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Management of the hemophilic pseudotumor of the abdomen: A rare pathological entity

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ABSTRACT

INTRODUCTION: Hemophilic pseudotumor is a rare complication that occurs in patients with severe hemophilia. Results from multiple episodes of bleeding into the bones and soft tissues.
PRESENTATION OF CASE: A 31 years old male patient, with severe hemophilia A. Diagnosed with an abdominal tumor 10 years ago during routine screening, that progressively grew to encompass the entire abdominal area, with symptoms of intestinal obstruction.
DISCUSSION: Hemophilic pseudotumor appears as a painless tumor of slow growth that can compress vital organs producing bone destruction, muscle and skin necrosis. The tumor may have fistulas or break spontaneously.
CONCLUSION: The abdominal hemophilic pseudotumor is a rare pathological entity, with few reports worldwide, but must be considered in hemophilic patients with a well documented abdominal tumor.

1. Introduction

Hemophilia A and B are linked to disturbances caused by a deficiency of factors VIII and IX, respectively.[1] Hemophilia A is a congenital disease transmitted by the X chromosome with a recessive trait, characterized by a deficiency in the production of factor VIII.[2] The incidence of Hemophilia A is 10–20 cases per 100,000 people.[3] Clinical manifestations vary depending on the severity of the disease. A rare complication is the hemophilic pseudotumor, also known as hemophilic cyst.[4] The hemophilic pseudotumor affects 1–2% of patients with a severe disease,[5] frequently associated with a traumatic injury. The hemophilic pseudotumor develops from repeated episodes of bleeding, either from fracture sites or bleeding or subperiosteal hemorrhage of any soft tissue.[1] The inadequate reabsorbed blood becomes an encapsulated area of blood and necrotic tissue. These lesions grow over time, causing symptoms of compression.[6] The interior of the pseudotumor consists of blood products at different stages of development, surrounded by a fibrous capsule containing hemosiderin-laden macrophages.[5] It appears as a painless tumor of slow growth that can compress key organs causing bone destruction, muscle and skin necrosis. The tumor may have fistulas or break spontaneously.[7]

There are very few cases reported in the literature involving the abdomen. Most studies describe those involving the musculoskeletal system. The objective of case presentation is to describe a patient with a hemophilic pseudotumor of the abdomen, and to review the literature on hemophilic pseudotumor.

2. Clinical case

This is a case of a 31 year old male, diagnosed with severe hemophilia A at 2 years of age, treated with factor VII 2000 IU weekly since, whit a family history of 2 cousins with hemophilia (no other specified). In September 2011, he underwent drainage of a hematoma of the left pelvic limb. In October 2011 he underwent an embolization of a bleeding blood vessel on the lower left limb. Repeat hemarthrosis of the left hip with avascular necrosis of the femur. Blood transfusion on multiple occasions. In August 2012 had a hematoma in the left thigh surgically removed. In September 2012 presented upper gastrointestinal bleeding requiring hospitalization with remission of the symptoms.

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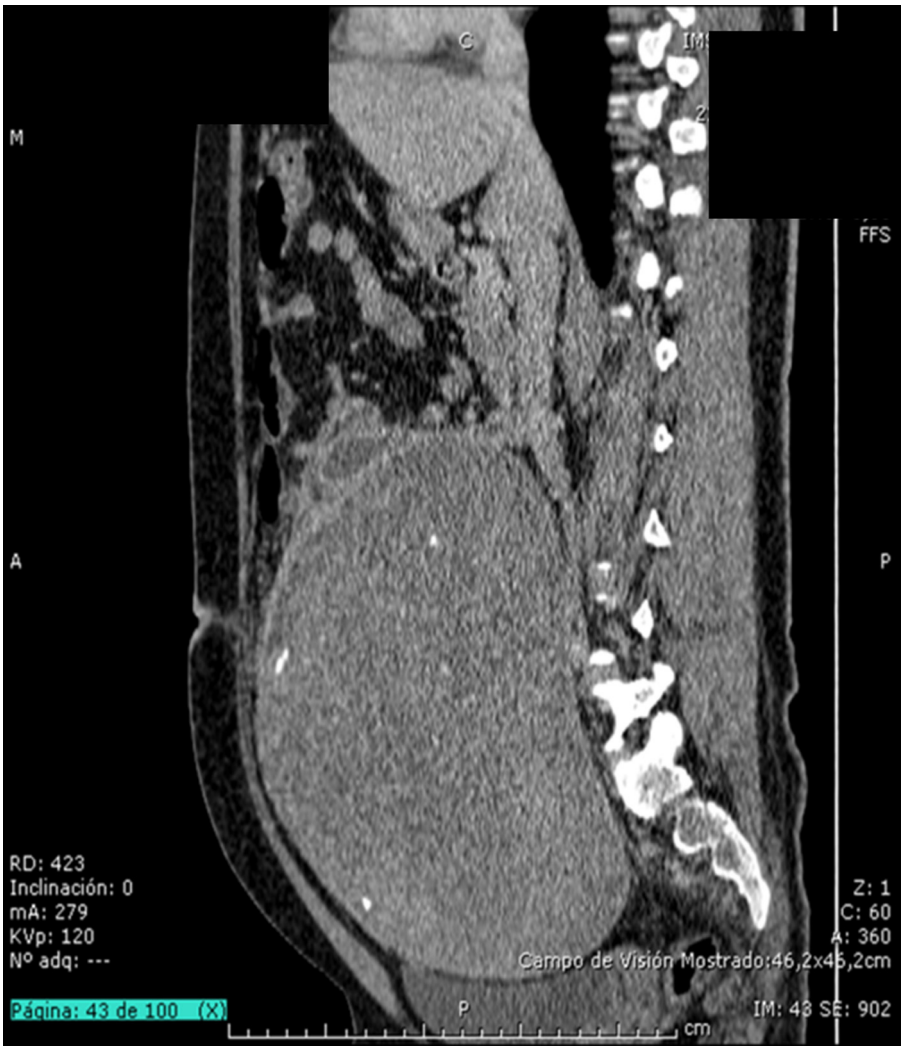

Fig. 1. CT scan of the lesion. Coronal plane.

2.1. Present illness

An abdominal tumor diagnosed 10 years ago during routine screening that progressively grew to encompass the entire abdominal area, with no associated symptoms. During his last hospital admission, hematology referred him to general surgery due to bowel obstruction symptoms, and to evaluate surgical treatment of a pelvic tumor referred as a possible teratoma. A CT scan of the abdomen (Figs. 1 and 2) was performed, showing a tumor in the pelvis with clear borders, 241 mm × 192 mm of diameter, from the pelvis and projected into the bladder, rectum and adjacent structures, including the kidneys. The lesion had irregular wall thickness, with a density of 195 Houndsfield units in its interior, with heterogeneous enhancement and vascularity present within; with tomographic diagnosis of abdominal pelvic teratoma.

It was programmed for elective surgical removal. Surgical procedure was performed on October 2012, an exploratory laparotomy with no complications.

The patient underwent surgery with surgical findings that included a cyst-like tumor of 40 cm × 30 cm displacing loops of small bowel, colon and bladder, with adhesions to the great omentum, small intestine, descending colon and bladder. Macroscopically appeared as an old, organized, and calcified clots with a volume of 5000 ml. Histopathological report of hematic material with hemosiderin-laden macrophages.

3. Discussion

The hemophilic pseudotumor of the abdomen is a rare but often disabling condition, potentially fatal in patients with severe hemophilia.[8] Diagnosing a hemophilic pseudotumor with invasive techniques such as, aspiration and biopsy are not advisable due to increased risk of complications (hemorrhage, infection).[5] Imaging techniques of which MRI is preferred, allows recognition of blood products in various stages of evolution. Ultrasonography (USG) shows a central anechoic region with increased echoes behind the lesion due to enclosed fluid in the pseudotumor. Computed tomography (CT) identifies the thick pseudocapsule, but cannot differentiate a hematoma from a chronic abscess. CT is particularly helpful in the evaluation of bone, whereas MRI is superior to CT for delineating soft tissue.[9]

At the moment, surgical excision of pseudotumor is the preferred treatment by many authors. However, there are instances where surgical extraction of the lesion is not feasible. In such situations, radiotherapy and arterial embolization should be considered either alone or in adjunction to surgery.[6] The decision to

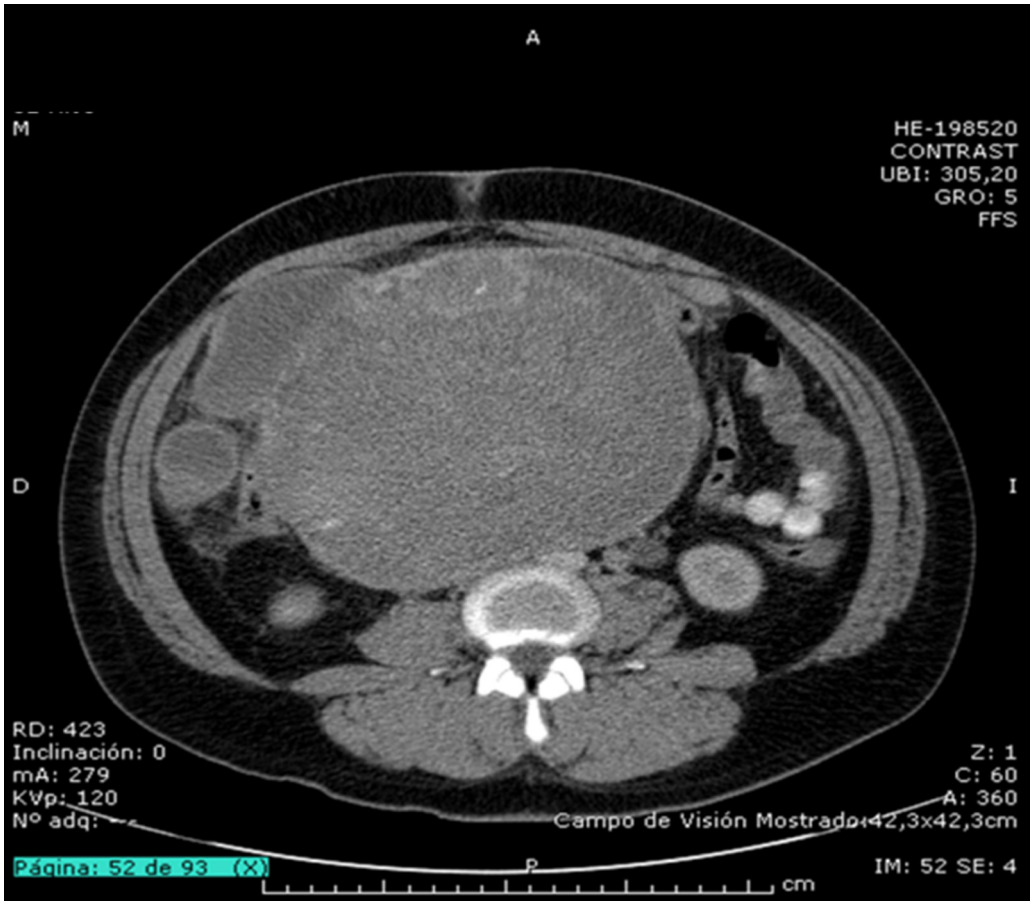

Fig. 2. CT scan of the abdomen. Sagittal plane.

operate on this patient was made based on the gradual increase in the tumor size, the symptoms that the displayed by the patient. In this patient, the first surgical intervention was performed without major intraoperative complications. This type of procedure is associated with complications such as bleeding, bowel perforation, and damage to nearby structures due to firm vascular, and nervous adhesions. Surgical resection after performing arterial embolization to reduce the vascularization of the pseudotumor is a good alternative, thereby reducing the size of the pseudotumor and the risk of bleeding complications during surgery, at best about 2 weeks prior to surgery. This time lapse will allow for mass shrinkage but is insufficient for vessel restoration.[10,11] It is a rare pathological entity, but it must be considered in the hemophilic patient with a long-standing abdominal tumor and trauma history, to plan appropriate treatment.

4. Conclusions

The abdominal hemophilic pseudotumor is a rare pathological entity, with few reports worldwide, but must be considered in hemophilic patients with a well documented abdominal tumor, as a complication of the hematologic disease. Signs and symptoms of compression should be evaluated by CT or MRI because its early diagnosis is crucial for evaluation and proper surgical planning, and treatment in conjunction with other specialties.

Conflict of interest statement

None of the authors have conflict of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and case series and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

The study conception and design, as well as writing the manuscript works were all equally distributed among all the authors.

Key learning points

- Hemophilic pseudotumor etiology.
- Hemophilic pseudotumor management.

References

1. Pakala A, Thomas J, Comp P. Hemophilic pseudotumor: a case report and review of literature. *Int J Clin Med* 2012;**3**:229–33.
2. Mathew M, Goel G, Kurien A. Digital pseudotumor associated with Von Willebrand disease. *Internet J Hematol* 2009;**5**:2.
3. Van der Bom JG, ter Avest P, Van den Berg HM, Psaty BM, Weiss NS. Assessment of incidence of inhibitors in patients with haemophilia. *Haemophilia* 2009;**15**:707–11.
4. Garcia-Perez R, Torres-Salmeron G, Sánchez-Bueno F, Garcia-Lopez A, Parrilla-Paricio P. Intraabdominal hemophilic pseudotumor: case report. *Rev Esp Enferm Dig* 2008;**100**:275–80.
5. Stafford JM, James TT, Allen AM, Dixon LR. Hemophilic pseudotumor: radiologic–pathologic correlation. *Radiographics* 2003;**23**:852–6.
6. Espandar R, Heidari P, Rodriguez-Merchan C. Management of haemophilic pseudotumors with special emphasis on radiotherapy and arterial embolization. *Haemophilia* 2009;**15**:448–57.
7. Uriza LF, Berdugo A, Partija RT, Blanco G. Pseudotumor hemofílico. Reporte de caso. *Univ Méd* 2006;**48**:71–4.
8. Rodriguez-Merchan EC. Haemophilic cysts (pseudotumours). *Haemophilia* 2002;**8**:393–401.
9. George H, Marvin SG, Abdelwahab IF. Hemophilia: evaluation of musculoskeletal involvement with CT, sonography, and MR imaging. *Am J Roentgenol* 1992;**158**:119–23.
10. Rodriguez-Merchan EC, Jimenez-Yuste V. The role of selective angiographic embolization of the musculo-skeletal system in haemophilia. *Haemophilia* 2009;**15**:864–8.
11. Ahuja SP, Sidonio Jr R, Raj AB, Bertolone SJ, Silverman C, Antekeier DP, et al. Successful combination therapy of a proximal haemophilic pseudotumour with surgery, radiation and embolization in a child with mild haemophilia A. *Haemophilia* 2007;**13**:209–12.