalized in another institution for about 4 days and had been scheduled for laparoscopic cholecystectomy, but it was cancelled due to the presence of a mass around the gallbladder that involved the liver. Physical examination revealed pale facies, symmetrical thorax (clear and ventilated lung fields), abdominal distension, pain on deep palpation in the upper abdomen, positive Murphy's sign, and increased hydroaeric noises on auscultation. No megalies were observed.

Vital signs on admission were: blood pressure of 120/80 mmHg, temperature of 38°C, oxygen saturation of 98%, pulse of 80 bpm, and respiratory rate of 19 per minute. Blood count test reported leukocytes 19.05 x103/ul; neutrophils 82.3%; 9% lymphocytes; red blood cells 3.78 M/ul; hemoglobin 10.1 g/dl; hematocrit 29.9%; platelets 626 x103/ul; bilirubin: total 0.49 mg/dl, direct 0.21 mg/dl and indirect 0.28 mg/dl; electrolytes: sodium 147 mEq/L, potassium 3 mEq/L and chlorine 99 mEq/L; gamma glutamyl transferase 120 M/L; alkaline phosphatase 107 U/L; Amylase 102 U/L; lipase 142 U/L, and PCR 47.69 mg/L. After the tests were performed, imaging was requested, revealing the presence of the large gallstone in the second part of the duodenum (Figure 1).

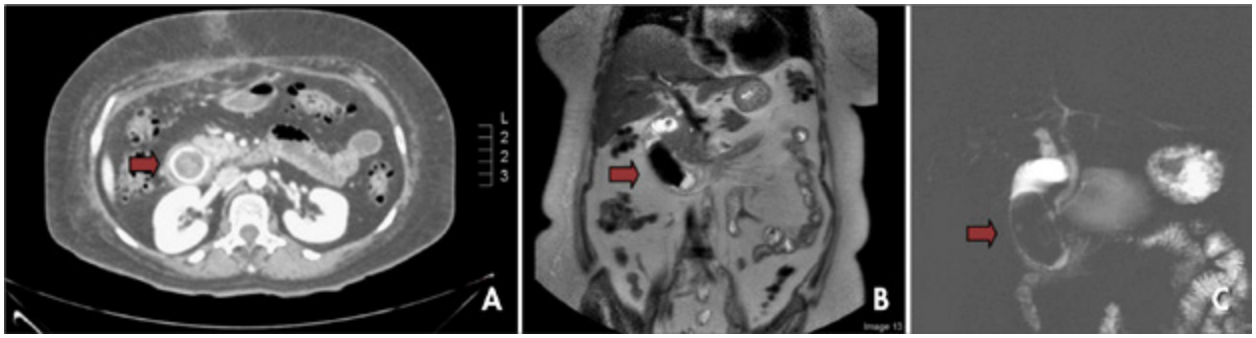


Figure 1. A) Three-phase computed tomography of the abdomen in venous phase; B) and C): T2-weighted SPAIR magnetic resonance cholangiography.

Source: Own elaboration based on the data obtained in the study.

The patient underwent surgery, during which the abdominal cavity was approached by exploratory laparotomy, observing a plastron in the gallbladder topography that involved the greater omentum, the duodenum and the hepatic flexure of the colon. An exploration was made from the angle of Treitz to the duodenum, palpating a 10x5cm stone tumor, which was manually taken to the jejunum at 50cm from the Treitz angle (Figure 2).

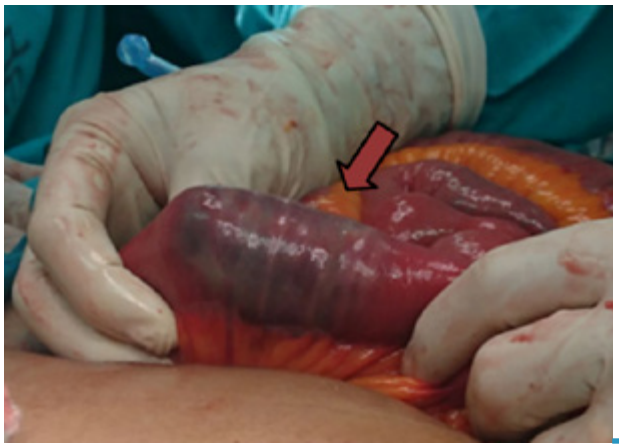


Figure 2. Hard mass found in the second part of the duodenum.
Source: Own elaboration based on the data obtained in the study.

Once the stone was taken to the jejunum at 50 cm from the Treitz angle, a longitudinal enterotomy, parallel to the major axis of the stone, was performed (Figure 3A). The stone was extracted (Figure 3B), and suturing was performed transversely with prolene 3/0.

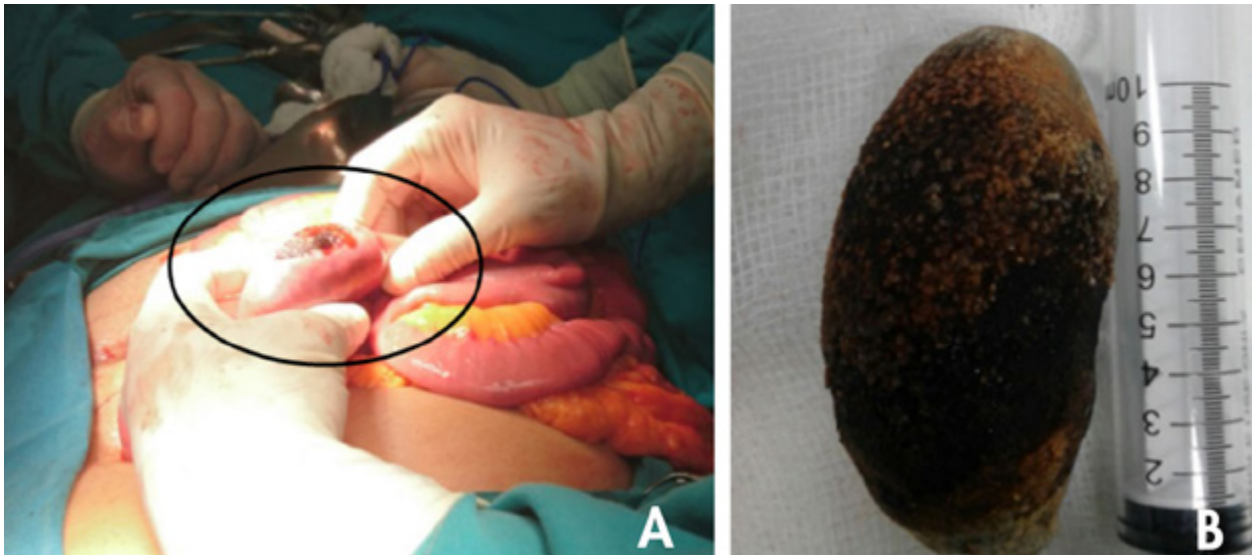


Figure 3. A) Enterotomy parallel to the major axis of the stone (circle); B) Extraction of the biliary stone, with dimensions of 10x5cm.
Source: Own elaboration based on the data obtained in the study.

Within the first two post-operative days, the patient did not present any complications. On the third day, he was able to pass flatus and evacuate the bowel, so he started oral refeeding with clear liquids. On the next day, the patient was able to start a general diet that was satisfactorily tolerated. He did not report any symptoms or fever, so he was discharged on the fifth day. Outpatient postoperative controls were scheduled at 15 days. A second surgery was not considered since there was no evidence of any intestinal obstruction complication that required a new hospitalization.

DISCUSSION

Currently, biliary lithiasis is one of the most common digestive diseases, with prevalence between 10% and 20% in western countries, being more frequent in seniors and women. Although most biliary stones are clinically silent, symptoms or complications appear in 20% of cases. The most frequent location is the gallbladder (85%), but they can be found anywhere in the biliary tract. (7) Cholecystoduodenal fistula and GI are the least frequent complications. (8)

The formation of the fistula between the gallbladder and the lumen of the digestive tube may be related to a chronic inflammatory process, which produces less vesicular arterial flow and venous and lymphatic drainage. This situation may cause an increase of the vesicular intraluminal pressure that would favor necrosis and fistulization of the biliodigestive barrier. Cholecystoenteric fistulas occur in 0.3-0.5% of patients with cholelithiasis, and most of them are cholecystudymal (60%), cholecystocolonic (17%), cholecystogastric (5%) and choledochoduodenal (5%). (9)

BS is a rare and high risk complication of cholecystitis that affects only 0.9-3.2% of patients with a history of gallstones. (10) Its clinical picture is nonspecific and its most common symptoms are epigastric pain, nausea and vomiting. In the most severe cases, upper gastrointestinal bleeding can occur due to the erosion of the duodenal mucosa as a consequence of gallstone migration. (11) The most frequent complication is electrolyte imbalance. (12)

The diagnosis of this disease is delayed because it can be mistaken with other causes of gastroduodenal obstruction. For this reason, differential diagnosis should be made to rule out bezoar, tumors, peptic stenosis and superior mesenteric artery syndrome, among others. (13) An intestinal obstruction occurs when the size of the gallstone is at least 2.5cm in diameter, but, in general, for BS to occur, the dimensions must be larger. (14)

Since the symptoms are nonspecific, a high level of clinical suspicion is required to achieve a diagnosis. Complementary exams can be useful; for example, abdomen radiography can show the Rigler triad (aerobilia, dilated small bowel loops with air-fluid levels and ectopic biliary lithiasis), which is pathognomonic of GI, although it is observed in less than 15 % of the cases.

In case of diagnostic doubts, imaging tests can be used, such as upper gastrointestinal endoscopy, which allows direct visualization of the stone causing obstruction in 69% of patients. Also, abdominal ultrasound can reveal gallstones in 75% of BS patients, of which 23% can be visualized in the duodenum, while pneumobilia can be seen in 45%. Computed tomography can find perivesicular inflammatory changes, pneumobilia, cholecistoduodenal fistula, filling defects, level of

obstruction and features of the gallbladder. Finally, magnetic resonance cholangiography can be useful to describe fluids and stones. Other echographic signs are contraction of the gallbladder, thinning of the wall of the gallbladder and dilatation of the duodenum. (4,15,16)

The ideal treatment of BS is to resolve the digestive obstruction and the biliary disease during the same surgery through gastrotomy or enterotomy, gallstone extraction, cholecystectomy and fistula repair. However, most authors recommend a two-stage treatment, resolving first the obstructive disease and then addressing the biliary condition, if necessary. Considering that patients are usually old, with an important comorbidity and poor general conditions that may lead to an emergency, long surgical times are not recommended, therefore, only gastrotomy or enterotomy and gallstone extraction should be performed. (17)

Another treatment option is oral endoscopy after performing endoscopic mechanical lithotripsy and the subsequent oral extraction of the fragments. Regarding lithiasis impact in the terminal ileum, cases of extraction by colonoscopy have been described. (18) Lithiasis size is an important factor to consider, since stones >2.5cm are difficult to extract by endoscopy, so there is also risk of impaction in the esophagus. (5)

CONCLUSIONS

BS is a rare case of GI caused by gastric obstruction due to the presence of one or more gallstones in the duodenum secondary to a cholecystoduodenal fistula, which occurs more frequently. This is a late diagnosis disease that requires ancillary techniques such as radiography, ultrasound, abdominal CT, magnetic resonance imaging and upper endoscopy. The ideal treatment is to remove the stone endoscopically, but most authors of the analyzed reviews prefer a two-stage intervention, resolving the intestinal obstruction first and then the fistula.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Rodríguez-Hermosa JJ, Codina-Cazador A, Gironés-Villa J, Roig-García J, Figa-Francesch M, Acero-Fernández D.** Íleo biliar: resultados de análisis de una serie de 40 casos. *Gastroenterol Hepatol.* 2001;24(10):489-94. http://doi.org/f2ks55.
2. Puri V, Lee RW, Amirlak BA, Lanspa SJ, Fitzgibbons RJ Jr. Bouveret syndrome and gallstone ileus. *Surg Laparosc Endosc Percutan Tech. 2007;17(4):328-30. http://doi.org/bhv339.*
3. **Englert ZP, Love K, Marilley MD, Bower CE.** Bouveret syndrome: gallstone ileus of the duodenum. *Surg Laparosc Endosc Percutan Tech. 2012;22(5):301-3. http://doi.org/f4b9v3.*
4. **Doycheva I, Limaye A, Suman A, Forsmark CE, Sultan S.** Bouveret´s síndrome: case report and review of the literature. *Gastroenterol Res Pract.* 2009;2009:914951. http://doi.org/dcg8mn.
5. Nickel F, Müller-Eschner MM, Chu J, von Tengg-Kobligk H, Müller-Stich BP. Bouveret's syndrome: Presentation of two cases with review of the literature and development of a surgical treatment strategy. *BMC Surg.* 2013;13:33. http://doi.org/gbdqvr.
6. **Mengual-Ballester M, Guillén-Paredes MP, Cases-Baldó MJ, García-García ML, Aguayo-Albasini JL.** Hemorragia digestiva y obstruc-

ción intestinal alta como presentación del síndrome de Bouveret. *Cir Cir.* 2011;79(6):557-9.

7. **Horton J, Bilhartz L.** Litiasis biliar y sus complicaciones. In: Feldman M, Sleisenger MH, Friedman LS. Enfermedades gastrointestinales y hepáticas. Fisiopatología, diagnóstico y tratamiento. 7th ed. Buenos Aires: Editorial Médica Panamericana; 2004. p. 1126-1152.
8. **Lobo DN, Joblin JC, Balfour TW.** Gallstone ileus: Diagnostic pitfalls and therapeutic successes. *J Clin Gastroenterol.* 2000;30(1):72-6.
9. **Liew V, Layani L, Speakman D.** Bouverets syndrome in Melbourne. *ANZ J Surg.* 2002;72(2):161-3. http://doi.org/fh4fzt.
10. **Stinton LM, Shaffer EA.** Epidemiology of Gallbladder Disease: Cholelithiasis and Cancer. *Gut Liver.* 2012;6(2):172-87. http://doi.org/cj3k.
11. **Sánchez-García M, Tomoiu IG, Reina-Escobar D, Delgado-Casado JA.** Causa infrecuente de obstrucción intestinal. *Semergen.* 2015;41(4):18-20. http://doi.org/f2tffm.
12. **Alvarez-Chica LF, Bejarano-Cuéllar W, Rojas-Cardozo OL.** Ileo biliar y sindrome de Bouveret. Lo mismo pero distinto: descripción de dos casos y revisión de la literatura. *Rev. Col Gastroenterol.* 2010;25(1):86-93.
13. **Báez-García JJ, Martinez-Hernandez P, Iriarte-Gallego G.** Síndrome de Bouveret: reporte de un caso. *Rev Gastroenterol Mex.* 2009;74(2):118-21.
14. **Vidal O, Seco JL, Álvarez A, Triñanes JP, Serrano LP, Serrano SR.** Síndrome de Bouveret: cinco casos. *Rev Esp Enferm Digest.* 1994;86:839-44.
15. **Cappell MS, Davis M.** Characterization of Bouveret's syndrome: A comprehensive review of 128 cases. *Am J Gastroenterol.* 2006;101(9):2139-46. http://doi.org/drzmqq.
16. **Marchall J, Hayton S.** Bouveret's syndrome. *Am J Surg.* 2004;187(4):547-8. http://doi.org/b3b4w4.
17. **Blanco-Gnzález FJ, Artigas-Martín JM, Fuertes-Ferández MI, Hernando-Almudí E, García-Calleja JL, del Río-Marco F.** Síndrome de Bouveret: diagnóstico preoperatorio por imagen. *Cir Esp.* 2000;67(6):619-20.
18. **Nuño-Guzmán CM, Marín-Contreras ME, Figueroa-Sánchez M, Corona JL.** Gallstone ileus, clinical presentation, diagnostic and treatment aproach. *World J Gastrointest Surg.* 2016;8(1):65-76. http://doi.org/cj3n.

https://doi.org/ideaspilarojas@gmail.com

RECURRENT NEUROCYSTICERCOSIS OF THE FRONTAL LOBE. CASE REPORT.

Keywords: Neurocysticercosis; *Taenia solium*; Frontal lobe; Colombia.
Palabras clave: Neurocisticercosis; *Taenia solium*; lóbulo frontal; Colombia.

Ángela Medina-Ortega
David López-Valencia
Medical Program- Faculty of Health Sciences
Universidad del Cauca
- Popayán - Colombia.
Centro de Estudios en Microbiología y Parasitología (CEMPA)
- Faculty of Health Sciences - Universidad del Cauca
- Popayán - Colombia.

Luis Reinel Vásquez-Arteaga
Centro de Estudios en Microbiología y Parasitología (CEMPA)
- Faculty of Health Sciences - Universidad del Cauca
- Popayán - Colombia.
Department of Internal Medicine - Faculty of Health Sciences
-Universidad del Cauca - Popayán - Colombia

Corresponding author
David López Valencia. Medical Program,
Faculty of Health Sciences, Universidad del Cauca.
E mail drdlopez1@gmail.com

Received: 22/06/2017 **Accepted:** 09/01/2018

ABSTRACT

Introduction: Neurocysticercosis (NCC) is the result of ingestion of pork tapeworm eggs (*Taenia solium*) from an individual with taeniasis (taeniasis/cysticercosis complex). This disease causes the highest helminthic-related morbidity and mortality rates due to its deleterious effects on the central nervous system. 80% of the cases can be asymptomatic and 20% show non-specific clinical manifestations.

Case presentation: The following report presents the case of a patient with headache, dromomania, intracranial hypertension syndrome, and cognition and gait impairment. A brain CT showed a right frontal subcortical cyst and bilateral frontoparietal calcified nodules. Neurocysticercosis of the frontal lobe was suspected as the main diagnosis considering the clinical manifestations, anamnesis and local epidemiology.

Discussion: *T. solium* reinfection in the right frontal lobe was suspected in this patient due to perilesional edema, calcified nodules randomly distributed on the imaging and the information supplied by his relatives during anamnesis. Symptoms and signs of NCC depend on localization, number, dimensions, cysticercus stage (vesicular, colloidal, granular-nodular and calcified nodule), genotype and immune status of the host. Between 60 and 90% of cysticerci are mainly observed in the brain parenchyma, but other less frequent localizations include ventricles, subarachnoid space, eyes, meninges and spinal cord.

Conclusions: It is important to know and educate the community about the life cycle of parasites, epidemiology, prevention measures and clinical manifestations of neurocysticercosis in order to make a timely diagnosis and administer an effective treatment.

RESUMEN

Introducción. La neurocisticercosis (NCC) es causada por la ingesta de huevos de la tenia del cerdo (*Taenia solium*) provenientes de un individuo con teniosis (complejo teniasis-cisticercosis). Esta enfermedad produce la mayor morbimortalidad por sus efectos dañinos sobre el sistema nervioso central. El 80% de los casos pueden ser asintomáticos y el 20% restante presenta manifestaciones clínicas que son inespecíficas.

Presentación del caso. Paciente con cefalea, tendencia dromomaníaca, síndrome de hipertensión intracraneal, deterioro cognitivo y alteración de la marcha. Se realizó una tomografía computarizada que reveló un quiste subcortical frontal derecho y calcificaciones frontoparietales bilaterales. Como diagnóstico principal se sospechó NCC del lóbulo frontal por su cuadro clínico, anamnesis y epidemiología regional.

Discusión. Se sospecha que el paciente cursaba con reinfección por *T. solium* en el lóbulo frontal derecho por la presencia de edema perilesional, los nódulos calcificados distribuidos aleatoriamente en la imagen y la información suministrada por sus familiares durante la anamnesis. Los signos y síntomas de la NCC dependen de la ubicación, el número, las dimensiones, los estadios del cisticerco (vesicular, coloidal, granular-nodular y nódulo calcificado), el genotipo y el estado inmune del hospedero. El 60-90% de los cisticercos se ubican en el parénquima cerebral, siendo menos frecuentes las ubicaciones ventricular, subaracnoidea, ocular, meníngea y medular.

Conclusiones. Es importante educar a la comunidad, por un lado, sobre el ciclo de vida del parásito *T. solium* y su epidemiología y, por el otro, acerca de las medidas de prevención y las manifestaciones clínicas de la NCC, esto con el fin de realizar un diagnóstico oportuno y un manejo efectivo.

INTRODUCTION

Neurocysticercosis (NCC) is the most frequent helminth infection of the central nervous system and the main cause of acquired epilepsy worldwide (30%). (1-3) NCC occurs when an individual ingests the eggs of the *Taenia solium* tapeworm (carried by pigs), previously expelled in the feces of another individual with teniosis. (4)

Currently, there are 50 million people affected by NCC around the world, which makes it an endemic disease in Colombia and other Latin American countries. (5) In 2010, the World Health Organization (WHO) listed NCC as an unattended zoonosis in permanent expansion, thus turning it into a serious public health issue in developed countries due to migratory phenomena. (3,6)

Flórez-Sánchez *et al.* (7) found that the prevalence of seropositivity for cysticercosis varies in the Colombian population (0.53-40.19%) and that the Vaupés department has the highest seroprevalence rates.

The objective of this article is to promote knowledge about the heterogeneous manifestations of neuroinfection by *T. solium* and highlight the importance of the prevention measures against the disease.

CASE PRESENTATION

A 51-year-old male patient from the rural area of Caldono (Cauca, Colombia), farmer, of low socioeconomic status and with basic primary education was admitted to a hospital in Popayán in November 2015, describing a single NCC event successfully treated with albendazole 2 years before. The reason for consultation was the impossibility of walking by himself. During anamnesis, his relatives reported frequent consumption of undercooked pork, lack of sewage service and lack of knowledge of proper hand washing by the patient.

The patient presented with a clinical picture of 8 months of evolution consisting of progressive gait impairment, loss of sphincter control, left hemiparesis and headache. On admission, he did not obey orders, presented dromomania, inappropriate language, dysarthria, cognitive deterioration, space-time disorientation, verbal-motor automatisms (he constantly said "I am alone"), pathological palmomental reflexes and bilateral prehension.

The neurology service requested a computed tomography (CT) that revealed a dilation of the supratentorial ventricular system and a right frontal subcortical cystic lesion that created a mass effect with midline shift. In addition, he presented perilesional edema and small residual bilateral frontoparietal calcifications, suggesting sequelae of NCC (Figure 1). Blood count, C-reactive protein (CRP) and renal function were normal. Pharmacological management was initiated with albendazole at an oral dose of 1600mg every 24 hours, dexamethasone 8mg IV every 8 hours, paracetamol at an oral dose of 1g every 8 hours and omeprazole at an oral dose of 20mg every 24 hours. The patient did not report any side effect caused by these drugs.

A week after admission, the patient presented with left hemiplegia and sialorrhea, and he did not have any verbal response. A craniotomy was performed to remove the cyst and conduct a histopathological study, while

a ventriculoperitoneal shunt was arranged to reduce intracranial pressure (Figure 2). Macroscopically, neurosurgery reported a frontal cyst of greenish content with walls strongly adhered to the parenchyma and the frontal horn of the lateral ventricle. Histopathological analysis confirmed the suspicion of NCC and reported reactive gliosis. The patient evolved satisfactorily, did not present any type of sequelae and was discharged.

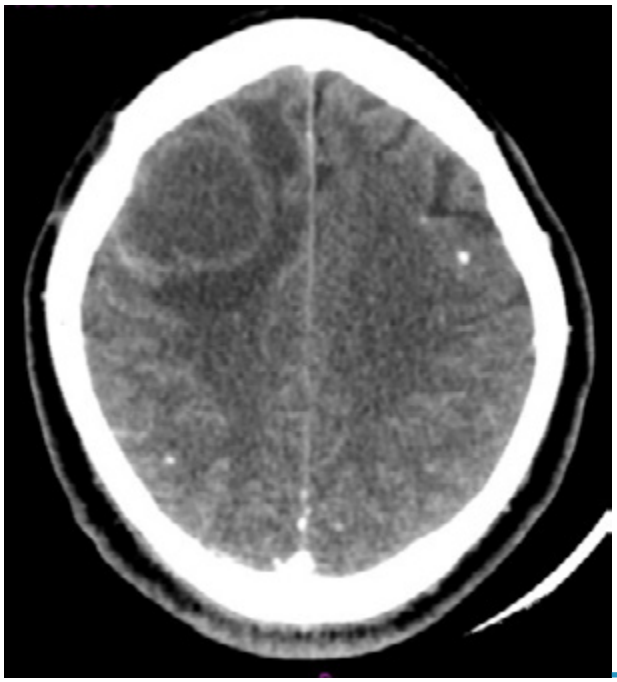


Figure 1. Computed tomography with right frontal subcortical cystic lesion, perilesional edema and calcified nodules.
Source: Own elaboration based on the data obtained in the study.

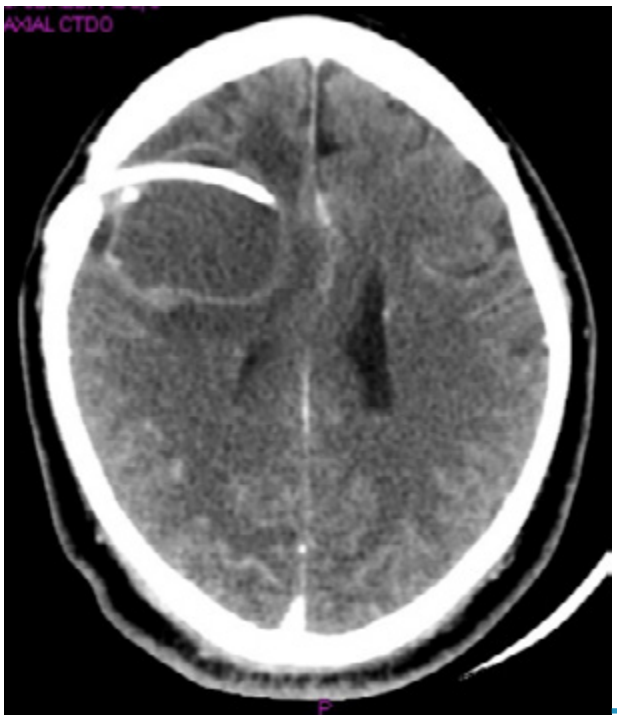


Figure 2. Computed tomography with right frontal subcortical cystic lesion, midline shift and ventriculoperitoneal shunt.
Source: Own elaboration based on the data obtained in the study.

Reinfection was suspected due to a previous history of NCC (a significant risk factor), the presence of calcified nodules in the imaging and regional epidemiology.

DISCUSSION

The patient presented with a frontal syndrome characterized by left hemiparesis, disobedience of orders, dromomania, cognitive impairment, space-time disorientation and verbal-motor automatism, which are related to cysticercosis cysts in the right frontal lobe. According to anatomical distribution, 60-90% of cysticerci are located in the cerebral parenchyma (8), being less frequent in the ventricles, subarachnoid space, eyes, meninges and spinal cord. (9)

When the *T. solium* larva is viable, initial neurological symptoms are caused by compression, in most cases. When the cysticercus dies, intense inflammation with exudate, periarteritis and endarteritis is usually observed, which can close the vascular lumen and impede the normal flow of cerebrospinal fluid, favoring the presence of hydrocephalus and intracranial hypertension. (10) Following the inflammatory response, the cysticercus usually reaches its final stage (nodular calcified), which has been associated with epilepsy. (10) However, the patient reported in this article did not develop hydrocephalus or epilepsy during his stay in the service.

Reinfection by *T. solium* in this patient was suspected due to the presence of calcified nodules and perilesional edema (11) —compatible with active cysticercosis— observed on the CT scan and based on the information provided by his relatives during anamnesis: history of NCC 2 years before, frequent consumption of undercooked pork, lack of drinking water and sewage services and lack of proper hand washing habits. (12,13) The patient and his relatives were given the pertinent recommendations and measures to prevent the disease: correct hand washing (soap and water or glycerinated alcohol) before and after consuming food and after contact with another person (4); adequate disposal of feces in the rural area; exclusive consumption of drinking water; environmental sanitation, and management of pigs under the current regulations. (9,14)

The signs and symptoms of NCC depend on the location, number, dimensions, cysticercus stage (vesicular, colloidal, granular-nodular and calcified nodule), genotype and immune status of the host. (4) Headaches and seizures are the most frequent clinical manifestations; visual or psychiatric disorders, hydrocephalus and meningitis may also occur. (3,15)

The prevalence of NCC is higher in rural areas, where people work with pigs and sanitary conditions are often deficient. (3,5,16) This disease compromises the health of those affected, the livelihood of agricultural communities and their economy due to absenteeism, increase in health costs and community stigmatization. (3,16) According to Flórez-Sánchez *et al.* (7), the prevalence of cysticercosis in Colombia ranges between 0.53% and 40.19% in Caldas and Vaupés, respectively. In Cauca, Vásquez-Arteaga *et al.* (14) found a seroprevalence of 55.2% in patients with neurological symptoms of five municipal hospitals. This information is very useful for the region, since the history of NCC and the neurological manifestations compatible with the disease make it necessary to discard it.

In southwestern Colombia, cysticercus is known as "*granalla*", "*granizo*", "*pepa*" or "*pepita*",

and is considered as a part of the pig. Actually, the inhabitants of this region think that they add a taste to the meat. In addition, knowledge on the life cycle of the parasite is deficient, which leads to difficulties when making promotion and prevention campaigns. (17) Understanding the life cycle and behavior of the teniasis/cysticercosis complex (TCC) is important when considering implementing public health policies and promotion and prevention campaigns.

Depending on the development stage of *T. solium*, if the cysticercus or eggs are swallowed, teniosis or cysticercosis, respectively, will occur (Figure 3). It should be noted that both diseases could occur simultaneously in the same individual. (9)

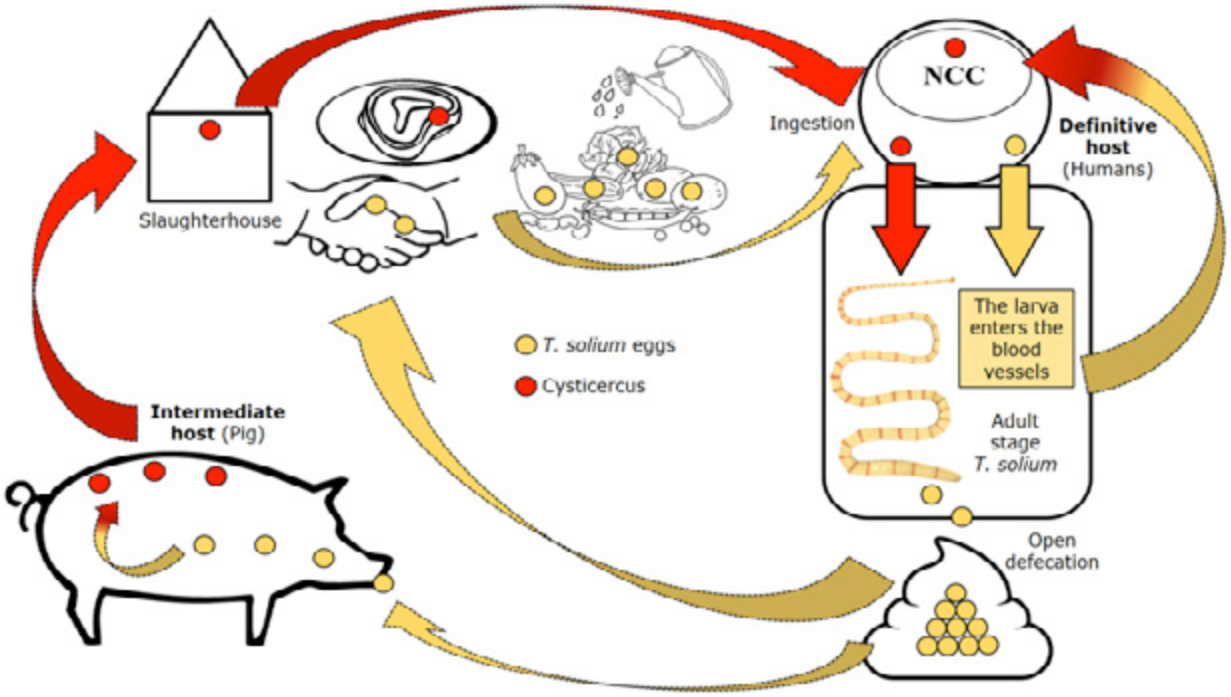


Figure 3. Life cycle of the teniasis/cysticercosis complex.
Source: Own elaboration based on the data obtained in the study.

As seen in Figure 3, humans develop teniosis when cysticerci are ingested. The larvae mature to the adult form of T. solium and release the eggs to the environment; then, pigs consume them and suffer from cysticercosis. When animals are slaughtered, if there are deficiencies in sanitary control, pork meat is commercialized and humans end up consuming cysticerci and developing teniosis. Similarly, the definitive host may present with cysticercosis when consuming food irrigated with water contaminated by the eggs of the adult tapeworm. Contact with sick individuals is an important way of contagion, being the main risk factor for TCC infection. (18)

This case shows strength in diagnosis, epidemiology and clinical foundation. The importance of anamnesis should be highlighted, since it provides a guide for patient's approach. In the same way, intervention for promotion and prevention is highlighted as relevant. However, this case did not include a

molecular test that identified IgM antibodies for *T. Solium* due to local limitations.

CONCLUSIONS

Making a timely diagnosis along the process (medical history, imaging and laboratory tests) is important when the history, signs and symptoms are compatible with NCC.

Providing comprehensive management to the patient, in this type of cases, is necessary, first, to carry out a complete cysticidal treatment and, second, to provide information to patients, relatives and the community in general about the prevention measures against NCC.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Del Brutto OH, Engel JJ, Eliashiv DS, Garcia HH.** Update on Cysticercosis Epileptogenesis: the Role of the Hippocampus. *Curr Neurol Neurosci Rep. 2016;16(1):1. http://doi.org/f8hmnn.*
2. **Sinha S, Sharma BS.** Neurocysticercosis: a review of current status and management. *J Clin Neurosci. 2009;16(7):867-76. http://doi.org/ckxxc4.*
3. Organización Mundial de la Salud. Teniasis y cisticercosis. Nota descriptiva. Nueva York: OMS; 2017 [cited 2017 Dec 17]. Available from: https://goo.gl/5rSTwS.
4. **López-Valencia D, Zúñiga-Cerón LF, Saavedra-Torres JS, Medina AP.** Neurocisticercosis, caracterización de una enfermedad desatendida y re-emergente. *Morfolia*. 2014 [cited 2018 Jan 30];6(3):42-60. Available from: https://goo.gl/PWgYDM.
5. **Ndimubanzi PC, Carabin H, Budke CM, Nguyen H, Qian YJ, Rainwater E, *et al.*** A systematic review of the frequency of neurocyticercosis with a focus on people with epilepsy. *PLoS Negl Trop Dis. 2010;4(11):e870. http://doi.org/bwtqb6.*
6. **Bock M, Garcia HH, Chin-Hong P, Baxi SM.** Under seize: neurocysticercosis in an immigrant woman and review of a growing neglected disease. *BMJ Case Rep. 2015;2015. http://doi.org/f8z49v.*
7. **Flórez-Sánchez A, Pastrán SM, Vargas NS, Beltrán M, Enriquez Y, Peña AP, *et al.*** Cisticercosis en Colombia. Estudio de seroprevalencia 2008-2010. *Acta Neurológica Colomb. 2013;29(2):73-86.*
8. **Mahale RR, Mehta A, Rangasetty S.** Extraparenchymal (Racemose) Neurocysticercosis and Its Multitude Manifestations: A Comprehensive Review. *J Clin Neurol. 2015;11(3):203-11. http://doi.org/cj2c.*
9. **López-Valencia D, Medina-Ortega A, Saavedra-Torres JS, Zúñiga-Cerón LF, Zamora-Bastidas TO.** Neurocysticercosis, unusual manifestations. Rev. *Fac. Med. 2016;64(3):561-4.* http://doi.org/cj2d.
10. **Botero D, Restrepo M.** Cisticercosis. In: Parasitosis humanas. 5th ed. *Medellín: Corpración para Investigaciones Biológicas; 2012. p.* 530-54.
11. **Jehangir A, Hwang E, Qureshi A, Jehangir Q.** Recurrent neurocysticercosis. *J Community Hosp Intern Med Perspect.* 2014;4(3). http://doi.org/cj2f.
12. **Pal A, Biswas C, Ghosh TR, Deb P.** A rare case of recurrence of primary spinal neurocysticercosis mimicking an arachnoid cyst. *Asian J Neurosurg.* 2017;12(2):250-2. http://doi.org/f955wc.
13. **Joshi KC, Singh H, Sakhuja P, Singh D.** Repeated hydrocephalus in recurrent intraventricular neurocysticercosis: An uncommon presentation. J Neurosci Rural Pract. 2013;4(1):87-9. http://doi.org/f4rsrd.

14. **Vásquez-Arteaga LR, Zamora-Bastidas TO, Vivas-Velásco VH, Giraldo-Forero JC, Casas-Zúñiga JC.** Epidemiología de la cisticercosis humana en pacientes de consulta neurológica en Popayán, Cauca, Colombia. *Medicina*. 2016;38(4):305-15.
15. **Garcia HH, Gonzalez AE, Evans CA, Gilman RH.** Taenia solium cysticercosis. *Lancet*. 2003;362(9383):547-56. http://doi.org/d5zfvn.
16. **Bhattarai R, Carabin H, Proano J V, Flores-Rivera J, Corona T, Flisser A, *et al.*** Cost of neurocysticercosis patients treated in two referral hospitals in Mexico City, Mexico. *Trop Med Int Health. 2015;20(8):1108-19. http://doi.org/f7tg57.*
17. **Agudelo-Flórez P, Restrepo BN, Palacio LG.** Conocimiento y prácticas sobre teniasis-cisticercosis en una comunidad colombiana. *Rev. Salud Pública*. 2009;11(2):191-9. http://doi.org/d4rqf5.
18. **Flisser A.** Cisticercosis: enfermedad desatendida. *Bol Med Hosp Infant Mex. 2011;68(2):138-45.*

https://doi.org/10.15446/cr.v4n1.66069

ENDOSCOPIC MANAGEMENT OF SINONASAL HEMANGIOPERICYTOMA: CASE REPORT AND LITERATURE REVIEW

Keywords: Hemangiopericytoma; Nasal cavity; Paranasal sinuses; Solitary fibrous tumors.
Palabras clave: Hemangiopericitoma; Cavidad nasal; Senos paranasales; Tumores fibrosos solitarios.

Francisco Rosa
José Gameiro dos Santos
Jorge Oliveira
Cecília Almeida e Sousa
Centro Hospitalar do Porto
Department of Otolaryngology
Head and Neck Surgery
Portugal

Corresponding author:
Francisco Rosa
Department of Otolaryngology
Head and Neck Surgery
Centro Hospitalar do Porto
Email: franciscorosa97@gmail.com

Received: 01/07/2017 **Accepted:** 13/12/2017

ABSTRACT

Introduction: Hemangiopericytoma is a rare vascular tumor of the sinonasal region, associated with epistaxis and nasal obstruction as the main symptoms. When located in this region, it has special clinical characteristics that differentiate it from others.

Case presentation: The following paper reports the case of a 43-year-old female patient presenting with right nasal obstruction and 6 months of evolution associated with mucopurulent rhinorrhea and recurrent right side epistaxis. Physical examination showed a right obstructive mass originating from the cribriform plate. Computed tomography of the paranasal sinuses revealed a complete blockage of the right nasal cavity by a homogeneous content, with soft tissue density and no evident contrast enhancement. The lesion extended superiorly to the cribriform plate but without intracranial or orbital extension. The patient was treated with endoscopic surgery and anatomopathological study revealed sinonasal hemangiopericytoma. The patient had complete remission and subsequent 3-year follow-up without recurrence.

Conclusion: The recommended treatment for hemangiopericytoma is total surgical excision with free margins. The results are generally good and the risk of recurrence seems to be associated with incomplete tumor excision. A literature review is presented and its main characteristics are discussed.

RESUMEN

Introducción: El hemangiopericitoma es un tumor vascular poco frecuente en la región nasosinusal que se asocia con epistaxis y obstrucción nasal como principales síntomas. Cuando se presenta en esta región, tiene características clínicas especiales que lo hace diferente de otras localizaciones.

Presentación del caso: A continuación se presenta el caso de una paciente de 43 años de edad, con síntomas de obstrucción en fosa nasal derecha y 6 meses de evolución asociados con rinorrea mucopurulenta y epistaxis recurrente del lado derecho. El examen físico mostró una masa obstructiva en el lado derecho con origen en la placa cribriforme. Una tomografía computarizada de los senos paranasales reveló un bloqueo completo de la cavidad nasal derecha por un contenido homogéneo, con densidad de partes blandas, sin captación de contraste evidente. La lesión se extendía hacia el lado superior de la placa cribriforme, sin extensión intracraneal u orbitaria. La paciente fue tratada con cirugía endoscópica y el estudio anatomopatológico reveló un hemangiopericitoma sinonasal. La paciente tuvo remisión completa y se realizó seguimiento por 3 años sin recurrencia.

Conclusión: El tratamiento recomendado para el hemangiopericitoma es la escisión quirúrgica total con márgenes libres, cuyos resultados son generalmente buenos. El riesgo de recurrencia parece estar asociado con una escisión tumoral incompleta. Se presenta una revisión de literatura, así como comentarios sobre sus características principales.

INTRODUCTION

Hemangiopericytomas are tumours of vascular origin that are rarely seen in the nose and paranasal sinuses. (1) Histological and biological differences may be observed between sinonasal hemangiopericytoma and its soft tissue counterpart. Its designation

as 'hemangiopericytoma-like' tumor implies that it is related to, yet distinct from, soft tissue hemangiopericytomas. (2,3)

From a therapeutic point of view, the mainstay of treatment is surgical excision with clear resection margins, as these tumors are relatively radioresistant. Nowadays, given the extraordinary development of endoscopic techniques, sinonasal hemangiopericytoma can be managed endonasally, with very few exceptions. (4)

CASE REPORT

A 43-year old Caucasian female patient, teacher, with no relevant medical history, presented to our hospital with complaints of right nasal obstruction of 6 months of evolution, associated with mucopurulent rhinorrhea and recurrent right side epistaxis. Physical examination showed a right obstructive mass originating from the roof of the nasal fossa (cribriform plate), which caused a deviation of the nasal septum to the left and a lateralization of the middle turbinate to the right (Figure 1A).

Computed tomography of the paranasal sinuses (CT-PS) revealed a complete blockage of the right nasal cavity by a homogeneous content, with soft tissue density, without evident contrast enhancement. This neoformation caused bulging with thinning and remodeling of the septum and the wall of the right maxillary sinus. The lesion extended superiorly to the cribriform plate but without intracranial or orbital extension, and posteriorly through the choana to the nasopharynx (Figure 1B). CT scan of the neck and thorax did not identify relevant alterations. A biopsy of the lesion was performed and the histological study revealed an inflammatory polyp with no signs of malignancy.

The patient underwent endoscopic surgery with en bloc resection of the lesion that originated in the cribriform plate of the ethmoid (Figure 1C), without sequelae associated with the procedure.

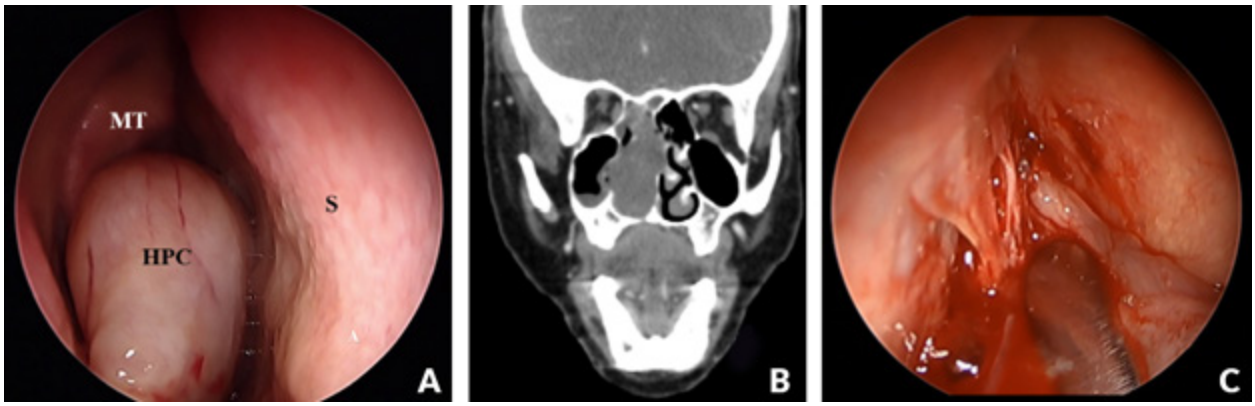


Figure 1. (A) Rhinoscopy: Obstructive mass originating from the roof of the right nasal fossa. HPC - Hemangiopericytoma, S - Nasal Septum, MT - Middle Turbinate; (B) CT paranasal sinuses in soft tissue window (coronal); (C) endoscopic surgery.
Source: Own elaboration based on the data obtained in the study.

The anatomopathological study of the surgical specimen revealed a tumor with morphological and histochemical characteristics compatible with sinonasal hemangiopericytoma and immunoreactivity to vimentin, CD34 and alpha-actin (Figure 2).

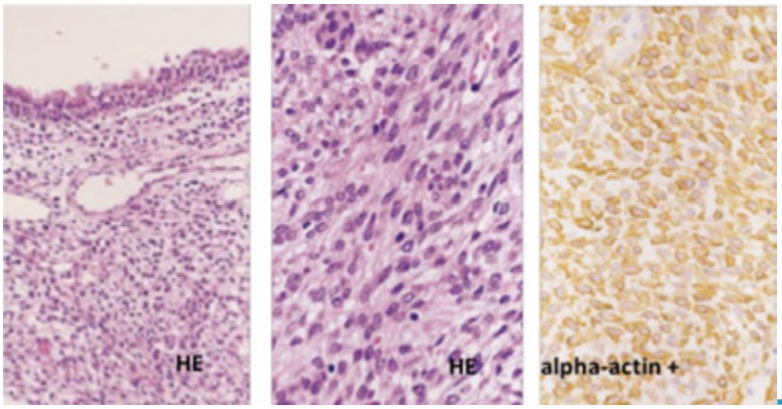


Figure 2. Anatomopathological study: Polyp lined by respiratory epithelium, expanded chorion by proliferation of cells of elongated nuclei, sometimes fusiform in appearance. Immunoreactivity for vimentin, CD34 and alpha-actin. Characteristics suggestive of sinonasal hemangiopericytoma. HE - Hematoxylin-Eosin.
Source: Own elaboration based on the data obtained in the study.

The patient underwent a 3-year follow-up, during which time she remained free of symptoms and showed no signs of local or metastatic recurrence on objective examination and CT-PS imaging (Figure 3).

DISCUSSION

Hemangiopericytoma, also known as extrapleural solitary fibrous tumor, is a rare tumor, initially described by Stout and Murray in 1942. Its origin is mesenchymal, originating from capillary pericytes and representing less than 1% of all vascular tumors and only 1 to 2% of soft tissue tumors. (1)

It can develop anywhere in the body, being more frequent in the limbs, retroperitoneum and skin. Only 15% of hemangiopericytomas develop in the head and neck region, more frequently in the nasal cavity and paranasal sinuses. (2) Several etiological factors have been proposed, including hypertension, hormonal or metabolic imbalance and trauma; however, the etiology of sinonasal hemangiopericytoma is still unknown. (5)

Its origin is usually benign but its biological behavior and natural history are still relatively unknown, with a potential risk of malignancy. Sinonasal hemangiopericytoma presents some histological and biological differences with respect to hemangiopericytomas in other places, and is often known as 'hemangiopericytoma-like' sinonasal tumor. Some authors suggest a similarity with glomus tumors. (2,3)

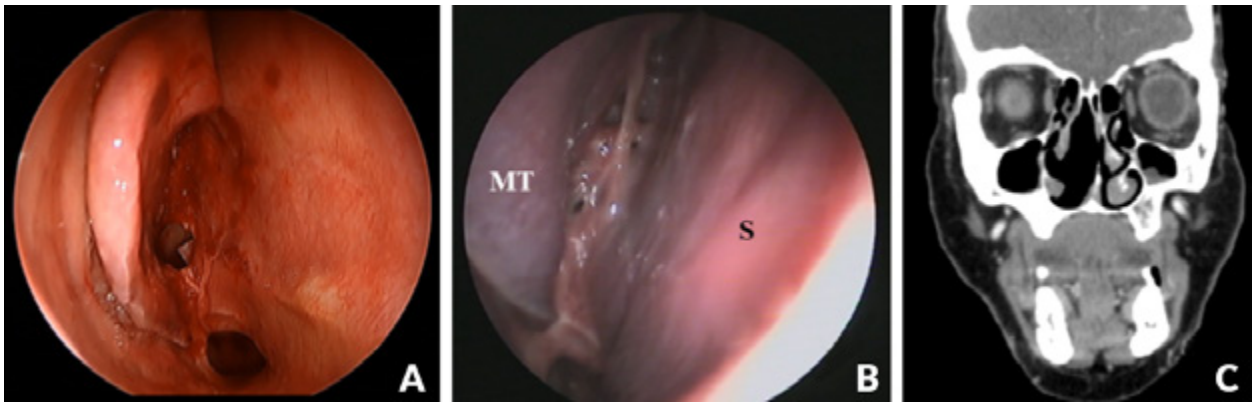


Figure 3. (A) After removing hemangiopericytoma; (B) Endoscopic examination without evidence of local recurrence (3-year follow-up). MT - Middle Turbinate; S- Nasal Septum; (C) CT paranasal sinuses in soft tissue window (coronal) - 3-year follow-up.
Source: Own elaboration based on the data obtained in the study.

Sinonasal hemangiopericytoma tends to be immunoreactive with vimentin, α-smooth muscle actin, and muscle specific actin. However, unlike lobular capillary hemangiomas and solitary fibrous tumors, it rarely stains positively for CD34, although staining for CD34 and S100 protein can be focally and weakly positive in a small percentage of tumors. (4,5)

These lesions occur mainly between the ages of 40 and 60, affecting both females and males. The most common initial presentation is nasal obstruction and recurrent epistaxis. The lesions are usually painless and the symptoms are originated by their growth and tumor compression. (3)

The initial diagnostic assessment must include endoscopic and neuro-radiological evaluation with CT and magnetic resonance imaging (MRI). CT imaging demonstrates tumor involvement of the soft tissue in the nasal cavity and paranasal sinsuses, with bone destruction observed in large tumors. MRI shows sinonasal hemangiopericytoma as a solid mass with isotense signals on contrast-enhanced T1 imaging, which is useful for differentiating it from inflammatory fluid caused by sinus obstruction. (6)

Imaging evaluation allows the characterization of tumor extension and its relation with adjacent structures, namely, intracranial and intraorbital components. (5) MRI is superior to CT mainly for the assessment of tumor relation with vascular structures, being important for surgical planning. Chest CT is recommended for assessment of distant metastases that may occur by hematogenic/lymphatic dissemination to the lung. (6) When necessary, additional investigation can be performed through angiography (with preoperative embolization) in larger and strongly vascularized lesions. (7)

Biopsy of sinonasal lesions, which are suspected of being vascular tumors, is not routinely performed by our department in the outpatient clinic. Nevertheless, in this case, a biopsy was performed considering that physical examination and CT scan did not present strong evidence of a vascularized lesion. Based on the biopsy results, only sinonasal polyposis was suspected initially, therefore, no preoperative magnetic resonance imaging was performed. Although this is a limitation, it had no influence on the procedure and surgical outcome, since complete resection of the lesion was achieved without associated complications and without recurrence after 3 years of follow-up.

Sinonasal hemangiopericytoma is a benign lesion with a low risk of malignancy (<10%). Malignant lesions have an increased risk of recurrence and are associated with cellular pleomorphism, moderate to severe nuclear atypia, bone invasion, and tumor necrosis. Some authors state that the presentation of hemangiopericytoma at the sinonasal level is less aggressive but locally recurrent. (6,8). The treatment of choice is endoscopic surgical resection with free margins, with positive margins being the main positive predictive factor for recurrence of hemangiopericytoma. High recurrence rates are probably associated with the difficulty of total excision of the tumor at this location. (9)

Adjuvant radiotherapy is a second-line treatment, indicated for lesions with malignant characteristics and incomplete surgical resection/unresectable tumors. The use of chemotherapy is controversial and its efficacy has not been proven. Prognosis is usually good if complete surgical excision of the primary tumor is achieved, with a 5-year survival rate of 89-100%. (1) However, long-term follow-up is necessary given the potential risk of malignancy and tumor recurrence. (4)

With less than 200 cases of sinonasal hemangiopericytomas reported in the litera-

ture, only limited assumptions can be made about the tumor. (10, 11) This is another case of sinonasal hemangiopericytoma to add to the small but growing body of literature on this disease. Historically, open surgical methods for tumor extirpation have been considered as standard treatment, although endoscopic resection has increased in recent decades. (12,13) This clinical case demonstrates that endoscopic resection is a safe, viable and reasonable alternative and is currently the gold standard of treatment.

CONCLUSION

Hemangiopericytoma is a rare tumor in the sinonasal region with usually benign etiology. Adequate surgery with free resection margins is the treatment of choice. Nowadays, with very few exceptions, resection should be conducted endoscopically. Patient outcome is generally good, and the risk of recurrence seems to be related to complete resection.

FUNDING

None stated by the authors.

CONFLICT OF INTEREST

None stated by the authors.

REFERENCES

1. **Stout AP, Murray MR.** Hemangiopericytoma: a vascular tumor featuring Zimmermann´s pericytes. *Ann Surg.* 1942;116(1):26-33. http://doi.org/djpbvb
2. **Compagno J.** Hemangiopericytoma-like tumors of the nasal cavity: a comparison with hemangiopericytoma of soft tissues. *Laryngoscope.* 1978;88(3):460-9. http://doi.org/cj4f39
3. **Li XQ, Hisaoka M, Morio T, Hashimoto H.** Intranasal pericytic tumors (glomus tumor and sinonasal hemangiopericytoma-like tumor): report of two cases with review of the literature. *Pathol Int.* 2003;53(5):303-8. http://doi.org/d4d3sz
4. **Simmonds JC, Rebeiz EE.** Surgical resection of sinonasal hemangiopericytoma involving anterior skull base: Case reports and literature review. *Am J Otolaryngol.* 2017;38(1):87-91. http://doi.org/f9fv9j
5. **Shobha BV, Shivakumar BN, Reddy S, Dutta N.** Sinonasal hemangiopericytoma: A rare case report with review of literature. *J Oral Maxillofac Pathol.* 2015;19(1):107. http://doi.org/cjx7
6. **Bignami M, Dallan I, Battaglia P, Lenzi R, Pistochini A, Castelnuovo P.** Endoscopic, endonasal management of sinonasal haemangiopericytoma: 12-year experience. *J Laryngol Otol.* 2010;124(11):1178-82. http://doi.org/fm7rvx
7. **Ledderose GJ, Gellrich D, Holtmannspötter M, Leunig A.** Endoscopic Resection of Sinonasal Hemangiopericytoma following Preoperative Embolisation: A Case Report and Literature Review. *Case Rep Otolaryngol.* 2013;2013:796713. http://doi.org/f98qk8
8. **Thompson LD, Miettinen M, Wenig BM.** Sinonasal-type hemangiopericytoma: a clinicopathologic and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. *Am J Surg Pathol.* 2003;27(6):737-49. http://doi.org/cbm3k6
9. **Tessema B, Eloy JA, Folbe AJ, Anstead AS, Mirani NM, Jourdy DN, *et al.*** Endoscopic management of sinonasal hemangiopericytoma. *Otolaryngol Head Neck Surg.* 2012;146(3):483-6. http://doi.org/cpksh6
10. **Duval M, Hwang E, Kilty SJ.** Systematic review of treatment and prognosis of sinonasal hemangiopericytoma. *Head Neck.* 2013;35(8):1205-10 http://doi.org/f5b4kg
11. **Al Saad S., Al Hadlaq R., Al-Zaher N.** Glomangiopericytoma (Hemangiopericytoma) of the maxi-

llary sinus and sinonasal tract. *Hematol Oncol Stem Cell Ther.* 2017; 10(2):96-98 http://doi.org/cjx8

12. **Dahodwala MQ, Husain Q, Kanumuri VV, Choudhry OJ, Liu JK, Eloy JA.** Management of sinonasal hemangiopericytomas: a systematic review. *Int Forum Allergy Rhinol.* 2013; 3(7):581-7 http://doi.org/f5bqxk
13. **Peyvandi A, Naghibzadeh B, Roozbahany NA.** Sinonasal Hemangiopericytoma: A Case Report. *Iran J Med Sci.* 2015; 35(3), 251-253.

https://doi.org/10.15446/cr.v4n1.67067

CERVICAL LYMPHANGIOMA IN ADULTS: CASE REPORT AND CURRENT TREATMENT

Keywords: Adult; Lymphangioma, Cystic; Sclerotherapy; Picibanil.
Palabras clave: Adulto; Linfangioma quístico; Escleroterapia; Picibanil.

Enrique Cadena-Piñeros
Head and Neck Unit
- Instituto Nacional de Cancerología -
Bogotá D.C. - Colombia.
Department of Surgery -
Universidad Nacional de Colombia -
Faculty of Medicine - Bogotá D. C. - Colombia.
Otolaryngology Unit - Clínica de Marly
- Bogotá D.C. - Colombia.
Head and Neck Surgery Unit
- Clínica de Marly - Bogotá D.C. - Colombia

Andrés Rojas Gutiérrez
Head and Neck Unit
- Instituto Nacional de Cancerología -
Bogotá D.C. - Colombia.

Corresponding author:
Enrique Cadena. Head and Neck Unit
- Instituto Nacional de Cancerología - Bogotá D.C. -
Colombia. E-mail enriquecadena2005@yahoo.com.

Recibido: 15/08/2017 **Aceptado:** 18/01/2018

RESUMEN

Introducción: Los tumores quísticos del cuello son inusuales en los adultos. Sin embargo, se pueden encontrar metástasis a ganglios, quistes branquiales, quistes tiroglosos, linfangiomas, entre otros. Clínicamente, estos últimos son masas blandas de crecimiento lento que se localizan en diferentes espacios del cuello.

Reporte de caso: Se reporta un caso de linfangioma en una mujer de 36 años, quien consultó por masa lateral derecha del cuello con evolución de 20 días sin síntomas sistémicos asociados. A pesar de que la recaída es frecuente, la paciente fue tratada con cirugía exitosa sin evidencia de recidiva durante 12 meses de seguimiento.

Discusión: Cuando se presentan en niños, los tumores quísticos del cuello se pueden convertir en urgencias quirúrgicas debido a obstrucción de la vía aérea; no obstante, en los adultos solo producen deformidad de contorno y rara vez requieren una intervención apremiante, lo que permite conductas conservadoras como la observación, el drenaje repetido o la escleroterapia. Esta última puede hacerse con el OK-432 (Picibanil); sin embargo, la cirugía es una buena opción de tratamiento sin estar exenta de complicaciones.

Conclusión: Se realizó revisión de las diferentes opciones de tratamiento y se concluyó que la resección quirúrgica de los linfagiomas continúa siendo la opción más adecuada para el manejo de esta compleja lesión del cuello.

ABSTRACT

Introduction: Cystic tumors of the neck are rare in adults. Some of them include metastatic nodes, branchial cysts, thyroglossal cysts and lymphangiomas, among others. Clinically speaking, lymphangiomas are slow-growing soft masses located in different spaces of the neck.

Case report: This paper reports the case of a 36-year-old woman presenting with lymphangioma, who consulted due to a right lateral mass in the neck of 20 days of evolution without associated systemic symptoms. Although relapse is frequent, the patient was successfully treated with surgery, without evidence of recurrence at 12 months of follow-up.

Discussion: When cystic tumors of the neck occur in children, surgical urgencies may arise due to obstruction of the airway. However, lymphangioma in adults only produce contour deformity and rarely require urgent intervention, which allows for conservative management such as observation, repeated drainage or sclerotherapy that can be done using OK-432 (Picibanil). Nevertheless, surgery remains a good treatment option, but some complications may occur.

Conclusion: Different treatment options were reviewed, which led to conclude that surgical resection of lymphangiomas continues to be a good treatment for this complex neck lesion.

INTRODUCTION

Cystic lesions of the neck are rare and difficult to interpret for clinicians, since they can be benign or malignant pathologies. The most common location is the posterior triangle, where inflammatory, metastatic adenopathies or lymphoproliferative diseases can occur. In addition, congenital malformations such as branchial cysts, hemangiomas and lymphangiomas

may be observed in this area. (1) Branchial cleft cysts and hemangiomas are easily diagnosed in children under 2 years of age due to their clinical characteristics. These lesions are observed as a mass that usually does not involve inflammatory changes and presents a slow and painless growth or lymphangiomas, which are rare after the third decade of life. (2-5)

The current trend to treat lymphangiomas is different; conservative interventions such as sclerotherapy, now performed with new substances, have relegated surgery to the background due to the high rate of relapse or persistence. (4,5) Considering that lymphangiomas are rare in adults (4) and that the patient treated was a 36-year-old woman who underwent a complete surgical resection, a review of the subject is presented with a description of the different treatment options.

CASE PRESENTATION

A 36 year-old female patient, Caucasian, from the city, consulted due to a right lateral mass in the neck of 20 days of evolution without associated systemic symptoms, dysphonia, dysphagia or pain. No personal or family history of importance in relation to the current disease and no history of neck trauma or infection of the head and neck area were described. On physical examination, a right lateral soft mass of 6cm was found, which was tender on palpation and with displacement and lateralization of the sternocleidomastoid muscle (Figure 1).

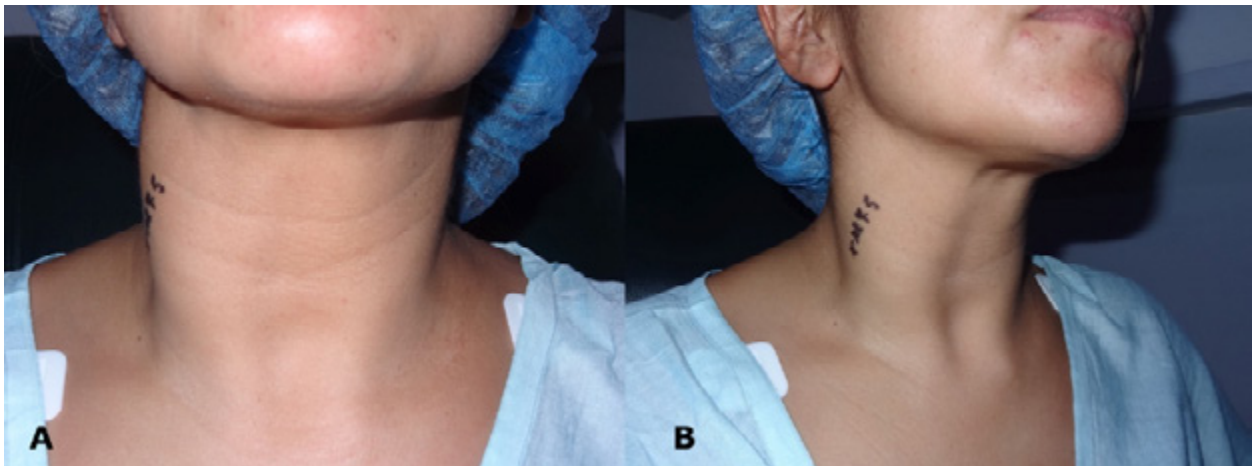


Figure 1. Deformity of the right contour of the neck. A) Anterior aspect of the neck; B) Lateral aspect of the neck.
Source: Own elaboration based on the data obtained in the study.

A contrast study of the neck using computed tomography (CT) showed a cystic lesion with thin walls of 55x21x43mm, which compressed the internal jugular vein, occupied the right III and IV levels and was clearly delimited by vascular and muscular structures (Figure 2).

Fine needle aspiration (FNA) helped to diagnose cyst content with lymphoid tissue lining, which could be lymphocele or lymphangioma (cystic hygroma). Due to the lack of diagnostic clarity, a surgery was performed 15 days later. During the procedure, a medial cystic mass was found in the sternocleidomastoid muscle adhered to the internal jugular vein and to the phrenic, hypoglossal, spinal and vagus nerves. Finally, the lesion was completely resected along with the entire capsule (Figure 3).

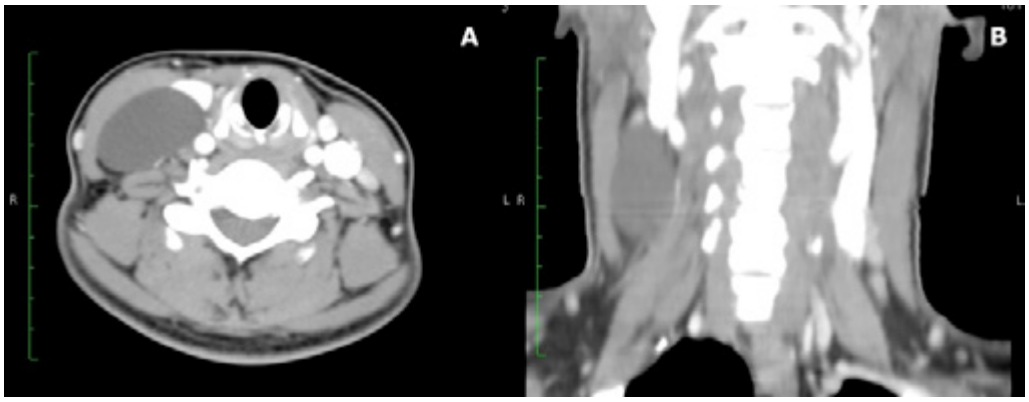


Figure 2. Contrast computed tomography of the neck. A) Thin-walled cyst medial to the sternocleidomastoid muscle; B) Cystic lesion occupying the right III and IV levels.

Source: Own elaboration based on the data obtained in the study.

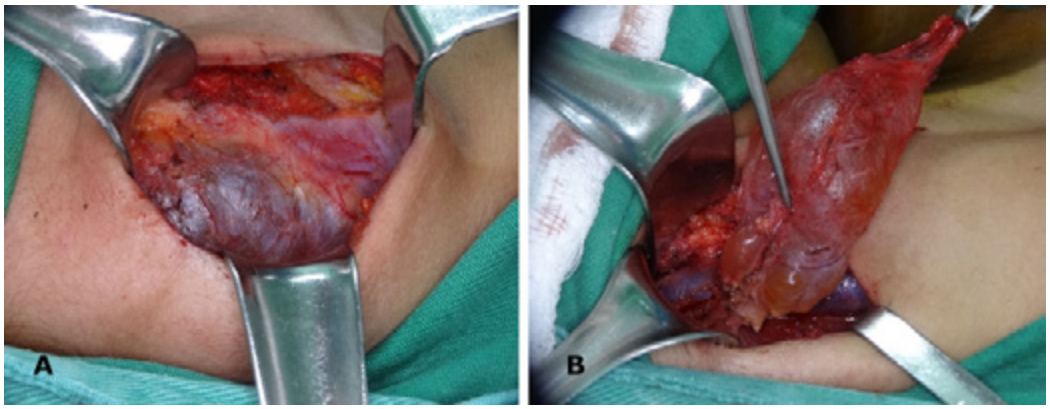


Figure 3. Intraoperative appearance. A) Transverse incision of the neck. A deep lesion of the sternocleidomastoid muscle was observed along with junctions to the internal jugular vein and accessory nerve. B) Lymphangioma was dissected from the internal jugular vein.

Source: Own elaboration based on the data obtained in the study.

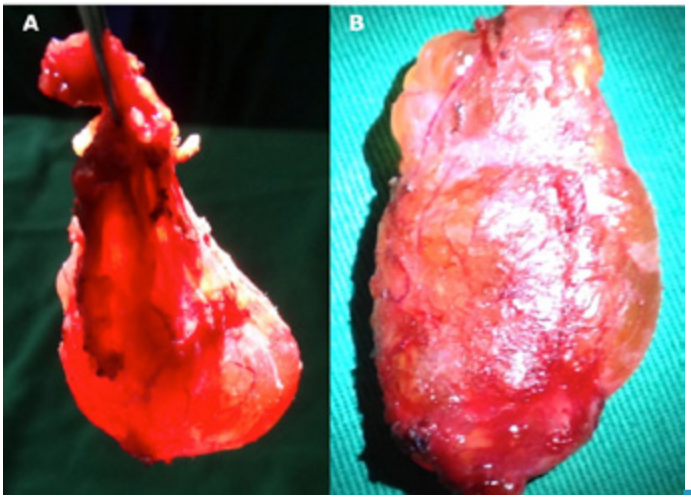


Figure 4. Surgical specimen: transillumination revealed a thin-walled cyst.

Source: Own elaboration based on the data obtained in the study.

Transillumination of the surgical specimen revealed a thin-walled, cystic-like mass with citrine material content, which correlated with the diagnosis of preoperative FNA (Figure 4).

The final pathology reported a cystic mass of 6.9x3.2x3cm, with smooth external surface and a clear liquid content. Additionally, the smooth inner surface was accompanied by an adipose tissue of 2.5x1.8x1cm with another tissue of 1.5x0.6x0.5cm. The final diagnosis of the pathology was lymphangioma and five reactive lymph nodes, negative for tumor infiltration. The patient did not present any complication during the postoperative period, nor motor deficit related to the dissected structures, nor evidence of relapse or persistence of the lesion after 12 months of follow-up. No complaints were made by the patient, who reported excellent functional and cosmetic results.

DISCUSSION

Lymphangiomas, also known as cystic hygromas, are congenital malformations that are usually diagnosed at birth and can cause airway obstruction, thus constituting a medical emergency. (6) These lesions are considered vascular anomalies of the lymphatic system and are classified as hamartomas based on pathology findings. (7) Different etiological and pathogenic factors have been described as causal, including traumatic, infectious and chronic inflammatory factors, as well as alterations in embryological development. The latter includes sequestration of lymphatic tissue, defects in the fusion of the venous system, and lymphatic obstruction that causes its expansion and obstruction. (8) In adults, late proliferation of cellular nests of the lymphatic system may appear due to stimulation caused by trauma or infection. (9,10)

Lymphangiomas correspond to 25% of all vascular tumors in children and adolescents; 70% of cases are found in the head and the neck. In 2006, Thompson (11) proposed classifying them into cystic (cystic hygroma), capillary or cavernous, but there are different histological classifications:

Colbert *et al.* (12) classified them into *macrocysts*, formerly known as cystic hygromas, and in *microcysts*, which may occur simultaneously in a lesion.

Bhayya *et al.* (13) classified them in *lymphangioma simplex*, composed of small thin-walled lymphatics; *cavernous lymphangioma*, comprised of dilated lymphatic vessels with surrounding adventitia, *cystic lymphangioma*, consisting of a large lesion, with macroscopic lymphatic spaces surrounded by fibrovascular tissue and smooth muscle, and *benign lymphangioendothelioma*, lymphatic channels that seem to be dissected through dense collagen bundles.

In 1995, de Serres *et al.* (14) proposed a classification according to anatomical location: *Stage I*, unilateral infrahyoid lesion; *Stage II*, unilateral suprahyoid lesion; *Stage III*, unilateral suprahyoid and infrahyoid lesion; *Stage IV*, bilateral suprahyoid lesion, and *Stage V,* bilateral suprahyoid and infrahyoid lesion.

Based on the last classification system, the case presented here corresponds to stage I (right lateral extension from a horizontal plane to the hyoid bone and to the superior clavicular ridge). This made dissection and complete resection easier, without evidence of residual tissue on the surgical bed.

The diagnostic studies used to determine cystic tumors of the neck include high resolution ultrasound, which can prove the presence of a cystic lesion with multiple septa and without internal vascular flow that allows differentiating them from mixed vascular lesions. However,

CT and magnetic resonance imaging (MRI) determine more precisely the relationship of the mass with the adjacent structures. The T1 and T2 sequences of the MRI facilitate the visualization of cleavage planes with the muscles and can define whether it is a vascular malformation. (12)

Fageeh *et al.* (15) proposed observation, percutaneous drainage, carbon dioxide laser (CO_2) and Nd-YAG laser, diathermy and surgical resection as management options. Recently, Miceli & Stewart (16) recommended sclerotherapy with doxycycline or radiotherapy as treatment options. Observation may be indicated for patients younger than 3 years who do not have obstruction of the airway and have lesions <4cm, since they could present spontaneous regression. (15,17)

Surgical resection has traditionally been advocated as the best treatment option, but in cases where the lesions extend into deep neck spaces, as the floor of the mouth or parapharyngeal space, complete removal may be difficult to achieve without damaging the nervous and vascular structures. This situation has led to seek alternatives such as drainage and sclerotherapy with different substances such as tetracycline, bleomycin and triamcinolone. Radiofrequency ablation is another therapeutic option to treat this entity. (12) Complications and associated morbidity can include infections, tissue necrosis, cranial nerve lesions, vascular thrombosis and even endocrine disorders. (6,15)

Ogita *et al.* (18) described the use of a streptococcal preparation OK-432 (Picibanil) as a safe alternative to perform sclerosis in cystic lesions of the neck, considering that this substance produces fibrosis secondary to inflammatory and cicatricial changes with the consequent contraction of the lymphangioma. (18) OK-432 is a lyophilisate made of group A *Streptococcus pyogenes* incubated with penicillin, which is initially used as immunotherapy for cancer in cases of pleural and peritoneal carcinomatosis. (19)

These authors used this preparation in 83 patients with benign cystic lesions of the neck; 12 of them had lymphangiomas, achieving complete reduction in 67% and almost complete in 33% of the cases. Only some patients developed minor complications such as fever between 37.5°C and 38°C for a few days after the injection, which was controlled with antipyretics and without antibiotics. (19)

Unilateral lesions composed of microcysts, delimited and extended to one or two compartments of the neck, are candidates for surgical resection, as they may be completely resected. However, the lesions located near the floor of the mouth or the parapharyngeal space are more likely to be partially resected. (6) In this patient, with no family history or genetic alterations, the lymphangioma was secondary to interruption of lymphatic drainage, probably due to previous neck trauma that went unnoticed. Similar to Gow *et al.* (6), for this case, surgical resection of the mass was decided because the CT showed a single cyst circumscribed to the III and IV right levels of the neck (Figure 2).

Although, it extended to level III, IV and Vb, it was possible to remove the mass without breaking the cysts (Figure 3). Macroscopic examination showed that the lesion had a clear content that allowed transillumination, confirming a lymphangioma (Figure 4).

CONCLUSIONS

Radiological images such as neck CT, MRI and neck ultrasound with FNA are of great help for diagnosing and planning the management of lymphangiomas of the neck.

Clinical suspicion of lymphangioma helps to establish the treatment for a patient with soft mass and radiological images suggestive of neck cysts. However, the definitive diagnosis is only obtained with the final pathology.

In adult patients, conservative observation treatment or sclerotherapy with different preparations such as picibanil are management options, especially when dealing with a complex lesion extended to the parapharyngeal space, submandibular space, pterygoid fossa or floor of the mouth.

Surgery is recommended for patients with *lymphangioma simplex* stage I, with single or multiple cysts that extent to one or two levels of the neck, and radiological planes well defined by images.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Brea-Álvarez B, Roldan-Fidalgo A.** Quistes en el triángulo cervical posterior en adultos. *Acta Otorrinolaringol Esp.* 2015;66(2):106-10. http://doi.org/f2rvqc.
2. **Derin S, Sahan M, Dere Y, Çullu N, Sahan L.** Cervical Cystic Hygroma in an Adult. Case Reports in Pathol. 2014;2014:209427. http://doi.org/gb99bt.
3. **Gaddikeri S, Vattoth S, Gaddikeri RS, Stuart R, Harrison K, Young D, *et al.*** Congenital cystic neck masses: embryology and imaging appearances, with clinicopathological correlation. Curr Probl Diagn Radiol. 2014;43(2):55-67. http://doi.org/cj35.
4. **Rolekar NG, Shah DK.** Recurrent cystic lymphangioma of the neck in an adult: A case report of rare entity. *Int J Med Sci Public Health.* 2014;3(2):243-5. http://doi.org/cj36.
5. **Costache A, Dumitru M, Tweedie D, Sarafoleanu C, Anghel I.** Adult cervical lymphangioma - ultrasonography, surgical removal, and pathology results. Case report. *Med Ultrason.* 2015;17(3):411-3. http://doi.org/cj37.
6. **Gow L, Gulati R, Khan A, Mihaimeed F.** Adult-onset cystic hygroma: a case report and review of management. *Grand Rounds.* 2011;11:5-11. http://doi.org/c7k5tw.
7. **Rey-Marcos M, Martín-Sánchez V, Cordero-Civantos C, Blanco-Pérez P, Batuecas-Caletrío A, Tapia-Risueño M.** Linfangioma quístico en el adulto. Rev. Soc. Otorrinolaringol. Castilla Leon Cantab. La Rioja. 2012;3(36):291-6.
8. **Wiegand S, Eivazi B, Barth PJ, von Rautenfeld DB, Folz BJ, Mandic R, *et al.*** Pathogenesis of lymphangiomas. Virchows Arch. 2008;453(1):1-8. http://doi.org/c6pjts.
9. **Suk S, Sheridan M, Saenger JS.** Adult lymphangioma: a case report. *Ear Nose Throat J.* 1997;76(10):881-3.
10. **Aneeshkumar MK, Kale S, Kabbani M, David VC.** Cystic lymphangioma in adults: can trauma be the trigger? *Eur Arch Otorhinolaryngol.* 2005;262(4):335-7. http://doi.org/dcms3t.
11. **Thompson L.** Lymphangioma. *Ear Nose Throat J.* 2006;85(1):18-9.
12. **Colbert SD, Seager L, Haider F, Evans BT, Anand R, Brennan PA.** Lymphatic malformations of the head and neck-current concepts in management. *Br J O Maxillof Surg.* 2013;51(2):98-102. http://doi.org/cj39.
13. **Bhayya H, Pavani D, Avinash-Tejasvi ML, Geetha P.** Oral lymphangioma: A rare case report. *Contemp Clin Dent.* 2015;6(4):584-7. http://doi.org/cj4b.
14. **de Serres LM, Sie KC, Richardson MA.** Lymphatic malformations of the head and neck. A pro-

posal for staging. Arch Otolaryngol Head Neck Surg. 1995;121(5):577-82. http://doi.org/dp89pq.

15. **Fageeh N, Manoukian J, Tewfik T, Schloss M, Williams H, Gaskin D.** Management of Head and Neck Lymphatic Malformations in Children. *J Otolaryngol.* 1997;26(4):253-8.
16. **Miceli A, Stewart KM.** Lymphangioma. In: StatPearls. Treasure Island: StatPearls Publishing; 2017 [cited 2018 Feb 1]. Available from: https://goo.gl/7iBg5s.
17. **Shumaker PR, Dela Rosa KM, Krakowski AC.** Treatment of Lymphangioma Circumscriptum Using Fractional Carbon Dioxide Laser Ablation. *Pediatr Dermatol.* 2013;30(5):584-6. http://doi.org/f49g5p.
18. **Ogita S, Tsuto T, Tokiwa K, Takahashi T.** Intracystic injection of OK-432: a new sclerosing therapy for cystic hygroma in children. *Br. J. Surg.* 1987;74(8):690-1. http://doi.org/dxjpfb.
19. **Ohta N, Fusake S, Watanabe T, Ito T, Aoyagi M.** Effects and mechanism of OK-432 therapy in various neck cystic lesions. *Acta Otolaryngol.* 2010;130(11):1287-92. http://doi.org/d74qrb.

https://doi.org/10.15446/cr.v4n1.67960

COLONIC ATRESIA IN A NEWBORN. CASE REPORT

Keywords: Intestinal atresia; Colon; Colostomy.
Palabras clave: Atresia, Colon, Colostomia.

Diego Montenegro. MD
Fundación Universitaria Juan N Corpas
- Bogotá, D.C. – Colombia.

Silvia Aragón
Juan Valero
Pediatric Surgery Service - Faculty of Medicine
Universidad Nacional de Colombia
- Bogotá D.C. - Colombia.
Pediatric Surgery Unit -
Fundación Hospital Pediátrico la Misericordia
- Bogotá D.C. - Colombia.

Corresponding author:
Diego Armando Montenegro,
Fundación Universitaria Juan N Corpas
- Bogotá, D.C. – Colombia.
Email: diego-montenegro@juanncorpas.edu.co.

Received: 26/09/2017 **Accepted:** 09/03/2018

ABSTRACT

Introduction: Colonic atresia is the least common type of intestinal atresia; however, it must be suspected in patients with partial or complete intestinal obstruction, failure to pass meconium, vomit and abdominal distension. Good prognosis has been described in patients with timely treatment.

Case report: This paper reports the case of a newborn patient presenting with vomit, abdominal distension, failure to pass meconium and a rapid progressive clinical deterioration. A colonic atresia was found during exploratory laparotomy, which required a temporary colostomy due to the discrepancy of the proximal and distal calibers. Subsequently, colonic anastomosis was performed using a protective colostomy that was finally closed. This patient had a good post-operative recovery.

Conclusion: Colonic atresia must be considered as an important cause of distal intestinal obstruction in pediatric patients and, therefore, it should always be suspected. Ruling out other associated abnormalities is also recommended, as well as performing a rectal biopsy for Hirschsprung's disease to avoid complications.

INTRODUCTION

Colonic atresia is a rare pathology (1) observed in 1.8%-15% cases of intestinal atresia. (2,3) Its incidence is estimated between 1:1 498 (4) to 1:66 000 live births. (5) The clinical presentation can be described as a partial or complete intestinal obstruction, associated with failure to pass meconium, vomit and abdominal distension, depending on the type of atresia. Abdominal x-ray is the mainstay for diagnosis; however the surgeon may also consider a barium enema.

In newborns with colonic atresia, other malformations have been reported, including gastroschisis, small bowel atresia, omphalocele, anorectal malformations, ocular and facial malformations, common bile duct cysts, musculoskeletal abnormalities (6), and Hirschsprung's disease. The last condition must be discarded by rectal biopsies at onset of symptoms. (7)

Colonic atresia has the most favorable prognosis for intestinal atresias. (8) The mortality reported is less than 10% (6), although Dalla Vecchia *et al.* reported zero mortality in a series of nine cases. (8) Nevertheless, mortality may increase to 100% if treatment is delayed beyond 72 hours, considering the risk of intestinal perforation and peritonitis. (6)

CLINICAL CASE

A 3 day-old female newborn, with a gestational age of 40 weeks and a history of self-limited polyhydramnios observed in a third-trimester ultrasound, was referred to the Fundación Hospital Pediátrico la Misericordia (HOMI). Her parents were of the lower middle class, with no specific ethnic group.

In her first day of life, the newborn failed to pass meconium and barely tolerated breastfeeding; however, she was discharged from another hospital. On her second day of life, the patient presented with vomit of alimentary content and oral intolerance. The clinical picture then progressed towards abdominal distention, vomiting of fecal content, deterioration of general state and respiratory failure that required invasive mechanical ventilation. The patient was transferred to our center.

On admission at HOMI, the patient presented with markedly distended abdomen (Figure 1) with absent bowel sounds, normal position of the anus and no genital abnormalities.

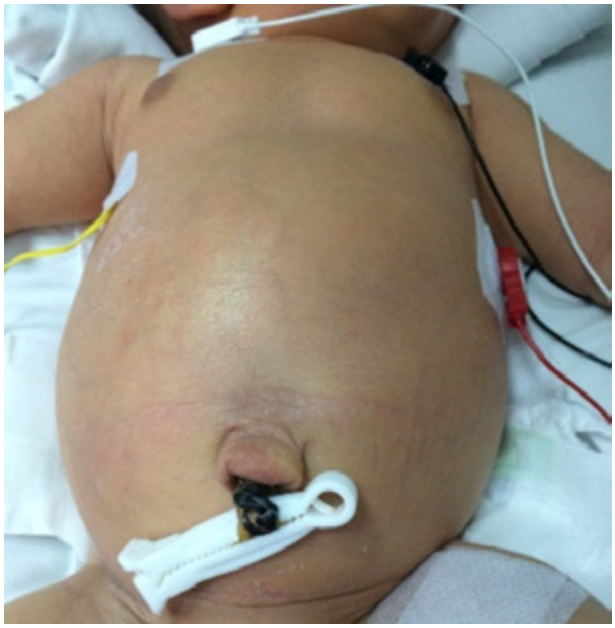


Figure 1. Severe abdominal distension before surgery.
Source: Own elaboration based on the data obtained in the study.

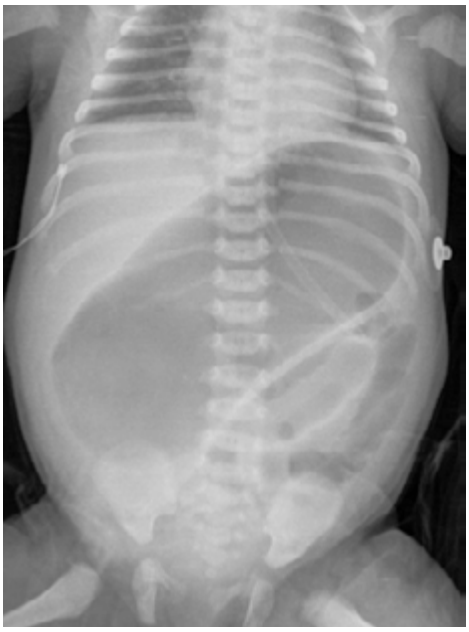


Figure 2. Significant colon distension.
Source: Own elaboration based on the data obtained in the study.

Invasive ventilation was initiated and a nasogastric tube was passed in-situ. An abdominal x-ray showed a large dilated loop of bowel (Figure 2), without a double bubble sign, ruling out duodenal atresia. Based on those findings, in her third day of life, an exploratory laparotomy was performed. A right colon atresia type III (Grosfeld classification) (11) was identified, with a difference of proximal distal caliber from 10 to 1 (Figure 3). A derivative Hartmann's colostomy was performed, given the difference of caliber size that did not allow primary anastomosis. Rectal biopsies were taken, reporting ganglion cells and ruling out Hirschsprung's disease.

Colostomy was closed after 11 months, during which time instillations of saline solution were made through the rectum to stimulate the growth of the hypotrophic intestine. A barium enema was made before the procedure (Figure 4).

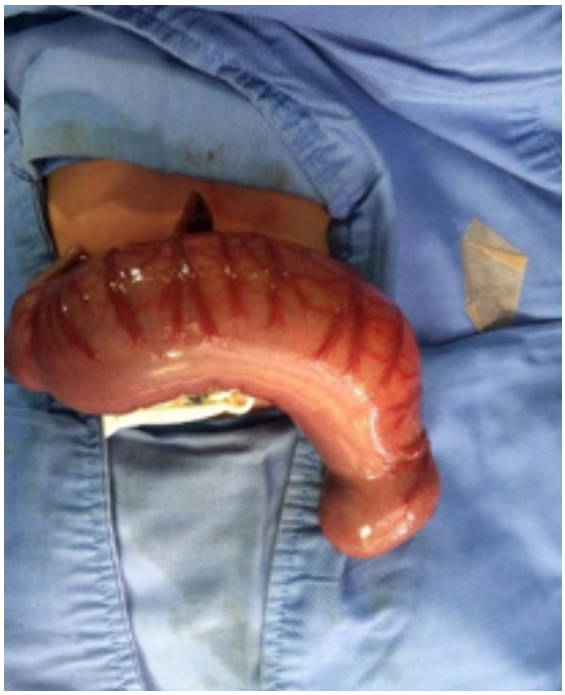


Figure 3. 3A. Colon atresia type III.
Source: Own elaboration based on the data obtained in the study.

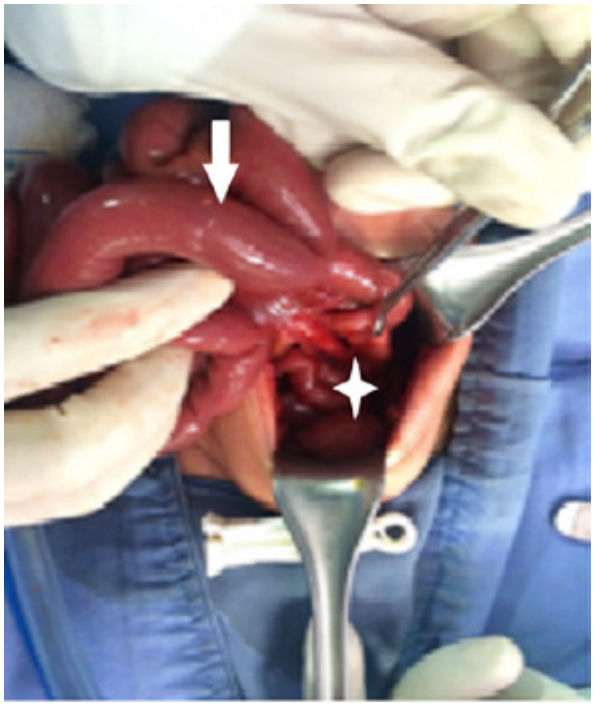


Figure 3. 3B. Evidence of difference in approximate distal 10-1 caliber.
(*) Distal, (Arrow) Proximal.
Source: Own elaboration based on the data obtained in the study.

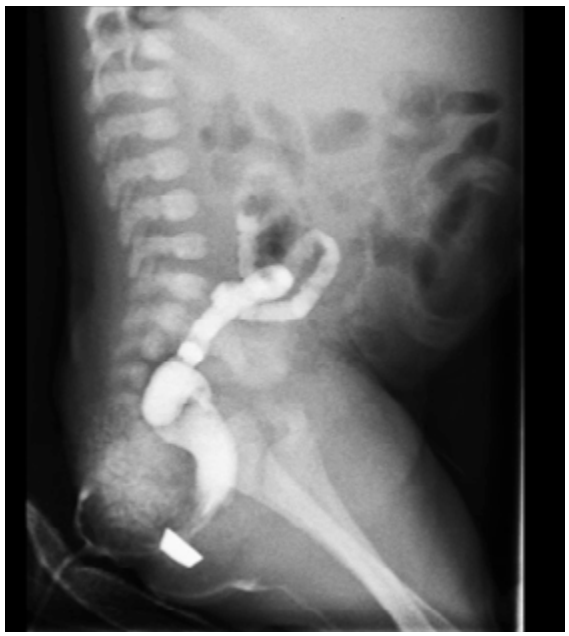


Figure 4. Hypoplasia of distal colon.
Source: Own elaboration based on the data obtained in the study.

During surgery, a discrepancy of calibers from 5 to 1 was observed and lateral anastomosis with a mechanical lineal suture was performed, leaving a colonic lateral window diversion proximal to the anastomosis, which is useful in cases of extreme size discrepancy. (12) During the third surgery, the protective colostomy was closed, observing hermetic anastomosis, with no stenosis. Post-surgical evolution was good, and the patient was discharged three days after surgery. No other abnormalities associated with this patient were observed. After 2 years of follow-up, no complications and daily normal evacuations were reported.

DISCUSSION

Since this condition was first reported in 1673 by Binniger, colonic atresia has had different types of approaches. At the beginning of the last century, Gaub described the first case of a survivor treated with colostomy, and fifty years later, Potts reported the first case of a successful primary anastomosis. (8) Due to the rarity of this pathology, many case reports are available but few studies have investigated this condition.

The etiology of the disease corresponds to an extrinsic obstruction of the mesenteric vessels during fetal development. The causes of this alteration are diverse and have been described in the literature as internal hernias, volvulus, intussusception, and associations to gastroschisis strangulation. (6,8)

As in this case, most cases have been reported in full-term pregnancies (8,9), and some cases have been associated to the first degree of consanguinity, which could imply a possible genetic causal relationship. (6,8)

Both Louws and Grosfeld classifications (11) are still used to subdivide anatomy and to define the treatment and prognosis of the

patients. In this regard, three main types of colon atresia are identified, while a fourth type relates to multiple atresias. Type I atresia is related to an obstructive membrane or intraluminal septum, with intact intestinal wall and mesentery; type II makes reference to blind loops separated by a fibrous cord and without mesentery alteration; type III atresia presents separate blind intestinal loops with a V-defect in the mesentery. (10,11)

The highest incidence of intestinal atresia lesions is found near the splenic flexure. The ascending colon is an uncommon site of presentation (10), and is even less common in relation to the type III pattern (8), as is the case of the patient presented in this paper.

Prenatal diagnosis is of great importance since ultrasound examination can show a relatively characteristic image (8) that may allow a faster management with fewer complications associated with delayed diagnosis. Abdominal x-ray is the initial examination that may confirm clinical impressions.

The differential diagnosis of colon atresia includes acquired colonic stenosis that may be secondary to necrotizing enterocolitis (NEC) or infectious diseases; meconium plug syndrome in the context of Hirschsprung's disease or cystic fibrosis, and small left colon syndrome. (6)

Barium enema is particularly useful to identify the anatomy and configuration of the colon, as well as the level of atresia and other abnormalities. (13) During the procedure, physicians should be very attentive of intraluminal pressure, as it may lead to perforation. (8) Therefore, in some cases, it can be performed only post-operatively, using a water-soluble contrast agent.

Changes in surgical management have been reported in the literature, since previous recommendations included performing primary anastomosis depending on the location of the atresia with respect to its proximity to the splenic flexure. The current recommendation is to perform primary anastomosis regardless of the location. (6)

Cox *et al.* suggest that it is safe to perform primary anastomosis only if there is a maximum 3:1 caliber difference. (6) In this case, the difference of caliber between the proximal and distal segment was 10:1 and, therefore, initial management with derivative colostomy was chosen. Instillations of saline solution were administered through the anus to promote the growth of the distal hypotrophic colon. Despite this, during the second surgery, a discrepancy of 5:1 caliber was identified. In consequence, anastomosis was performed and a protective lateral window diversion was made proximal to it.

CONCLUSION

Although colon atresia is a rare disease, it must be considered as an important cause of distal intestinal obstruction in newborn patients. Early treatment is imperative as mortality of this disease increases considerably 72 hours after birth.

The discrepancy between the proximal and distal diameters should be considered during the procedure to decide whether to perform primary anastomosis. Considering the evidence of the association of colonic atresia with other gastrointestinal abnormalities, like Hirschsprung's disease —although the incidence reported is only 2%—, performing rectal biopsies during the initial intervention is recommended to discard this pathology, taking into account the risks of closing a colostomy in an aganglionic colon. (14) Once the early treatment of the disease is done, good prognosis and low mortality have been reported.

CONFLICT OF INTEREST

None stated by the authors.

FUNDING

None stated by the authors.

REFERENCES

1. **Dalla Vecchia L, Grosfeld J, West K, Rescorla F, Scherer L, Engum S.** Intestinal Atresia and Stenosis. *Archives of Surgery.* 1998;133(5).
2. **Freeman N.** Congenital atresia and stenosis of the colon. *British Journal of Surgery*. 1966;53(7): 595-599.
3. **Powell R, Raffensperger J.** Congenital colonic atresia. *Journal of Pediatric Surgery.* 1982;17(2):166-170.
4. **Sturim H, Ternberg J.** Congenital atresia of the colon. Surgery. 1966;59(3):458-464.
5. **Davenport M, Bianchi A, Doig C.** Colonic atresia: current results of treatment. *Journal of the Royal College of Surgeons of Edinburgh.* 1990;35(1):25–28.
6. **Hsu C, Wang S, Houng J, Chiang P, Huang C.** Congenital Colonic Atresia: Report of One Case. *Pediatrics & Neonatology.* 2010;51(3):186-189.
7. **Diaz D, Eftekhari K.** Atresia of the Colon Associated with Hirschsprung's Disease. *Archives of Iranian Medicine.* 2015;18(5):322.
8. **Etensel B, Temir G, Karkiner A, Melek M, Edirne Y, Karaca *et al.*** Atresia of the colon. *Journal of Pediatric Surgery*. 2005;40(8): 1258-1268.
9. **Rescorla F, Grosfeld J.** Intestinal atresia and stenosis: analysis of survival in 120 cases. *Surgery*. 1985;98(4):668 -676.
10. **Mansoor H, Kanwal N, Shaukat M.** Atresia of the ascending colon: A rarity. *APSP Journal of Case Reports.* 2010;1(1):3.
11. **Grosfeld J, Ballantine T, Shoemaker R.** Operative management of intestinal atresia and stenosis based on pathologic findings. *Journal of Pediatric Surgery.* 1979;14(3): 368-375
12. **Peña A, Bischoff A.** Surgical Treatment of Colorectal Problems in Children [recuerso electronico]. Cham; 2015
13. **Cox S, Numanoglu A, Millar A, Rode H.** Colonic atresia: spectrum of presentation and pitfalls in management. A review of 14 cases. *Pediatric Surgery International.* 2005;21(10):813-818.
14. **El-Asmar KM, Abdel-Latif M, El-Kassaby AA, Soliman MH, El-Behery MM.** Colonic atresia: association with other anomalies. J Neonat Surg. 2016; 5:47.

Universidad Nacional de Colombia
Bogotá Campus ~ Bogotá D. C., Colombia
Faculty of Medicine ~ Building 471 ~ Office 219
Telephonic numbers: 316 5000 ext. 15161 ~ Bogotá, D. C., Colombia
Web: http://revistas.unal.edu.co/index.php/care
email: casereport_fmbog@unal.edu.co