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CASE REPORTS

PARACOCCIDIOIDOMICOSIS

pulmonar asociada a choque séptico en un paciente inmunocompetente.

Editores: Edith Ángel Müller
Bibiana Jeannette Escobar
David Rincón Valenzuela

Centro Editorial
Facultad de Medicina
Sede Bogotá

UNIVERSIDAD NACIONAL DE COLOMBIA

Editorial:

CASE REPORTS

José Ricardo Navarro-Vargas
Universidad Nacional de Colombia
Bogotá Campus
Faculty of Medicine
Department of Surgery
Bogotá, D.C. – Colombia

Corresponding author
José Ricardo Navarro-Vargas
Departamento de Cirugía
Facultad de Medicina
Universidad Nacional de Colombia
Bogotá. Colombia. Email: jrnavarrov@unal.edu.co

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Observing and recording that which may be relevant and disseminating it through a concrete and well-written text are the teachings of Charles Darwin (1809-1882) and William Osler (1849-1919). Darwin published one of the greatest scientific books of all time, On the Origin of Species (1859), which was the result of multiple observations written down by the author on a daily basis. For his part, Osler was an outstanding doctor, a great semiologist, the father of modern medicine, and did not waste a moment to write down his scientific concerns and make them public.

Evidence-based medicine displaced case series and case reports in the medical literature. However, according to Vandenbroucke, "all kinds of research have a place in science, as well as their own space", in other words, they must be understood as complementary to medical research. Reporting particular or rare diseases is one of the objectives of case reports, but it is not the only one, since many of them have served as the basis for the development of major controlled clinical trials that have led to discover and describe new diseases, pharmacological side effects (beneficial or deleterious), the mechanisms of diseases, to provide education and promote medical audit, and to recognize rare manifestations of certain diseases.

Some researchers consider case reports as the first line of evidence-based medicine research. For example, finding a strange coagulation property in a person coming from a family that has had multiple venous thrombosis events led to discover activated protein C resistance, which is the most frequent cause of the congenital anomaly that leads to deep vein thrombosis: Factor V Leiden.

Case reports foster new ideas, new proposals and new theories; they allow to induce, infer, and even develop new projects from amazement, since a control group represents the expected course of the disease regarding the occurrence of a new phenomenon. All this makes it necessary to review the literature and make new associations, new interpretations, generate new knowledge.

For example, the case of Mary Mallon (better known as Typhoid Mary) was groundbreaking, since she was the first patient diagnosed as a healthy carrier of *Salmonella Typhi* in North America. However, thanks to the report of 53 infected patients and 3 deaths who had contact with her through her services as a cook, the necessary associations could be established from a public health perspective and she was left in quarantine (at that time, it was the best treatment to prevent the transmission of contagious diseases).

In order to prepare a good case report, it is necessary to ask the following questions: why exactly is this observation important? What teaching does it bring? Do you object to any previous evidence? Is it against some scientific concept or current evidence? Is there any unexpected association? Was it a provoked observation that led to study a mechanism? Can this mechanism be generalized? Is it a rare entity that, if reported, brings a great benefit to the scientific community and to patients? Once these questions have been answered, it is recommended to follow the IMRaD format (Introduction - Methods - Results –and - Discussion), although it is not always the most appropriate method to report a case or series of cases.

This issue presents case reports as broad as medicine itself: spontaneous pneumomediastinum, Ekbom's syndrome, appendiceal cystic dilation, pulmonary paracoccidioidomycosis and septic shock in an immunocompetent patient, congenital laryngeal saccular cyst,

splenic rupture associated with thrombocytopenic purpura due to mononucleosis infection, dissection of ascending aorta in a patient with Marfan syndrome, and vesicular agenesis and choledocholithiasis.

As you can see, case reports will continue to give us a lot to talk and teach about, I would say, forever, since they are the starting point for macro studies done with the best available clinical evidence.

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PULMONARY PARACOCCIDIOIDOMYCOSIS ASSOCIATED WITH SEPTIC SHOCK IN AN IMMUNOCOMPETENT PATIENT. CASE REPORT

Keywords: Paracoccidioidomycosis, Fungus, Paracoccidioides, amphotericin B, Sepsis, Klebsiella pneumoniae.
Palabras clave: Paracoccidioidomicosis; Hongo; Paracoccidioides; Anfotericina B; Sepsis; Klebsiella pneumoniae.

Freddy Mauricio Quintero-Álvarez
Juan Pablo Báez-Duarte
Jessica Paola Montes-Ortíz
Sergio Andrés Mendinueta-Giacometto
Emergency Service
Hospital Universitario de Santander
Bucaramanga – Colombia.

José Mauricio García-Habeych
Department of Internal Medicine
Faculty of Health
Universidad Industrial de Santander
Bucaramanga – Colombia.

Corresponding author:
Freddy Mauricio Quintero-Álvarez.
Email: freddyquintero05@gmail.com

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ABSTRACT

Introduction. Paracoccidioidomycosis (PCM) is a chronic granulomatous disease caused by the dimorphic fungus known as Paracoccidioides brasiliensis. This entity compromises mainly the lungs, but can spread to other organs, with particular trophism, through oral mucosa, adrenal glands, lymph nodes, among others.

Case presentation. This paper reports the case of a male patient with pulmonary PCM treated at the Hospital Universitario de Santander. The patient was admitted with initial suspicion of active pulmonary tuberculosis due to the presence of multiple cavitations and nodules of random distribution in the lung parenchyma observed in the chest tomography, and subsequent isolation of yeasts compatible with Paracoccidioides. Amphotericin B deoxycholate was administered without favorable outcomes and development of septic shock by extended spectrum Klebsiella pneumoniae. In spite of multi-conjugate antibiotic management, the patient presented multiple organ failure syndrome with fatal outcome at 21 days of hospitalization.

Conclusion. Pulmonary PCM is an endemic disease that leads to an inadequate immune response of the host that —along with risk factors such as smoking, alcohol abuse, malnutrition and low socioeconomic status— facilitates the onset of life-threatening infections or coexisting diseases. Timely diagnosis based on early clinical suspicion potentially influences the patient's survival.

INTRODUCTION

Paracoccidioidomycosis (PCM), also known as South American blastomycosis, was first described in 1908 by Adolfo Lutz. This is a chronic granulomatous disease caused by the dimorphic fungus known as Paracoccidioides brasiliensis. It is endemic in Latin America, with predominance in Brazil —with the highest incidence in the southeast of the country— followed by Venezuela, Colombia, Ecuador and Argentina. (1). The dimorphic fungus grows as a yeast in the tissues of the host and in cultures at 36-37°C, but it develops as a slow growing mold at temperatures <28°C (2).

This disease compromises mainly the lungs, but can spread to other organs, with particular trophism, through oral mucosa, adrenal glands, reticuloendothelial system, skin and bones. (3) This paper reports a case of pulmonary PCM in an immunocompetent patient —a rare disease in Colombia— who was diagnosed in a tertiary care hospital in Santander, Colombia, and had a fatal outcome.

CASE PRESENTATION

67-year-old mestizo male from the rural area of the municipality of Aratoca (where temperatures vary between 16°C and 26°C), who worked as a farmer (mostly in coffee crops), with unclear pathological history of epilepsy without treatment, chronic smoking associated with regular consumption of alcoholic drinks, and without exposure to individuals with a history of tuberculosis.

The patient visited a primary health center referring symptoms of 4 months evolution including productive cough with mucopurulent expectoration, progressive dyspnea even when putting small efforts, fever, chills and unintentional progressive weight loss. He stated that the symptoms exacerbated 7 days before with hemoptoic cough and evening diaphoresis. The patient was referred to a secondary

care health center where a chest x-ray was performed, showing abundant alveolar opacities in both pulmonary fields, formation of diffuse pneumatoceles, and signs of air trapping (Figure 1). A sputum KOH test was performed, reporting a double refractory wall of yeast with intracytoplasmic vacuoles with multiple or chain budding. The results were compatible with paracoccidioides, so he was assessed by the internal medicine service and referred to the Hospital Universitario de Santander due to the high risk of ventilatory failure.

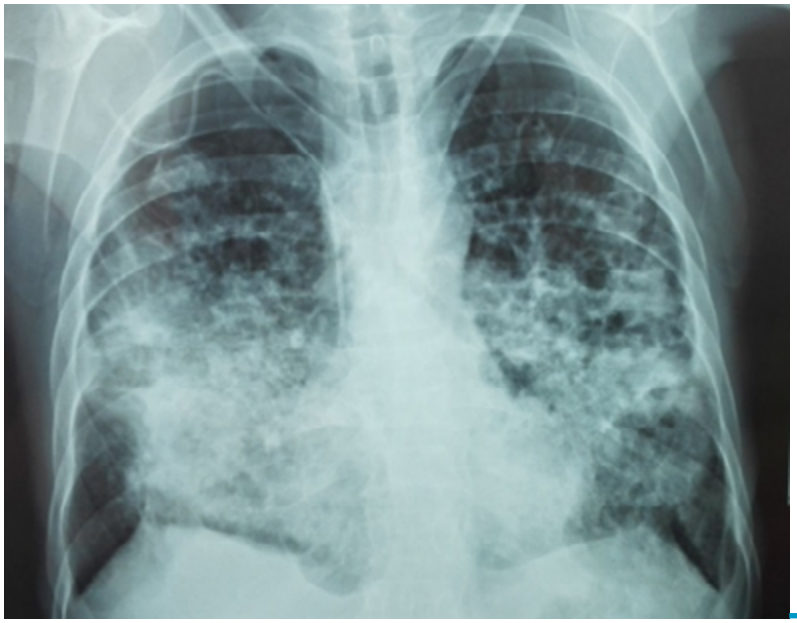

Figure 1. Chest x-ray with abundant alveolar opacities in both lung fields with formation of pneumatoceles.
Source: Own elaboration based on the data obtained in the study.

On physical examination at admission, the patient presented with respiratory distress at rest despite supplementary oxygen with a mask and Ventury mask with a 50% inspired fraction, significant reduction in muscle mass, vital signs with normal blood pressure, a heart rate of 106 bpm and respiratory rate of 23 breaths per minute. He did not have a feverish state at the time of assessment, but there were intercostal retractions with diminished respiratory sounds on auscultation and rhonchi with diffuse rales.

High-resolution computed tomography (HRCT) of the chest was performed (Figure 2), which showed multiple cavitations, mostly thin-walled, and randomly distributed nodules with a predominantly right pleural effusion.

The initial tests ruled out infection by human immunodeficiency virus through fourth-generation ELISA technique. Three serial sputum smears were performed, which reported negative results for acid-alcohol resistant bacillus (BAAR). In addition, a polymerase chain reaction (GenXpert) was performed for each bacilloscopy sample, yielding negative results for BAAR as well. A sputum KOH test was performed again, which confirmed the presence of blastoconidia related to *P. brasiliensis.*

Similarly, leukocytosis, progressive thrombocytopenia and deterioration of renal function were documented (Table 1). Blood gas tests were made, showing hypoxemia with hypercapnia associated with mixed acid-base disorders due to normochloremic metabolic acidosis, metabolic alkalosis and respiratory alkalosis (Table 2).

During hospital stay, the patient was assessed by pneumology, indicating treatment

with amphotericin B deoxycholate at a dose of 0.7-0.8 mg/kg/day. On day 12 of hospitalization, the patient presented increased respiratory function and worsening of hypoxemia (Table 2), invasive mechanical ventilatory support requirement, hypotension refractory to fluid management, and vasopressor support with norepinephrine was initiated. In addition, control laboratories reported significant leukocytosis and thrombocytopenia without bleeding (Table 1). Bronchial secretion and blood cultures reported growth of *Klebsiella pneumoniae* with antimicrobial resistance pattern of extended-spectrum beta-lactamase (ESBL), for which antibiotic treatment with Meropenem was prescribed.

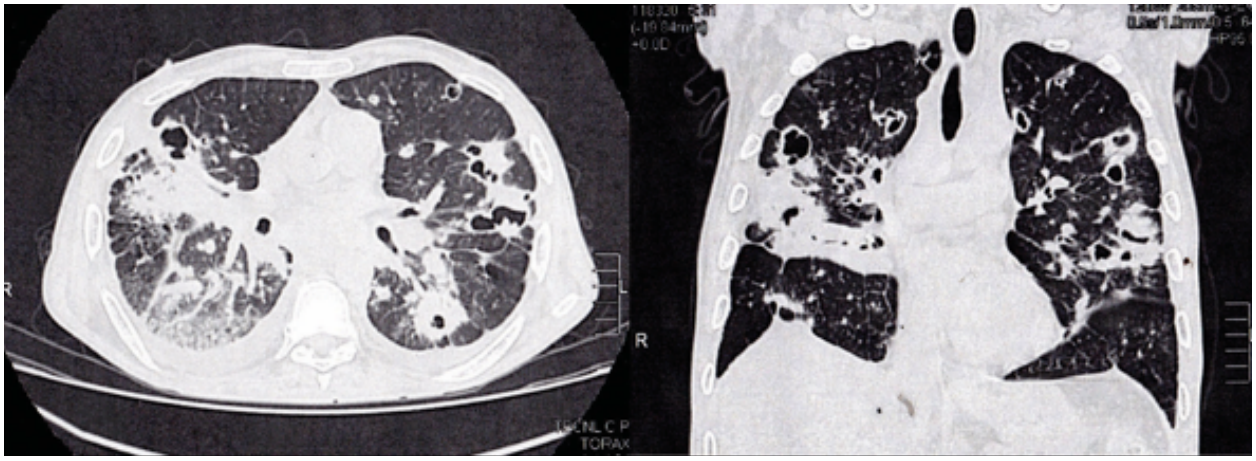

Figure 2. High-resolution computed tomography of the chest.
Source: Own elaboration based on the data obtained in the study.

Table 1. Laboratory tests.

Day	WBC (5-10) x 10^3/uL	Platelets (150-450) x 10^3/uL	BUN (8-23) mg/dL	Creatinine (0.67-1.17) mg/dL
1	17.5	480	10	0.34
2	10.6	338	23	1.24
3	10.5	320	29	1.24
4	10.4	307	30	1.03
5	5.8	283	30	0.89
6	11.3	234	30	0.89
7	15.4	215	27	0.85
8	22.2	205	23	0.93
9	19.7	186	23	0.93
10	49.7	68	13	0.92
11	67	28	17	1.55
12	31.4	10	20	2.25
13	23.8	10	19	2.22

Continúa en la siguiente página.

Day	WBC (5-10) x 10^3/uL	Platelets (150-450) x 10^3/uL	BUN (8-23) mg/dL	Creatinine (0.67-1.17) mg/dL
14	14.4	16	21	2.06
15	22.9	44	22	1.76
16	15.6	47	24	1.7
17	16	90	31	1.44
18	12.5	99	30	1.22

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

Table 2. Arterial blood gases.

Day	pH (7.35-7.45)	PO2 (75-100) mmHg	PCO2 (38-42) mmHg	FiO2	PaFi (>300)	SO2 (94-100) %	HCO3 (22-28) mEq/L	Lactate (0.5-2) mmol/L
8	7.40	78	32.1	50%	156	96	19.5	1.8
9	7.40	99	32.7	50%	198	98	20.0	Not taken
10	7.25	83	38.1	70%	118	94	16.4	5.6
11	7.22	107	42.0	80%	134	97	17.1	2.9
12	7.23	100	40.2	70%	143	97	16.7	3.6
13	7.27	94	40.1	70%	134	97	18.0	2.7
14	7.27	79	47.5	90%	87	95	21.7	2.2
15	7.26	148	47.2	100%	148	99	21.1	2.4
16	7.23	119	55.0	70%	170	98	22.9	2.2
17	7.20	103	62.4	70%	147	97	24.1	2.6

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

The subject was taken to a medical-scientific autopsy, in which histopathological studies were carried out, including cuts of the lung parenchyma (Figure 3), showing the presence of granulomas with abundant multinucleated Langhans giant cells and refractory rounded structures in the cytoplasm. Some of the latter had multiple budding and were compatible with *P. brasiliensis* through periodic acid-Schiff (PAS) staining and methenamine silver stain (GMS). This finding was also observed in lymphatic and hematopoietic system cuts, while no foci of pneumonia caused by *K. pneumoniae* were identified. The renal parenchyma showed diffuse and generalized necrosis of the epithelium of the proximal tubules, secondary to tissue hypoperfusion and triggered by septic shock.

DISCUSSION

This paper reported a case of PCM in a patient with undefined structural lung disease, with subsequent complication due to septic shock associated with *K. pneumoniae*. This disease is found in Latin America, with 80% of the cases reported in Brazil, followed by Colombia, particularly Santander and Norte de Santander, which are considered endemic areas. The national incidence ranges from 0.1 to 2.4 per million inhabitants and mortality is approximately 10-15% (4-7).

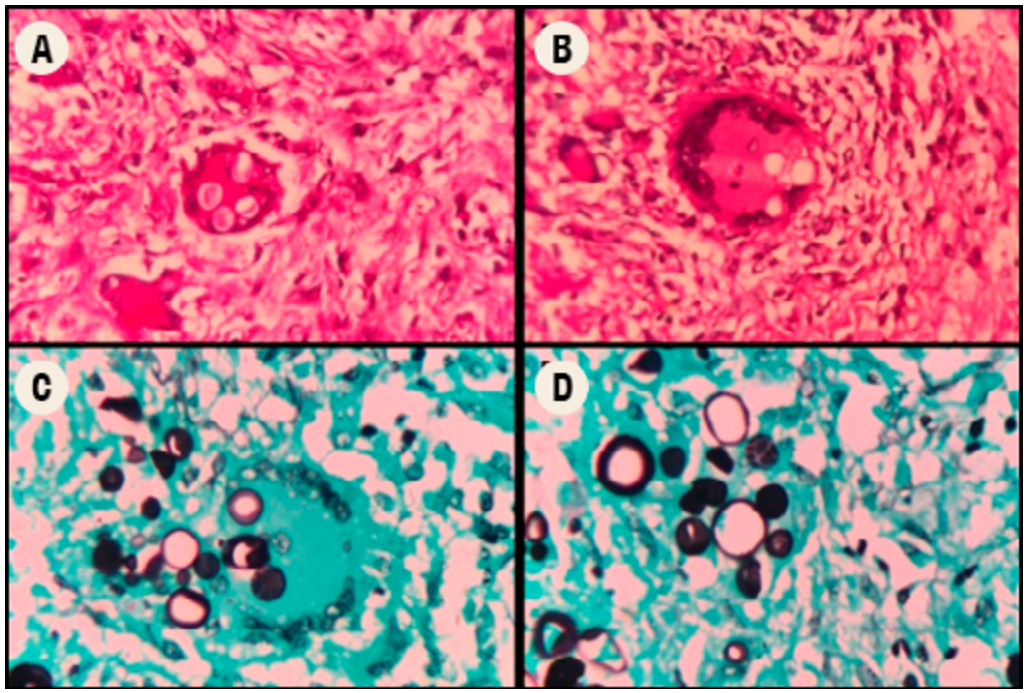

Figure 3. Lung histopathology. A and B: periodic acid-Schiff (PAS) staining; C and D: coloration with methenamine silver stain (GMS) corresponding to P. brasiliensis.

P. brasiliensis is a dimorphic fungus (2) that affects mostly individuals who are engaged in agricultural activities because of the manipulation of soils that generate aerosols with spores of this fungus, which end up being inhaled by the farmers. In addition, a greater incidence is observed in the male gender, with a male-to-female ratio of 2:1 (3). This difference may be caused by the fact that men are more dedicated to agricultural activities and the protective effect of estrogen in women, thus avoiding the transition of the fungus from mycelium to yeast (2). The transmission from person to person of this mycosis has not been documented, and tobacco and alcohol consumption has been associated with the risk of PCM infection (8).

PCM is a chronic systemic invasive mycosis that, like tuberculosis, mainly affects the lungs; both diseases can coexist in 15-20% of cases (9,10) with varying degrees of parenchymal injury and tendency to fibrosis (2). Given this possible coexistence, tuberculosis was one of the first pathologies to be discarded in this patient, besides an immunodeficiency state as a trigger of the infection.

Moreover, the association with other pulmonary structural processes, such as chronic obstructive pulmonary disease and lung cancer, has been described (11). In this case, the association of PCM, in the absence of tuberculosis, and the bacteremia by *K. pneumoniae* was striking. Although the concurrent process of tuberculosis and pulmonary PCM (11) and pulmonary tuberculosis and bacterial pneumonia have been documented (12), the association of pulmonary PCM and *K. pneumoniae* infections is not frequent. In a series of reference cases, only one presented the concomitant process (13). Although this patient did not comply with the description of the CD40L phenotype and HIV infection

was ruled out, the presence of structural lung disease should be highlighted, as well as his malnutrition status, smoking and active alcohol consumption, hospital stay —which favors nosocomial infections—, and his exposure to drugs with potent adverse effects such as amphotericin B —which favors a possible poor immune response.

The presence of non-modifiable risk factors such as age and sex, together with modifiable factors such as type of job, smoking and consumption of alcoholic drinks, associated with a low socioeconomic level and high degree of malnutrition conditioned a depressed immune response and subsequent infection by *P. brasiliensis* (11). However, mycosis per se modulates the host's innate and acquired immune response, so that overlapping or co-existing infections may be facilitated.

Chest scans are the main tool to suspect this infection. Findings are nonspecific and include diffuse micronodular infiltrates predominantly in the middle zone of the lung, cavitations and tumor masses (5,14,15). Chest radiography shows mainly interstitial opacities (nodular or reticular), and when caverns are observed, pulmonary tuberculosis is the main differential diagnosis. (15,16). In high-resolution chest tomography, untreated PCM findings are characterized by attenuation of the ground-glass lung parenchyma associated with small centrilobular nodules, cavity nodules, large nodules, and scar emphysema (5,15,16), being the peripheral and the posterior distributions in the lung predominant (16). The reversed halo sign is observed in about 10% of cases (16).

Considering the wide differential diagnosis (17) and the clinical-radiological dissociation (18), a rapid confirmation of the diagnosis is required to initiate treatment. These tools include microbiological, immunological and molecular evaluation methods (19). The reference diagnostics are:

- Pulmonary tuberculosis
- Histoplasmosis
- Systemic lupus erythematosus
- Hodgkin lymphoma
- Cutaneous and mucosal Leishmaniasis
- Wegener's granulomatosis
- Actinomycosis
- Blastomycosis

Microscopic evaluation of sputum or compromised tissues allows rapid diagnosis. The use of routine stains, such as potassium hydroxide (KOH), calcofluor for wet mounts and Grocott-Gomori staining or periodic acid-Schiff (PAS) for smear (20), allows to visualize spherical or double-walled yeasts, from 30µ to 60µ in diameter, with multiple budding (14). Digested or concentrated sputum can be positive in 60-70% of chronic PCM cases (15). Cultivation of the fungus on Sabouraud dextrose agar medium is ideal for isolation, but may take 20 to 90 days (14,15,21). In certain cases, microbiological documentation of the presence of PCM is not possible, so, in case of clinical and radiological suspicion, the use of serological methods is necessary.

The detection of antibodies against the gp43 antigen is carried out by means of an immunodiffusion reaction, which is positive in 90% of the patients prior to the eradication treatment (19,22,23). In HIV patients, results should be interpreted with caution due to cross-reaction with *Histoplasma capsulatum* (24). Another option is to detect the p27 antigen, with a sensitivity and specificity of almost 100% (5,20), which avoids cross reaction (20). Molecular methods based on the polymerase chain reaction technique are not commercially feasible yet (20).

The treatment of *P. brasiliensis* differs from other invasive fungi due to its high sensitivity to different antifungal medications; the selection should be made according to the severity of the disease (Table 3).

Table 3. Treatment options for pulmonary paracoccidioidomycosis.

Drug	Dose	Interaction	Adverse effects
Trimethoprim/ Sulfamethoxazole	480-960 mg every 8-12 hours	Phenytoin	Leukopenia, megaloblastic anemia, thrombocytopenia
Ketoconazole	200-400 mg/day	Astemizole, fexofenadine, loratadine	Itching, vomiting, nausea, anorexia
Itraconazole	100-400 mg/ day	Cisapride, quinidine, diazepam, digoxin, indinavir, ritonavir, sulfonylureas	Nausea, vomiting, increased serum transaminases, hypokalemia, hypertriglyceridemia and hyperuricemia.
Fluconazole	300-400 mg/ day	Cisapride, cyclosporine, rifampicin, rifabutin, sulfonylureas, theophylline	Headache, nausea, vomiting, abdominal pain, diarrhea
Amphotericin B	1-3 mg/kg/day	Cyclosporine and aminoglycosides	Fever, dyspnea, bronchospasm, redness, tachycardia, acute kidney injury

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

In cases of mild to moderate infection, the therapeutic option is itraconazole. The literature reports a 90% cure rate with relapses up to 15%. One treatment option is to combine trimethoprim and sulfamethoxazole (25). In case of serious infections, the use of intravenous agents is indicated, being amphotericin B in its conventional or lipid formulation the preferred choice (14,15,19) —as administered to this patient— evaluated at a dose of 1-3 mg/kg/day. Similarly, in order to prevent relapse, long-term treatments with sulphonamides or azoles are indicated (15).

Although PCM is an endemic entity with high incidence and mortality, few studies have been carried out to define the appropriate therapeutic option (15). In addition, both the adverse effects and the interaction of medications that may affect its effectiveness must be considered (10).

CONCLUSIONS

Pulmonary paracoccidioidomycosis is an endemic disease in Santander, although it is underdiagnosed due to the presence of more common pathologies and similar clinical characteristics such as tuberculosis and histoplasmosis. For this reason, late diagnosis is frequent in most of the affected individuals. Unfortunately, mycosis modulates the immune, innate and acquired response of the host, thus facilitating the onset of superimposed or co-existing infections, whose complications such as sepsis and secondary multiple organ failure eventually lead to a fatal outcome.

The timely diagnosis of PCM is of utmost importance, considering that the ideal diagnosis is achieved through microscopic evaluation of the sputum or detection of gp43 antibodies. Pharmacological treatment, despite being

aggressive and considering that the disease usually occurs in immunologically compromised patients, could have a favorable impact on the reduction of complications and mortality in this type of patients.

CONFLICT OF INTERESTS

None stated by the authors.

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SPLENIC RUPTURE ASSOCIATED WITH THROMBOCYTOPENIC PURPURA CAUSED BY INFECTIOUS MONONUCLEOSIS. CASE REPORT

Palabras clave: Infección; Virus de Epstein-Barr; Púrpura; Rotura de bazo.
Keywords: Infection; Epstein-Barr virus; Purpura; Spleen Rupture.

Alicia Santa Cortés-González
Verónica García-Torres
Pediatric Service - General Hospital of Zone Number 11- Instituto Mexicano del Seguro Social.
Faculty of Medicine - Universidad Veracruzana - Xalapa - México

Nataly Yazmín Cortés-Trujillo
Pediatric Service - General Regional Hospital Number 36 - Instituto Mexicano del Seguro Social - Puebla - México
Faculty of Medicine - Universidad Popular Autónoma de Puebla - Puebla - México

Rocío Maily Vázquez-Martínez
Faculty of Medicine - Universidad Nacional Autónoma de México - México D.F. - México
Instituto de Ciencias de la Salud - Universidad Autónoma del Estado de Hidalgo - Pachuca - México

Uziel Suarez-Cruz
Faculty of Medicine - Universidad Veracruzana - Xalapa - México
Pediatric Service - General Hospital of Zone Number 36 - Instituto Mexicano del Seguro Social - José Cardel - México

Corresponding author:
Alicia Santa Cortés-González.
Email: aliciasantac@hotmail.com

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ABSTRACT

Introduction. Splenic rupture associated with thrombocytopenic purpura caused by infectious mononucleosis is extremely rare. The evolution of patients with infectious mononucleosis associated with Epstein-Barr virus is favorable, self-limiting and does not require specific therapeutic interventions. The symptoms are well tolerated and have a low frequency of complications.

Case presentation. Female 12-year-old patient presenting with diffuse abdominal pain, distension, nausea, tegument pallor and unquantified fever for two days. Upon admission to the emergency department, hemodynamic decompensation, purpuric lesions and ecchymosis in the limbs were observed. Laboratory and cabinet studies were carried out to confirm anemia, thrombocytopenia and splenic hematoma. Finally, an exploratory laparotomy was performed considering the possibility of hemoperitoneum.

Results. The patient presented with splenomegaly, broken subcapsular hematoma, bleeding of 4000mL and accessory spleen lobe with splenic rupture.

Conclusions. Spontaneous splenic rupture is a rare but possible complication of infectious diseases. However, its association with thrombocytopenic purpura is extremely rare.

RESUMEN

Introducción. La rotura esplénica asociada a la presencia de purpura trombocitopénica causada por mononucleosis infecciosa es extremadamente rara; la evolución de los pacientes con mononucleosis infecciosa asociada al virus de Epstein-Barr es benigna y autolimitada y no requiere intervenciones terapéuticas específicas. El cuadro es bien tolerado y tiene una baja frecuencia de complicaciones.

Presentación del caso. Paciente femenino de 12 años de edad con dos días de evolución de dolor abdominal difuso, distensión, náuseas, palidez de tegumentos y fiebre no cuantificada, quien a su ingreso al servicio de urgencias muestra datos de descompensación hemodinámica, lesiones purpúricas y manchas equimoticas en extremidades. Se realizan estudios de laboratorio y gabinete que confirman anemia, trombocitopenia y hematoma esplénico, por lo que se practica laparotomía exploradora ante la posibilidad de hemoperitoneo.

Resultados. La paciente presenta esplenomegalia, hematoma subcapsular roto con sangrado de 4000mL y lóbulo accesorio de bazo con rotura esplénica.

Conclusiones. La rotura espontánea del bazo es una complicación infrecuente pero posible en enfermedades infecciosas; sin embargo, su asociación a purpura trombocitopenica es extremadamente rara.

INTRODUCTION

Splenic rupture associated with infectious mononucleosis (IM) occurs in 0.1-0.5% of cases and is a very rare complication. Its association with thrombocytopenic purpura is extremely rare, since it only occurs in 0.2-0.6% of the total cases. The causative agent is the Epstein-Barr virus (EBV) with positive heterophile antibodies (1,2).

The following case is presented considering the low incidence of this condition and its

aggravating influence on a disease that is usually benign.

CASE REPORT

Female 12-years-old mestizo patient from Perote, Mexico, the third child of a healthy 33-year-old mother, eutocic delivery, without perinatal complications. She was a high school student, of low socio-economic conditions and fed with an adequate quantity of food, with predominance of carbohydrates and vegetables. The onset of symptoms occurred 2 days before consultation, presenting with diffuse abdominal pain, distension, nausea, tegument pallor and unquantified fever; in consequence, she was taken to the emergency service of the General Hospital of Zone Number 11 of Instituto Mexicano del Seguro Social in Xalapa, Mexico.

Physical examination revealed tegument pallor, hyperemic oropharynx, tonsil hypertrophy, cervical adenitis, cardiopulmonary signs without compromise, distended abdomen painful on palpation predominantly in left flank with splenomegaly 3-3-3 below costal margin, no hepatomegaly on palpation, peritoneal irritation, diminished peristalsis, petechiae on the limbs and ecchymosis, no edema and good capillary refill. Medical history revealed a traumatic brain injury a year earlier when another child threw a stone that accidentally hit her while they were playing. During that episode, the girl presented with a subarachnoid hematoma that was drained without complications, and was prescribed a treatment with magnesium valproate. Furthermore, she had a history of recurrent upper respiratory infection; the last episode occurred three weeks before developing the current symptoms.

On admission to the emergency service, complementary tests were performed, reporting Hb 7g/dl, Ht 22%, thrombocytopenia 94 000/mm3, leukocytes 5 900/mm^3, neutrophils 2 700/mm^3, and lymphocytes 2.66/mm^3. Liver function tests reported TGO 66 UI/L, TGP 94 IU/L, LDH 1 375 IU/L, prolonged prothrombin time PT of 17.1 seconds, and aPTT 29.5 of seconds. Abdominal ultrasound revealed hepatopathy of origin to be determined, free fluids in cavity greater than 100mL, enlarged spleen (138x74mm) with hypoechoic area in the upper pole suggesting hematoma and accessory splenic lobe. The abdominal X-ray showed distended colon and an image suggestive of splenic hematoma already confirmed by ultrasound.

During her stay in the emergency room, a hematologic assessment was requested, revealing non-immunological thrombocytopenia secondary to hypersplenism. The patient was also assessed by general surgery when hemodynamic instability and hemoglobin decrease were observed, leading to decide an exploratory laparotomy before a possible hemoperitoneum occurred, and was transferred to the operating room.

Hemoperitoneum was observed during the surgery, so 4000mL of blood were aspirated and systematic cavity revision was initiated. Splenomegaly (20x10cm), broken subcapsular hematoma and splenic accessory lobe were found (Figure 1). Splenectomy was performed in which broken subcapsular hematoma was observed (Figure 2).

Three units of erythrocyte concentrates (750mL) were transfused urgently and cefotaxime and metronidazole were administered. Pathology reported chronic passive splenic congestion and subcapsular hematoma.

The Paul-Bunnell test, which yields a positive result when it detects heterophilic antibodies in the blood and a negative result when the suspicion of MI and the investigation of heterophile antibodies are persistent, was positive

in this patient, so it was not necessary to look for specific EBV antibodies. The evolution of the patient was satisfactory, the lesions in the lower limbs disappeared, as well as the symptoms. Vaccination against pneumococcus and influenza was applied without complications, ferrous fumarate supplement was administered for 6 months, hospital discharge was given 5 days after the surgery, and the patient was followed by external consultation at one the month, when an adequate health status was observed.

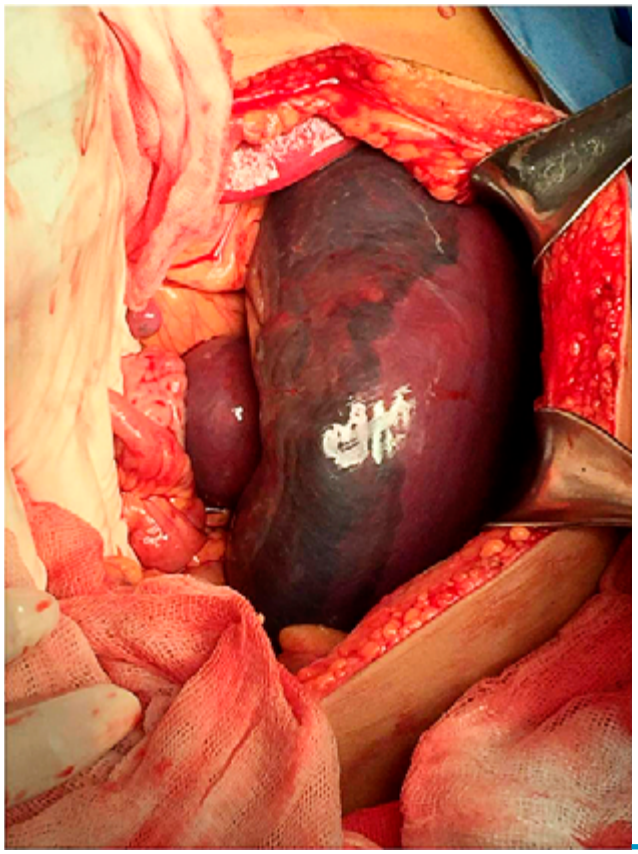

Figure 1. Subcapsular hematoma and splenic accessory lobe.
Source: Own elaboration based on the data obtained in the study.

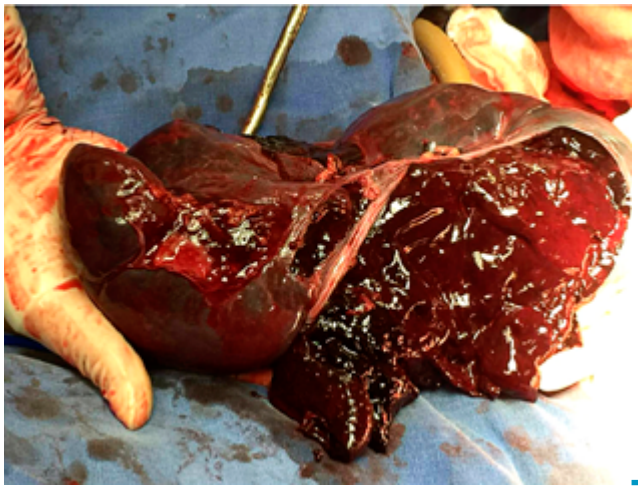

Figure 2. Splenic rupture.
Source: Own elaboration based on the data obtained in the study.

DISCUSSION

Patients with infectious mononucleosis associated with EBV do not require specific therapeutic interventions. This infection occurs at any age, but most cases associated with this virus occur during adolescence or early adulthood. Therefore, the age of the patient has a profound influence on the clinical expression of the infection. Young children tend to present signs such as dermatitis, neutropenia or pneumonia more frequently, while heterophile antibodies are negative in about half of the cases. As age increases, the disease develops more symptoms, with an increase in serological positivity. At 25 years of age, most people are seropositive and not susceptible to reinfection (3).

The typical clinical manifestations of MI are pharyngitis, fever and lymphadenopathy. In more than 50% of cases, pharyngitis is the most frequent symptom and can be severe. The onset may be abrupt, but usually prodromal symptoms occur such as chills, diaphoresis, low-grade fever, anorexia, and malaise (4,5). Fever occurs in more than 90% of patients with MI, usually showing peaks during the afternoon with values of 38-39°C, although they can be up to 40°C; in most cases, the fever resolves within a period of 10 to 14 days (6,7,8).

Dermatitis can be macular, erythematous, petechial, scarlatiniform, urticarial, herpertiform or similar to erythema multiforme, mainly involving the upper limbs and the trunk, and is observed in 5% of patients (9,10). The symptoms are well tolerated and have a low frequency of complications. Most cases have a benign course and patients should receive only symptomatic management and be warned of potential hematological, neurological, cardiac, respiratory, dermatological, renal, splenic, hepatic, immunological and gastrointestinal complications, which, in general, are very rare (11,12).

MI diagnosis is clinical, but laboratory tests are essential to confirm the cause or determine the diagnosis in atypical presentations. The association of heterophile antibodies response during the acute phase of MI was observed by Paul and Bunnell in 1932. They stated that, after infection, 85-90% of the patients produce IgM heterophile antibodies, which agglutinate when they are mixed with ram or horse red blood cells. This procedure was called the Paul-Bunnell test; it was the first serological test developed for this disease, and is currently used to establish a diagnosis. Heterophile antibodies receive this name because they can react with antigens from unrelated species (8,13).

The patient was assessed according to her age group (adolescence) and presented the most symptomatic form of the disease with increased serological positivity, and dermatological and hematological complications with purpuric lesions and ecchymosis in the lower limbs due to a low platelet count. Moreover, she presented thrombocytopenic purpura secondary to infection. Immunological balance breakdown between B lymphocytes and the subtypes of T lymphocytes observed in this patient may lead to the appearance of antibodies against platelets that would explain the purpura. (14).

Trombocytopenia associated with mononucleosis may promote bleeding due to the decrease of the manganese dismutase enzyme. Also, protection against free radicals decreases because of the antibodies, altering the capillary endothelium and increasing the fragility of the spleen, leading to hypersplenism (15).

Subsequently, the patient presented hemodynamic instability with shock, anemia and splenic rupture with ruptured subcapsular hematoma that may have been preceded by an

intermittent subcapsular hemorrhage. Splenic rupture is rare and presents as a picture of acute abdominal pain (12); in consequence, an emergency splenectomy was recommended for this patient.

Most cases of infectious mononucleosis are benign and self-limited, and resolve spontaneously within 2-3 weeks (5,10).

CONCLUSION

Spontaneous splenic rupture is a rare complication, but possible due to infectious diseases, being malaria and infectious mononucleosis the most frequent. Appropriate identification and treatment of this emergency requires a multidisciplinary approach and rapid action due to its high morbidity and mortality rates.

CONFLICT OF INTERESTS

None stated by the authors.

FUNDING

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AGENESIS OF THE GALLBLADDER AND CHOLEDOCHOLITHIASIS. CASE REPORT

Keywords: Gallbladder, abnormalities; Choledocholithiasis; Common bile duct diseases; Laparoscopy; CPRE (MeSH).
Palabras clave: Vesícula biliar, Anomalías; Coledocolitiasis; Conductos biliares, anomalías; Laparoscopia; ERCP (DeCS).

Daniel Rodrigo Riaño Pinto
Departamento de Cirugía
Facultad de Medicina
Universidad Nacional de Colombia
Sede Bogotá – Colombia

Corresponding author:
Daniel Rodrigo Riaño Pinto
Department of Surgery - Faculty of Medicine -
Universidad Nacional de Colombia
Bogotá - Colombia.
Email: drrianop@unal.edu.co

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ABSTRACT

Introduction. The most frequent elective surgery in General Surgery is the gallbladder surgery (cholecystectomy)in General Surgery in adults. There are many abnormalities of the gallbladder and the common bile duct. The most uncommon case is gallbladder agenesis. It could be difficult even for a experimented surgeon. It´s the most erratic biliar duct malformation, and there are near 500 cases reported.(1)

Case presentation. We present a case report of a 44 years old female patient, with abdominal pain in right superior quadrant, history of jaundice and acholia, with higher hepatic enzymes and direct bilirubin, with high probability of Choledocholithiasis. The images had not finding of the gallbladder (ultrasonography, Magnetic Resonance). The endoscopic retrograde colangiopancreatography (ERCP) was done, without removal of lithiasis and it used stent.

Finally we did laparoscopy common bile duct exploration, and the surgery confirmation of agenesis of the gallbladder, with mecanic lithotripsy, and the success with total resolution of the patology in the posterior medical control.

Conclusion. Agenesis of the gallbladder is a rare pathology that not many surgeons have the opportunity to treat. However, a surgeon must be prepared for any malformation and anatomical variant.

RESUMEN

Introducción. Una de las cirugías electivas que más desarrolla el cirujano general en adultos, es la colecistectomía. Sin embargo, el cirujano debe estar preparado para múltiples hallazgos, entre ellas las malformaciones. El caso más exótico que puede encontrar el mismo, es la agenesia de la vesícula biliar, el cual puede desorientar completamente a un cirujano incluso experimentado, debido a que es la malformación con más baja incidencia de las vías biliares y sólo hay cerca de 500 casos reportados en la literatura.1

Presentación del caso. Se presenta el caso de una paciente de 44 años, con cuadro clínico de dolor abdominal en cuadrante superior derecho, historia clínica de ictericia y acolia, con elevación del perfil hepático (hiperbilirrubinemia directa) y alta probabilidad de coledocolitiasis. En los estudios imagenológicos (Ultrasonografia y Resonancia Nuclear Magnética de Vías biliares), no hubo hallazgo de vesícula biliar.

Por ende, se realizó la colangiografía pancreática retrograda endoscópica (CPRE) en la cual no se logró la extracción de cálculos, y requirió uso de Endoprótesis. Finalmente, el tratamiento derivó a exploración de Vías biliares por laparoscopia, en dónde se confirmó el hallazgo de agenesia de vía biliar sospechado por la Resonancia Magnética y ecografías previas, se realizó entonces litotripsia mecánica dirigida con resolución completa del cuadro clínico. Y seguimiento posterior exitoso, con mejoría de la sintomatología inicial de la paciente.

Conclusión. La agenesia vesicular una patología extraordinaria que incluso el cirujano general no se pueda encontrar alguna vez en su vida. Sin embargo, este debe estar preparado para todas las malformaciones y variantes anatómicas.

INTRODUCTION

Agenesis of the gallbladder is the least frequent malformation of the bile duct, with a

variable incidence between 0.01 and 0.06%, according to the literature, although it may be lower, with a ratio women to men of 3:1 (1). This condition was first described by Lemery in 1701, although other authors cite Bergman in 1702 as the first (2). The literature shows about 400 case series in total, which is why this case and literature review are relevant.

Agenesis of the gallbladder is not an isolated malformation, since studies show great association with other cardiovascular, genitourinary and central nervous system conditions in up to 15-33% of the cases, specifically pulmonary agenesis, tetralogy of Fallot, and anomalies in the limbs and in the genitourinary tract (1,3-6).

Most case series have been presented by Kumar, who classifies patients with agenesis of the gallbladder into three groups: a) asymptomatic patients (35%) diagnosed after performing studies to establish another cause; b) symptomatic patients (50%), of which 33% have a dilated primary bile duct and 33% lithiasis in the main bile duct, and c) children with more complex congenital anomalies (15%) such as lung agenesis, tetralogy of Fallot, and abnormalities of the limbs or the genitourinary system, which are often incompatible with life (7).

Moreover, if information relates only to symptomatic gastrointestinal cases, the statistics become more revealing, since 34% of them present with acid-peptic disease, and 50% are symptomatic with respect to pathology of the bile duct (7). Additionally, if only patients with biliary tract symptoms are considered, 33% present with dilation of the bile duct and 33% with choledocholithiasis, while the remaining cases report sphincter of Oddi dysfunction. With this in mind, it is possible to conclude that vesicular agenesis is a risk factor for such pathologies, as some authors have suggested (1,6-9).

CASE PRESENTATION

44-year-old woman from Bogotá D.C., mestizo, who was initially treated in the outpatient clinic for pain in the epigastrium and in the right hypochondrium, exacerbated by the intake of fatty foods, with approximately one year of evolution The symptoms led to conclude a clinical picture suggestive of vesicular lithiasic pathology, so an ambulatory ultrasound was performed, showing no gallbladder, an intrahepatic and extrahepatic bile duct of normal caliber, and a 6 mm common bile duct. A subsequent esophagogastroduodenoscopy discarded acid-peptic disease, so a magnetic resonance cholangiography (MRC) was requested.

Two weeks after the initial consultation, the patient was admitted to the emergency department due to a clinical picture of three days of evolution consisting of abdominal pain in the epigastrium, which radiated to the back, and subjective fever. No emesis nor irritating urinary symptoms were observed, but choluria without acholia.

The patient presented the following surgical history: tubal ligation and right hip osteotomy. Upon physical examination, mild jaundice and pain were observed in the right hypochondrium and the epigastrium, without a positive Murphy sign nor signs of peritoneal irritation.

Paraclinical tests yielded the following results: leukocytes: 13 395, N% 87; Hb: 14; platelets: 344 000; amylase 34 U/L; total bilirubin: 4.66 mg/dL; direct bilirubin: 4.09 mg/dL; indirect bilirubin: 0.57 mg/dL; alkaline phosphatase: 1292 IU/L.

For its part, MRC showed generalized dilation of the intrahepatic and extrahepatic bile ducts, common bile duct with an average dilatation of 1 cm, with presence of hyperintense faceted images in T2. Gallbladder was not identified (Figure 1).

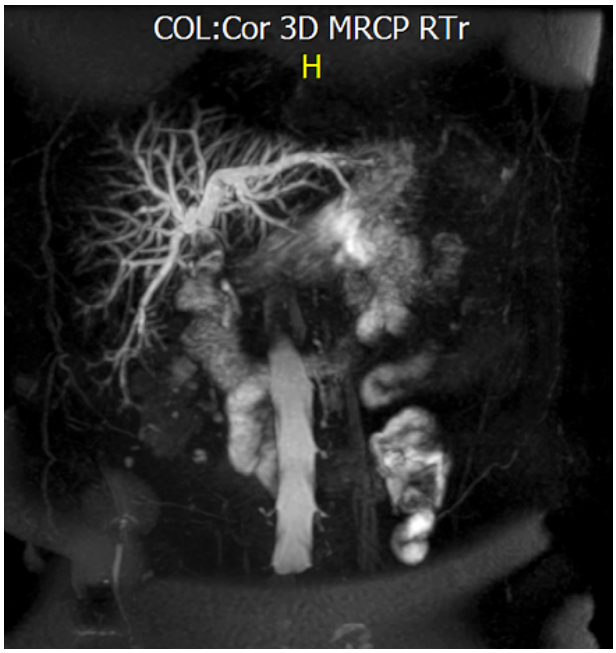

Figure 1. MRC - 3D reconstruction of the coronal section in the bile duct. Note the absence of gallbladder.
Source: Own elaboration based on the data obtained during the study.

Since several aspects indicated a high probability of choledocholithiasis, an endoscopic retrograde pancreatic cholangiopancreatography (ERCP) was performed, revealing a dilated, tortuous, and impacted intrahepatic bile duct, with no possibility of extraction, therefore, placing a stent was decided. Consequently, the case was taken to a surgical board to perform laparoscopic bile duct exploration.

During the procedure, the following findings were observed: absence of gallbladder, edematous biliary tract with duodenum adhesion, iatrogenic duodenum lesion of 5 mm (during the release of adhesions) and choledocholithiasis.

The procedure was performed by plastron adhesiolysis, primary duodenorrhaphy with absorbable suture by laparoscopy, and bile duct exploration by laparoscopy plus mechanical lithotripsy.

In the postoperative period, the patient presented a good evolution, with normalization of the liver profile and successful discharge after five days. Ambulatory follow-up showed an asymptomatic patient, without episodes of choluria, acholia or pain, so it was concluded that pain was completely resolved. Likewise, the patient showed tolerance to food; however, the patient did not attend subsequent follow-up sessions.

DISCUSSION

Agenesis of the gallbladder is a rare pathology that not many surgeons have the opportunity to treat. However, a surgeon must be prepared for any malformation and anatomical variant; actually, Skandalakis (10) points out vesicular triplication, which includes, agenesis of the gallbladder and how to act when it is suspected.

It should be noted that before 1950, ultrasound studies were not available, so cholelithiasis was diagnosed when oral cholecystography showed that the gallbladder was excluded (it was not observed in radiological images). When the patient underwent cholecystectomy, surgeons were surprised to notice the absence of the gallbladder.

Fortunately, today surgeons can plan procedures in an optimal manner, since ultrasound has a performance close to 80 or 90%, and is sufficient in most cases. However, in a case such as the one presented here, in which a scleroatrophic vesicle, chronic cholecystitis and acalculous cholecystitis were identified — the latter using tomographic studies—, agenesis of the gallbladder cannot be disregarded. In these cases, other specialized methods such as MRC (Figure 2), endoscopic ultrasonography, ERCP, among others, can be helpful. (9).

Before ultrasounds, Frey performed the largest amount of surgeries involving vesicular agenesis, establishing a triad for its diagnosis

under the following criteria: absence of inflammatory signs or fibrosis in the vesicular bed, complete dissection looking for an ectopic gallbladder (this dissection was extensive, including dissection of the left hypochondrium), and intraoperative cholangiography (6).

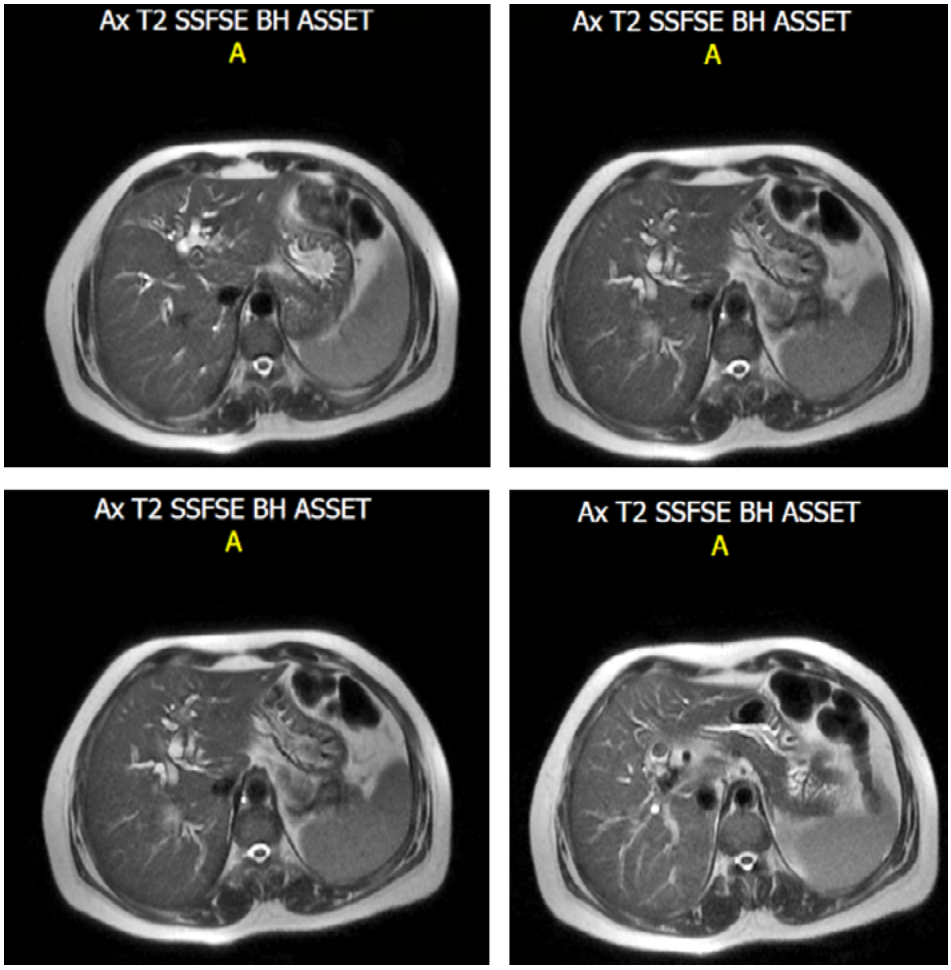

Figure 2. MRC sequence in T2 of the bile duct. Note the absence of gallbladder.
Source: Own elaboration based on the data obtained in the study.

However, this procedure was done before the laparoscopic era and the arrival of MRC. Therefore, today it is considered that, during the surgical act, fibrous remnant or scar should be sought; the vesicular ectopic position should be discarded (intrahepatic, adhered to the left lobe, falciform ligament, retroperitoneal or in the anterior abdominal wall), and, according to the ability or criterion of the surgeon, intraoperative cholangiography should be performed, although it is not mandatory, as stated by Frey (11).

However, despite the fact that postoperative ERCP/MRC should inevitably be performed, in this case, MRC should always be considered, since agenesis is an exotic pathology and the infrequent vesicular positions mentioned above can be overlooked (3 -5,7-14), thus avoiding, in the first instance, the risks of ERCP when there is no lithiasis in the

primary bile duct (choledocholithiasis) (Figure 3). However, in the presence of agenesis, ERCP has been reported as not effective due, in part, to dysfunction of the sphincter of Oddi, so many patients need surgery and lithotripsy, as happened in this case (15).

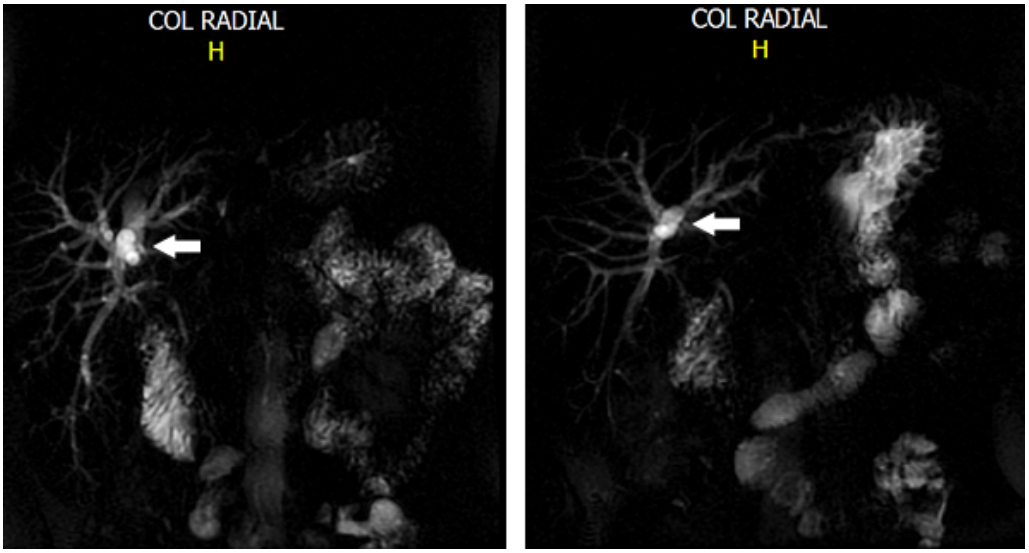

Figure 3. 3D reconstruction of the bile duct. Note the multiple hyperintense images, the major ones (yellow arrow) correspond to lithiasis of the bile duct.
Source: Own elaboration based on the data obtained in the study.

Finally, the safety vision of Strasberg (16) is recommended, since it is the most effective way to prevent bile duct injury as exposed in said reference.

Unfortunately, long-term follow-up was not possible, since the patient, as mentioned before, did not attend the subsequent controls, which is common in the Colombian context.

CONCLUSIONS

Although it is not possible to make generalizations based on the current evidence, other authors report two more cases in the region (17); therefore, the following steps are advisable when the intraoperative gallbladder cannot be identified:

- Looking for the ectopic gallbladder
- Looking for a scar in the liver bed that suggests agenesis
- Not manipulating excessively the bile duct
- Performing intraoperative cholangiography depending on the medical center, preference and experience of the surgeon
- Unfailingly performing a postoperative MRC

CONFLICT OF INTERESTS

None stated by the author.

FUNDING

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INFORMED CONSENT:

The patient signed an informed consent during hospitalization for the completion of this report.

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CONGENITAL SACCULAR CYST OF THE LARYNX. CASE REPORT AND LITERATURE REVIEW

Palabras clave: Quistes; Laringe; Ablación por catéter; Tratamiento de radiofrecuencia pulsada; Neonatos.
Keywords: Cysts; Larynx; Catheter Ablation; Pulsed Radiofrequency Treatment; Newborn.

Ricardo Enrique Guerra, MD
Department of Otolaryngology
- Hospital Infantil Universitario de San José -
- Fundación Universitaria de Ciencias de la Salud -
Sede Bogotá - Bogotá D.C. - Colombia

Mauricio Puerta, MD
Diana Patricia Anzola, MD
- Fundación Universitaria de Ciencias de la Salud -
Sede Bogotá - Bogotá D.C. - Colombia

Corresponding author:
Diana Patricia Anzola.
Fundación Universitaria de Ciencias
de la Salud - Sede Bogotá
Bogotá D.C. Colombia.
Correo electrónico: danzola@fucsalud.edu.co

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ABSTRACT

Introduction: Congenital saccular cyst is a rare but benign lesion, caused by a dilated laryngeal sac full of mucus that does not communicate with the laryngeal lumen. Its definitive treatment is surgical according to the literature.

Objective: To review the literature and report a case of congenital laryngeal saccular cyst, as well as its treatment by endoscopic approach and radiofrequency, which is most easily found in our country.

Materials and methods: Presentation of a case report and literature review in PubMed and Tripdatabase using the described keywords.

Results: This is a rare condition with an incidence of 1.82 cases per 100 000 live births. Diagnosis is achieved by laryngeal endoscopy, images or clinical review. The case reported here corresponds to a newborn patient with respiratory distress and stridor, who was diagnosed with laryngeal saccular cyst that was resected surgically by means of endoscopy and radiofrequency, with no subsequent recurrence.

Discussion: Understanding this disease is highly important to achieve proper diagnosis and provide treatment using the resources available in our country, such as radiofrequency.

Conclusions: Despite the lack of case reports, knowing the characteristics of congenital saccular cyst is necessary to indicate proper treatment based on the available resources. It is possible to perform endoscopic resection of this lesion if it is <3cm by means of radiofrequency, which is a safe and effective method.

RESUMEN

Introducción. El quiste sacular congénito es una lesión rara, pero benigna, dada por la dilatación del sáculo laríngeo que se llena de moco y que no se comunica con el lumen laríngeo. Según la literatura, su tratamiento definitivo es quirúrgico.

Objetivo. Realizar una revisión de la literatura y el reporte de un caso de quiste sacular laríngeo congénito y su tratamiento mediante abordaje endoscópico y uso de radiofrecuencia.

Materiales y métodos. Se realizó la presentación del caso clínico y se llevó a cabo una revisión de la literatura en las bases de datos de Pubmed y Tripdatabase usando palabras claves descritas.

Resultados. Esta patología es escasa, con una incidencia de 1.82 casos por 100 000 nacidos vivos. El diagnostico se realiza por medio de endoscopia laríngea, imágenes o revisión de la clínica. El caso expuesto es de una paciente recién nacida que presenta estridor y dificultad respiratoria y es diagnosticada con quiste sacular laríngeo, el cual es resecado de forma quirúrgica por medio de endoscopia con uso de radiofrecuencia. El procedimiento da como resultado la no reaparición del quiste.

Conclusiones. Pese a no existir muchos reportes de caso, hay que conocer las características del quiste sacular congénito para poder realizar el tratamiento adecuado con los recursos disponibles. Es posible realizar resección endoscópica de esta lesión si es <3cm por medio de radiofrecuencia, un método seguro y eficaz.

INTRODUCTION

Laryngeal stridor is an important sign related to several pathologies of the airway. When it occurs in neonates and is associated with respiratory distress, it must be treated urgently. One of its causes, although rare, is congenital laryngeal saccular cyst (1); however, differential diagnoses include more frequent pathologies such as laryngomalacia, tracheomalacia, subglottic stenosis, vocal cord paralysis, laryngocele, laryngeal membrane, among others (2).

Saccular cyst is a benign supraglottic lesion that is usually unilateral, does not have an opening to the laryngeal ventricle and is greater than 1cm (3). This lesion corresponds to 25% of all laryngeal cysts (4) and to 2-5% of all benign laryngeal lesions (5,6). Since this is a very rare pathology and there is no exact data on its incidence, although some authors state that it is 1.82 per 100 000 live births (7), while others state that it is less than 1 per 300 000 live births (8).

CASE PRESENTATION

Full-term newborn patient with adequate weight for gestational age, delivered via cesarean section and with polyhydramnios and APGAR 10/10. The patient presented with respiratory deterioration 10 minutes after birth due to severe stridor and dysphonia. Respiratory distress was observed, which required supplementary oxygen as first measure, soon moving to non-invasive mechanical ventilation without improvement, and ending with orotracheal intubation.

Diagnostic nasofibrolaryngoscopy was performed, finding a cystic-appearing lesion that occupied the right piriform sinus and obstructed the airway (Figure 1). Contrast computed tomography (CT) of the larynx with 3D reconstruction under sedation was requested to assess the extension of the lesion. A right cystic-appearing lesion (1.3x1.2x2.1cm) was observed, which compromised the hypopharynx, pyriform sinus, glottis and right supraglottic space, and caused loss of definition of laryngeal ventricle and right vocal structures with wall enhancement (Figure 2). Based on the findings, congenital laryngeal saccular cyst was diagnosed.

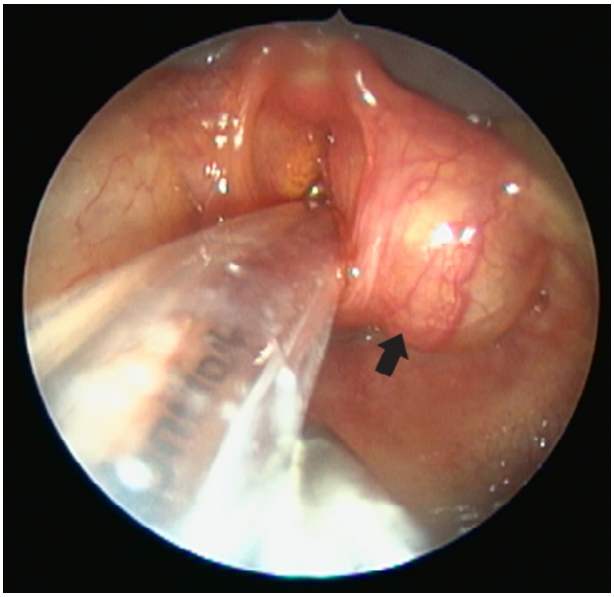

Figure 1. Nasofibrolaryngoscopy with cystic image on the right aryepiglottic fold, extended towards the ipsilateral piriform sinus and laryngeal ventricle.

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

Surgical approach was performed by means of microlaryngoscopy and endoscopic resection of the laryngeal lesion with radiofrequency (laryngeal electrode tip MC/AC/BC 403, 100-300V, 100-500 kHz). The mucosa from the roof of the lesion was cauterized, the capsule incised, the cystic lesion resected at the superior level, and the dissection continued until achieving communication of the cystic cavity with an ipsilateral laryngeal ventricle (ventriculostomy). Pre-surgical findings

included a cystic lesion that compromised the right aryepiglottic fold and extended to the posterior ipsilateral cricopharyngeal region — rejected by the right ventricular band until the laryngeal ventricle—, healthy vocal cords and free subglottis.

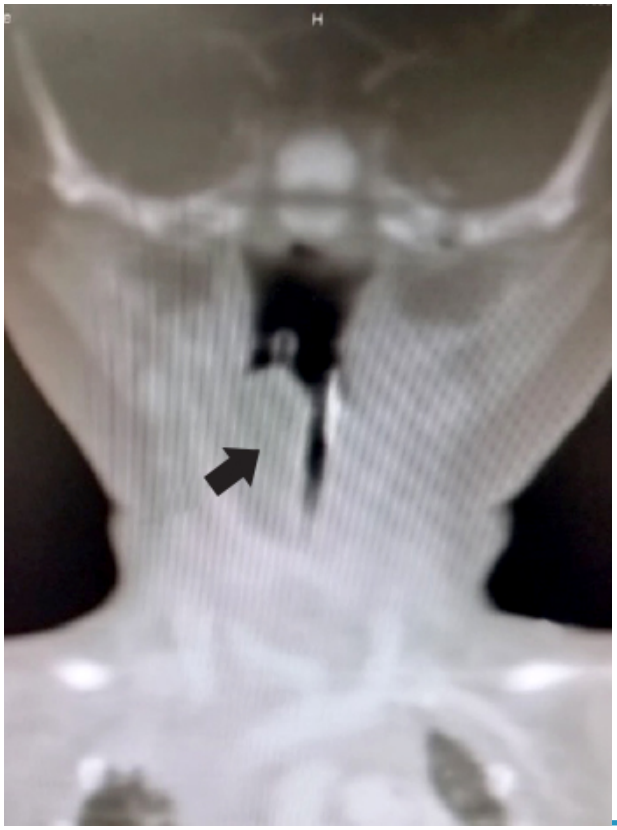

Figure 2. Contrast coronal CT scan of the neck showing a cystic lesion extending from the piriform sinus to the trachea with a significant decrease in the caliber of the airway.
Source: Own elaboration based on the data obtained in the study.

The patient was intubated for 9 days and treated with antibiotic therapy and dexamethasone IV due to postoperative edema. On day 10, adequate extubation tolerance was achieved. Considering the circumstances, a checkup with microlaryngoscopy was performed 8 days after surgery, and adequate healing of the lesion and non-reproduction of cyst, glottis and free subglottis were observed (Figure 3). A postoperative control was performed after a year using nasofibrolaryngoscopy without new surgical interventions, no recurrence of the cyst, and adequate epithelization and laryngeal development.

DISCUSSION

The saccular cyst consists of a dilation of the laryngeal ventricle filled with mucus that does not communicate with the laryngeal lumen (9). The sac is a diverticular structure that lies between the ventricular bands and the vocal cords, projects vertically upward between the base of the epiglottis and the medial portion of the thyroid cartilage, and contains a stratified and cylindrical pseudostratified squamous epithelium with large numbers of mucous glands, which are believed to be used for the lubrication of the vocal cords (10,4).

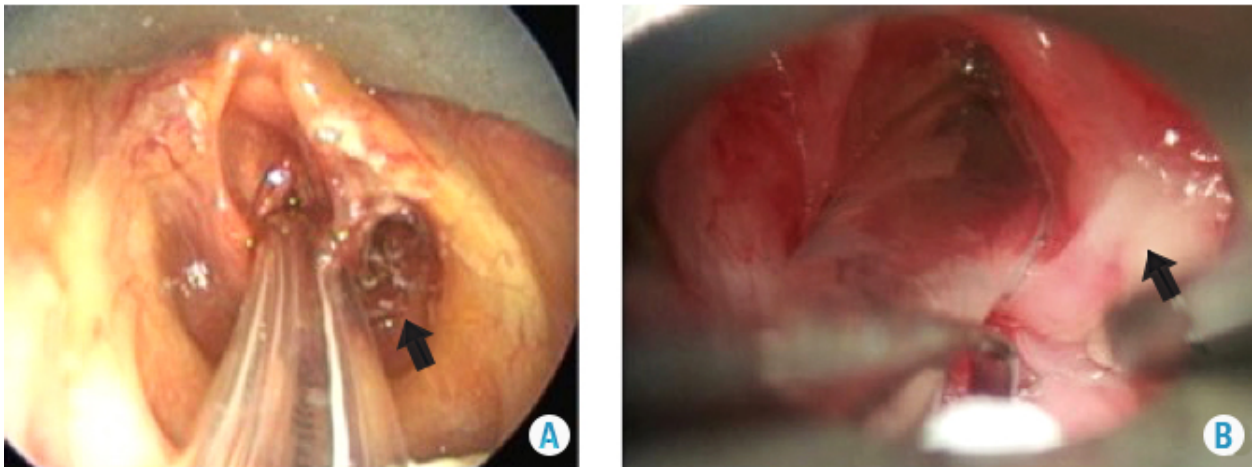

Figure 3. Microlaryngoscopy A) Immediate postoperative period after marsupialization and resection of saccular cyst. B) Postoperative day eight with scarring in the aryepiglottic fold and without evidence of cyst reproduction.
Source: Own elaboration based on the data obtained in the study.

The main symptom of this pathology is stridor at birth that can conceal laryngomalacia or congenital vocal cord paralysis, which appear with stridor in the first weeks of life (11). Other symptoms depend on the size of the lesion and its location; for example, dysphagia may occur if the cyst occupies the hypopharynx (12) or if there is progressive respiratory distress until total airway obstruction, cyanosis, apnea, hoarse cry and low-pitched cry, especially with changes of position or agitation. If the cyst is on the right side, the symptoms are accentuated in the contralateral ulnar position (4).

Two types of saccular cysts are described in the literature: anterior and lateral. Anterior cysts are characterized by a submucosal mass dependent on the false vocal cord that protrudes through the anterior portion of the ventricle, while lateral cysts, usually the most common, occupy the entire ventricular band and exit the pharynx through the mucosa of the aryepiglottic fold (13).

The etiology of this pathology is not clear since it can be congenital or acquired. The first is caused by obstruction or atresia of the sac, which, depending on its location, will make the cyst more extensive or not. This obstruction can be caused by the abnormal migration of the tissue from the fourth branchial arch generating the cystic formation (14) or by the isolation of the cells of the sac due to the abnormal migration of mesenchymal cells through persistent fetal vessels in the larynx. The second may be a consequence of sac obstruction secondary to neoplasia, trauma or inflammation with subsequent fibrosis (15).

Most of the literature found on congenital saccular cyst is based on case reports; in the most extensive study, data on 20 cases were collected in a period of 15 years (16), which makes the low incidence of this pathology evident. This study found that the incidence in the Hospital Infantil Universitario de San José in the last 5 years has been 1 for every 22 318 live births. Most of the cysts are lateral with an average size of <3cm, as the one described in this article (3,7).

Diagnosis can be made through imaging studies such as lateral radiography of soft tissues of the neck, where a sac full of mucus

is visualized in the supraglottic region; using CT or nuclear magnetic resonance is also possible. Several authors state that diagnostic confirmation is achieved through direct visualization of the lesion, either by direct or optical fiber laryngoscopy (17,15), as described here.

The treatment of this pathology is mainly surgical and includes the aspiration of the cystic content with needle and marsupialization, and even cyst excision via endoscopy or external approach (17,18). The first two are considered insufficient treatments because, in the first case, the mucosa glands of the cyst continue to secrete mucus, thus generating reappearance, and in the second, multiple procedures are required due to cyst recurrence that can easily generate laryngeal stenosis with subsequent tracheotomy (8,19). The excision of the cyst by microlaryngoscopy and external approach are the treatments with the highest success rates according to the literature. However, some authors, such as Ward *et al.* (12), recommend resection by means of an external approach as the only definitive and safe treatment for the non-formation of laryngeal stenosis, given the poor manipulation of the laryngeal mucosa.

There are case reports such as Khodaei *et al.* (17) and Danish *et al.* (5) which establish microlaryngoscopy with CO2 laser as the first line of management and the most used treatment. Lateral cervicotomy through the thyroid membrane is used for complex, recurrent or> 3cm cysts, since they can increase anesthesia time and pose a higher risk of superior laryngeal nerve injury (8,19,20). Another endoscopic approach that has shown lower morbidity is CO2 laser vestibulectomy, since it reduces surgical time, avoids vascular or superior laryngeal nerve damage and accelerates tissue recovery. (20).

The definitive management, as stated in the literature, is microlaryngoscopy, since most reports and case series use CO_2 laser for endoscopic resection (3,19,20). In Colombia, this laser is not available in all hospitals, so the procedure, in this case, was performed using radiofrequency. Of all the articles reviewed, only the series by Kumar *et al.* (16) used this resection method and demonstrated that it is safe and effective because it improves surgical precision and the healing time of the mucosa, minimizes bleeding in the surgical site, preserves the surrounding tissues, and only 1 of 6 cases require a second surgery (16). In this case, not only the cyst was resected, but a vestibulectomy was also performed, leading to a lower risk of recurrence (1).

Despite the low frequency of this pathology, it is important to know and be able to diagnose and treat it safely and effectively with the resources available in most hospitals in Colombia. This is useful to avoid significant compromise of the patient's airway.

CONCLUSIONS

It was possible to resect a laryngeal sacular cyst smaller than 3cm in a safe and effective manner through microlaryngoscopy and radiofrequency, despite the lack of case reports regarding the use of this method.

CONFLICT OF INTERESTS

None stated by the authors.

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SPONTANEOUS PNEUMOMEDIASTINUM. CASE REPORT

Palabras clave: Enfisema mediastiníco, Enfisema subcutáneo.
Keywords: Mediastinal Emphysema; Subcutaneous Emphysema.

Laura Marcela Velásquez Gaviria, MD • Andrés Fernando Rodríguez Gutiérrez, MD
Sebastián Felipe Sierra Umaña, MD • Andrés Garcés Arias, MD
Department of Internal Medicine – Faculty of Medicine –
Universidad Nacional de Colombia
Bogotá – Colombia

Laura Salazar Franco • Luis David Sáenz Pérez
Sebastián Salinas Mendoza • Cristian Alejandro Castillo Rodríguez
Medical Program – Faculty of Medicine –
Universidad Nacional de Colombia
Bogotá – Colombia

Diego Fernando López Donato, MD
Department of Radiology
Faculty of Medicine
Universidad Nacional de Colombia
Bogotá – Colombia

Luisa Fernanda Patiño Unibio, MD
Department de Internal Medicine
Faculty of Medicine
Pontificia Universidad Javeriana
Bogotá – Colombia

Corresponding author:
Sebastián Felipe Sierra Umaña
Universidad Nacional de Colombia
Bogotá – Colombia.
Email: sfsierrau@unal.edu.co

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ABSTRACT

Introduction: Spontaneous pneumomediastinum (SPM) is defined as the presence of air in the mediastinum. It is a rare entity considered benign and self-limiting, which mostly affects young adults. Its diagnosis is confirmed through clinical and radiological studies.

Case description: 21-year-old male patient with cough and greenish expectoration for four days, associated with dyspnea, chest pain, fever and bilateral supraclavicular subcutaneous emphysema. Chest X-ray suggested pneumomediastinum, which was confirmed by tomography. The patient was hospitalized for observation and treatment. After a positive evolution, he was discharged on the sixth day.

Discussion: SPM is a differential diagnosis in patients with chest pain and dyspnea. Its prevalence is lower than 0.01% and its mortality rate is low. It should be suspected in patients with chest pain and subcutaneous emphysema on physical examination. Between 70 and 90% of the cases can be identified by chest X-ray, while confirmation can be obtained through chest tomography. In most cases it does not require additional studies.

Conclusion: SPM is a little known cause of acute chest pain, and rarely considered as a differential diagnosis; it is self-limited and has a good prognosis.

INTRODUCTION

SPM is defined as the presence of air in the mediastinum without an apparent secondary cause (1). It is rare, benign and self-limiting, and affects mostly young adults with an average age of 25 years (2), ranging between 13 to 35 (3); a study by Cáceres *et al.* (4) reported a similar incidence between men and women. In 1944, Macklin *et al.* suggested that SPM originates after an alveolar rupture caused by increased intrathoracic pressure, with subsequent passage of air into the interstitium and bronchovascular tissues of the tracheobronchial tree (5).

The most frequent symptoms are chest pain, dysphagia, persistent cough and dyspnea, while risk factors include chronic obstructive pulmonary disease, asthma, and tobacco and illicit drugs use. In addition, precipitating factors such as nausea, vomiting, cough, upper respiratory tract infection and strenuous physical exercise have been observed (3). SPM cases have also been reported as complications of pneumonia by influenza A (H1N1) in children, mainly during the pandemic period of this infection in 2009 (6).

The goal of treatment is to control symptoms and may require observation. The length of hospital stay varies from a few hours to several days (2,4). This article presents a SPM case in a young adult.

CASE DESCRIPTION

21-year-old male patient from Garagoa (Boyacá), resident of Bogotá D.C. Colombia, an industrial automation student, mestizo, socioeconomic stratum 3, who presented a clinical picture of four days of evolution consisting of cough with greenish expectoration, dyspnea, chest pain, and unquantified fever. On physical examination he did not have respiratory distress and his vital signs were normal. Bilateral supraclavicular subcutaneous emphysema, decreased vesicular murmur and bilateral intermittent wheezing were identified; no other abnormal findings were observed. The patient had no relevant medical history.

Based on the clinical and epidemiological characteristics, an acute respiratory infection of viral origin was considered; in addition, due to the presence of subcutaneous emphysema and alterations in pulmonary auscultation, spontaneous pneumothorax was suspected. Leukocytosis with neutrophilia and mild oxygenation disorder was found in the requested paraclinical exams (Table 1), while left chest and left supraclavicular soft tissues were observed on the chest radiograph (Figure 1).

Table 1. Paraclínical exams.

	On admission	Control at 72 hours
Hematogram	Leukocytes 14670 cell/mm^3 Neutrophils 13670 cell/mm^3 Hemoglobin 17 g/dL Hematocrit 48% Platelets 257000 cells/mm^3	Leukocytes 12150 cell/mm^3 Neutrophils 8240 cell/mm^3 Hemoglobin 16.7 g/dL Hematocrit 47.7% Platelets 264000 cell/mm^3
Arterial blood gas	pH 7.43 PO2 55.8 mmHg FiO2 0.24 PCO2 35.6 mmHg PAFI 232.7 HCO3 23.3 mmol/L BE -0.4 mmol/L	pH 7.43 PO2 62.6 mmHg FiO2 0.21 PCO2 33.7 mmHg PAFI 297 HCO3 22.3 mmol/L BE - 0.6 mmol/L

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

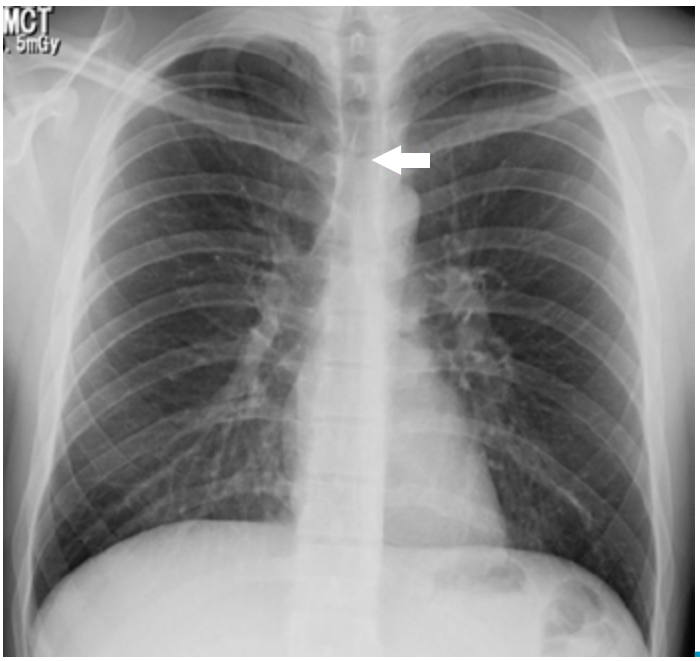

Figure 1. PA chest x-ray: pneumomediastinum, delimitation of anatomical structures allowing a neat visualization of its contours (arrow).
Source: Own elaboration based on the data obtained in the study.

Later, a chest tomography was performed, which showed air in the anterior, middle, posterior and superior mediastinum, reaching the lower neck (Figure 2). Due to the absence of risk factors related to secondary causes, SPM secondary to an acute respiratory infection of viral origin was diagnosed; the patient was maintained under observation, and treatment including oxygen through nasal cannula, respiratory therapy, analgesia and rest was indicated.

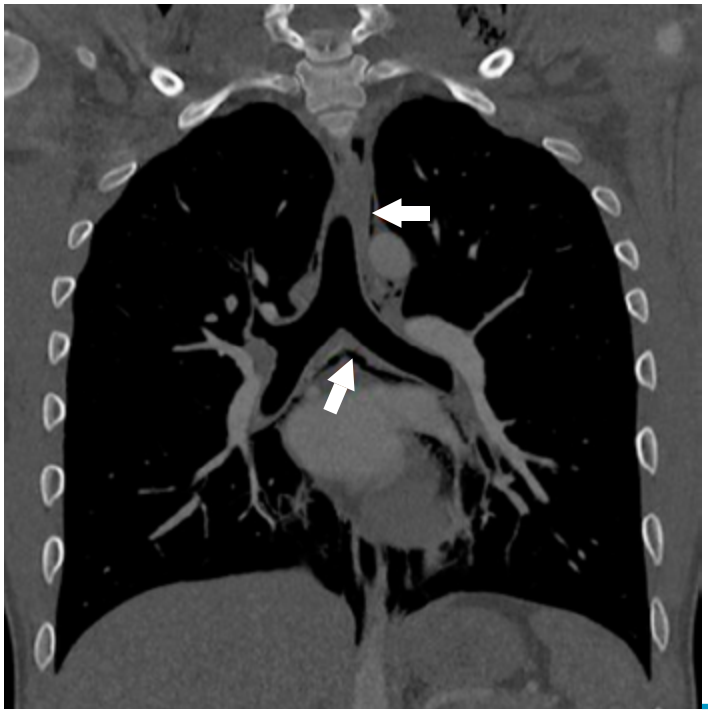

Figure 2. Chest tomography, coronal plane: pneumomediastinum, presence of infracarinal and paratracheal air (sepia arrow). Left supraclavicular subcutaneous emphysema is also observed.
Source: Own elaboration based on the data obtained in the study.

The patient improved during follow-up, period in which leukocytosis and oxygenation disorder were corrected (Table 1), and was discharged after six days of hospitalization. Outpatient radiographic monitoring was requested and he was given recommendations and warning signs. The patient did not present adverse drug reaction or other events during hospitalization.

DISCUSSION

Pneumomediastinum was first reported in 1819 by René Laennec while spontaneous pneumomediastinum was described in 1939 by Louis Hamman (7). Its incidence is less than 0.01% and has a recurrence rate of 1.6% per year (8,9). SPM is a differential diagnosis in patients with chest pain and dyspnea, and is believed to be caused by alveolar rupture due to increased intraalveolar pressure (1,10); therefore, its association with pneumothorax is frequent, being found in 32% of patients (11). In 44% of cases, patients have a history of congestive lung disease, such as asthma, chronic obstructive pulmonary disease, interstitial disease, pulmonary fibrosis, pneumonitis, among others (11).

The mean age at diagnosis is 25 years (11), similar to that of patients with spontaneous pneumothorax (9). In 34% to 49% of the cases, precipitating factors, such as in-

haled drug abuse, acute respiratory infection, vomiting, asthmatic crisis and intense exercise are observed (9,11).

The most common clinical manifestations include chest pain (68-78.1%), dyspnea (28.1-44%), sore throat (14.1-28%) and cervical pain (54.7%) (9,12). Furthermore, subcutaneous emphysema is the most frequent symptom in about 40 to 100% of patients (9,12,13); in contrast, Hamman's sign (systolic crackle heard with a stethoscope at the left sternal border) is found in only 20% of cases (14,15).

Its presentation is usually masked because of the low specificity of the symptoms and the lack of knowledge of this entity (16). The diagnosis is made based on clinical manifestations and radiological confirmation, in addition to searching for triggers (4,17). 79% of the patients are diagnosed in the emergency room, 11% in the critical care unit, 2% during hospitalization, and 8% in outpatient consultations (11).

Radiological studies of the thorax are important in the evaluation and exclusion of secondary causes (9), and are sufficient to confirm the diagnosis (18). Not all patients with pneumomediastinum require contrast radiographic imaging, which is reserved for patients who are suspected of having a tracheobronchial or esophageal injury, especially when vomiting, dysphagia, known gastrointestinal disease, trauma, fever, hemodynamic instability, pleural effusion or pneumoperitoneum are involved (19).

Around 70% to 90% of SPM cases can be identified by chest X-ray (20). The presence of mediastinal air creates an interface with the anatomical structures that allows to visualize its contours neatly. Radiological signs depend on the quantity and location of the air (21): when it surrounds the vascular structures, the ring sign and the tubular artery sign appear. The delimitation of the inner and outer wall of the bronchus is possible due to the presence of intra and extraluminal gas, generating a double wall sign. The continuous diaphragm sign is caused by air posterior to the pericardium.

Other radiological signs include subcutaneous emphysema, radiolucent lines in the upper mediastinum, pneumoprepericardium, "Naclerio V", extrapleural air sign and, thymic wing sign caused by the delimitation of the thymus in children (16,21). Chest tomography delimits the extension of the pneumomediastinum, and provides information about its etiology and differential diagnoses (21,22).

In most cases studies that look for secondary causes are unnecessary, since, in general, there are no alterations of the respiratory or digestive tracts. Advanced diagnostic procedures, restricting diet, administering antibiotics and prolonging hospitalization stay are not appropriate measures (19). SPM has a good prognosis and can be treated conservatively (18), which has shown good results in different studies (2,9,19,23). Such treatment consists of analgesia, rest, oxygen and bronchodilators (24).

In theory, oxygen supplementation is of great importance for treatment, regardless of the presence of an oxygenation disorder, since it increases the pressure of nitrogen diffusion in the interstitium and promotes the absorption of free air (16) accelerating the resolution time.

The mean time of hospitalization is 4.6 days (9) and its management in a critical care unit is unnecessary unless required or in cases in which esophageal rupture is highly suspected (19). Once the patient is discharged, radiological follow-up can be performed until full resolution (16).

The case described here corresponds to a patient, whose epidemiological, clinical

and radiological characteristics are the most frequently reported in the literature. This is a typical case that contributes to the diagnostic approach in young patients who present chest pain on arrival to the emergency room. It is important to mention that this case had several limitations, including the lack of microbiological isolation of the germ responsible for the acute respiratory infection, radiological control, and information on outpatient follow-up to objectify the resolution of pneumomediastinum. However, this report is important because it illustrates a radiologically confirmed clinical case of a rare disease causing chest pain.

CONCLUSION

SPM is a rare entity that requires high clinical suspicion for both diagnosis and radiological confirmation. Its treatment is symptomatic and has a good prognosis. SPM should be considered as a differential diagnosis in patients with chest pain.

CONFLICT OF INTEREST

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Hospital Universitario Nacional de Colombia. Bogotá, Colombia.

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ASCENDING AORTIC DISEASE IN A PATIENT WITH MARFAN SYNDROME

Palabras clave: Aneurisma de la Aorta; Cirugía torácica; Aorta Torácica; Disección aórtica aguda, Marfan.
Keywords: Aortic Aneurysm; Thoracic surgery; Thoracic aorta; Acute Aortic Dissection, Marfan.

Edison Ricardo Espinoza Saquicela
General Surgeon
Cardiothoracic Surgery resident
Instituto Nacional de Cardiología Ignacio Chávez
Universidad Nacional Autónoma de México
Mexico City – México

Stefanía del Cisne Serrano Olmedo
General Surgeon
Urology resident. Hospital Juárez de México
Universidad Nacional Autónoma de México
Mexico City - México

Corresponding author:
Edison Ricardo Espinoza Saquicela
Instituto Nacional de Cardiología Ignacio Chávez
Universidad Nacional Autónoma de México
Ciudad de México. México
Email: enzo35467a@hotmail.com

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ABSTRACT

Introduction. Acute thoracic aortic dissection is caused by a tear in the intimal lining of the aorta, and is a symptom of acute aortic syndrome. The dissection allows the blood to pass through the rupture and separates the tunica intima from the tunica media or the tunica adventitia, creating a false intravascular light. Early diagnosis directly affects the chances of survival, since it is a medical emergency that can lead to death, even with optimal treatment.

Case description. The following report presents the case of a 26-year-old man with a history of Marfan syndrome, retrosternal lancinating pain, nausea, vomiting, and medium effort dyspnea that evolved to orthopnea, perioral cyanosis, murmur of aortic insufficiency and mitral systolic murmur. Complementary studies (chest x-ray, electrocardiogram, angiography, tomography, and echocardiogram) were performed, obtaining a diagnosis of Stanford type A ascending aortic dissection. Surgical treatment was indicated to replace the aortic root using a composite prosthesis and Bentall and De Bono coronary reconstruction. During the procedure, right coronary button destructuration occurred, so it was necessary to perform a venous bypass with a left internal saphenous venous hemoduct. Weaning extracorporeal circulation was achieved and then low expenditure of refractory character (despite vasopressor support at maximum dose), refractory ventricular fibrillation and asystole were observed. The patient did not recover and died as a consequence of acute transoperative myocardial infarction.

Conclusion. The treatment for ascending aortic dissection remains a therapeutic challenge. Timely diagnosis is directly related to life expectancy in patients who suffer from this condition, hence the importance of proper diagnosis and management.

RESUMEN

Introducción. La incidencia de síndrome de Marfan que ha sido reportada a nivel mundial es de 1 en 5000 casos, de los cuales aproximadamente el 80% o más desarrollan complicaciones cardiovasculares. La disección aórtica torácica aguda requiere una rotura en la íntima de la aorta, que forma parte del síndrome aórtico agudo. En la disección, la sangre pasa a través de la rotura y separa la íntima de la media o la adventicia, lo que crea una falsa luz intravascular. Un diagnóstico temprano incide directamente en las posibilidades de supervivencia, pues se trata de una emergencia médica que puede llevar a la muerte, incluso con un tratamiento óptimo.

Descripción del caso. Se presenta el caso de un hombre de 26 años con antecedente de síndrome de Marfan, dolor retro esternal lancinante, náusea, vómito, disnea de medianos esfuerzos que evolucionó a ortopnea, cianosis peribucal, soplo de insuficiencia aórtica y soplo sistólico mitral. Se realizaron estudios complementarios (radiografía de tórax, electrocardiograma, angiografía, tomografía, ecocardiograma) y se le diagnosticó disección de aorta ascendente tipo A Stanford, por lo que se decidió iniciar tratamiento quirúrgico mediante reemplazo de la raíz aórtica mediante prótesis compuesta y reconstrucción coronaria tipo Bentall y de Bono. En el procedimiento se presentó desestructuración de botón coronario derecho, por lo que fue necesario realizar puente venoso con hemoducto venoso de safena interno izquierdo. Se logró destete de circulación extra corpórea y luego se observó bajo gasto de carácter refractario (pese a soporte vasopresor

a dosis máxima), fibrilación ventricular refractaria y asistolia; el paciente no mostró recuperación y falleció como consecuencia de un infarto agudo de miocardio transoperatorio.

Conclusión. El tratamiento de la disección aórtica de aorta ascendente sigue siendo un desafío terapéutico. El diagnóstico oportuno tiene relación directa con la esperanza de vida de quienes lo padecen, de ahí su importancia diagnóstica y su manejo.

INTRODUCTION

Marfan syndrome is a rare connective tissue disorder that equally affects men and women (autosomal dominant), with an approximate incidence rate of 1 in 5 000 people. It is characterized by an alteration in the production of fibrillin due to a mutation in the FBN1 gene of chromosome 15, which weakens the connective tissue of the body, whether tendinous, ligamentous, cartilaginous, vascular or heart valves. (1). It is worth noting that not all patients with this syndrome develop the same symptoms or severity (allelic heterogeneity) (2). Since the aorta wall is weak, a dilatation of the aorta occurs leading to an aortic aneurysm. In addition, if there is a rupture in the vascular wall, a double lumen may be formed, with the consequent passage of blood, which is called aortic dissection.

Acute thoracic aortic dissection is the result of a tear in the wall of the aorta that allows blood flow between its layers separating them, which leads to the appearance of a double light within the same vessel, causing the blood flow through a false light to exert an occlusive effect on the light itself and condition the flow to the vessels that derive directly from the aorta. The importance of early diagnosis directly affects the chances of survival, since it is a medical emergency that can lead to death, even with optimal treatment.

The Stanford classification is used to better understand aortic dissection. It is divided into types A and B. Group A includes the dissections that involve the ascending aorta, which is why they are considered to pose higher risk because the dissection and false light that is generated can rapidly affect the coronary ostium and compromise the blood flow to the myocardium, leading to a massive and fulminating infarction, hence the importance of timely diagnosis and appropriate surgical treatment.

In many cases, if the aortic root is dilated, and depending on the degree of the dilation, the apparatus of the aortic valve is affected, which implies a backward flow of blood to the ventricles with the consequent decrease in the volume of ventricular ejection. To compensate this anomaly, the cavities of the heart increase their size, in other words, a cardiac dilation occurs.

The estimated mortality rate for type A aortic dissection is directly related to the time of evolution, being of 1% per hour within the first 24 hours, 29% at 48 hours, 44% after a week, and reaching 50% at two weeks. Therefore, emergency surgery is the best option.

Despite the advances for early diagnosis and the innovations in surgical management, type A aortic dissection of the ascending aorta has a high morbidity and mortality both in the short term and in the long term: at the hospital level, it ranges between 15% and 35%, with a 5-year survival rate in 65-75% of cases.

Type B dissections involve the aorta posterior to the left subclavian artery, with no compromise of the ascending aorta. Usually, its management is not urgent, although it is equally

important since relevant organs may also be affected, in which case other therapeutic options should be analyzed.

CASE PRESENTATION

26-year-old male, mestizo, single patient from Guayaquil (Ecuador), with a basic level of education, a history of Marfan syndrome and hypertension in treatment, who attended consultation reporting a clinical picture of 48 hours of evolution characterized by intense lancinating retrosternal pain and medium effort dyspnea that evolved to orthopnea. The vital signs on admission obtained after physical examination were: BP: 120/50 mmHg, HR: 108/min, RR: 22/min, oxygen saturation: 95%, temperature: 35.4°C, and height: 180 cm. He presented with disproportionate upper limbs in relation to thoracoabdominal structure, pale skin, aranodactyly, perioral cyanosis, and piriform thorax with sternal depression. Auscultation revealed systolic and diastolic murmur and mitral systolic murmur.

Complementary diagnostic exams

1. Chest x-ray: cardiomegaly, mediastinal widening at the expense of ascending aorta (Figure 1a).
2. Electrocardiogram: sinus tachycardia, left ventricular hypertrophy with signs of diastolic overload.
3. Echocardiogram: significant aortic valve regurgitation with severe dilatation of the valve ring.
4. Computed tomography angiogram of the chest: aortic root of 90.2 mm, ascending aorta of 80.8 mm, double lumen observed at aortic root level correlating with aortic dissection (intimal flap) with visible leakage of contrast material towards false lumen. Normal mitral, tricuspid and pulmonary valves; insufficient tricuspid aortic valve and increased pulmonary systolic pressure (60 mmHg) (Figures 1b and 2).

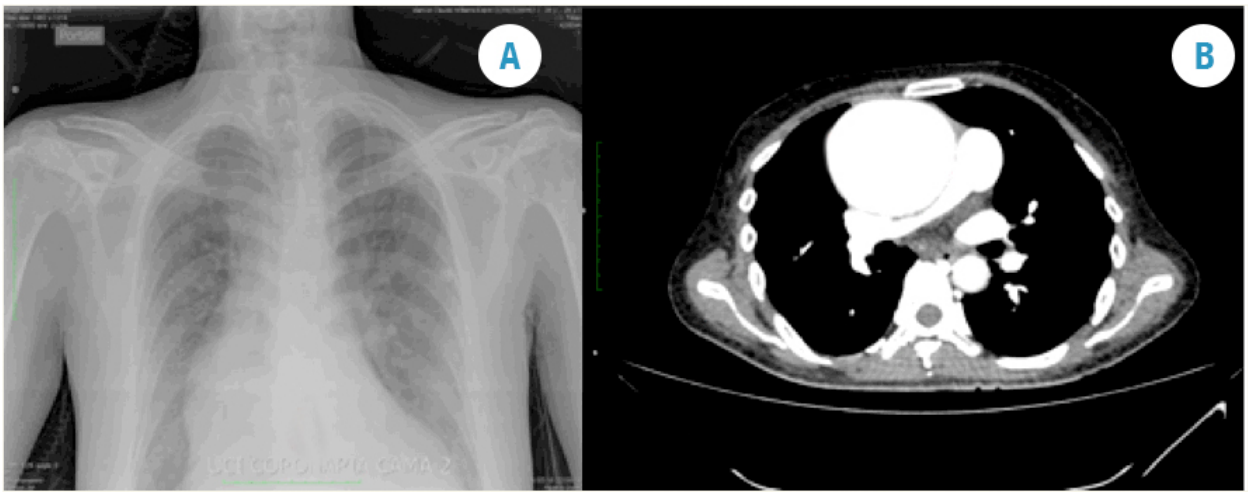

Fig 1A. Posteroanterior chest radiograph. Grade 4 cardiomegaly and mediastinal widening at the expense of ascending aorta.
Fig 1B. Computed tomography angiography: sagittal section. Aortic root of 90.2 mm, ascending aorta of 80.8 mm, insufficient tricuspid aortic valve and increased pulmonary systolic pressure (60 mmHg).
Source: Own elaboration based on the data obtained in the study.

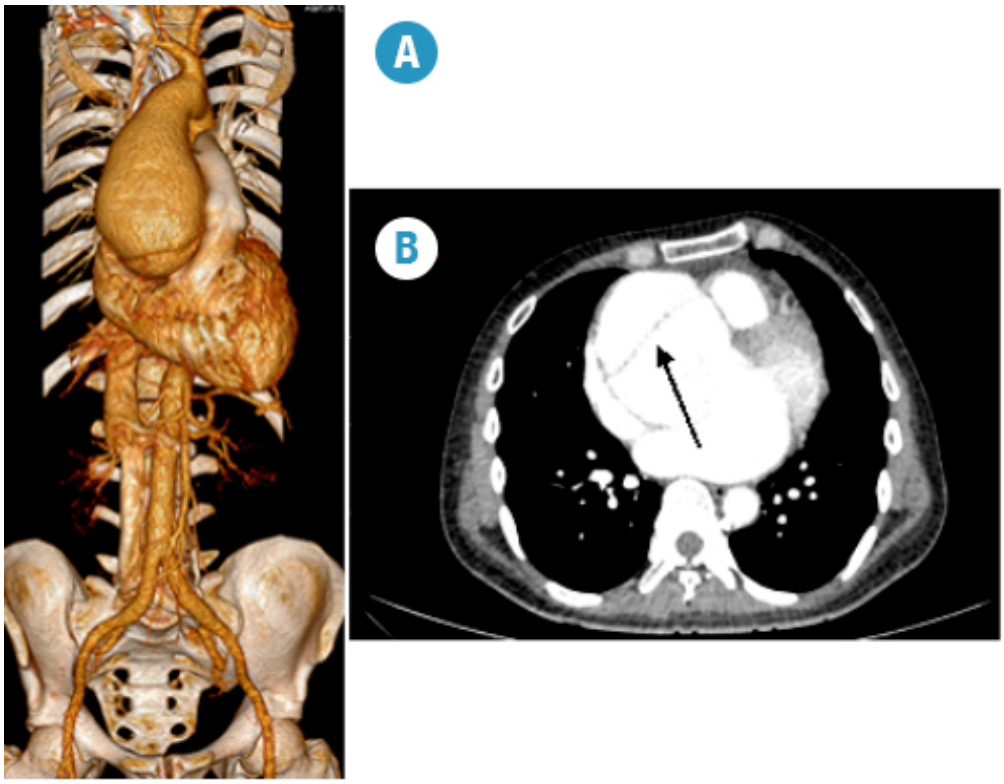

Fig 2A. 3D reconstruction of computed tomography angiography of the chest. Dilation of the ascending aorta. Aortic root 90.2 mm, anteroposterior diameter of ascending aorta of 80.8 mm.
Fig 2B. Arrow. Intimal flap.
Source: Own elaboration based on the data obtained in the study.

Evolution

One of the most important aspects is the length of hospital stay, since this is directly related to morbidity and mortality. In this case, approximately 48 hours passed between the completion of the complementary examinations, the confirmation of Stanford type A ascending aortic dissection as definitive diagnosis, and the decision to start surgical treatment through Bentall and De Bono surgery, which represented an increase of about 29% in the mortality rate.

The clinical management of the patient was holistic, and one of the parameters actively treated was blood pressure through carvedilol 6.25 mg VOc/12h, which kept the values within normality ranges.

Surgical procedure

Once the patient was under general anesthesia and strict asepsis, proximal and distal control of the right axillary artery was performed by cannulation for extracorporeal circulation (ECC), followed by medial stereotomy, exposing the pericardial sac through an inverted T incision, which exposed the ascending aortic aneurysm from the aortic annulus, with a diameter greater than 100 mm that deformed the base of the heart and extended to the distal third of the ascending aorta. Cardiomegaly IV/IV was found in both right and left cavities, along with global dilation of the ventricles and intraoperative pulmonary artery pressure over 60 mmHg according to Swan-Ganz catheter measurements.

The extracorporeal circulation entrance was then isolated by aortic clamping and the dissected aortic wall was opened longitudinally, which allowed to identify false and true lumen. Then, through a partial resection of the dissection focus, the distal end of the aorta was verified and the false lumen was sealed with BioGlue 1. Resection of the tricuspid aortic valve was continued, isolating left and right coronary buttons and implanting a valved tube sj # 27. Once the normal function of the prosthetic discs was confirmed, the left and right coronary button reimplant was placed and its permeability was checked. Immediately, distal anastomosis was performed from the valved tube to the distal ascending aorta. Finally, a prosthetic neo-ostium venous bypass implant was placed since the right coronary button was unstructured with suture dehiscence, which caused an acute intraoperative myocardial infarction.

Upon removing ECC, acute right ventricular distension and sustained ventricular fibrillation were observed, which required defibrillation (20-30 joules) together with antiarrhythmic therapy. Finally, arrhythmia was controlled. The patient presented three episodes of ventricular fibrillation which responded to electrical cardioversion.

ECC removal was attempted on three occasions, but low expenditure was observed despite maximum inotropic support, then the patient suffered refractory ventricular fibrillation and later asystole despite an epicardial pacemaker and died. The aortic clamping time was 150min and ECC time, 305 min (Figures 3, 4 and 5).

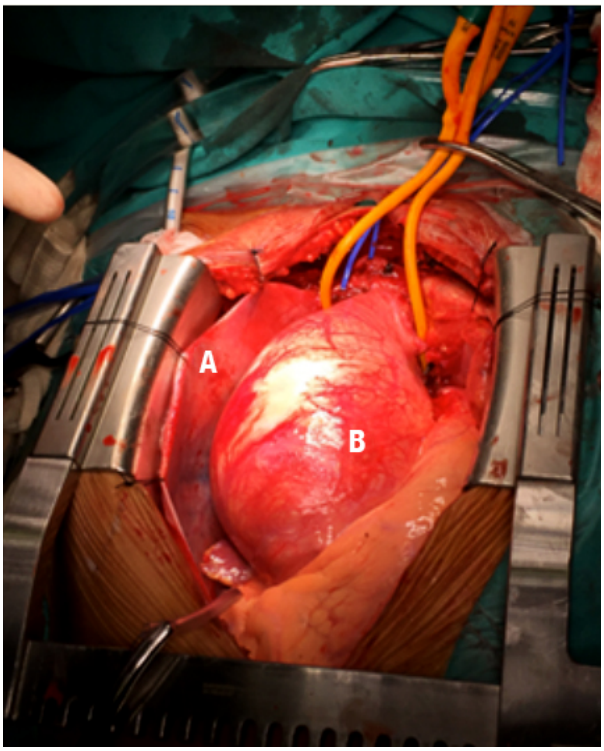

Fig 3. Surgery. Exposure by median sternotomy.
A. Pericardial sac. B. Aneurysmal ascending aorta.
Source: Own elaboration based on the data obtained in the study.

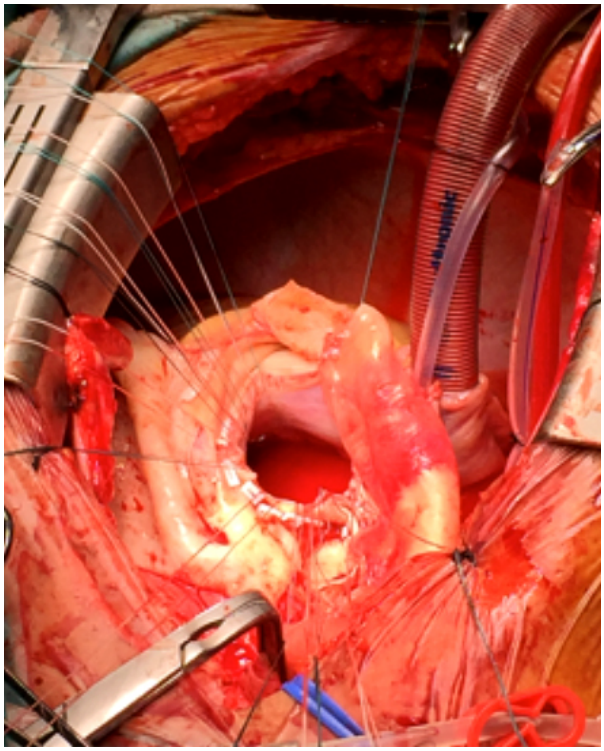

Fig 4. Exposure of the aortic ring with appropriate fixation points for subsequent placement of prosthetic material.
Source: Own elaboration based on the data obtained in the study.

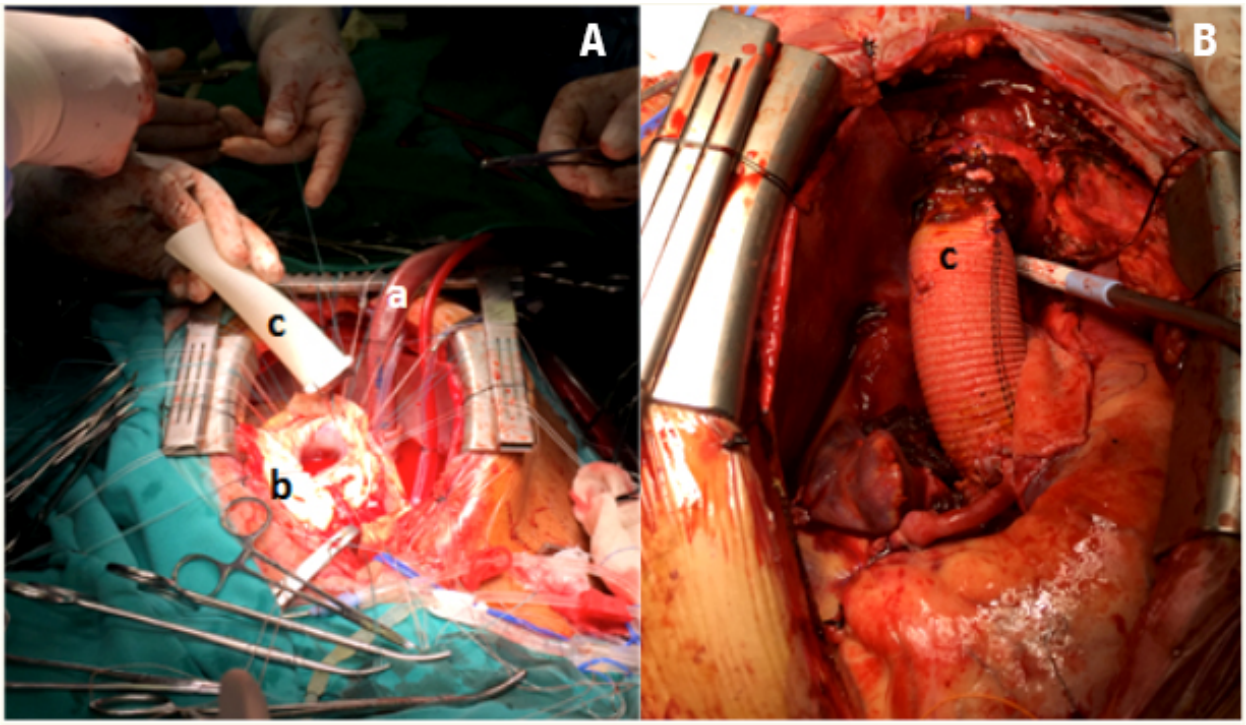

Fig 5A Exposure of the aortic ring with the appropriate fixation points after prosthesis placement.
Fig 5b. Replacement of the aortic root using composite prosthesis and Bentall coronary reconstruction. a: extracorporeal circulation system. b: aortic ring with corresponding fixation points. c: prosthetic material.
Source: Own elaboration based on the data obtained in the study.

DISCUSSION

The Marfan syndrome is an autosomal dominant pathology with a relatively low frequency, which tends to cardiovascular complications in most cases. The evolution of Marfan syndrome leads to an aneurysmal dilation of the aorta, which may be added to a rupture of its wall with the subsequent development of aortic dissection. In this case, a type A dissection was diagnosed based on the Stanford classification, the most frequently used nowadays, or DeBakey type II (3).

The ascending aortic aneurysms have a relatively low prevalence, as well as an important variability that can be accompanied by an aortic dissection or rupture of the aorta, increasing its mortality rate to almost 100%. The complications observed in the patients who suffer from this condition are closely related to the diameter of the aorta, its growth rate and the time of evolution. In vessels with a diameter greater than 5 cm, the risk of rupture and dissection increases by up to 45% (4). The symptomatology is variable and depends on the type of aneurysm, as well as on its location and relationship with neighboring structures. An aortic aneurysm can cause fatigue, hoarse or strained voice and pain in the left shoulder. If it is accompanied by aortic dissection, it can cause intense lancinating pain in the anterior chest, which usually radiates towards the back in the interscapular region.

Aortic dissection is characterized by the creation of a false lumen in the middle layer of the aortic wall. It is classified according to

the presence and location of the first tears, as well as the retrograde or anterograde extension of the dissection (5).

If coronary vessels are compromised since birth, associated acute coronary syndrome symptoms are added, which constitutes an emergency and modifies both the management and the necessary times.

Opting for surgical treatment depends on several factors and choice is based on the anatomical conditions of the aorta, the underlying disease, the risk of anticoagulation, the age of the patient, the presence of an active infection, among others. Several surgical techniques have been developed that reflect the evolution in the management of the ascending aortic aneurysms; each has its own advantages, limitations and risks. The replacement of the aortic root and the ascending aorta with valved tubular grafts, known as the Bentall procedure, along with annuloaortic ectasia is the most appropriate choice of the treatment for this condition. (6).

The literature reports that the Bentall procedure has a low morbidity and a mortality of 13%, being septic shock and ventricular fibrillation the most common causes of death (7). These figures vary considerably depending on the moment when surgery is performed: if surgery is scheduled, mortality rates range between 1.7% and 17.1%, but if surgery is the consequence of an emergency, the values increase to between 23% and 50% (7). The survival rate at 5 years ranges between 73 and 92%, while survival at 10 years is between 60 and 73% (8). However, surgical mortality can vary greatly depending on the hospital center, the experience of the medical team, the available resources and the heterogeneity of the patients (9). It is worth noting that dissection distal to the operated segment persists in 60% to 75% of the patients who undergo a dissection of the ascending aorta (10).

Due to its comorbidity rates and its degree of evolution, Bentall procedure is the most favorable choice for this type of patients. In this case, the procedure consisted in performing a prosthetic replacement that compensated aortic valve deficiency as a result of the large dilatation of the aortic annulus and subsequent replacement of the damaged aortic wall. Finally, the aortic buttons were reimplanted to the prosthesis.

However, there are other surgical options. For example, David's technique, developed by the Canadian surgeon Tirone David in 1992, achieves a greater stabilization of all the components of the root of the aorta, which requires the proper function of the aortic valve (6). In this case, the patient had a large dilatation of the aortic annulus that made this procedure difficult to use.

It is important to consider the need for developing new therapeutic options for this type of patients in order to improve their survival rates and quality of life.

CONCLUSION

The case reported here demonstrates that the treatment of patients with Marfan syndrome should be done from a multidisciplinary and joint approach, where timely diagnosis together with follow-up by specialized services and a qualified team allow a timely action and, thus, avoid that the patients with this condition look for care when it is already at a late stage.

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INFORMED CONSENT

The authors met the regulations on of anonymity of the case study.

CONFLICT OF INTEREST

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APENDICULAR CYSTIC DILATATION. CASE REPORT

Palabras clave: Apendicitis; Mucocele; Neoplasias del apéndice.
Keywords: Appendicitis; Mucocele; Appendix neoplasms.

Irlanda Moyota, MD • Lúver Macías, MD
Raúl León, MD • David Yépez, MD
Hospital Luis Vernaza
Department of General Surgery
Guayaquil – Ecuador

Corresponding author:
Irlanda Moyota Paguay.
Loja No. 700 y Escobedo,
Postal code EC090103.
Guayaquil, Ecuador.
Email: iris84mo@gmail.com

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RESUMEN

Introducción. El mucocele es una dilatación del apéndice vermiforme que se caracteriza por su contenido mucoide viscoso. La incidencia de esta entidad es baja —entre el 0.2-0.3% y el 0.7% del total de las apendicectomías— por lo que se considera un caso raro, el cual afecta más a mujeres que a hombres, con una relación 4:1. Con frecuencia, el diagnóstico es incidental y su manejo es quirúrgico.

Caso clínico. Paciente de 69 años de edad, de sexo masculino, que acude a emergencias debido a un cuadro clínico de dolor abdominal de 5 días de evolución en la fosa iliaca derecha. El dolor se acompaña de anorexia, náuseas y alza térmica no cuantificada. El examen físico revela signos de Mc Burney y Blumberg positivos e irritación peritoneal, por lo que se realiza apendicectomía más rafia de base apendicular. Entre los hallazgos intraoperatorios se observó tumoración apendicular con base ancha y de paredes gruesas, y contenido mucoide. El estudio histopatológico mostró una estructura de apéndice cecal con necrosis coagulativa en mucosa y pared, así como mucosa con depósito focal de material mucoide. El paciente fue dado de alta a los 8 días sin complicaciones.

Conclusión: No existen estudios específicos que lleven a predecir el mucocele apendicular, ni estudios que alerten sobre la futura aparición del mismo; no obstante, conocer sus características es útil para sospecharlo en casos con cuadros similares.

Debido a sus complicaciones, es necesario considerar controles radiológicos con un menor intervalo de tiempo e, incluso, un tratamiento quirúrgico precoz, con el objetivo de evitar complicaciones tales como obstrucción intestinal, hemorragia o pseudomixoma peritoneal.

ABSTRACT

Introduction. Mucocele is a dilatation of the vermiform appendix characterized by viscous mucoid material secretion. Its incidence is low —0.2-0.3% to 0.7% of the total of the appendectomies—, therefore, it is considered as a rare entity, which affects mainly women with a ratio of 4:1. Diagnosis is often incidental, and its management is surgical based on histology.

Clinical case. 69-year-old male patient who presented with abdominal pain of 5 days of evolution in the right iliac fossa, accompanied by anorexia, nausea and unquantified fever. Physical examination revealed positive Mc Burney's and Blumberg's signs, indicating peritonism. Appendectomy and appendiceal raffia were performed using the Parker-Kerr technique. Intraoperative findings included an appendicular tumor with a thick base and mucoid content. The histopathological study showed a cecal appendix structure with coagulative necrosis of the mucosa and the wall, as well as mucosa with focal deposit of mucoid material. The patient was discharged after 8 days without further complications.

Conclusion: Studies on appendicular mucocele are scarce, and due to its complications, radiological controls at a shorter time interval, and even early surgical treatment, are necessary to avoid complications such as intestinal obstruction, peritoneal bleeding or pseudomyxoma.

INTRODUCTION

Carl Freiherr von Rokitansky, Austrian anatomo-pathologist, first described appendiceal mucocele in 1842 (1). It is defined as a cystic dilatation of the vermiform appendix, whose causes include benign and/or malignant processes. It has a low incidence estimated at around 0.3% of appendectomies (2,3). From a histological perspective, it can be classified into four types: simple mucocele (accumulation of mucus in the appendicular cavity with obstruction), focal or diffuse hyperplasia of the mucosa, mucinous cystadenoma, and mucinous cystadenocarcinoma; when a spontaneous perforation of the appendix occurs, pseudomyxoma peritonei can be observed (4).

The clinical picture is not necessarily characteristic, since it can be asymptomatic and, in some cases, it is incidentally discovered through radiological-endoscopic studies or during surgical procedures. Half of the cases present pain in the right iliac fossa (5). This can be a benign or malignant process, so each case must be individualized to know its nature. Complications are rare and include intestinal obstruction or digestive bleeding, being peritoneal pseudomyxoma the most severe form, which occurs when mucinous material disseminates in the peritoneum.

The preoperative diagnosis is essential for planning the surgery and includes imaging techniques such as ultrasound and contrasted tomography (CT) of the abdomen and pelvis, which allow a better characterization of the tumor. An ultrasound can confirm the cystic aspect, while abdominal and pelvic CT scan with contrast defines the wall, the presence of calcifications and the density of the content. Other tests available, although less useful for diagnosis, are barium enema and colonoscopy (6,7).

The treatment indicated for appendiceal mucocele is the surgical removal of the appendix, even if the affected mucocele compromises the terminal ileum or the cecum. Cystadenocarcinoma cases with mesenteric involvement or an adjacent organ require hemicolectomy (8).

The objective of this work is to report a case of this rare pathology, discuss its particular characteristics and present a brief literature review.

CLINICAL CASE

69-year-old mestizo, male patient, without a relevant medical history, who attended the emergency room with a clinical picture of 5 days of evolution characterized by colicky abdominal pain of slight intensity, which progressed to moderate, at the right iliac fossa level. The pain was accompanied by nausea, malaise and unquantified fever.

The patient reported that he was unable to pass flatus and was constipated, which led him to self-medicate with oral analgesics. After realizing that the picture did not improve, he decided to visit the emergency room of the Luis Vernaza Hospital. Physical examination showed the following vital signs: blood pressure 140/80 mmHg, heart rate 103 bpm, SpO2 98%, temperature 37.5°C, conscious state, orientation in space, time, and person, afebrile and semimoist mucous membranes. The abdomen was soft, depressible, and painful in the right iliac fossa, with positive McBurney's point and Blumberg's sign. The results of the laboratory tests were: leukocytes 10,440/ul, segmented of 76.1%, and CRP: 249.97 mg /L.

An abdominal ultrasonography was performed, which showed a dilatation of the cecal appendix of 93.9mm x 35.9mm, thickening of the wall and laminar fluid in the right iliac fossa (Figure 1).

An exploratory laparotomy was decided with conventional appendectomy plus raffia with appendicular base, similar to the Parker-Kerr technique. Findings included appendiceal tumors with a wide base of thick walls and mucoid content. The samples taken were sent to pathology (Figure 2).

The macroscopic study revealed a cecal appendix of 8x6x5cm, appendiceal space reaching 4.5cm in diameter, and serous surface completely occupied by a whitish plate with fibrinous appearance. The parietal bone thickness ranged between 0.2 and 1 cm (Figure 3).

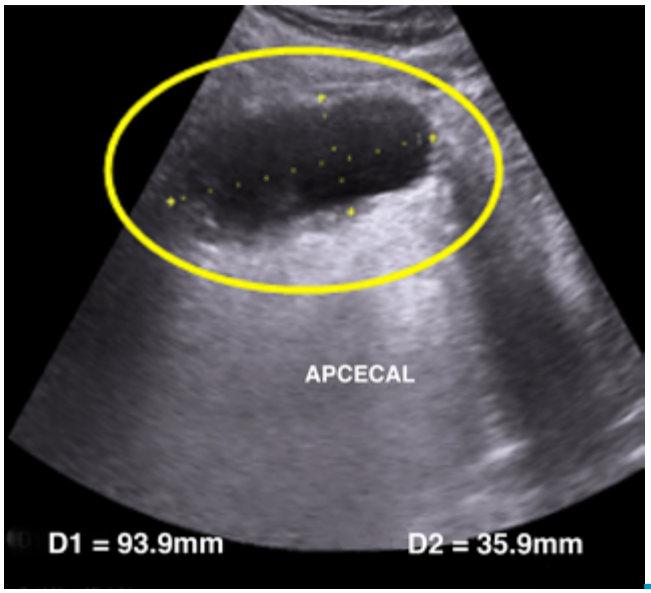

Figure 1. Abdominal ultrasound: dilatation of the cecal appendix (circle).
Source: Own elaboration based on the data obtained in the study.

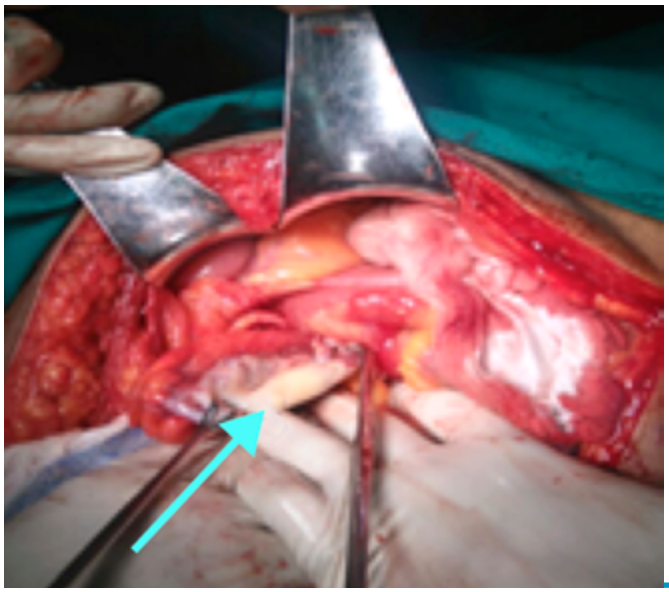

Figure 2. Sample of dilated cecal appendix compatible with mucocele.
Source: Own elaboration based on the data obtained in the study.

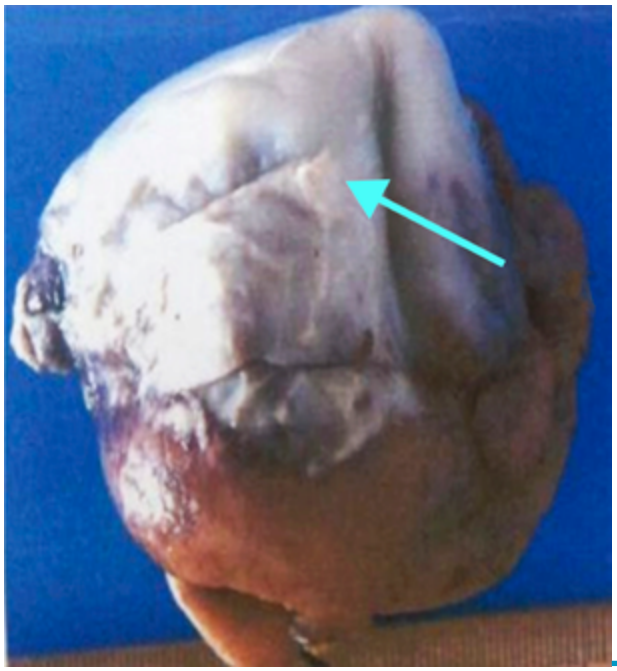

Figure 3. Macroscopic piece
Source: Own elaboration based on the data obtained in the study.

The histopathological study showed a cecal appendix with coagulative necrosis in the mucosa and the wall, mucous membrane with focal mucoid deposit that did not invade the parietal thickness, fibrinonecrotic and hemorrhagic exudate that extended to serosa and periapendicular mesentery, and multiple microabscesses. The final diagnosis was acute gangrenous appendicitis and appendiceal mucocele. Finally, the patient was discharged after 8 days, without further complications.

DISCUSSION

Appendiceal mucocele is a cystic dilatation of the appendiceal lumen with accumulation of mucus. It is caused by an obstruction of the appendix due to the excessive accumulation of mucus with abnormal dilatation. However, the term mucocele does not describe the potential aggressiveness or biological behavior and, therefore, is a descriptive term that does not have a diagnostic character and does not correspond to a defined clinical-pathological entity.

Appendiceal mucocele is a rare pathology, found in 0.2% to 0.3% of appendectomies, which corresponds to 8% of the total appendiceal tumors, with a higher incidence in women (ratio 4: 1) and average age of presentation of 55 years (9,10).

This report describes the case of 69-year-old male patient, that is, in the seventh decade of life, which coincides with the literature. The pathology report revealed simple mucocele, which is found in 20% of cases (11). The histopathological result described acute gangrenous appendicitis and acute fibrinopurulent periappendicitis, appendiceal mucocele and omentum without histopathological alterations.

The form of presentation is non-specific and incidentally appears in 25% of cases. The most frequent symptoms include abdominal pain (27%), palpable abdominal mass (50%), weight loss (13%), nausea or vomiting

(9%), and less frequently, dysuria, hematuria or complications such as perforation (12), bleeding, intussusception or torsion (13).

This patient presented a picture similar to acute appendicitis with pain in the right iliac fossa, anorexia, nausea, and positive Mc Burney' point and Blumberg's sign, which led to a differential diagnosis of acute appendicitis, which is very frequent.

Laboratory tests may be useful to determine an infectious disease, but not to suspect mucocele. For this reason, it is advisable to perform imaging tests, including ultrasounds, which, in this case, served to observe an elongated and tubular structure (also known as chicken drumstick) compatible with the appendix, with a diameter greater than 1.5cm, a thin wall and calcifications (14). Nuclear magnetic resonance (NMR) is also useful for studying this picture. In this case, ultrasound showed a dilated tubular image, with a diameter of 35mm, which led to suspect appendiceal mucocele, although the result was not conclusive.

The treatment for this entity is surgical by laparotomy or laparoscopy; the careful handling of the piece is very important to avoid its rupture and consequent dissemination (pseudomyxoma peritonei). When only the appendix is affected without local or cecal invasion, it can be treated with appendectomy and mesoappendix excision, but if the caecum or an adjacent organ is involved, a right hemicolectomy should be performed (5).

Since the condition of the patient resembled an acute abdomen of 5 days of evolution, an exploratory laparotomy was performed, in which a dilated tumor-like appendix was found, as well as necrosis in its lower third and broad base, which led to suspect mucocele. The caecum was in good condition, so a conventional appendectomy was performed. The patient was finally discharged at 8 days without further complications.

The final result was mucocele without histopathological alteration of the omentum. Based on this finding, periodic check-ups by external consultation were indicated without the need to perform any other surgical procedure until now.

CONCLUSION

Mucocele has a low incidence of 0.2-0.3% among the total of appendectomies. The treatment is surgical due to the potential risk of malignant transformation and to prevent other complications such as rupture and dissemination. It is important to consider the diagnosis prior to surgery and to perform a careful resection, either by laparoscopy or laparotomy.

CONTRIBUTION OF THE AUTHORS

The author and co-authors participated throughout the process of preparation of this case report.

CONFLICTS OF INTEREST

None stated by the authors.

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DELUSIONAL INFESTATION. EKBOM'S SYNDROME IN A 53-YEAR OLD WOMAN. CASE REPORT

Keywords: Case Report; Delusional parasitosis; Ekbom delusory parasitosis; Delusory parasitosis.
Palabras clave: Reporte de caso; Delirio de parasitosis; Delirio de parasitosis de Ekbom.

Mario Javier Olivera MD
Instituto Nacional de Salud
Parasitology Department
Bogotá – Colombia

Hugo Paez Ardila MD
Eliana Maldonado Lara MD
Universidad del Rosario
Infectology Department
Bogotá – Colombia

Julián Felipe Porras Villamil MD
Gabriela Andrea López Moreno MD
Christian Camilo Toquica Gahona MD
Medical Doctor
Universidad Nacional de Colombia
Faculty of Medicine Medical School
Bogotá – Colombia

Corresponding author:
Mario Javier Olivera.
Instituto Nacional de Salud.
Avenida calle 26 No. 51-20 – Zona 6 CAN.
Bogotá, D.C. – Colombia
Email: moliverajr@gmail.com

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ABSTRACT

Introduction: Delusional infestation is a rare psychiatric disorder defined as a condition in which the patient has the unshakable belief and perception of being infested with parasites. Its treatment is difficult, and frequently includes antipsychotic medications (such as olanzapine or aripiprazole). Non-pharmacological treatment, particularly psychotherapy, can be used for less severe cases. Dermatologists and psychiatrists must take a multi-disciplinary approach (preferably in a psycodermatology dedicated clinic) since this type of patients sometimes refuse treatment.

Case description: A 53-year-old female businesswoman describes a clinical history of five years of visual hallucinations, depressive symptoms, and generalized pruritus, along with the use of toxic substances to "clean" her skin and cloths. She reports similar symptoms in some relatives but they were not evaluated. Blood tests and analyses of the "specimen" brought by the patient were performed, yielding negative results. The patient had never been assessed by any specialist, and showed disoriented during the consultation. Follow-up was not possible due to the reluctance of the patient to follow the indications and seek psychiatric treatment. Moreover, the patient did not respond to further communication attempts.

Discussion: Delusional infestation is an uncommon disease that endangers the patients and the people around them. Its treatment is difficult and long, and not conducting proper follow-up is a great risk. Its prevalence and incidence is variable and generally unknown. It can affect the patient, their next of kin, pets or the environment, and the "pathogen" can be a living organism or an inanimate object.

Conclusion: This case is important as it shows the hardships of treatment, adequate follow-up and care, as well as the need to improve how these patients are approached. Additionally, both classical and uncommon signs and symptoms could be observed as the patient stated that her relatives were affected (possible delusional infestation by proxy).

CASE DESCRIPTION

Delusional parasitosis (DP), delusional infestation (DI) or Ekbom's syndrome is an uncommon but not rare (1-3) psychiatric disorder in which patients have a fixed, false belief that they are infected or infested with parasites or other living organisms (4-6). This condition may lead to self-mutilation (5) or affect other members of the family (7). It was first described more than a hundred years ago (8), and is classified as a persistent delusional disorder in ICD-10 (9) or as a delusional disorder of the somatic type in DSM-V (10), although much is left to understand about this disease. The prevalence is estimated between 0.18 and 4.2 per 100,000 (11,12), with an incidence of 1.9 per 100,000 (12). However, its variation is wide: in the United Kingdom, the estimated incidence is about 4.9 per million (11).

Delusional infestation by proxy is estimated in approximately 5-15% of the cases (7, 13). Its onset has been associated with changes in the glucose metabolism of the thalamus and the left putamen, and with alterations in the dopaminergic neurotransmission of the striatum, again in the left putamen (14). There is also evidence of abnormal frontolimbic brain activity (15) and abnormal grey and white matter volume (16,17).

It generally affects women over 50 (30-60) years of age (6, 18), with a mean age of

onset at 56.9 years (19). It can be classified as primary or secondary; the primary form of the disease does not present an organic or psychiatric underlying cause (Table 1) (6,20,21), and patients are otherwise mentally healthy (6,22). A really uncommon form of this disease is DI by proxy, which is a shared psychosis referred as folie à deux or folie à trois, where patients believe that other individuals or pets are infested rather than themselves (18,23,24). This presentation is more frequent in veterinary practice (18).

Table 1. Drugs or conditions associated with the onset of secondary delusional infestation.

Drugs or conditions	References
HIV	(4)
Neuropsychiatric drugs	(25, 26)
Dialysis	(20, 27)
Neuropsychiatric diseases such as schizophrenia and depression	(20, 28)
Stroke	(20)
Psychotropic drugs	(20, 29)
Dementia	(20)
Renal disease	(30)
Iatrogenic	(31, 32)
Occupational hazard for entomologist and healthcare workers	(33)

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

The duration of the disease ranges from months to years (34). The most affected organ is the skin but other parts of the body can be affected as well (21,35). For its diagnosis, delusion must have a duration of at least 1 month, but patients must remain highly functional (1) and present with two main symptoms: 1) the strong belief of being infested despite medical evidence shows otherwise (100% of the patients), and 2) abnormal sensations as if an infectious agents caused them (88%) (6,36,37). Considering that the delusion of being infected can vary in intensity (38), in less severe cases the belief can be wavered but not reduced (39). Some of the symptoms reported are listed in Table 2 (1,4,5,6,12,19,20,40,41).

Table 2. Possible symptoms of delusional parasitosis described by patients or relatives.

Symptoms described by patients	Symptoms described by relatives
Pruritus	Psychosis
Poor sleep	Confusion
Tactile hallucinations	Strange behavior
Self-mutilation or self-damage	
Visual hallucinations, and other types of hallucinations	
Delusional ideas	
Dysphoria	
Disturbed reasoning and judgement	
Formication and other sensations of movement under the skin	
Intrusive and non- reducible belief of infestation	
Proof of infestation (specimen or matchbox sign)	
Use of toxic products topically or orally to “treat” the condition	

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

Atypical manifestations appear when the “pathogens” are relatively large, the environment is infested rather than the individual, or the infestation is caused by inanimate objects (3,6,42). The infesting species range from unspecific living beings to microorganisms and small animals. These specimens can be “stored” and handled without disgust (6,43), and are treated as trophies instead; the spec-

imen, however, should be examined (43). This is known as the "Matchbox sign", which is characterized by the patient collecting an inert substance (for example, dead skin) in a container, stating that it contains living specimens of the parasite in different stages of development (12).

The delusion can result in damage to the skin, hair, eyes and other family members, as they try to "clean" the infestation with dangerous substances or elements including fire and electricity (6,44,45). This condition can be affected by a large array of complications as listed in Table 3 (6,12,35).

Table 3. Possible complications of delusional parasitosis

Erosions
Excoriations
Ulcers
Lichenification
Chronic irritant contact dermatitis
Lichen simplex chronicus
Prurigo nodularis
Lichen amyloidosis
Corneal abrasions
Secondary infections
Increased mortality

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

The recommended treatments are based on anti-psychotic medication such as risperidone, pimozide (36,46), olanzapine, amilsupride, quetiapine and aripiprazole (47-50). Atypical anti-psychotics have a more favorable side-effect profile (50). The therapeutic effect may be observed between one to ten weeks (47). Lepping et al. (51) assessed the efficacy of this drugs, reporting that typical and atypical anti-psychotics achieved a remission proportion of 60-100%. In less severe cases, non-pharmacological treatment, such as psychotherapy, can be used (50,52). Other non-pharmacological therapies include: neurosurgery, transcutaneous electric stimulation and electroconvulsive therapy (50), with a less than optimal success rate (36,50).

Joint management by psychiatrists, psychologists and dermatologists is required (28,53-55), preferably in a clinic dedicated to psychodermatology (56,57). Adequate treatment leads to remission in 75% of the cases, although 25% of those patients may relapse and require longer therapy (58). Treatment should be introduced after obtaining a complete medical history and a systematic evaluation (54), specially to discard secondary causes or differential diagnosis (59-63) (Table 4). Offering antimicrobial or anti-parasitic drug trials reinforces the delusional ideas and is not recommended (6).

Table 4. Differential diagnosis of delusional parasitosis.

Hypocondriasis circumscripta
Dermatitis artefacta
Skin picking disorder
Chronic pruritus
Morgellons disease

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

Three groups of patients have been suggested for classification purposes: 1) patients with hypochondriac traits, 2) patients with paranoid symptoms and without hypochondriac apprehensions, and 3) patients with hypochondriac and paranoid traits (64). Most patients have other associated psychological disorders including anxiety, depression and appearance-related concerns, among others

(37, 65). More information can be obtained in the review made by Freudenman (6).

CASE DESCRIPTION

Female, 53 year-old, Hispanic businesswoman who attends, by her own volition and alone, an outpatient medical appointment at the Parasitology Department of Instituto Nacional de Salud (National Health Institute), referring a clinical history of 5 years characterized by the perception of macroscopic parasites that crawl over her body, biting her face, head and anterior thorax, and leaving white eggs which evolve to brown adults in about eight days. She also referred generalized pruritus and shows excoriations due to scratching (Figures 1 and 2), which she "treated" with Vicks Vaporub, domeboro (calcium acetate and aluminum sulphate), crotamiton and Canesten (clotrimazole) cream. At some point, she also used thinner and varsol to wash her clothes and skin.

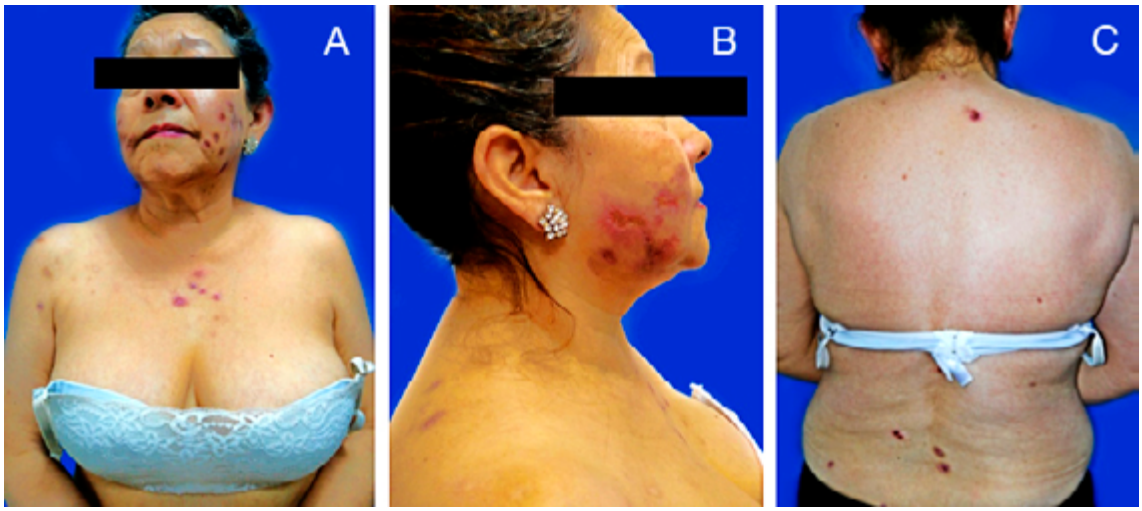

Figure 1. A. Front of the patient. The lesions can be observed in the cheeks, arms, and chest. B. Profile of the patient portraying a close up of the cheek lesions. C. Back of the patient showing the extent of the lesions.
Source: Own elaboration based on the data obtained in the study.

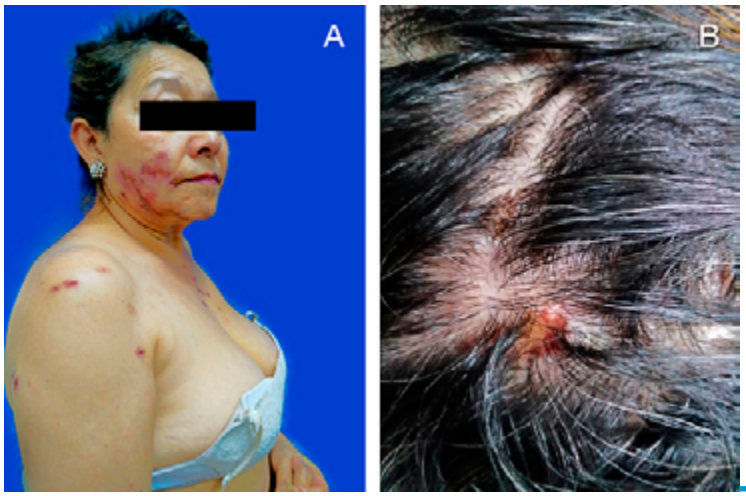

Figure 2. A. Profile of the patient showing the lesions on both cheeks B. Picture showing some of the multiple lesions on the scalp.
Source: Own elaboration based on the data obtained in the study.

The patient takes to the appointment, in a small plastic jar, pieces of skin and coagulated blood which she says contain the eggs and two adults (Figure 3). This specimen is handled without disgust or contempt. She also describes how one of her sons have seen the parasites fly after one of the eggs hatched, while other members of the family do not have parasites but suffer from pruritus. Unfortunately, the relatives were not present during the interview and refused to be interviewed or examined afterwards, so this may be another delusory idea of the patient.

She denies travelling to other places in the past five years. Medical background includes diabetes mellitus diagnosed twelve years ago, and pharmacological background includes metformin 850mg/day, glibenclamide 5mg/day, ivermectine 51 drops (which she has used in repeated doses since the onset of the disease), difenhidramine 50mg every 8 hours and, occasionally, amoxicillin 500mg every 12 hours; she denies using other medications. Her gynecological background is G4P3C0V3.

Physical examination revealed an arterial pressure of 100/60, heart rate of 84, respiratory rate of 18, temperature at 35.5°, weight 51.3kg, height 145cm, multiple excoriations on the scalp and neck, and cicatrized lesions in the inferior third of the face, chest and back. The patient had normo-reactive isocoria and eye bags; the rest of the physical examination was normal.

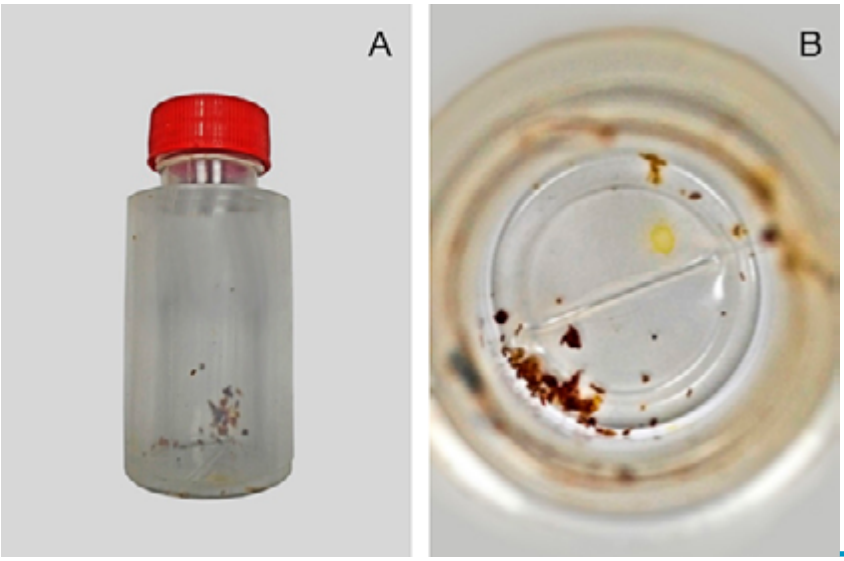

Figure 3. A. Jar brought to the appointment by the patient. B. Close up of the jar contents.
Source: Own elaboration based on the data obtained in the study.

Regarding mental aspects, the patient had a negative attitude, was passive, cried easily, but had good visual contact with the interviewer. She had a slight psychomotor retardation and a sad attitude, with anxiety traits. Delirious thoughts were present, as well as sad mood, with semi-structured suicidal ideations, non-existent introspection, uncertain prospection, and good interpersonal relationships. Her speech was not slurred, and her behavior was organized and non-catatonic. Even though she was depressed, she did not show any signs of alogia or affective flattening, and the depressive symptoms started long after the delusions. She did not hear voices.

Samples were taken, and cultures from feces and lesions were obtained, yielding negative results for microorganisms. Hemo-

gram was normal, tests for liver and renal function were normal, and glycemic control was optimal. Toxicological exam was normal. Complete studies of the skin and fecal matter were made, yielding negative results as well. The specimens brought by the patient were analyzed providing a negative result. The patient was examined for needle marks which were absent. She did not present alterations in the rhinal mucosa nor respiratory or cardiac problems in the physical exam or medical tests.

Due to the absence of criteria to diagnose other psychiatric disorders or substance abuse or physical disease, the patient met the criteria of a delusional disorder of the somatic type (at least 1 month of delusions, visual hallucinations consistent with the disease, social function not markedly impaired, and brief mood episodes compared to the delusional period), thus Ekbom syndrome was diagnosed. The patient did not accept psychiatric treatment, and neither dermatology nor psychiatry services were able to assess the patient. Follow-up was not possible due to the refusal of the patient to start psychiatric treatment, even when it was suggested and advised for her depression.

Even though the patient did not accept treatment or referral, medical indications and recommendations were given. Prognosis is not optimal without pharmacological and dermatological follow-up or treatment, therefore, it is highly unlikely that the condition of the patient will be resolved.

Written informed consent was obtained from the patient for the publication of this case and the photographs.

DISCUSSION

Ekbom's syndrome, better referred as delusional infestation, is an uncommon disease that generally affects woman above 50 years of age. Physicians must be cautious when approaching this kind of patients, considering the words used and even the proposed therapies, in order to generate a good patient-physician relationship and help patients to understand the importance of treatment with a psychiatrist. Not conducting proper follow-up is dangerous; for instance, this case is a prime example of that, since the patient presented a typical primary delusional infestation that not only affects her life and self-esteem, but her family's life as well. Here, the information provided by the patient allowed to infer (although it was not confirmed) that some members of her family may suffer from delusional infestation by proxy. This presentation is similar to other cases reported in the literature (63).

Treatment is difficult and must be carried out based on an interdisciplinary approach. One way of improving treatment is through the use of psycodermathology, which is a new subspecialty emerging from dermatology, and includes interventions such as psychoeducation and cognitive behavioral therapy or joint interviews with dermatologists and psychiatrists (66) that have proven to be cost-effective (57).

Patients generally blame parasites, but can also consider other small organisms or inanimate objects. This condition can affect the individual, someone or something close to the individual or the environment, and there are even cases of shared psychosis.

Usually, sick patients are not violent, but they can be a threat to themselves and others since they may use dangerous substances during the "cleansing" process, and may even mutilate themselves. This case, as other cases (37), shows the relationship between underly-

ing psychiatric diseases and DI, and the long process involved to diagnose this disease. Physicians must first discard and find evidence that the patient is not infested, looking for symptoms or signs of underlying psychiatric disorders or the use of psychotropic drugs or conditions associated with this disease as well. Considerations about pets or individual victims of DI by proxy and their protection are outside the scope of this report.

Some of the most important aspects of this study include, first, initial assessment and referral, even though the patient did not follow the instructions, and second, the thorough clinical and physical examination alongside a complete laboratory analysis done to the patient. Some of the limitations are that the patient was never assessed by a specialist, that the outcome of the case is unknown, and that the relatives could not be evaluated, therefore, delusional infestation by proxy could not be confirmed.

CONCLUSION

This case clearly exposes the hardships of treating this type of psychiatric patients, as well as the difficulties for adequate follow-up and care. A better way of following and delivering care to these patients could possibly include domiciliary visits or clinics dedicated to psycodermatology. However, this case is important since it shows both classical and uncommon signs and symptoms, that the disease can affect patients and possibly their relatives as they can also suffer from similar symptoms (delusional infestation by proxy), and that they should also be included in the treatment. A comprehensive approach to the family and its involvement should be considered when approaching this type of patients.

PATIENT'S PERSPECTIVE

Follow-up was difficult, but during the few interviews that could be conducted, she expressed that her situation was dire, that she was desperate. Her mood had not improved and the negative thoughts were worse over time. At first, her work nor her relationships were affected, but they were compromised as the disease progressed, to such an extent that she was seeking advice from multiple physicians, but she thought that they did not believe her and never returned.

TRANSPARENCY

The authors declare that all the information contained in these pages is true, honest and transparent, that no important aspect of the case was omitted, and that every relevant characteristics or differences have been exposed.

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CONFLICT OF INTERESTS

None declared by the authors.

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