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REPORTES DE CASO

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Editorial:

CASE REPORT: WHY, WHAT FOR AND HOW

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THE IMPORTANCE OF CASE REPORTS

When analyzing Medicine based on the scientific method, a transversal dialectical process becomes evident, since its construction as a science is founded on the fact that knowledge is constantly challenging itself through experimentation and new observations. Such dialectical phenomenon is not the only one that defines Medicine as a science: readiness to create new hypotheses about health-disease processes that cannot be explained with current knowledge is also part of this.

This search for truth mechanisms in Medicine is especially applicable when its object of study is the biological machine that makes up humans. This conception of Medicine —which could be called positivist— is limited and impractical if applied in isolation, although there are strong attempts to clearly define its object of study from reductionism because of the complexity of humans as a biological phenomenon and as a social, ethical and political entity.

However, the positivist approach to Medicine is, at the same time, one of its fundamental components. In turn, case reports become a powerful investigative tool for such approach and a communication mechanism for clinical practice; it is the most efficient way of detecting atypical events in health-disease process, either generated by novelty or by peculiarity **(1)**.

That each tool and research approach are applicable and are limited must be clear **(1)**. Evidence-based Medicine (EBM), as a substantial instrument for clinical practice, emerges and is defined by the intention to support diagnostic and therapeutic clinical decisions, and to determine the prognosis of the disease **(2)**. Nevertheless, defining EBM only based on randomized clinical trials is a mistake; therefore, suggesting that clinical cases are excluded is false. Thus, clinical cases can also be, sometimes, decisive evidence with the highest quality **(3.4)**.

Although clinical decision is the ultimate goal of medicine, it is not its only goal. The discovery of new diseases, how they generate and the behavior of a healthy body are also research objectives for Medicine. In that sense, randomized clinical trials lose effectiveness to detect new diseases or its variants, since their focus is on population, and inferential and descriptive statistics are their backbone. In consequence, they have little power to discover rare or novel variations of the disease, because they are intended to study trends and average values of morbid phenomena.

Thus, case reports —understood as a way of communicating that which is atypical and new— contributes substantially in areas where large-scale population studies have limitations **(5)**. Hence, the role of case reports in positivist medicine, as described above, is sufficient to demonstrate their importance, but this is not its only use. Table 1 shows other scenarios in which case reports have an important role.

Evidence of the potential usefulness of case reports in various scenarios and their resurgence as an investigative method is found in the significant increase of specialized journals about the topic in the last decade **(6)**.

Table 1. Usefulness of case reports in medical and related sciences.

Study of health and disease
Recognition of new diseases
Description of atypical variants of known diseases
Report of rare diseases

Study of health and disease
Atypical associations of symptoms or signs Proposal of hypotheses about the mechanism of diseases Study of the physiology and anatomy of the healthy body
Proof of concept in diagnosis and therapeutics
Proposals of new diagnostic tools Presentation of novel therapeutic tools
Epidemiological surveillance
Drug monitoring and reporting of adverse events in medical interventions Report of beneficial side effects of interventions
Pedagogical usefulness
Medical education through emphasis on important clinical lesions Presentation of useful images in medical training
Public health analysis
Comparisons of medical care forms in different settings Study of health inequities

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

CLARIFYING OBJECTIVES

The first step to report a case is the result from a thorough clinical work summarized in:

- Committed, rational and systematic patient care.
- Willingness to observe.
- Willingness to study and deepen medical education.
- Capacity to be surprised.
- Reflective, critical and self-critical attitude.

Case reports, rather than a form of publication, are the result of a deep commitment of the physician to the patient and to study, which leads to determine the novelty and merit of sharing a particular clinical phenomenon, since it could be the input to deepen the knowledge in medicine (e.g. the atypical presentation of a disease, the results of a therapy, the usefulness of a diagnostic method, among others).

Based on this, when a case report is proposed, clarifying the objectives of the report must be sought. In this sense, the following fundamental questions are proposed whenever a case report is intended:

What do I want to communicate as novel or useful for Medicine? What implications does the observation that I want to report have? Does it contradict current knowledge on certain pathology? Does it expand what is currently known about this disease? Is it something anomalous, unexplained or unexpected? Does it exemplify and clarify a disease?

Based on the clarity given by the answers to these questions, exposure and discussion of the clinical case should be developed, since they are the guidelines to reach the goal, transmit a clinical observation and propose its implications.

HOW TO REPORT A CASE

Judicious clinical practice and clear objectives are two of the pillars of case report. The third is the way how information is communi-

cated, the accuracy of the case report, which will define its usefulness for the knowledge of the disease. This, if done properly, will allow comparison with other cases or make it part of the foundation of other research methodologies through which the validity of the hypothesis of the case reported can be assessed **(5.7)**.

Also, various organizations and medical journals have realized the need to generate quality standards in the way how cases are reported. One of the most important initiatives is the development of CARE consensus and guidelines (acronym for CAse REport), formally published in 2013 in various journals **(8)**. This guideline is accepted by Revista Case Reports and is recommended as a guideline for case reports.

With this in mind, the suitability of a clinical case report is based on three characteristics:

- Transparency
- Full descriptions
- Precision

Transparency involves ethical and epistemological commitment, by those who report, to exposing the case fully and truthfully, including limitations, omissions and errors. This principle determines the compression of medical practice as an imperfect process, with, frequently, limited resources and proneness to error as any human activity. Similarly, understanding the imperfection of clinical practice is seen as an invitation to continuous improvement and transparent case report is an ideal strategy for this.

On the other hand, complete and accurate descriptions are necessary for the case to provide sufficient tools which help corroborating or rejecting hypotheses derived from observation **(9)**. It includes, therefore, adequate sociodemographic identification of the patient, account of symptoms and background, physical examination, reasoning and diagnostic strategies, therapeutic approach, patient monitoring and, finally, outcome of the case. In addition, the limitations of the case should be explained, discussion about it should be established, hypotheses defined and conclusions and derived lessons proposed. CARE guide presents each of these aspects summarized in the items presented in Table 2.

Table 2. CARE Checklist - 2016.

Topic	Item	Checklist item description
Title	1	The words “case report” should be in the title along with the area of focus
Keywords	2	Four to seven key words—include “case report” as one of the key words
Abstract	3a	Background: What does this case report add to the medical literature?
	3b	Case summary: chief complaint, diagnoses, interventions and outcomes
	3c	Conclusion: What is the main “take-away” lesson from this case?
Introduction	4	The current standard of care and contributions of this case—with references (1-2 paragraphs)
Timeline	5	Information from this case report organized into a timeline (table or figure)
Patient information	6a	De-identified demographic and other patient or client specific information
	6b	Chief complaint—what prompted this visit?
	6c	Relevant history including past interventions and outcomes

Physical Exam	7	Relevant physical examination findings
Diagnostic	8a	Evaluations such as surveys, laboratory testing, imaging, etc.
Assessment	8b	Diagnostic reasoning including other diagnoses considered and challenges
	8c	Consider tables or figures linking assessment, diagnoses and interventions
	8d	Prognostic characteristics where applicable
Interventions	9a	Types such as life-style recommendations, treatments, medications, surgery
	9b	Intervention administration such as dosage, frequency and duration
	9c	Note changes in intervention with explanation
	9d	Other concurrent interventions
Follow-up and outcomes	10a	Clinician assessment (and patient or client assessed outcomes when appropriate)
	10b	Important follow-up diagnostic evaluations
	10c	Assessment of intervention adherence and tolerability, including adverse events
Discussion	11a	Strengths and limitations in your approach to this case
	11b	Specify how this case report informs practice or Clinical Practice Guidelines (CPG)
	11c	How does this case report suggest a testable hypothesis?
	11d	Conclusions and rationale
Patient perspective	12	When appropriate include the assessment of the patient or client on this episode of care
Informed consent	13	Informed consent from the person who is the subject of this case report is required by most journals
Additional information	14	Acknowledgement section; Competing Interests; IRB approval when required

Source: (10).

According to this, we would like to invite our readers to conduct clinical observation and develop the capacity to be surprised which, through judicious and routine clinical practice, will allow realizing new, contradictory or unexpected phenomena that are a powerful fuel for the progress of Medicine. Then, case reports will be the log that allows communicating such observations.

Likewise, standardization of the way to report clinical cases is urgent, where transparency and accuracy allow approaching the truth in medicine for the sake of scientific accuracy and reproducibility. The CARE guide is proposed as an ideal tool for this purpose.

Finally, an invitation to enjoy this new issue of the journal is presented. We would like to congratulate all authors for their courage and hard work. You, along with readers, are the essence of this publication.

Thank you for your contributions.

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CASE REPORTS
REPORTES DE CASO

Case report:

ACUTE GENERALIZED EXANTHEMATOUS PUSTULOSIS RELATED TO PHENYTOIN ADMINISTRATION

Palabras clave: Erupciones por medicamentos; Pustulosis exantematica aguda generalizada; hidantoínas.
Keywords: Drug eruptions, Acute Generalized Exanthematous Pustulosis, hydantoins.

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SUMMARY

The occurrence of acute generalized exanthematous pustulosis adverse reactions to medication administration is becoming more frequent. This article reports the case of a 78-year-old woman who attended the clinic with generalized papules and pustules on the scalp, trunk and limbs, with a concordant histology study and who was diagnosed with acute generalized exanthematous pustulosis (AGEP) associated with the use of phenytoin, a medication that may cause different skin reactions and that has been previously related to this disease. The patient was treated with systemic steroids and the disease had a satisfactory outcome.

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INTRODUCTION

Adverse drug reactions are common in in-patients and can happen in several ways, for example as the emergence of inflammatory lesions on the skin **(1)**. Sometimes the cause of the disease can be determined from the morphology of primary lesions. Presence of pustules, in particular, may help to guide the diagnosis of some reactions to medications **(1)**.

From the presence of pustules it is possible to make an acute generalized exanthematous pustulosis (AGEP) diagnosis, a condition related to the administration of some medications such as anticonvulsants, antimalarials and antibiotics **(2)**. This paper presents a case of AGEP associated with the use of phenytoin.

CASE REPORT

A 78-year-old woman from Bogotá, Colombia, with seven days of course of the disease and who had pruritic erythematous lesions in her trunk and scalp. She was experiencing fever from the fifth day, which is why she was hospitalized.

The patient had a history of a two-month-old stroke associated with a seizure she suffered two weeks prior to her hospital admission. Due to this seizure episode, the patient was administered oral phenytoin 200mg/day. She had been previously under pharmacological management for dyslipidemia and dyspepsia with acetylsalicylic acid 100mg/day, lovastatin 40mg/day and omeprazole 20mg/day.

Physical examination of the patient revealed erythematous plaques with fine scale, as well as superficial pustules on the scalp, chin, trunk, and thighs (see Figure 1). In addition, there were no signs of lymphadenopathy and lesions were easy to detach by using curettage. Likewise, a 38.1°C temperature was found in one of her records during hospitalization.

Blood tests showed a complete blood count with hemoglobin 9.7g/dl, hematocrit 30.0%, leukocytes 13950 mm3/dl, neutrophils 81%, lymphocytes 15%, eosinophils 1% and platelets 107000 mm3/dl, whereas transaminases and creatinine studies were normal. The haematoxylin and eosin histopathology report of a skin biopsy allowed identifying epidermis with basal layer vacuolation and subcorneal pustules, as well as dermal edema and dense superficial perivascular lymphocytic infiltrate with presence of eosinophils (see Figure 2). By relating findings made on the pustular skin to its presence in subcorneal histology with measured fever and leukocytosis an AGEP diagnosis was made, thus phenytoin administra-

tion was suspended, since it was considered as a possible cause of the reaction, instead a prednisolone dose of 0.5mg/kg administered orally during seven days was added to the patient's treatment. Once phenytoin was suspended, pustules and fever disappeared at the second day, while the erythema lasted 10 days, then skin symptoms started to decrease until they disappeared after a month, as evidenced in the dermatology outpatient visit.

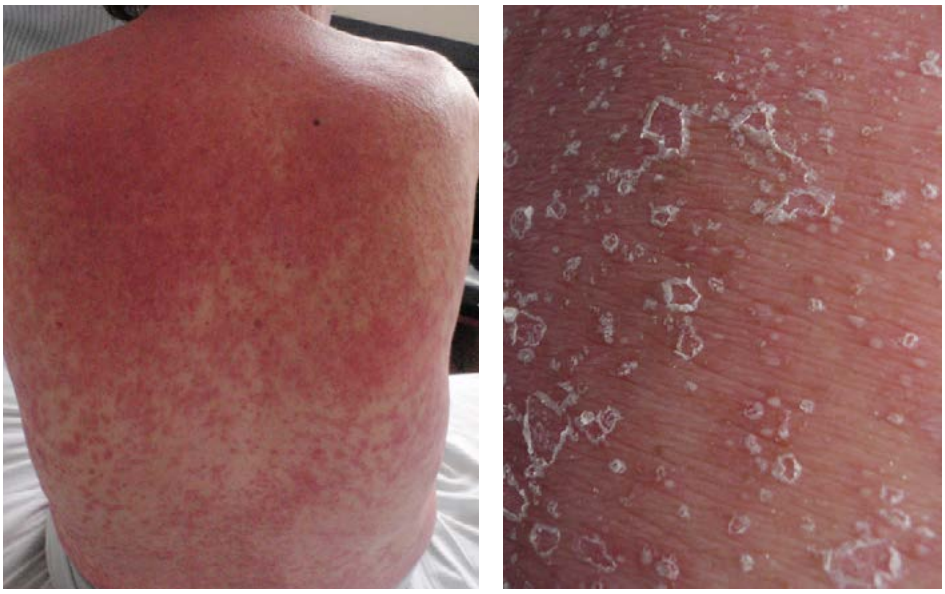

Fig 1. Dorsum. An erythematous base plaque and small pustules are observed when looking closer.
Source: Images obtained from the data collected in the study.

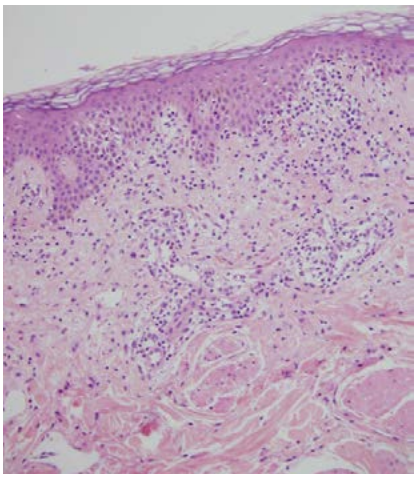

Fig 2. 10x magnified haematoxylin and eosin stain histopathology. Epidermis with presence of orthokeratosis and foci of spongiosis and superficial epidermal and superficial perivascular lymphoid infiltrate with eosinophils are observed.
Source: Image obtained from the data collected in the study.

DISCUSSION

AGEP type adverse drug reaction was first described in 1968 when studying patients suffering psoriasis with a clinical suspicion of pustular disease. Its name was established from the French translation made in 1980, distinguishing it from acute generalized pustulosis, a postreptococcal infection **(2-3)**.

Prevalence of AGEP by age or sex has not been determined and there are few reports describing it in children **(3-5)**. Despite this, it is thought that it has a prevalence of 1 to 5 per million people annually, although it is likely this disease is underdiagnosed **(2)**. In up to 90% of the cases of AGEP that have been reported the disease has been associated with a medication. In addition, there seems to be a predisposition to this condition in haplotypes like HLA-B51, HLA-DR11 and HLA-DQ3, although there is no a specificity according to the type of drug **(5)**. On the other hand, the increase of the expression of Fas, p53 and bcl-2, which leads to keratinocyte apoptosis, could cause AGEP, although a delayed hypersensitivity reaction has also been involved **(5)**.

Additionally, in the literature on this pathology there are studies describing some antibacterial agents, mainly macrolides and penicillins, as well as several antimycotics **(6-9)**; likewise, a large variety of medications, including some antihypertensives (calcium antagonists, angiotensin-converting enzyme inhibitors), antiarrhythmics, anticonvulsants, antidepressants and anxiolytics, even acetaminophen, have been reported **(6)**. The clinical profile of this disease occurs from one to three weeks after the medication has been administered, however in the case of antibiotics the occurrence average time is 2.5 days, while for other medications is 18 days **(6)**. It is important to note that, although less frequently, AGEP has been related to viral and bacterial infections and ultraviolet light exposure **(2,5,6)**.

Symptoms usually found in this clinical profile include fever, asthenia and adynamia. The main cutaneous manifestation of the disease is the presence of pustules, however AGEP clinical picture starts with edematous alike erythematous macules that extend mainly in intertriginous areas **(2,6)**. Generally, pustules emerge in these areas, they do not have a follicular pattern and, sometimes, they can come together, which causes a false Nikolsky sign **(2,9)**. Another characteristic of AGEP is the presence of lymphadenopathies **(2)**.

Systemic involvement is reflected in the appearance of neutrophilic leukocytosis, although eosinophilia may occur in one third of cases. Aminotransferases elevation is mild, less than the double of the normal value, whereas in the creatinine clearance process a 30% reduction occurs **(2,6)**.

However, analyzing AGEP through histopathology does not allow the medical doctor to make a diagnosis at the disease acute moment, which delays initial treatment; but histopathology findings such as the appearance of subcorneal and intraepidermal pustules with peripheral spongiosis may help in making the diagnosis. The dermis may also be affected by a superficial perivascular infiltrate containing lymphocytes, neutrophils and, to a lesser extent, eosinophils. Some mild vasculitis changes, as well as necrotic keratinocytes may also happen **(2,6)**.

Sidoroff *et al.* **(2)** suggest five diagnostic criteria for this disease:

1. Multiple pustules, from tens to hundreds, on an erythematous base.

2. Compatible histopathological changes.
3. Fever higher than 38 °C.
4. Neutrophil count higher than 7000 mm3.
5. Spontaneous resolution in 15 days.

In their research, in order to determine the diagnosis, Sidoroff *et al.* **(2)** created a table that includes the following items: morphology (pustules, erythema, distribution and scaling); course of the disease (acute onset, fever, spontaneous resolution, mucosal involvement and neutrophil count increase higher than 7000 mm3), and histological findings (neutrophil exocytosis, papillary edema and spongious changes). Nevertheless these items are complemented by an unpractical score in terms of daily clinical practice **(1,2)**.

Patch tests and lymphocyte transformation tests have proved useful to determine the agent involved in the disease, achieving a positivity rate of up to 80%; besides they make clear the process in the pathogenesis of T cells **(3)**.

The course of the disease implies that its clinical scenario should disappear between 4 and 10 days, with fever and lymphadenopathy being the first symptoms to do so. On the other hand, pustules usually heal spontaneously at approximately nine days resulting in scaling after their resolution **(2,6)**.

AGEP is a disease difficult to diagnose in its initial stage and can be confused with an infectious process. Similarly, its initial development may resemble that of a DRESS (drug rash with eosinophilia and systemic symptoms) hypersensitivity reaction to medications, where there is low or null presence of pustules. Furthermore, clinical presentation of pustular psoriasis is difficult to differentiate, but usually this is a long course disease that has frequent relapses, while subcorneal pustular dermatosis has a less acute development **(1,6)**. Likewise, Reiter's disease differs in joint involvement, which is not found in AGEP, and acneiform eruptions, which also have pustules, generally triggered by previous use of corticosteroids, which is a situation that constitutes the treatment of AGEP, rather than its cause **(3)**.

Despite there are vasculitis features in AGEP, it should be histologically differentiated from vasculitis purpura. Really severe cases may look like toxic epidermal necrolysis when pustules coincide with skin scaling **(1)**.

As it happened in the case reported here, the use of phenytoin has also been described in other AGEP cases where idiosyncratic adverse skin reactions to medications are reported, including DRESS-type hypersensitivity, maculopapular exanthema, Steven Jonhson syndrome and necrolysis toxic epidermal **(10,11)**. In addition, there are cases reporting cross-reactivity between phenytoin and carbamazepine and between phenytoin and phenobarbital with skin manifestations **(12)**. Nonetheless, there is only one case in the literature that reports the association of AGEP with this medication **(13)**.

CONCLUSION

Quitting the use of the medication is fundamental in the treatment for AGEP. In addition, using systemic corticosteroids may be appropriate in cases with hepatic or systemic involvement. The administration of antipyretics is adequate based on the symptomatology of the patient **(1)**.

AGEP is an adverse drugs reaction that usually finds its resolution after stopping the use of the medication causing it. There are different drugs related to its onset, including phenytoin, as seen in this case.

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Case report:

IDIOPATHIC MECONIUM PERITONITIS AS A SURGICAL EMERGENCY: A CASE REPORT IN A NEWBORN

Palabras clave: Meconium; Peritonitis; Cyst; Obstruction; Postoperative; Ileus; Digestive System Abnormalities; Prenatal Diagnosis.
Keywords: Peritonitis; Meconio; Quistes; Obstrucción intestinal; Ileus; Complicaciones postoperatorias; Anomalías del sistema digestivo; Diagnóstico prenatal.

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SUMMARY

Meconium peritonitis is a rarely described condition that is typically found in fetal and perinatal patients. It manifests in different ways and requires an early diagnosis. In this case report, a clinical suspicion of the disease was made at the antenatal period through an obstetric ultrasound scan showing an abdominal mass in both the bottom and top right quadrants. The patient initial symptoms were those of acute intestinal obstruction. Nevertheless, these symptoms improved after surgical approach was timely performed. Based on this outcome, it can be concluded that a favorable outcome to this pathology depends on its early diagnosis, since an early diagnosis decreases morbidity and mortality chances.

INTRODUCTION

Meconium peritonitis is a non-infectious chemical inflammatory response secondary to intestinal perforation in the fetal period **(1)**. It manifests in several ways, including cyst or meconium pseudocyst **(2)**, where the most common complication is intestinal obstruction, a surgical emergency that rapidly alters the patient's general condition, since its symptoms include emesis, oral feed intolerance and severe dehydration, therefore it requires a rapid and specialized approach, particularly in this age group **(3)**.

In this case, multiples studies and an early diagnosis allowing the anticipation of possible complications were required. Likewise, an analysis of the patient's clinical condition was necessary to decide aspects such as when to perform the surgery, pre-surgical studies, surgical approach and postoperative actions that prevented the development of pathologies such paralytic ileus.

CLINICAL CASE

A 19-day-old female patient from Manizales, Colombia, who was referred by her pediatrician due to a prenatal diagnosis consisting of an ultrasound showing abdominal mass with a fixed bowel loop compatible with possible meconium peritonitis. The patient had a left femur fracture secondary to perinatal trauma caused by fetal extraction through caesarean section. There were not complications during the term of pregnancy and, apart from folic acid during the first trimester, the mother did not consume any medication. The patient was asymptomatic in the outpatient evaluation, thus her case was referred to the surgical board of the Fundación Santa Fe de Bogotá, who decided to schedule a mass resection surgery.

When the girl was 24 days old, her mother took her to the emergency department due to abdominal distention and several emetic episodes. After performing the patient's physical assessment it was determined that she was active, her general condition was regular and that she had a left femur immobilization harness (fracture caused by labor dystocia). She had a distended abdomen, painful on palpation, with a mass located in her upper abdomen. The mass size was 5cm x 5cm approximately and it was delimited, solid and fixed. Later, this finding was confirmed through an abdominal ultrasound. In addition, it was observed that the size of the mass did not increase: it remained at 42 mm x 41 mm. Finally, an abdominal x-ray showed a calcified mass with signs of possible partial intestinal obstruction (see Figure 1).

Due to the peritonitis acute clinical manifestation, the patient was sent to the operating room where the surgeons found an abdominal mass of approximately 7 x 7 cm with two cavities in the right hemiabdomen that adhered

to the parietal peritoneum. The largest portion consisted of a solid mass of apparently meconium content, whereas the small portion was cystic, with non-fetid fibrinous membranes (see Figures 2a, 2b and 2c). Additionally, several adhesions in jejunoileal loops of bowel (see Figure 2d) and a 0.3 cm diameter antimesenteric intestinal perforation located at 10 cm from the ileocecal valve were observed. Finally, a 0.5 cm high broad-based Meckel's diverticulum located at 20 cm from the ileocecal valve was also found (see Figure 2e).

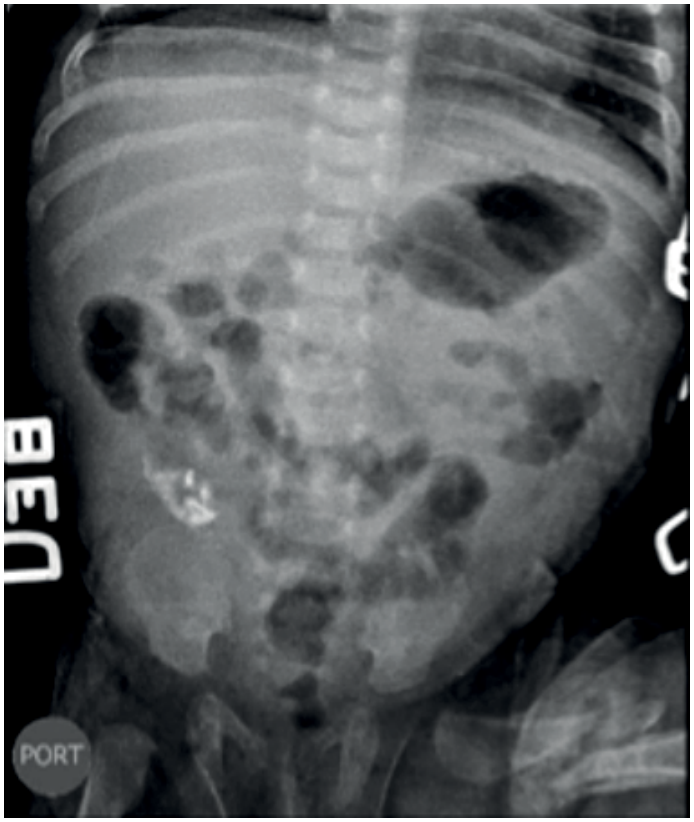

Fig 1. Abdominal x-ray. 42 mm x 41 mm calcified mass with signs of possible partial intestinal obstruction.
Source: Images obtained from the data collected in the study.

Taking these findings into account, a complete resection of the mass, a release of intestinal adhesions and a suture of the intestinal perforation, in a transverse direction, were made. Because of the size of the Meckel's diverticulum its resection was not deemed suitable. Subsequently, pathology samples were taken and the patient was transferred to the pediatric intensive care unit, where she started receiving parenteral nutrition.

The follow-up abdominal x-ray showed a smaller amount of gas in the gastric chamber, as well as residual heterogeneous calcifications in the right hemiabdomen (see Figure 3). Pathology results allow identifying "an ileal perforation with dystrophic calcification, adhesions of richly vascularized fibrous tissue

that was chronically inflamed, a peritoneal cyst associated with mixed inflammation and fibrosis, and meconium peritonitis", which led to dismiss an intestinal duplication diagnosis.

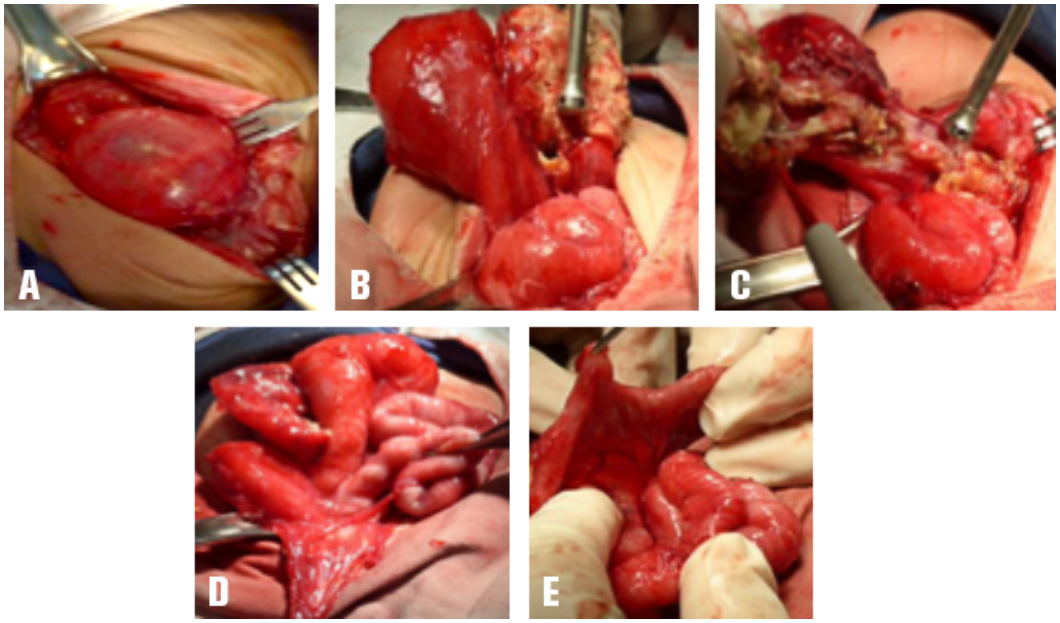

Fig 2A, 2B and 2C. 7 x 7 cm mass with two cavities: the first cavity is large and solid and has meconium content, while the second is cystic and has non-fetid fibrinous membranes. Fig 2D. Jejunoileal adhesions. Fig 2E. Antimesenteric intestinal perforation found 10 cm away from the ileocecal valve

Source: Images obtained from the data collected in the study.

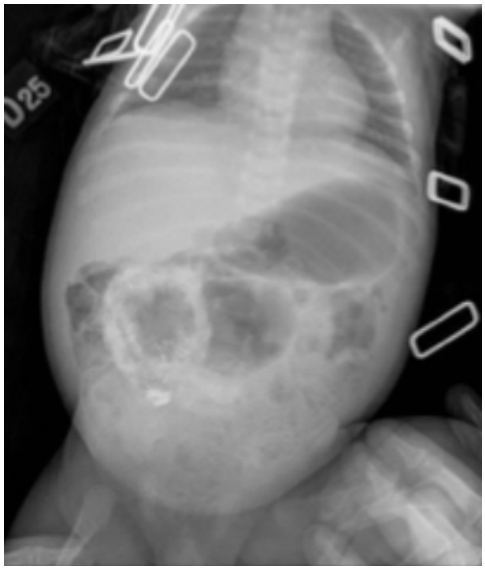

Fig 1. Postoperative abdominal x-ray taken 24 hours after the surgery was performed.

Source: Images obtained from the data collected in the study.

In the postoperative period, the patient had a four days development ileus, which is an expected outcome from the surgical intervention and the pathology she underwent. In the fifth postoperative day she successfully tolerated oral feeding and in the sixth postoperative day she had her first liquid bowel movement, so it was decided to quit parenteral nutrition and keep feeding her orally. Since there were no complications during her recovery, the patient was discharged in the seventh postoperative day.

DISCUSSION

Fortunately, the case reported here had an early diagnosis that was made in utero at week 24 of gestation from the identification of a calcified mass, which is a pathognomonic sign of meconium peritonitis **(3)**. This allowed the pediatric surgery unit to have a prior knowledge of the case, which in turn made possible choosing an appropriate treatment plan. Furthermore, since prenatal and postnatal images of the patient were available it was possible to define the surgical approach to be performed, as well as the location and size of the mass.

The positive aspect of the physical examination was the presence of a palpable abdominal mass, which is something unusual and is commonly associated with symptoms such as abdominal distension, ascites, and intra-abdominal calcifications, among others **(4)**. On the other hand, despite scheduling a mass resection surgery because of the asymptomatic condition of the patient, the procedure was performed as a surgical emergency due to the manifestation of acute obstructive symptoms that developed in a short time. However, the identification of the obstructive condition and its prompt medical-surgical management allowed an adequate progress and favorable development of both the surgical intervention and the postoperative recovery.

As for surgical management, a pseudocyst mass compatible with a meconium cyst was found. The mass was associated with a 0.3 cm diameter ileal perforation with necrosis in its borders and did not have underlying vascular involvement, thus resecting a part of the ileum was not necessary, a typical complication in this pathology **(5,6)**, obtaining a better prognosis. It is important to emphasize that during the course of the disease, an added left femur fracture was always considered. Fortunately, such fracture did not cause any complications. Findings made during the surgical procedure suggested a possible intestinal duplication, since the walls of the mass resembled those of the intestine, but this option was later discarded by the pathology report.

Meconium peritonitis may be secondary to several etiologies, including intestinal atresia, volvulus, congenital flange, and meconium plug syndrome **(2,7)**, the latter being the most common cause. A low percentage of these patients show a clinical picture of idiopathic origin, also known as simple meconium peritonitis **(5)**, which, after discarding intestinal duplication, is the most probable diagnostic suspicion for the case reported here. This condition has been associated with transient vascular insufficiency or hypoxia of the fetal bowel, as well as with certain fetal intestinal infections, however such associations have not been yet fully established **(6,5)**.

Regarding medical-surgical management in this case, surgical intervention instructions should be emphasized, including intestinal obstruction or persistent drainage of meconium to the abdominal cavity, which can lead to progressive abdominal distention, respiratory distress, or sepsis **(7)**. Therefore, the surgical

emergency instruction reported here was appropriate. At the beginning of the clinical picture, resecting the mass, due to its size rather than to gastrointestinal symptoms, was considered as an appropriate option in the context of an asymptomatic patient, given the risk of complications.

In conclusion, meconium peritonitis is a condition poorly described in the literature. In addition, it has a broad spectrum of both etiologies and probable short-term complications **(1)**. Its clear method of diagnosis allows the medical staff to provide a rapid and adequate management **(3)**. This case report shows how meconium peritonitis complications can develop acutely and unexpectedly. Fortunately, complications experienced by the patient had a favorable outcome since they were treated in a timely manner, which substantially decreased the probability of morbidity and mortality.

CONFLICTS OF INTEREST

The authors of this study state there were not conflicts of interests in writing this case report up (fees, personal benefits) or personal conflicts (directed towards a service or research unit) between authors and medicine manufacturers or surgical material suppliers.

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ACUTE IDIOPATHIC PULMONARY HEMORRHAGE IN INFANTS. REPORT OF TWO CASES AND LITERATURE REVIEW

Palabras clave: Hemosiderosis, Enfermedades pulmonares, Lactante. (DeCS).
Keywords: Hemosiderosis; Lung diseases; Infant. (MeSH).

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ABSTRACT

Acute idiopathic pulmonary hemorrhage is a rare life-threatening disease in children. The classic triad includes hemoptysis, anemia and respiratory distress. Since clinical presentation may vary, diagnosis can be difficult. Severe respiratory distress, and ventilatory failure requiring mechanical ventilation are often present. Chest X-rays usually show unilateral or bilateral infiltrates, therefore, other causes of pulmonary hemorrhage must be excluded, since most of them correspond to systemic diseases. Treatment often requires intravenous steroids to solve the respiratory failure in most cases.

We present two cases involving infants treated at Hospital San José (a fourth level hospital in Bogotá, Colombia) with acute idiopathic pulmonary hemorrhage which required mechanical ventilation and responded to intravenous steroids. A literature review was conducted with special emphasis on clinical presentation, diagnosis and therapeutic approaches.

INTRODUCTION

Diffuse pulmonary hemorrhage is a rare disease in the pediatric population, originated in the pulmonary microvasculature and, in most cases, associated with systemic diseases. When presented in isolation and possible systemic causes are discarded, it is known as idiopathic pulmonary hemosiderosis or idiopathic pulmonary hemorrhage (IPH) (1).

According to the American Center for Disease Control (CDC), acute idiopathic pulmonary hemorrhage (AIPH) refers to a pulmonary hemorrhage episode in a previously healthy infant with no history nor other neonatal diseases that may be considered as the cause. AIPH is manifested as hemoptysis, epistaxis or bleeding in the airway, unrelated to bleeding in the upper respiratory tract or digestive tract. It must be associated with severe respiratory distress or ventilatory failure, and mechanical ventilation is also required; chest X-ray or CT should show unilateral or bilateral alveolar infiltrates (2). Data on incidence is scarce, and most information comes from case reports: the largest American series has 47 cases (3), followed by Japan with 39 cases (4), France with 25 and Sweden with 19 (5,6). On the other hand, there are no pediatric case reports in Colombia.

Two cases of previously healthy infants who developed AIPH, respiratory failure and required mechanical ventilation are presented. These cases were treated at the Pediatric Intensive Care Unit (PICU) at Hospital San José (HSJ), a fourth-level university hospital in Bogotá, Colombia, which also provided the necessary data from clinical charts. For the first case, consent was requested, and follow-up was done; in the second case, the patient could not be located for follow-up. Furthermore, a review of existing clinical literature was used to conclude that early diagnosis of this disease, which is life-threatening, allows an adequate therapeutic approach, which may reduce morbidity and mortality.

PRESENTATION OF THE CASES

Case 1

Male, 40-days-old patient from Bogotá, born at full term, with normal neonatal adaptation, and appropriate weight and size. He was admitted to the emergency room due to a

sudden onset of heavy nose and oral cavity bleeding, associated with marked pallor and loss of tone. He presented cardiorespiratory arrest, so resuscitation was performed for 10 minutes and tracheal intubation showed heavy bleeding. Conventional mechanical ventilation and inotropic support with epinephrine was initiated; after stabilization, he was transferred to the PICU.

Important history included pentavalent, pneumococcal, rotavirus and oral polio vaccination the same day in the morning, and a feverish peak was treated with acetaminophen. This was reported to Instituto Nacional de Salud (local government health institution), who, after a case review concluded that the clinical picture could not be related to vaccination, since no similar cases were found in the literature. Exposure to tobacco smoke was discarded, the residence site had drinking water and no outstanding humidity was reported. Additionally, family history was uneventful and physical examination findings showed no other bleeding sites nor signs of abuse.

Central venous blood gases showed anemia with hemoglobin of 7.7 g/dl; clotting times were prolonged, and partial thromboplastin time was 55.4s with control time 30.1s. Prothrombin time was 13.6s with control time 10.9s and fibrinogen was 530 mg/dl. Renal function and aminotransferases were normal, C-reactive protein was negative, electrolytes were normal, transfontanelar ultrasound was normal and chest x-ray showed alveolar opacities in patches in all four quadrants.

The patient was diagnosed with idiopathic pulmonary hemorrhage and severe anemia that required transfusion of packed red blood cells at a dose of 20 ml/kg; methylprednisolone 1 mg/kg was also administered intravenously every 6 hours. The patient improved, inotropic support was discontinued, ventilatory parameters were decreased and was extubated on the third day. The hemogram performed after transfusion was normal, showing leukocytes 9800cel/µl, neutrophils 7800cel/µl, monocytes 700cel/µl, lymphocytes 1800cel/µl, hemoglobin 12.5 g/dl, hematocrits 37.1%, MCV 91.4 fl, MCH 30.8 pg, RDW 148% and platelets 238000 cells/µl. During hospital stay, the patient had thrombin time 16.3s and no new bleeding episodes occurred. The slightly altered clotting times were interpreted as caused by a post arrest effect.

Methylprednisolone was administered for four days, followed by prednisolone orally at doses of 1 mg/kg/day, with a gradual dose reduction until discontinuation on day 10. During hospital stay, he developed a urinary tract infection by Citrobacter freundii, an extended-spectrum β-lactamase producing bacteria, associated with the urinary catheter used. Therefore, ertapenem 15 mg/kg/dose was administered every 12 hours for 10 days. The patient was discharged after 17 days of hospitalization with an order for urethrocystography, which was normal. The patient was subsequently evaluated in a follow-up clinic at the age of 39 months; he was found healthy, with a good nutritional condition, normal neurodevelopment status, and no new episodes of bleeding nor heart or lung disease.

Case 2

Nine-weeks-old female patient, from Bogotá, born vaginally, full term, with normal neonatal adaptation, and adequate weight and height for age. The patient was admitted to a sec-

ond-level hospital with a clinical picture of two days of Bristol stool type 1 with blood traces. During observation, she presented mild respiratory distress without other symptoms, so supplemental oxygen was supplied improving oximetry and breathing pattern. She suddenly presented respiratory arrest, and resuscitation and endotracheal intubation was performed, and was then referred to HSJ.

On admission to PICU, the patient was hypoxemic with severe oxygenation disorder, poorly perfused and heavy bleeding was seen during endotracheal intubation. Laryngoscopy showed infra-glottal bleeding; the endotracheal tube was changed and conventional mechanical ventilation with high parameters was used. Physical examination showed no other signs of bleeding nor abuse. An interview with the mother revealed breast and formula feeding, no exposure to tobacco smoke, no abnormal humidity at place of residence, availability of drinking water and no remarkable medical family history.

Laboratory test results showed severe normocytic, heterogeneous normochromic anemia with hemoglobin 6.3 g/dl, hematocrits 19%, mean corpuscular volume 90 fl, mean corpuscular hemoglobin 29.8 pg, mean corpuscular hemoglobin concentration 32.9 g/dl and red cell distribution width 14.1%. In addition, the patient presented leukopenia 1800 cells/ml and neutropenia 700 cells/µl, with normal platelet count, normal clotting time PT 11.5s with control time 10.5, INR 1.09 and PTT 30.5s with control time 30.5, elevated aspartate aminotransferase 179 U/L and alanine aminotransferase 191 U/L, hypoalbuminemia 1.9 g/dL and C-reactive protein 4.1 mg/dL. Chest X-ray showed diffuse alveolar involvement in the four quadrants, without air trapping nor pleural effusion. An echocardiogram was performed with normal results. After analyzing these findings, the diagnosis was: multilobar pneumonia, pulmonary sepsis and possibly, acute idiopathic pulmonary hemorrhage (Figure 1).

To treat this infection, piperacillin/tazobactam 300 mg/kg/intravenous dose was initiated. Due to a distributive shock, vasopressor support with noradrenaline was also initiated, and a packed red blood cells transfusion was given. Because of the torpid evolution and the possibility of idiopathic pulmonary hemorrhage, on the following day, methylprednisolone 1 mg/kg/intravenous dose was ordered every 6 hours. On the second day steroids use, a control X-ray was done. A significant improvement in alveolar involvement was found, requiring a decrease in ventilatory parameters. Vasoactive support was discontinued after completing four days in steroids (Figures 2 and 3).

During hospital stay, no signs of infection was found and both viral panel and blood cultures were negative, so piperacillin/tazobactam was discontinued. Extubation was done at day 5, but reintubation was necessary due to laryngotracheitis associated with mechanical ventilation. Eight days after admission, the patient was finally extubated with success and discharged on oxygen with nasal cannula due to desaturation at room air.

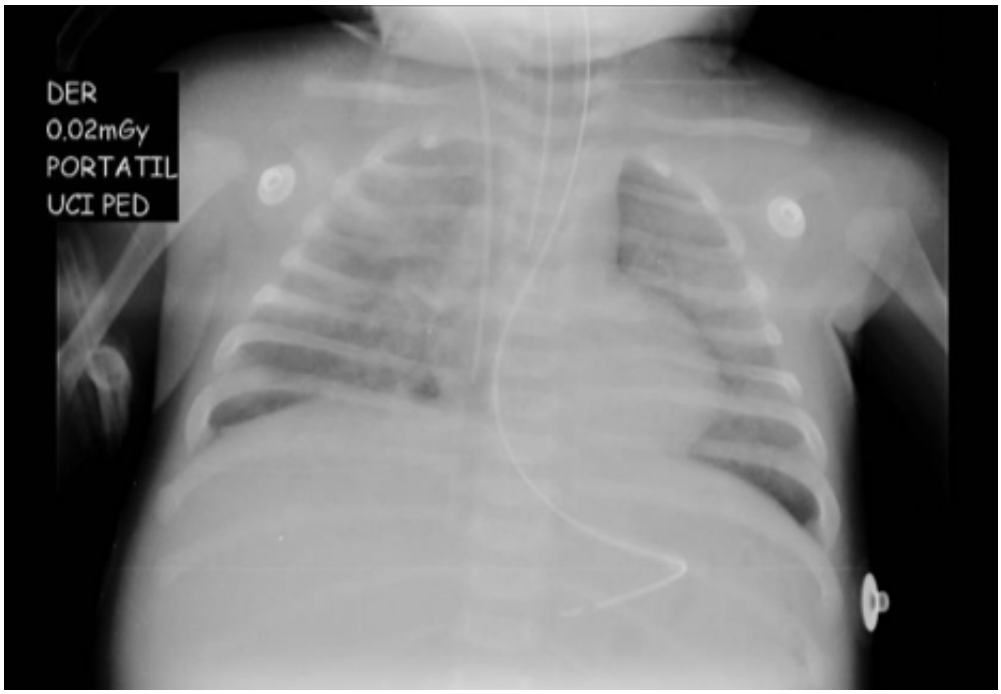

Fig 1. Chest x-ray, diffuse alveolar infiltrates.
Source: Own elaboration based on the data obtained in the study.

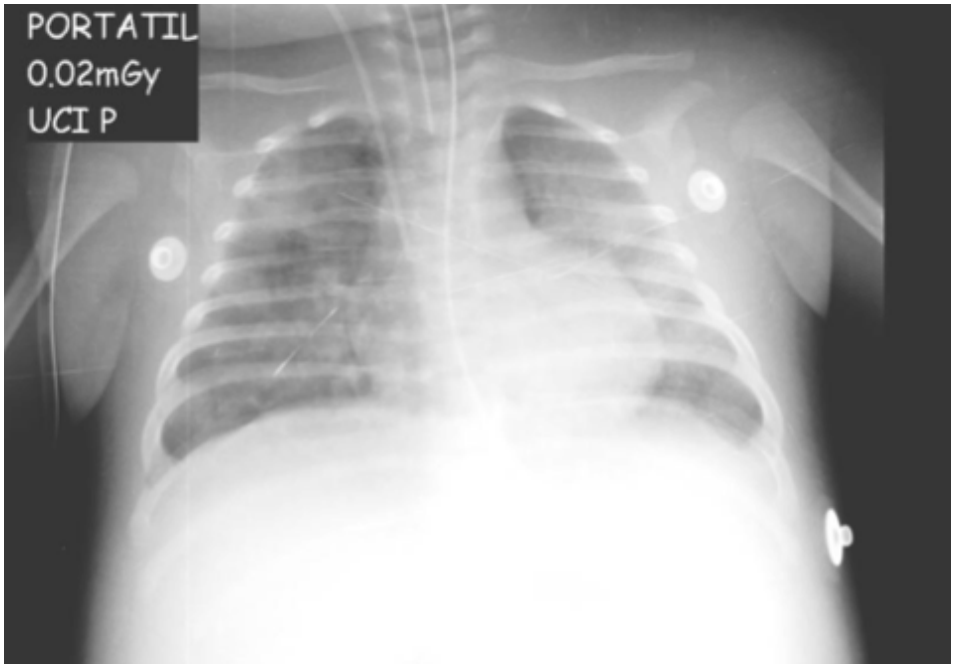

Fig 2. Chest x-ray: improvement in alveolar infiltrates after steroid use (2days).
Source: Own elaboration based on the data obtained in the study..

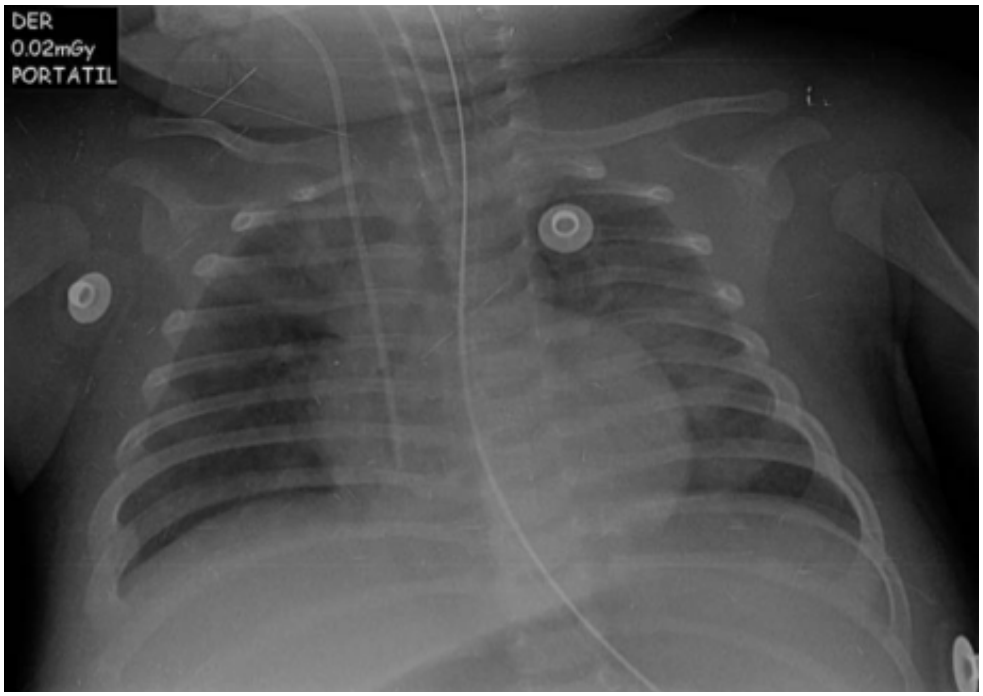

Fig 3. Chest x-ray: improvement in alveolar infiltrates after steroid use (4 days).
Source: Own elaboration based on the data obtained in the study.

DISCUSSION

Frequency

Pulmonary hemorrhage is exceedingly infrequent in children, so IPH is a rare cause of diffuse alveolar hemorrhage in this population. It is seen more often in children under the age of 10, mainly between 1 and 7, and without a clear impact of gender.

In Sweden, between 1950 and 1979, 10 cases were reported, representing an annual risk of 0.24 cases per million children (6). In Japan, for a period of 20 years (1974-1993), 39 cases were documented with an incidence of 1.23 cases/year per one million children (4). In the United States, initially, several outbreaks were reported: between 1992 and 1994, 7 cases of AIPH were documented in Chicago (3); between 1994 and 1997, an outbreak of 10 cases was identified in Cleveland, and, finally, 30 cases treated at Rainbow Babies and Children's Hospital between 1993 and 2000 (7) were reported. In France, 25 cases were reported from 1999 to 2012 (5).

Concepts and classification

Diffuse alveolar hemorrhage may result from immune processes such as vasculitis mediated by neutrophil cytoplasmic antibodies, anti-glomerular basement membrane syndrome, connective tissue diseases, or antiphospholipid syndrome. It could also be due to vasculitis mediated by IgA or cow's milk protein allergy, among others. Moreover, it may also be related to non-immune processes such as infection, heart disease, respiratory distress syndrome or acute coagulopathy. It has also been described as secondary to drugs and

toxics. When all the causes mentioned above have been ruled out, it is possible to diagnose idiopathic pulmonary hemorrhage. (8)

Risk factors and etiology

The IPH etiology is not yet clearly established. During the Cleveland outbreak, some risk factors, such as male sex, lack of breastfeeding, exposure to tobacco smoke and water pollution 6 months prior to the episode were reported. Exposure to fungi, especially to Stachybotrys chartarum toxin, was also suspected. However, CDC decided that establishing a causal relationship between Stachybotrys chartarum and IPH was not possible based on the existing evidence. It is worth considering that exposure to chemical agents has also been involved in other series (7).

Some authors have suggested that risk factors for sudden infant death syndrome (SIDS) may be related to IPH cases, because blood has been found in the airways of infants who died and were diagnosed with SIDS. A study in New Zealand found nosebleed in 15% of cases (9) and another study reported alveolar hemorrhage in 47% of autopsies (10). Nevertheless, pulmonary hemorrhage is common among autopsies in children, especially if cardiopulmonary resuscitation was performed. So it is difficult to conclude whether pulmonary hemorrhage was the cause of death or a marker for some other problem which resulted in death. (11)

Idiopathic pulmonary hemorrhage may be associated with celiac disease and this relationship is known as Lane-Hamilton syndrome. So far, there are few cases reported in the literature and only two theories have been proposed to explain the common pathogenetic pathways. The first suggests the presence of circulating immune complexes acting on the epithelial basement membrane, on the endothelial lung and on the enterocyte; the second proposes that this happens due to inadequate local immune response to T-cells in both entities during gluten intake in genetically predisposed individuals (12). In a cohort of 25 patients with IPH in France, antibodies specific to celiac disease samples were taken in 14 patients (56%), of which four (28%) were positive (5).

Clinic

IPH clinical presentation is characterized by the classic triad of hemoptysis, anemia and infiltrates on chest x-ray due to alveolar bleeding; however, these findings do not always occur simultaneously, which can delay diagnosis. When hemoptysis is severe, it can be life threatening, manifesting itself with breathing difficulty, abdominal pain, hepatosplenomegaly, leukocytosis (13) and bilateral alveolar or interstitial opacities in chest x-ray, which can lead to misdiagnose pneumonia (14).

The patients in this series presented an acute severe respiratory clinical picture; one case with the classic triad that allowed early diagnosis and the other, whose initial diagnosis was pneumonia, with anemia, interstitial opacities on chest x-ray and pulmonary bleeding during laryngoscopy. Moreover, AIPH was proposed for the second case based on torpid evolution of antimicrobial management as described in the literature. Unfortunately, the hospital lacked the necessary equipment to perform bronchoalveolar lavage, which is the ideal next step to complete the study.

Another form of presentation of AIPH is chronic pulmonary hemorrhage, manifested as iron deficiency anemia unresponsive to iron therapy, accompanied with respiratory symptoms such as coughing, wheezing, shortness

of breath, recurrent or chronic cyanosis, along with the appearance of swallowed blood, which can be confused with digestive tract bleeding (13). Recurrent bleeding episodes can cause progressive pulmonary fibrosis, thus, establishing an early treatment is important (5).

Diagnosis

AIPH is an exclusion diagnosis, therefore, in 2004, CDC established the criteria: a case is confirmed only when it occurs in infants younger than 1 year, with a gestational age at birth greater than 32 weeks, without neonatal medical history associated to pulmonary hemorrhage and with the following conditions (2):

1. Sudden onset of hemorrhage or evidence of bleeding in the airway, with signs such as epistaxis, hemoptysis and frank infra-glottic blood unrelated to medical procedures, or identification of more than 20% of hemosiderin-laden macrophages in bronchoalveolar lavage or biopsy. It is necessary to discard oropharynx and nasal bleeding.
2. Severe disease that leads to acute respiratory distress or respiratory failure, which should lead to hospitalization in a PICU or in a neonatal intensive care unit with intubation and mechanical ventilation.
3. Unilateral or bilateral diffuse pulmonary infiltrates on chest x-ray or chest CT documented in the first 48 hours of valuation.

For diagnosis of AIPH, physical abuse, disease with lung involvement at birth, history of bronchopulmonary dysplasia, congenital heart disease, pulmonary hypertension prior to endotracheal intubation or other diseases that could explain pulmonary hemorrhage, should be ruled out. Patients presenting with sudden pulmonary hemorrhage, with or without respiratory distress and with or without significant findings on chest x-ray, are probable cases (2).

Regarding the cases presented here, patients met the criteria: they were infants between 7.5 and 9 weeks of age, with evidence of airway bleeding, severe respiratory distress requiring endotracheal intubation, mechanical ventilation and bilateral infiltrates on chest x-ray. Given the favorable response to management, fibrobronchoscopy and bronchoalveolar lavage were not needed. No history of lung disease was found and heart disease was discarded.

In the first case, platelets were normal and clotting time was abnormal after the cardiac arrest. No new episodes occurred and, therefore, no further studies were conducted to verify bleeding disorders. In the second case, platelets and clotting times were normal; renal function was normal, there were no findings that indicated physical abuse and no significant gastrointestinal symptoms were found, hence, no digestive tract endoscopy was performed. Finally, no studies were conducted to find autoimmune diseases because of the patients' age and adequate evolution.

Investigation on AIPH patients should include studies for the diagnosis of autoimmune diseases, since some of them may initiate with pulmonary hemorrhage. In the French series with 25 IPH patients, 68% of them early on, and 6 more during follow-up, presented autoimmune antibodies; the most frequent were anti-smooth muscle antibody (50%), antinuclear antibodies (45%) and anti-neutrophil cytoplasmic antibodies (40%). These are related to vasculitis and systemic autoimmune diseases, whose identification and treatment are important for prognosis. It has further been described that one of every four patients surviving IPH is subsequently diagnosed with an autoimmune disease. Differential diagnoses and possible diagnostic aids are presented in Table 1.

Table 1. Differential diagnoses.

DIFFERENTIAL DIAGNOSES		
System or indication	Disorders found	Tests
Pulmonary	▪ Bronchopulmonary dysplasia ▪ Primary ciliary dyskinesia ▪ Bronchiectasis ▪ Cystic fibrosis ▪ Chronic aspiration ▪ Gastroesophageal reflux disease ▪ Diffuse alveolar injury	▪ X-ray or chest CT ▪ Bronchoscopy with bronchoalveolar lavage (BAL) to measure the percentage of hemosiderophages, lipid-laden macrophages and differential cells.
Cardiovascular	▪ Pulmonary hypertension ▪ Congenital heart disease ▪ Myocarditis ▪ Pulmonary vascular congestion ▪ Mitral stenosis ▪ Congestive heart failure ▪ Veno-occlusive disorders ▪ Hemangiomas ▪ Vasculitis	▪ Echocardiogram ▪ Immunological studies
Hematology	▪ Thrombocytopenia ▪ Acquired or congenital coagulopathies ▪ Disseminated intravascular coagulation	▪ Blood count ▪ Clotting times ▪ Reticulocyte count
Gastrointestinal	▪ Celiac Disease	▪ Transglutaminase immunoglobulin A and E
Immunological	▪ Heiner syndrome ▪ Wegener's granulomatosis ▪ Tuberous sclerosis ▪ Lymphangiomatosis or lymphangioleiomyomatosis ▪ Pulmonary-renal syndrome ▪ Systemic lupus erythematosus ▪ Goodpasture syndrome	▪ Test for milk cow protein allergy ▪ Immunoglobulin ▪ Anti-gliadin antibodies (Abs) ▪ Anti-endomysial antibodies (EMA) ▪ Anti-nuclear antibody ▪ Anti-dsDNA antibodies ▪ Anti-smooth muscle antibody ▪ Rheumatoid factor ▪ Anti-neutrophil cytoplasmic antibody ▪ Anti-glomerular basement membrane ▪ Complement ▪ Immune complex
Infectious	▪ Pulmonary or systemic infections	▪ Chest X-ray ▪ Cultures for bacteria, fungi and viruses in BAL ▪ Blood cultures
Physical abuse	▪ Suspected in repeated or unexplained trauma	▪ X-ray of long bones, rib and column grid

Source: Adapted from Taytard *et al.* and from 'Acute idiopathic pulmonary hemorrhage among infants. Recommendations from the working group for investigation and surveillance' (2,5).

Treatment

Management during the acute phase is supportive with mechanical ventilation, packed red blood cell transfusion if anemia is present, hemodynamic support and intravenous steroids. Methylprednisolone at a dose of 1 mg/kg every 6 hours or bolus in the first 3 to 5 days is used depending on the severity and the response. Then, prednisolone 1mg/kg/day is given; continuation or discontinuation is based on evolution. This medication can be used on outpatient daily, or with monthly boluses, according to clinical and laboratory findings. Steroids have shown to decrease the risk of pulmonary fibrosis (7). In life-threatening cases or with severe diffuse alveolar hemorrhage, which does not improve with intravenous steroids, the use of intrapulmonary recombinant factor VIIa has been described with good results in small case series (15).

In chronic cases with poor response to steroids due to steroid resistance or dependency, immunosuppressive agents such as azathioprine, hydroxychloroquine, methotrexate and cyclophosphamide have been used with variable results (7).

The outcome in these diseases may result in death; for example, in the Cleveland series, 5 out of 30 children died and in France, 2 out of 25 died during the acute phase of the disease. In the last series, the average follow-up time was 5.5 years, with good results in pulmonary function in 23 of them.

Patients in our series had a satisfactory clinical evolution: extubation was achieved early. One of them was discharged with oxygen by nasal cannula, and the other with oral steroid. During follow-up, the first case was assessed at 39 months of age and was healthy; the second case is unknown.

CONCLUSIONS

Acute idiopathic pulmonary hemorrhage is a condition that seriously threatens life, therefore, it is important to have a high index of suspicion in infants under one year of age, who were previously healthy, with a sudden onset of airway bleeding related to severe respiratory distress, with high mechanical ventilation requirement and chest x-ray infiltrates. Management with intravenous steroids allows successful bleeding resolution in most cases, so its administration must be timely. For this, the necessary tests must be performed to rule out other causes of diffuse alveolar hemorrhage whose treatment is different. Early diagnosis of this disease, which threatens life, allows adequate therapeutic approach, reducing its morbidity and mortality.

CONFLICT OF INTERESTS

None stated by the authors.

FUNDING

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CASE REPORTS
REPORTES DE CASO

NEVUS LIPOMATOSUS SUPERFICIALIS IN SACROCOCCYGEAL REGION. CASE REPORT IN AN INFANT

Palabras clave: Nevo; Lipoma; Hamartoma; Lipomatosis; Neoplasias cutáneas.
Keywords: Nevus, Lipoma, Hamartoma, Lipomatosis, Skin Neoplasms.

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ABSTRACT

Nevus lipomatosus superficialis is a benign tumor characterized by the presence of mature adipocytes located in the superficial layer of the dermis; its management is surgical and most of the time is reported in adults.

This case presents a malformation from birth in the sacrococcygeal region over the lateral line to the right, which is why, after obtaining imaging studies to rule out an associated hidden dysraphism, surgery was decided for total resection of the mass, without complications. Finally, the pathology report diagnoses nevus lipomatosus superficialis, which constitutes a rare and unusual case based on the differential diagnoses and presentation.

INTRODUCTION

Nevus lipomatosus superficialis is a benign lesion characterized by the presence of mature adipocytes in unencapsulated subcutaneous tissue **(1,2)**, which is relevant when differentiating this lesion from lipoma.**(2)**. Some authors consider it as a subtype of hamartoma, since its proliferation is associated with the presence of blood vessels in the dermis **(3)**.

This lesion was first described in 1921 by Hoffman & Zurhelle as an adipocyte malformation that appeared in several clustered nodes **(3)**, which could have a zosteriform arrangement. In 1950, Nikolowsky described a variant of the classical form **(3)** as a single pedunculated nodular lesion, also known as lipofriboma **(3)**.This paper presents the case of a patient who developed the second type of the lesion; since this is rarely described in children, a histological examination of the resected mass was necessary to differentiate it from other similar lesions such as lipomas and focal dermal hipopalsia, among others.

CLINICAL CASE

Female patient, 29 days old, from Bogotá, referred by her pediatrician due to a pedicle lesion in the sacrococcygeal region and no other history of importance. Since the lesion appeared in the midline, an MRI in the spine was performed, which discarded the presence of hidden dysraphism (Figure 1). During pregnancy, prenatal ultrasounds were normal, and the mother of the patient had no complications and did not require additional drug treatment; at the time of assessment, the mass had increased in size, but did not generate associated symptoms.

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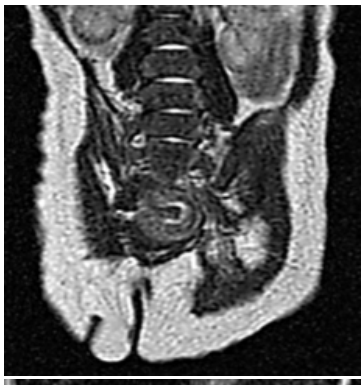

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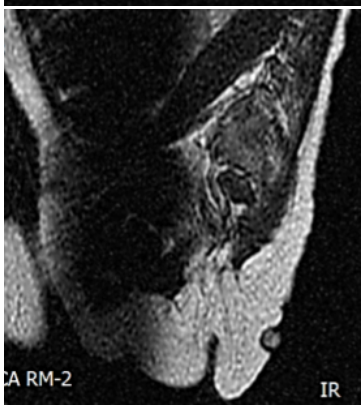

Fig 1. MRI of the lumbosacral spine.
A. Sagittal section: right paracoccygeal mass with broad based pedicle.
B. Coronal section: coccygeal region mass not connected to spinal cord
Source: Own elaboration based on the data obtained in the study.

Physical examination showed a well-defined mass in the previously described region, painless, without alterations in color, without renitency point or active secretion (Figure 2). A resection was scheduled in order to continue with histopathological studies.

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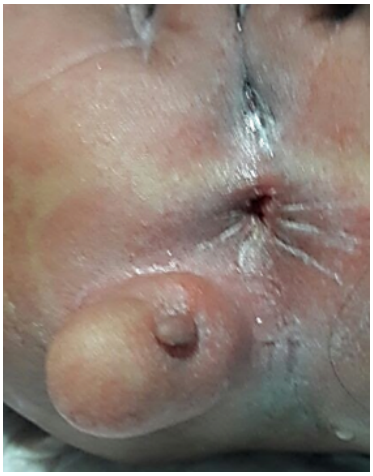

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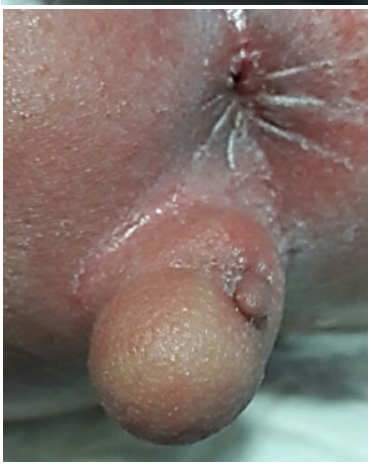
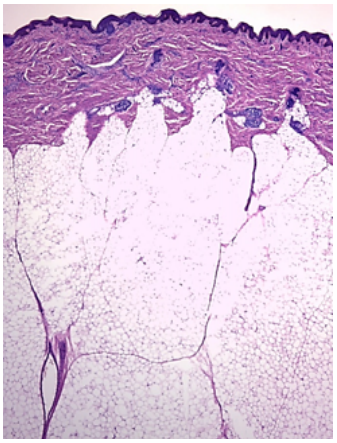


Fig 2. Mass of the patient at the time of the first consultation.
Source: Own elaboration based on the data obtained in the study.

The procedure was conducted two weeks after finding the unencapsulated, non-vascularized lipomatous mass. Since it presented a broad based pedicle, a skin plasty with simple skin flaps was necessary after completing the resection procedure; the mass was sent to pathology, which reported a mature adipose tissue of lobular appearance that completely occupied and expanded the superficial dermis with some fibrocollagenous interstitial tissue bands. The diagnosis of nevus lipomatosus superficialis or Hoffmann-Zurhelle was made based on these characteristics (Figure 3). Postoperatively, the patient had adequate cicatrization and no complications (Figure 4); during a follow-up at five months, no relapse or other alterations associated with surgery were reported.

A



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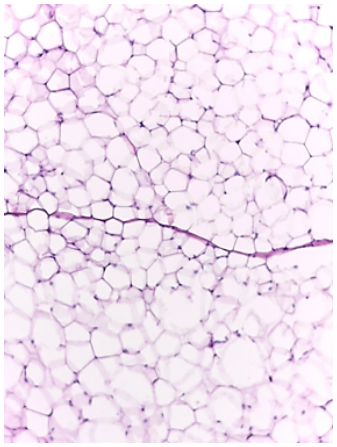

Fig 3. Histology of the resected mass.
A. Multiple partitioned lobes of lipomatous cells in epidermis.
B. Collagen band which groups lipomatous cells.
Source: Own elaboration based on the data obtained in the study.

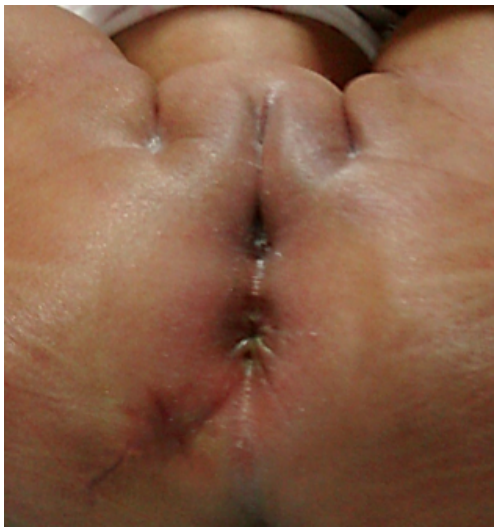

Fig 4. Tenth day after resection of the mass.
Source: Own elaboration based on the data obtained in the study.

DISCUSSION

The onset at birth of nevus lipomatosus superficialis and its location were noteworthy since, usually, they are a sign of hidden dysraphism in newborns **(4)**; given these specifications, using diagnostic imaging was necessary. The characteristics of the mass allowed a simple surgical approach and resection; it is important to highlight the size of the mass. Regarding the patient, a skin plasty had to be used to provide the best possible healing.

In general, lipomatous lesions in children are rare and their incidence is less than 10% **(3)**, while adults may show more than 50% **(3)**; among these lesions, nevus lipomatosus superficialis represents 2% **(3)**. The literature includes few reports of this entity not only because if its low incidence, but because it is underdiagnosed.

The pedicled lipofibroma differs from the classical form described by Hoffmann & Zurhelle **(3)**, which may appear at any site of the body and is unique, while histological features remain the same **(5)**. Differential diagnoses include focal dermal hypoplasia that has a distribution of collagen beams different from that of the lipomatous nevus, is horizontal simulating a scar and presents with other congenital malformations **(5)**. Segmental neurofibromatosis, which has similar lesions associated with light brown spots, is also included **(2)**.

Another differential diagnosis is hidden spinal dysraphism, which causes defects in the fusion of the neural tube and causes meningeal and spinal deformations, which lead to a progressive neurological deficit that can become severe **(4)**. Along with several entities such as tethered cord syndrome, diastematomyelia, meningoceles and myelomeningocele **(4)**, this condition is characterized by a cover skin lesion and associated skin disorders in the sacrococcygeal region on the midline, including dimples and dermal sinuses, connective nevi, localized hypertrichosis, hemangiomas, dyschromic lesions, lipomas, among others **(4)**. Based on these characteristics, this is the first differential diagnosis to consider when facing malformations in this location.

In conclusion, nevus lipomatosus superficialis has a low incidence in the pediatric population, is poorly described and its diagnosis can be only made through histopathology; treatment is surgical and consists of total resection. In this case, the unique pediculated variant was presented, which is less frequent than the classic one, in which, due to its location, other pathologies associated with the nervous system had to be considered in order to rule out serious neurological diseases. These diseases could be studied through imaging during the first weeks of the patient's life, so this report not only stands out because of the infrequency of the underlying pathology but also because of the differential diagnoses that had to be suspected.

CONFLICTS OF INTEREST

None stated by the authors.

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To Dr. Mariam Rolón, pathologist at Fundación Santa Fe de Bogotá.

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PRIMARY HEPATIC LEIOMYOSARCOMA. A CASE REPORT

Palabras clave: Leiomiosarcoma; Sarcoma; Neoplasias hepáticas; Hepatectomía; Actina de músculo liso.
Keywords: Leiomyosarcoma; Sarcoma; Hepatic neoplasms; Hepatectomy; Smooth muscle actin.

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ABSTRACT

Primary hepatic leiomyosarcoma is extremely rare among cases of liver tumors in adults, with an incidence of 0.1 and 1%. This paper describes the case of a 55 year-old man with a clinical evolution of five months consisting of abdominal pain, a large hard lump, weight loss, shortness of breath and fever.

A three-phase computed tomography (CT) showed a hypercaptive mass at its periphery during hypodense arterial phase at its center, located in segments V and VI, without a plane of separation of the liver. Due to the symptoms, the patient underwent an exploratory laparotomy, finding a cerebroid mass of 40 x 40 cm; a lumpectomy without hepatectomy was performed, leaving free surgical margins.

The diagnosis was largely made through a histopathological assessment, finding stromal multinucleated pleomorphic forms, desmin (+), SMA (smooth muscle actin) and MSA (muscle specific actin) (+), Ki67 (+) and negative for S100 (protein S100) and CD117 antibody, which confirmed the high grade pleomorphic leiomyosarcoma diagnosis. The patient was discharged 16 days after admission once his condition improved, and was referred to the oncology department for adjuvant chemotherapy.

Given the size of the mass, the prognosis was bleak, which left surgery as the only option to offer survival expectations through regulated or "atypical" hepatectomies along with safety margins and liver transplantation. With this in mind, the first option was chosen; six months after surgery, with clinical improvement and adjuvant therapy, the patient, still with unfavorable prognosis, remained stable attending multidisciplinary medical management controls.

INTRODUCTION

Primary hepatic leiomyosarcoma (PHL) is an extremely rare tumor. In 2011, the English literature reported less than 50 patients with this type of tumor **(1,2)**. To date, in the context of this study, there are no reports of similar cases in Ecuador, and by 2014, there were very few cases reported in Latin America **(3)**.

In adults, primary liver sarcomas are a group of rare tumors, representing between 0.1 and 1% of all existing malignant liver tumors in this population **(4)**. Usually, they develop in the uterus, the retroperitoneum, genital organs, lungs, liver and large vessels **(5)**; on the other hand, PHL generate in the smooth muscle cells of intrahepatic, bile ducts or round ligament vascular structures **(6)**.

Their unusualness, image manifestations and non-specific clinical presentation make early diagnosis difficult **(7)**. Currently, diagnosis can be made before surgery through cytology or image-guided percutaneous biopsy or after surgery **(8-12)**. However, the differential diagnosis between primitive or metastatic liver sarcoma may present difficulties sometimes **(9)**, reason why the anatomopathological study is fundamental. The histopathological diagnosis is characterized by the presence of diffuse infiltrates and uniform spindle-shaped cells with hyperchromatic nuclei **(8,9)**, while a positive reaction for desmin, SMA, MSA, Ki67 and a negative reaction for S100 and CD117 are observed in immunohistochemistry.

Resective surgery is the best treatment option for this condition; the same surgical principles of soft tissue sarcoma surgery should be kept, with margin cancer liver resection as the most appropriate choice or gold standard. Nevertheless, due to the advanced stage of the disease at diagnosis, lumpectomy or enucleation,

followed by treatment with adjuvant chemotherapy, may be another therapeutic approach to consider, even in cases with metastases **(10)**, leaving liver transplant as the last resort.

CLINICAL CASE

Male patient, 55 years old, professional working at an office with no relevant medical history, who denied using or consuming alcohol and tobacco, and reported a family history of diabetes *mellitus*, esophageal cancer and ocular melanoma. The clinical picture of the patient presented five months of evolution characterized by oppressive abdominal pain of moderate intensity, located in the right upper quadrant and umbilical region, accompanied by a rigid, hard abdominal mass of about 20 cm in diameter, painful on palpation (Figure 1), hyporexia and weight loss of about 45 pounds in 100 days, dyspnea on moderate efforts, afternoon fever one month prior to hospitalization, and stable vital signs.

Mild normocytic hypochromic anemia (hemoglobin: 10.1mg / dl), normal hepatic and cholestatic function, normal coagulation times and acute phase reactants without alterations were observed through tests. Also, normal results of tumor markers such as CEA, alpha-fetoprotein, CA 125, CA 15-3, CA 19-9 and CA 72-4 were obtained. Abdominal ultrasound showed a hypoechoic and vascularized tumor mass in the liver bottom of approximately 30 x 20 cm (Figure 2). Moreover, plain abdominal radiograph showed a radiopaque image in the hypochondria and right flank. Three-phase computed tomography angiography of the

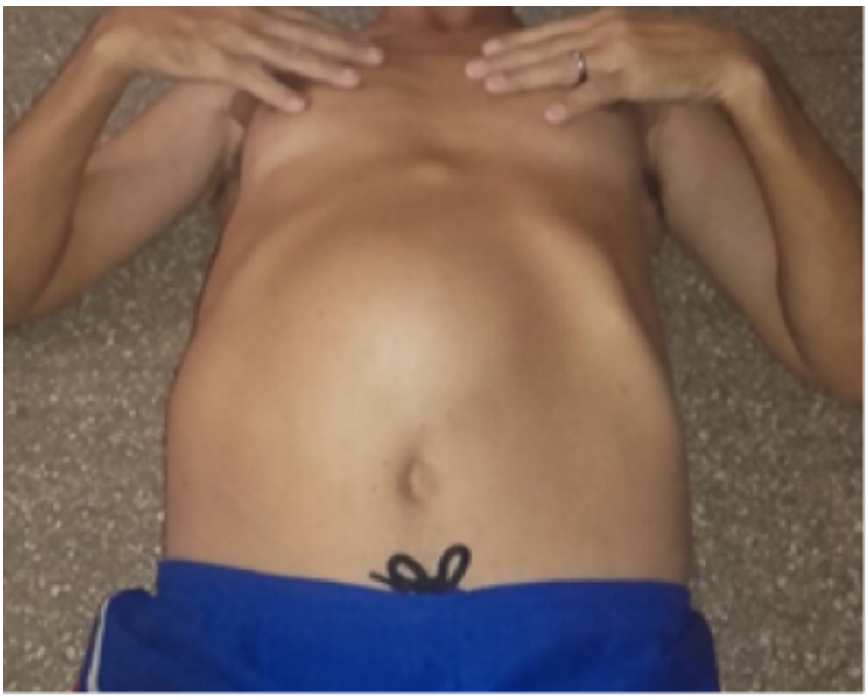

Fig 1. Patient's morphology. Large tumor mass in the right abdomen.
Source: Own elaboration based on the data obtained in the study.

abdomen showed a hypercaptive mass on its periphery, hypodense center in heterogeneous portal and late arterial phase, located in liver segments V and VI, without a plane of separation of the liver, in addition to retroperitoneal adenomegaly (Figure 3).

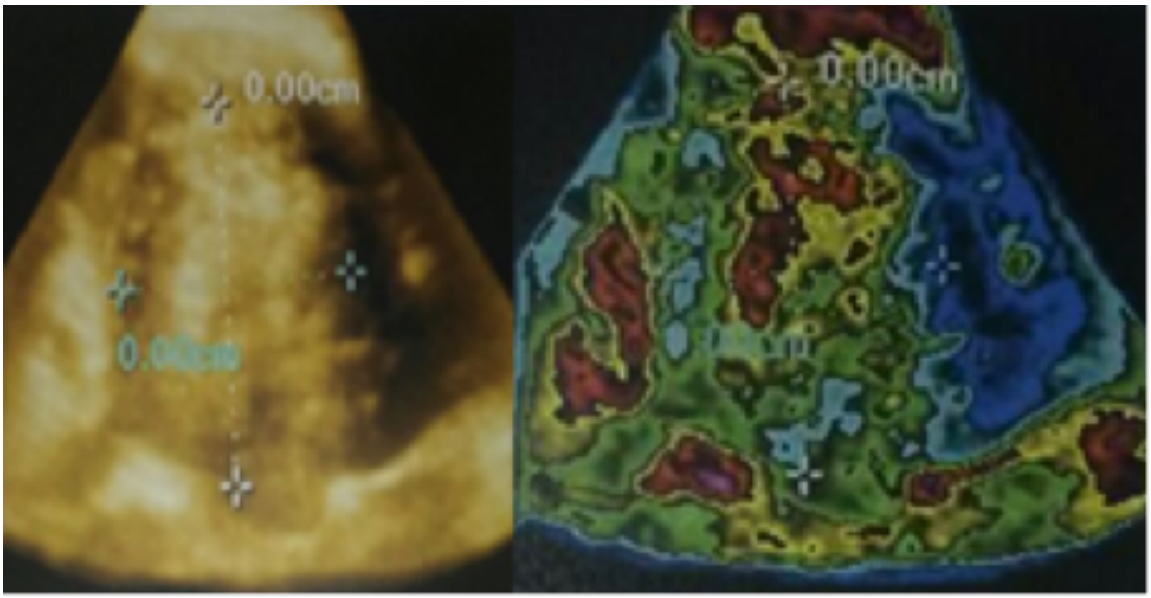

Fig 2. 2D abdominal ultrasound. Large, hypoechoic liver mass with vascularization.
Source: Own elaboration based on the data obtained in the study.

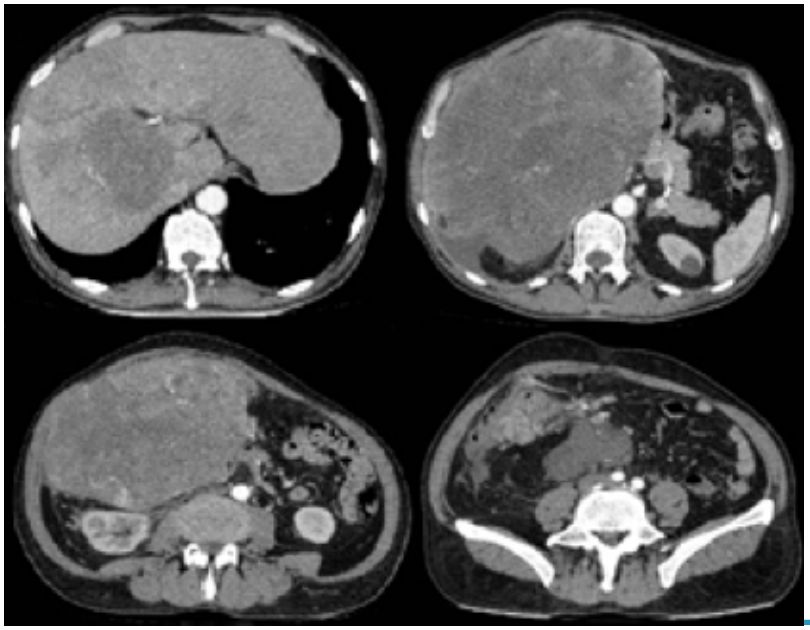

Fig 3. Three-phase tomography angiography. Large tumor mass in the right hepatic lobe, segments V and VI, which moves intra-abdominal organs.
Source: Own elaboration based on the data obtained in the study.

Finally, colonoscopy revealed normal-looking mucosa and decreased light at the ascending colon due to an apparent extrinsic compression. Exploratory laparotomy was performed to improve the clinical condition of the patient, finding a large, encapsulated, cerebroid tumor in liver bed, segments V and VI, of about 40x40 cm, displacing the right diaphragm and surrounding abdominal structures. A lumpectomy without hepatectomy was performed, with free surgical margins in sample freeze; also, neighboring structures were released and no masses, lymph nodes or metastatic seeding were identified with the naked eye. No intraoperative surgical complications occurred (Figure 4).

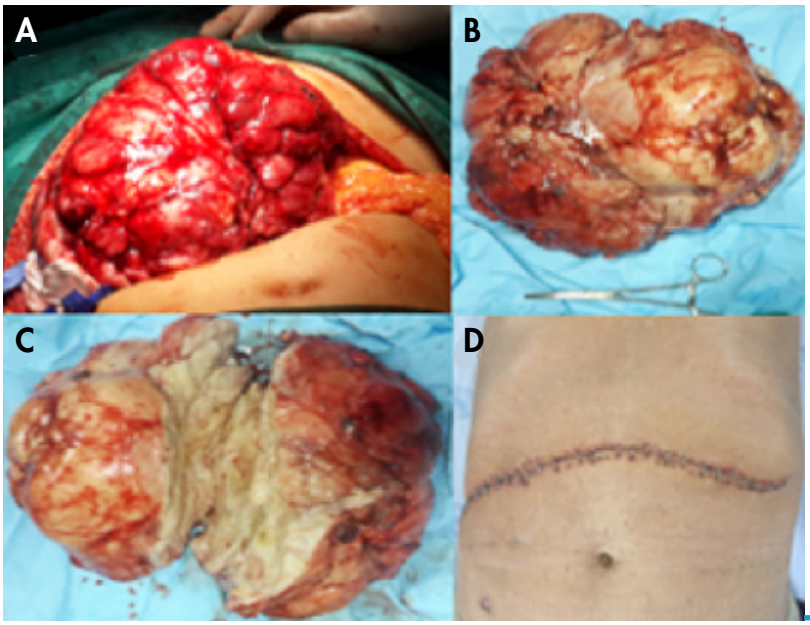

Fig 4. Visualization during exploratory laparotomy of primary, multilobulated, cerebroid, encapsulated hepatic leiomyosarcoma located in the right hepatic lobe.
Figure 4a. Tumoration dependent on hepatic segments V and VI.
Figure 4b. Tumor exposed once extracted from the liver.
Figure 4c. Longitudinal section of the tumor showing the macroscopic structure.
Figure 4d. Extended subcostal wound closure used in this surgical procedure.

Anatomopathological study (Figure 5) reported a malignant multilobulated tumor with the following characteristics: weight 4341 g; size 38 x 35 x 20cm; encapsulated, with irregular edges and cavitated; mesenchymal strain composed of spindle cells; with elongated core and numerous mitoses. Meanwhile, the immunohistochemical study showed positive desmin, positive SMA and MSA, positive Ki67 in 80% of cells and negative for S100 and CD117, confirming the diagnosis of high-grade pleomorphic leiomyosarcoma.

The patient was hospitalized for 16 days; after showing a favorable evolution, he was discharged and ordered monthly outpatient follow-up. He was also referred to the oncology department to initiate adjuvant chemotherapy every 25 days in three cycles, observing the clinical course and conforming to the requirements, with the possibility of subsequent radiotherapy to control clinical, imaging and laboratory parameters, which is currently pending. Six months after surgery, still with unfavorable prognosis, the patient was stable and attending multidisciplinary medical controls.

DISCUSSION

Clinical manifestations of PHL are not specific and, usually, tumors are asymptomatic

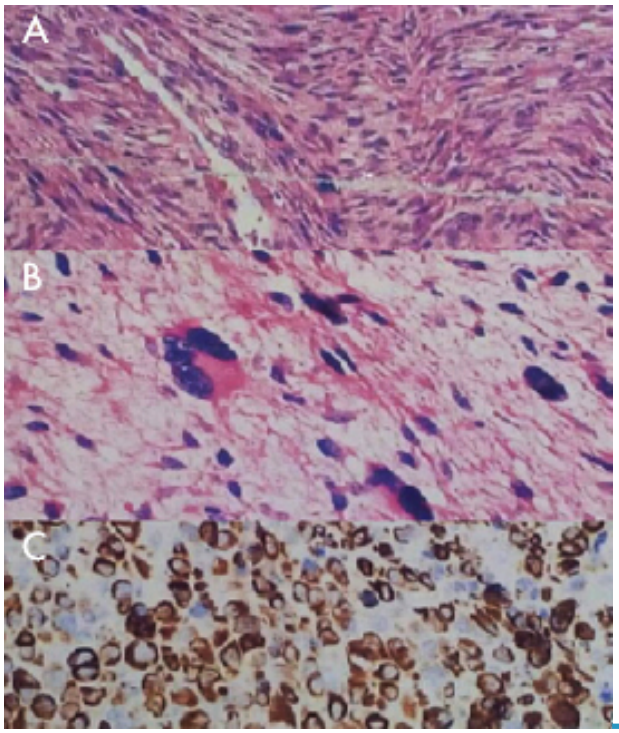

Fig 5. Histological and immunohistochemical study.
5.a. Fusiform muscle malignant cells with elongated core and necrosis areas, typical in this kind of tumor, are observed (staining with hematoxylin-eosin x100).
5.b. Presence of numerous mitosis, interwoven fascicles and pink cytoplasm with numerous pleomorphic multinucleated forms (staining with H&E x400).
5.c Immunohistochemistry reveals positive desmin sample with presence of typical smooth muscle cells (original magnification: x50).

before enlargement **(11,12)**. The average age of onset is between 40 and 50, with extreme ages of 22 and 77 **(13)**. The most common symptom is mild to severe pain in the upper abdomen, accompanied by weight loss, afternoon fever and asthenia **(14)**. Clinical examination allows identifying palpable abdominal mass or hepatomegaly in right-epigastric hypochondriac region. In general, non-specific increases in some biochemical function/liver damage parameters can be observed (bilirubin, alkaline phosphatase or transaminases) unlike specific tumor markers, as in this case, which does occur in sarcomas in other parts of the body.

On the other hand, imaging does not provide specific data **(16,17)**, because ultrasound shows a hypoechoic liver tumor, while three-phase CT shows a well-defined, heterogeneous, hypodense or isodense mass through central necrosis areas and peripheral enhancement, or as a thick wall cystic mass **(18)**, with an angiographic pattern of peripheral avascular mass or important pathological neovascularization **(19)**, superimposed on any liver tumor. Finally, through nuclear magnetic resonance, a heterogeneous area with hypointense lesion on T1 and hyperintense lesion on T2, with possible encapsulation, is observed **(2)**.

The most common site of tumor is the right lobe and metastases of about 40% at the time of diagnosis are common **(6)**, which is consistent with data reported here. Therefore, the differential diagnosis must be made between various types of benign and malignant hepatic solid tumors **(20,21)** such as hepatocarcinomas of different strain **(22)**, primitive or metastatic sarcoma **(8)**, and even sarcomas of the retrohepatic vena cava **(23)**.

Today, and in most case series **(20,24,25)**, the diagnosis of hepatic sarcoma can be established preoperatively, through image-guided percutaneous biopsy or cytology; however, if the liver damage appears to be malignant and

is considered resectable, diagnosis is made postoperatively, as in this case. Histopathological diagnosis shows four types of leiomyosarcomas: well differentiated, moderately differentiated, poorly differentiated and myxoid leiomyosarcoma **(26)**. In this case, the patient was classified as type 1, since immunohistochemical study showed positive desmin and SMA, but negative S-100, CD117 and NSE **(18,19)**, which is consistent with the parameters of this type of tumor and confirms PHL diagnosis.

As this was a large liver mass that generated large compression of neighboring structures and dyspnea, as well as peripheral vascularization, the possibility of image-guided percutaneous biopsy was discarded due to high risk of bleeding. Regulated or atypical hepatectomy with safety margins was the selected treatment. However, and due to the advanced evolutionary stage of the disease at diagnosis, enucleation followed by chemotherapy was an alternative treatment, which can also be used in metastases cases **(8)**; for this patient, this was the best option *a priori*, thus constituting an exceptional case with apparent absence of metastasis and free surgical margins.

Since this is the only treatment that allows prolonged survival expectations, its analysis reveals the main favorable prognostic factors: being younger than 50 years of age, early diagnosis with a size below 5 cm, tumor location, radical surgery with safety margins, adjuvant treatment with chemotherapy and, as a last resort, liver transplantation **(9)**. King *et al.* **(27)** describe cases with large tumors, which after five years had a survival rate of 18%, as well as cases with about 80% of survival at five years in the presence smaller tumors with clear margins. Also, Gates *et al.* **(28)** indicate that the combination of surgery with chemotherapy offers a median survival of 3.3 years.

PHL can present hematogenous metastases, mainly in the lung, followed by lymphatic and peritoneal paths. In this regard, Shivathirthan *et al.* **(29)** describe that the intermediate range in the identification of metastases between primary leiomyosarcoma and PHL was 29 months (range: 6-58 months). They also note that inoperability criteria may include extrahepatic spread of tumor, diffuse intrahepatic tumor and impaired liver function **(29)**.

The patient in this case has received three cycles of chemotherapy with ifosfamide and doxorubicin; he has also attended monthly clinical, laboratory and imaging follow-ups. However, the role of adjuvant therapy with chemotherapy/radiotherapy is not well defined yet because, despite the fact that chemotherapy with doxorubicin and ifosfamide shows a slow course of the disease and may prolong survival in resections with R1 stage, there is not sufficient evidence in unresectable tumors and metastases by PHL **(30)**.

The role of liver transplantation is still controversial, since it has low rates of survival and recurrence 95% before six months **(31)**.

CONCLUSIONS

PHL is an extremely rare tumor that, in most cases, is diagnosed in advanced stages, delaying treatment and worsening prognosis. Its finding should be suspected in patients with large tumor masses. However, despite having many advanced imaging studies, diagnosis is completely histopathological, whereas treatment is surgical, in most cases, depending on several factors. This case highlights surgical therapy and the rare diagnosis of this tumor.

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CONFLICTS OF INTEREST

None stated by the authors

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CASE REPORTS
REPORTES DE CASO

TRANSVAGINAL EVISCERATION AFTER ABDOMINAL HYSTERECTOMY. CASE REPORT

Palabras clave: Prolapso visceral; Histerectomía; Colostomía.
Keywords: Visceral Prolapse; Hysterectomy; Colostomy.

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ABSTRACT

Evisceration is a condition in which abdominal viscera protrude through an unnatural hole, with an incidence between 0.03 and 4.1%. This condition often occurs after an abdominal hysterectomy (47%), vaginal hysterectomy (29.4%) or laparoscopic approach (23.6%). It has the highest incidence in hysterectomized postmenopausal women, while the time interval between surgery and complication onset may vary from a few days to a few years. Moreover, in most cases, the eviscerated organ is the small intestine, which represents a surgical emergency. Transvaginal evisceration is a rare entity and is scarcely documented; the case of a 81-year-old patient with a history of abdominal hysterectomy, who attended the medical service after eight hours of evolution of a clinical picture characterized by pelvic pain and ejection of the large intestine (sigmoid colon) through the vaginal canal is presented here. The patient underwent an exploratory laparotomy, Hartmann colostomy, rectopexy to the promontory and restitution of traffic in a subsequent procedure; after presenting a satisfactory evolution, she was discharged.

INTRODUCTION

Intestinal transvaginal evisceration after hysterectomy is an unusual situation and little known by health professionals, as evidenced in the few cases published to date, hence the uniqueness and importance of the case reported here.

With an incidence between 0.03 and 4.1% (1), evisceration is a condition in which abdominal viscera protrude through an unnatural hole, leaving them exposed to external agents. In 2009, Partsinevelos *et al.* (2) published a review of 51 cases which found that the highest percentage of cases occurred after abdominal hysterectomy (47%), followed by vaginal hysterectomy (29.4%) and laparoscopic procedure (23.6%). Meanwhile, a study conducted in 2012 indicates a higher incidence of vaginal cuff dehiscence after undergoing laparoscopic hysterectomy (1.14%) than after undergoing abdominal (0.10%) and vaginal (0.14%) hysterectomy (3).

When analyzing the different laparoscopic approaches to hysterectomy, it is possible to observe that the incidence of evisceration is higher in total laparoscopic hysterectomy than in vaginal hysterectomy by laparoscopy; on the other hand, for laparoscopic hysterectomy, intracorporeal suturing of the vaginal cuff yields better results than vaginal suture (4).

The predisposing factors for this condition include pregnancy, pelvic surgery, multiparity, neuropathy, obesity, menopause and smoking, in addition to factors associated with the race of the patient (5); the time elapsed between surgery and complication onset may vary from 1 day to 25 years (6). The study required for diagnosis is clinical and is evaluated according to the symptoms presented, which consist of fullness sensation or perception of a foreign body in the vagina, and presence of a soft bulky mass, which often presents along with urinal symptoms.

Physical examination is key; if total prolapse is not evident, the patient is requested to push once to observe the protrusion of the organ (6,7,8). Vaginal evisceration is a surgical emergency that can be handled by vaginal or abdominal way, or both depending on the case.

CLINICAL CASE

81-year-old patient with a medical history of hypertension and diabetes mellitus. Surgical history includes abdominal hysterectomy plus colporrhaphy performed twice; the first

one was done due to genital prolapse two years before admission, and the second, four months before admission. Obstetric history includes four vaginal deliveries.

The patient attended medical consultation with a clinical picture of eight hours of evolution characterized by severe pain in the lower abdomen which radiated to the genital region, and by ejection of the sigmoid intestine through the vagina after performing Valsalva maneuver. Physical examination showed the following signs: blood pressure 150/80 mmHg; pulse 95/min; respiratory rate 22/min; temperature 36 °C, and saturation 98%.

The patient presented with dry oral mucosa, soft, pitting, painful superficial and deep palpation in the lower abdomen, mild pain, decreased bowel sounds, and no signs of peritoneal irritation. The mass protruding from the vagina, and corresponding to a segment of the large intestine (sigmoid) with discoloration, can be observed in Figure 1.

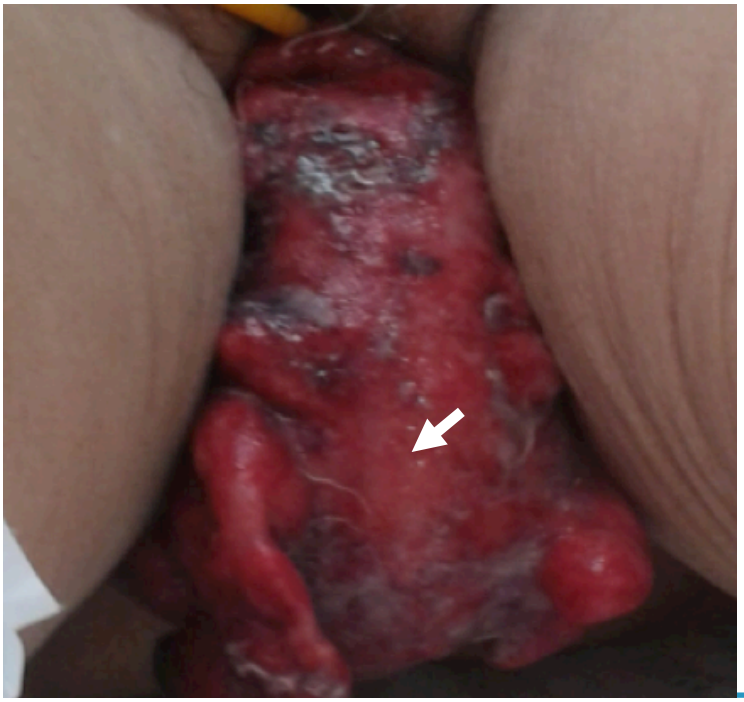

Figure 1. Sigmoid ejection through the vaginal orifice with color changes (arrow).
Source: Own elaboration based on the data obtained in the study.

The following results were obtained in the laboratory tests: blood count- leukocytes: 16,310/mm^3; neutrophils: 88.2%; hemoglobin: 12.5 g/dL; hematocrit: 38.4%; pH: 7.2 PCO_2:27.5 mmHg; HCO3:16.6 mEq/L; Na: 132 mEq/L; K: 3.9 mEq/L; Cl: 98 mEq/L; PCR: 8.99 mg/L; 3.0 mmol/L lactate. On the other hand, hydro-aerial levels were found in abdominal radiography. Diagnosis on admission: intestinal transvaginal evisceration.

The patient underwent an exploratory laparotomy and manual reduction of hernia. The most important findings were the presence of a sigmoid hernia ejected through vaginal orifice with color changes (Figure 2 and 3). A resection of approximately 20 cm in the devitalized sigmoid colon segment, Hartmann colostomy formation, with primary raffia in vaginal cuff without mesh placement, and rectopexy of the endopelvic fascia anterior to rectum were performed.

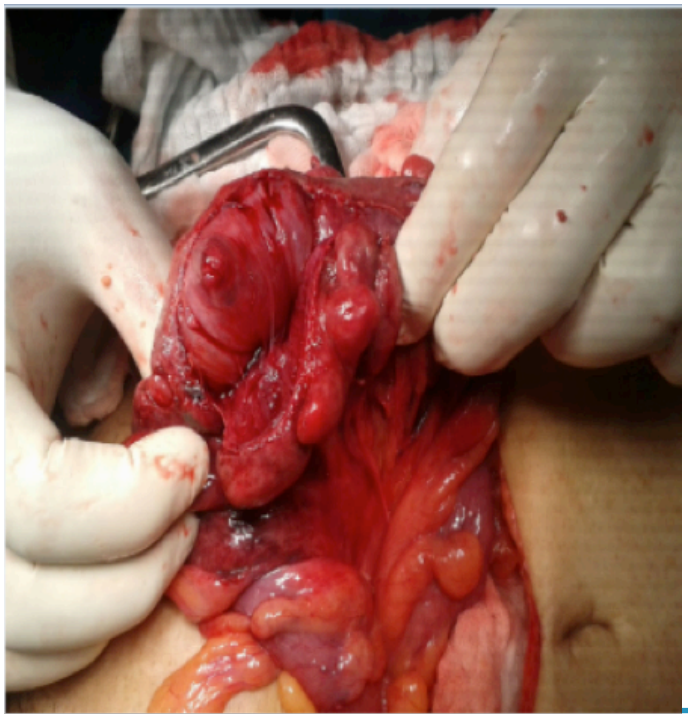

Figure 2. Sigmoid colon segment reduced, with color changes (arrow).
Source: Own elaboration based on the data obtained in the study.

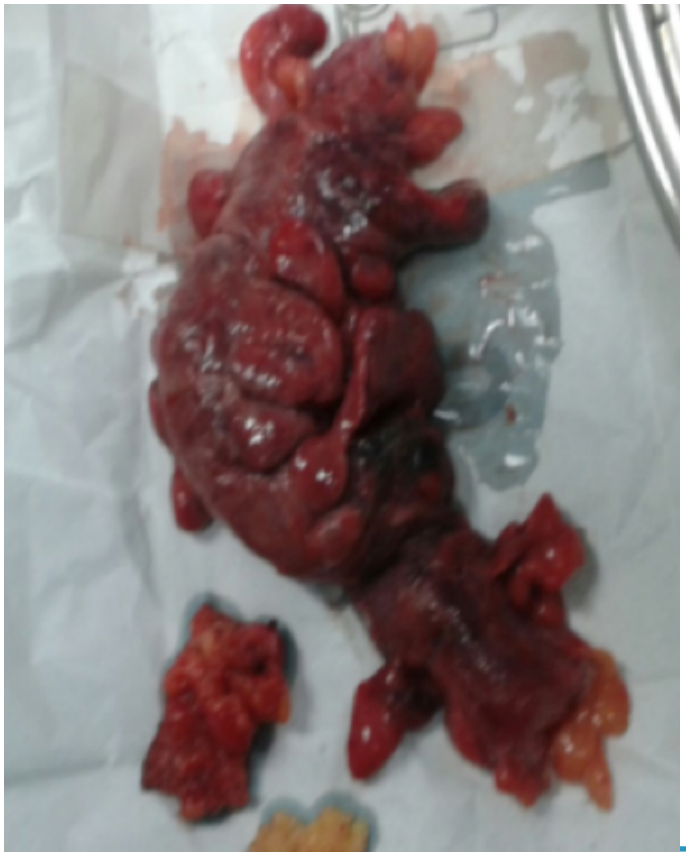

Figure 3. Resected segment of sigmoid colon.
Source: Own elaboration based on the data obtained in the study.

The patient was discharged 10 days after surgery, in good general condition. Six months after surgery, successful return of intestinal transit was performed.

DISCUSSION

Evisceration is a rarely reported complication; the terminal ileum is the organ that is primarily involved. Notwithstanding, there are cases that report omentum, fallopian tubes, appendix and ovarian cyst ejection (3.4). This case reports evisceration of the sigmoid colon.

The main factor associated with intestinal evisceration through vaginal cuff is vaginal, abdominal or laparoscopic hysterectomy, as it causes a shortening of the round and broad ligaments, leaving them without support, besides of cutting and leaving, with or without support, the uterosacral and cardinal ligaments. The loss of these supports relaxes the pelvic diaphragm and the perineal membrane (2,3,4).

Other associated factors include hypo-estrogenic state, which produces atrophy of the vaginal cuff; poor surgical technique; postsurgical infection or hematoma; intercourse before complete healing; Valsalva maneuvers; age; smoking; chronic treatment with corticosteroids; radiotherapy, and obesity. The time elapsed between surgery and the onset of complication varies as reported in the literature, and may be from 1 day to 25 years after surgery (6); in this case, 20 years passed.

The vaginal cuff prolapse is classified into three degrees (3):

- Grade I: does not reach the introitus
- Grade II: reaches introitus
- Grade III: goes beyond introitus.

The clinical picture is characterized by vaginal or pelvic pain, protrusion of tissue or lump through introitus, mass protruding through the vagina, back pain, urinary incontinence, and vaginal bleeding.

Surgery aims to reduce the intestine, the eventual resection of devitalized intestinal loops, and vaginal defect repair using stitches with non-absorbable material or by placing a non-absorbable polypropylene mesh (8). The approach may be vaginal, abdominal, or combined, depending on the conditions of the patient and bowel viability at the time of treatment (8,9).

According to the literature, the combined approach is considered as the most appropriate, especially when intestinal ischemia is suspected; it is also recommended for the proper evaluation and effective repair of the tissues involved, while repairing the pelvic floor during surgery (11). While other authors defend repair in a subsequent procedure (6), infection is reduced with the use of this approach, although surgical and anesthetic risk increase; however, treatment depends on the patient's hemodynamic status and pollution in the moment.

Nevertheless, other authors prefer primary repair (8-10,12). An effective technique for pelvic floor repair is McCall modified culdoplasty to prevent recurrence of enterocele; obliteration of recto-uterine pouch may also be useful (Moschowitz procedure) (8,10,12). In many cases, simple closure of the vaginal cuff is sufficient, but a non-resorbable polytetrafluoroethylene or polypropylene mesh could be placed to reinforce the suture.

In the case reported here, resection of the devitalized sigmoid segment was performed, followed by Hartman colostomy and sacra rectopexy in order to resolve the emergency, as well as associated comorbid factors. The restitution of intestinal transit was carried out in a subsequent procedure six months after resection.

CONCLUSION

Transvaginal evisceration is a highly unknown complication with low incidence in hysterectomised patients caused by the laxity of the tissues; therefore, early diagnosis and treatment are essential for preventing associated morbidity and mortality.

Proper treatment of pelvic dysfunctions and gynecological surgeries with a refined technique help avoiding, to a large extent, the onset of this condition and its impact on the organs of the abdominal cavity.

AUTHORSHIP CONTRIBUTION

The lead author and co-authors were involved during the entire process of this case report.

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CONFLICTS OF INTEREST

None stated by the authors.

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