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Case Report

Hemophilic Pelvic Pseudotumor: Imaging and Management

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Abstract

Hemophilic pseudotumor, although rare, can pose a significant and destructive complication in patients with hemophilia. While its incidence has been decreasing, we present a case involving a pelvic localization, highlighting the imaging characteristics observed in different imaging techniques, as well as our hybrid treatment approach. Given the non-specific radiological appearance, maintaining a high level of clinical suspicion is crucial for accurate diagnosis. Currently, surgical excision remains the preferred treatment for pseudotumors. However, there are situations where surgical removal may not be feasible. In such cases, a multidisciplinary team should carefully consider alternative approaches such as arterial embolization and radiotherapy. These interventions can be employed either as standalone treatments or as adjuncts to surgery, particularly in cases involving large pseudotumors located in the pelvic region. Endovascular embolization plays a pivotal role in reducing the size of the pseudotumor and minimizing the risk of bleeding complications during surgery.

Keywords: Arterial Embolization; Haemophilia; Pseudotumour Imaging

Introduction

Hemophilia-associated Pseudotumors (HPT) are infrequent but consequential occurrences in individuals with hemophilia A or B. Initially reported by Stalker in 1918, HPTs represent a rare source of morbidity and mortality within this patient population. Despite the increased utilization of recombinant factor concentrates in recent decades, the prevalence of HPT ranges from 1% to 2% among patients with moderate or severe hemophilia, with a notable history of trauma observed in 57-75% of cases [1]. HPTs arise as a result of recurrent bleeding episodes, manifesting as encapsulated hematomas at sites of bone fractures and soft tissue hemorrhages. Inadequate hematoma reabsorption leads to the formation of necrotic tissue surrounded by a fibrous capsule containing hemosiderin-laden macrophages [2]. This process culminates in a slow, painless growth that may exert pressure on adjacent organs, incite bone and muscle deterioration, and induce cutaneous necrosis [2].

While most reported cases primarily affect the musculoskeletal system, instances of abdominal involvement are exceedingly rare [2]. Radiological manifestations in the axial skeleton usually consist of lytic areas and new bone formation with calcifications in the adjacent soft tissues [3]. There are few descriptions in the literature of intra-abdominal hematomas without skeletal involvement, so we will detail their characteristics in CT and MRI as well as the embolization technique in our interventional vascular radiology unit in a patient with a large HPT.

Case Report

A 27-year-old male patient from Venezuela was diagnosed with hemophilia A at the age of two following a cephalohematoma. The patient did not receive inhibitor treatment in his home country. Genetic analysis revealed a mutation in exon 23 of the factor VIII gene: C. 6469 A>G p.Asn2157Asp. The patient presented to the emergency department of our hospital with left iliac

fossa abdominal pain and syncope at home. No fever or other accompanying symptoms were reported. Laboratory analysis revealed the following data (with reference values in brackets):

- Hemoglobin: 10.0 g/dL [13.0 - 18.0]
- Hematocrit: 30.6% [41.0 - 50.0]
- Platelets: 581.0 x 10^9/L [130.0 - 450.0]
- Prothrombin time (ratio): 1.45 [0.85 - 1.2]
- Activated partial thromboplastin time (ratio): 1.4 [0.81 - 1.3]
- APTT correction (ratio): 1.14 [0.96 - 1.3]
- Thrombin time (ratio): 1.0 [0.96 - 1.25]
- Fibrinogen: 822.0 mg/dL [170.0 - 470.0]
- Coagulative factor VIII: 60.0% [54.0 - 155.0]
- Chromogenic factor VIII: 72.0% [54.0 - 155.0]

A Computed Tomography (CT) scan revealed a large hematoma in the Retzius space (Figure 1) extending cranially towards the left iliopsoas muscle (Figures 2 & 3). Within the hematoma, extravasation of contrast in the portal phase was observed in the left suprapubic region, indicating active bleeding possibly originating from the left obturator artery. Hemostatic treatment with recombinant factor VIII (FVIII) was initiated, and the patient was transferred to the interventional vascular radiology unit. Pelvic arteriography and selective catheterization of the external iliac and left hypogastric arteries were performed, revealing a small point of contrast extravasation from an obturator branch of the inferior epigastric artery (Figure 4).

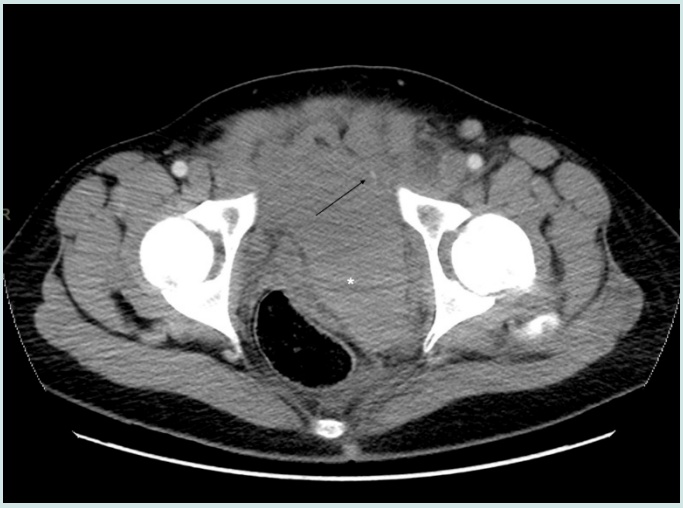

Figure 1: Emergency CT scan showing a great hematoma (asterisk) and a minimal contrast extravasation of contrast (black arrow) during portal phase in relation to active bleeding.

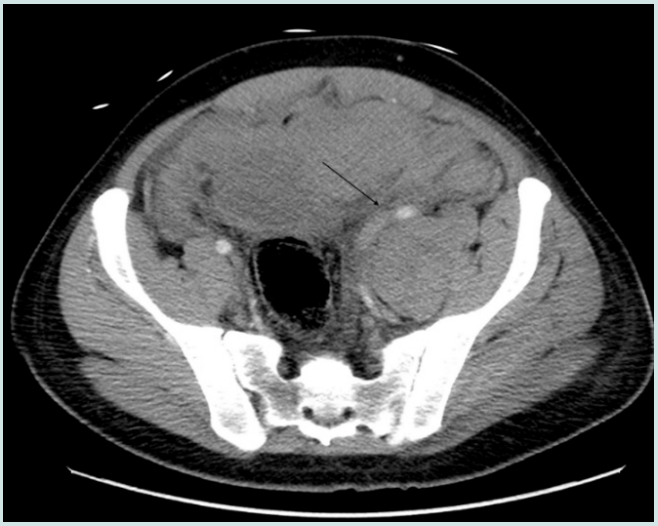

Figure 2: The same CT scan in a craneal image were we appreciate the extension towards the psoas muscle and the displacement of the iliac vessels (black arrow).

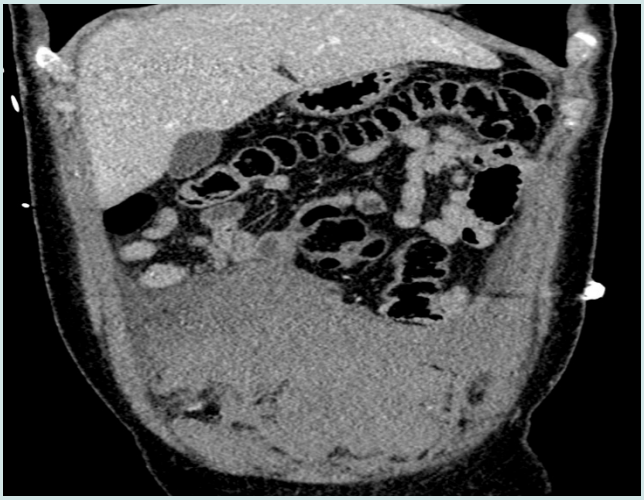

Figure 3: CT coronal oblique reconstruction.

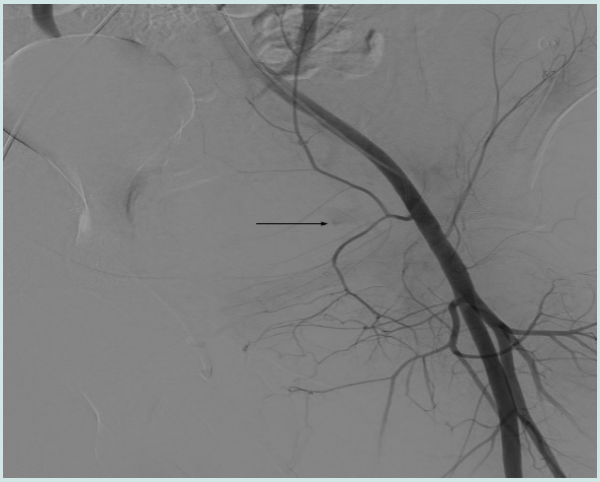

Figure 4: selective left hypogastric arteriography. A very small point of contrast extravasation is observed (black arrow), probably from a little obturator branch.

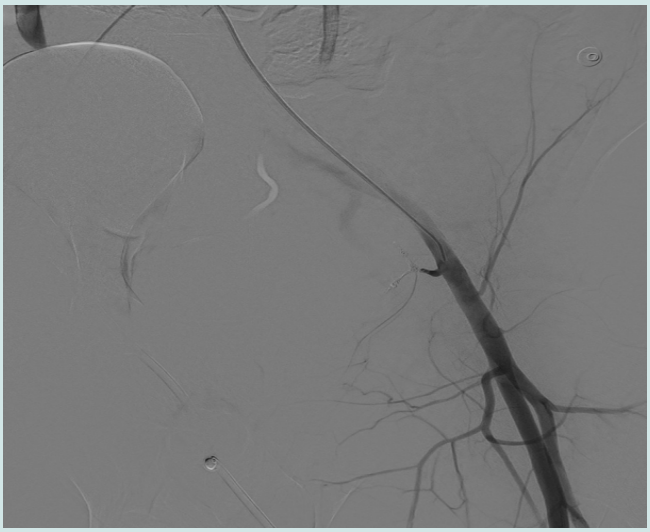

Figure 5: Final result after coil embolization of both obturator branch and inferior epigastric artery.

Considering the findings from the CT scan and angiography, embolization was performed using 2x3x20 mm Tornado coils (Cook Medical, Limerik, Ireland) in both the obturator and inferior epigastric arteries. Additionally, a 5 mm x 15 cm Concerto coil (ev3 Medtronic, the Netherlands) was used to embolize the common trunk of both arteries (Figure 5). Following the procedure, replacement therapy with recombinant factor VIII was continued, a packed red blood cell transfusion was administered, and empirical antibiotic therapy was initiated to prevent superinfection of the hematoma.

During the hospital stay, a follow-up CT scan was conducted to assess the evolution of the hematoma and its compressive effects on the abdomen, revealing only reactive lymph nodes. Magnetic Resonance Imaging (MRI) showed compression and displacement of the bladder to the right, extending into the left internal iliac chain and compressing the lumbar psoas and iliopsoas muscles. The lesion exhibited hypointense signal with a hyperintense peripheral halo on T1-weighted sequences (Figure 6), heterogeneous and hyperintense signal on T2-weighted sequences (Figure 7), restricted diffusion, and minimal peripheral enhancement in the dynamic contrast-enhanced study.

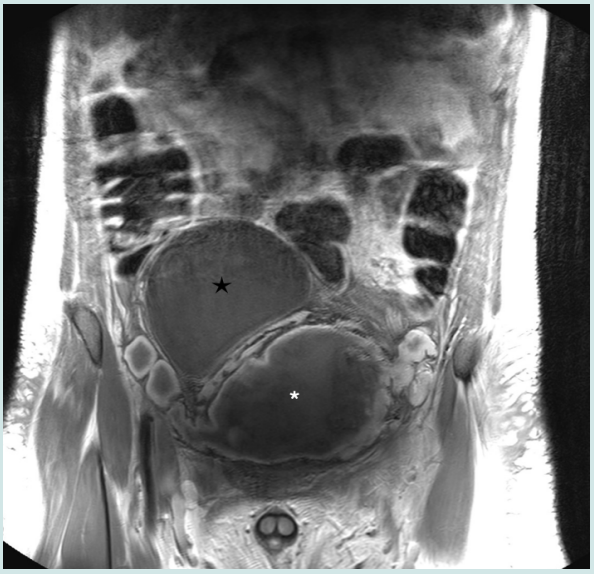

Figure 6: MRI image on T1-weighted sequences. Pseudotumor (asterisk) is hypointense with a hyperintense peripheral halo in relation to the capsule. The great displacement of the bladder is striking (star).

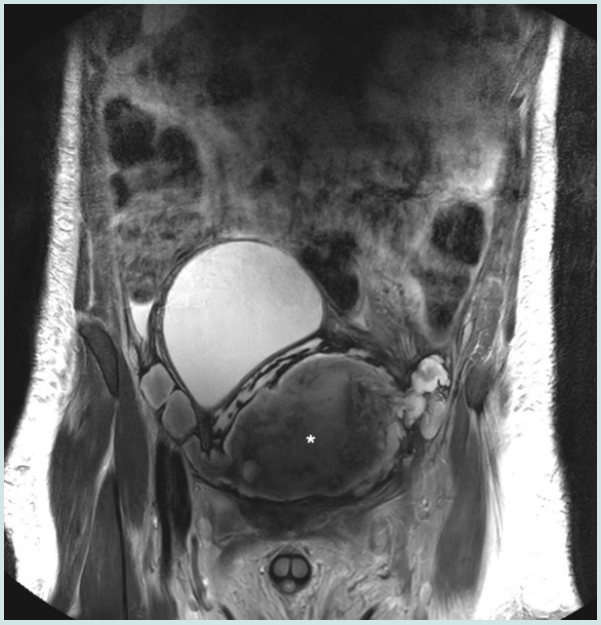

Figure 7: Coronal T2-weighted image were the lesion is now heterogeneous and hypointense (asterisk).

Bilateral hypervascularized retroperitoneal and iliac lymph nodes were also observed on the MRI, with some lymph nodes exhibiting hemorrhagic necrosis in the Retzius space and iliac regions. Differential diagnoses considered included neoplastic processes such as pheochromocytoma or paraganglioma, lymphoproliferative or infectious diseases, in addition to the hemophilic pseudotumor. After ruling out pheochromocytoma and germinal tumor, a decision was made in collaboration with the general surgery service to perform a biopsy/evacuation of the hematoma. During the intervention (Figure 8), excision of multiple lymph nodes was performed, and the hematoma was punctured, releasing old bloody material. A portion of the lesion (Figure 9) was sent to the pathological anatomy department for analysis, which revealed fibrosis and a xanthogranulomatous reaction in the tumor wall and iliac lymph nodes, along with hemosiderophages and giant cells consistent with fragments of a hemophilic pseudotumor.

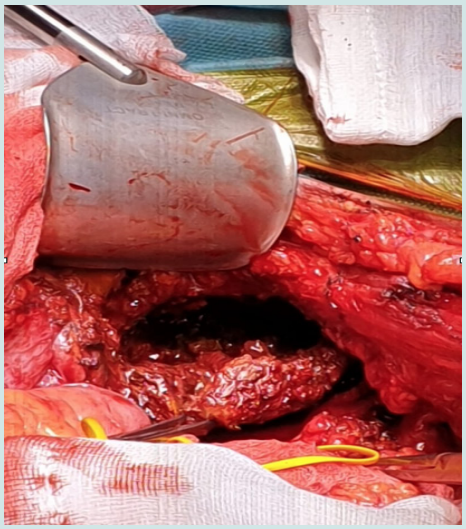

Figure 8: surgical median laparotomy access to Retzius space. The tumor is clearly observed.

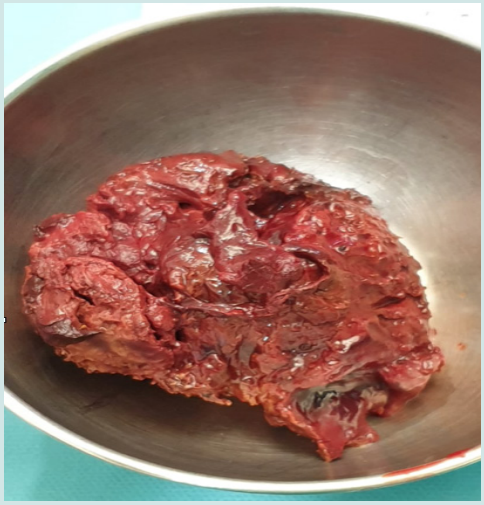

Figure 9: Part of the tumor erased. Its was hard to the touch and with multiple adhesions to the surrounding tissues.

In a multidisciplinary session involving the general surgery, vascular radiology, and urology services, it was decided to perform a second pre-surgical embolization to minimize the risk of bleeding during a subsequent intervention aimed at complete resection, scheduled for 10 days later. Seven days after the initial intervention, with FVIII levels above 30%, the patient experienced symptoms related to the psoas hematoma, confirmed by CT scan to have increased in size, although no active bleeding was observed. Another pelvic and selective arteriography of the left internal and external iliac arteries was performed. Recurrence of patency in the left inferior epigastric artery was noted, leading to embolization of its branches using 400-micron microspheres (LifePearl, Microvention, Terumo Europe, France) and multiple 2x5x50 mm Tornado coils (Cook Medical, Limerik, Ireland).

To precisely identify the feeding vessels of the pseudotumor capsule, C-arm cone-beam CT imaging was performed (Figure 10)

using an angiographic system (Allura Clarity, Philips Healthcare, Best, The Netherlands). Aberrant arteries with pathological uptake, likely responsible for the vascularization of the pseudotumor capsule (Figure 11) and shared vascularization with vesical and prostatic arteries, were observed (Figure 12). Embolization was carried out by infusing 400-micron microspheres (LifePearl, Microvention, Terumo Europe, France) into the areas with the greatest vascularization, and embolization with 2x3x20 mm and 2x4x40 mm Tornado coils (Cook Medical, Limerik, Ireland) was performed in the arteries sharing vascularization with the bladder.

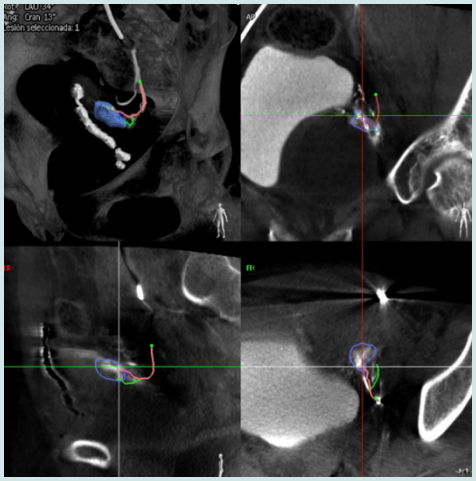

Figure 10: expert-CT tool after the cone-beam CT show us with colors the nutritional vessels of the tumor.

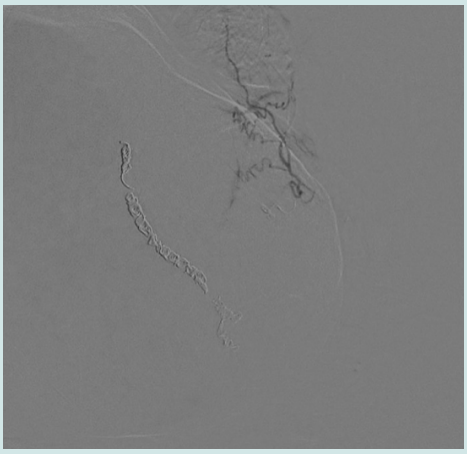

Figure 11: angiography demonstrating arteries feeding the pseudotumor capsule, in this case branches of the pudendal artery.

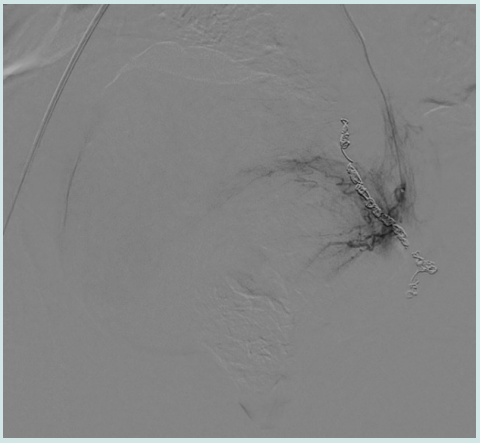

Figure 12: in super selective arteriographies some branches seems to share vascularization with vesical arteries.

The angiographic outcome demonstrated successful flow blockage in the treated arteries without any evidence of complications. A subsequent MRI, seven days after the second intervention, revealed a reduction in size compared to the previous CT scan for both the tumor in the Retzius space and the collection in the left lumbar psoas. Both lesions were well-defined and displayed the previously described signal characteristics. The lesion was subsequently excised through a median laparotomy approach. Adhesions between the anterior face of the bladder and the pseudotumor capsule, as well as the pubis, were observed. The release of the capsule from the pseudotumor was challenging, and during manipulation, the capsule was opened, releasing old blood. Complete excision of the lesion was performed. The postoperative period was uneventful, and the patient was discharged nine days after the intervention.

Discussion

Abdominal pseudotumors are rare but significant conditions that can have disabling and potentially life-threatening effects in patients with severe or moderate hemophilia [1-4]. Although they have also been reported in patients without hemophilia, such cases are uncommon [5]. Inadequate replacement treatment can contribute to the development of hemophilic pseudotumors, as observed in our patient who received suboptimal treatment for many years in his home country.

These pseudotumors are characterized as slow-growing masses with well-defined boundaries, containing clots in various stages of evolution and surrounded by a delicate capsule. They can be located intraosseously, subperiosteally, or within soft tissues, categorized as intramuscular or extramuscular [4,6-8]. Most cases documented in the literature predominantly affect the musculoskeletal system. Abdominal involvement is much rarer, and to our knowledge, all published cases thus far have been located in the iliac bones and iliopsoas muscles [7,8]. Nevertheless, pseudotumors can occur in any part of the body, and the symptoms and severity depend not only on their location and size but also on the structures they impinge upon [6]. As they expand, these masses can encompass adjacent structures such as nerves, blood vessels, and ureters. Although these structures become incorporated into the pseudotumor, they are not invaded; however, they can undergo pressure necrosis, including bone tissue [6].

Typically, patients exhibit a slow and stable disease progression until they experience sudden complications resulting from bleeding, as seen in the case we describe, or from other causes such as nerve or vascular compression, ureteral obstruction, intestinal obstruction, fistulization to the skin or large bowel, infection, or pathological fractures [4,6,7,9]. On Computed Tomography (CT), HPT appears as a low-density mass (10-35 Hounsfield units) that may contain calcifications, surrounded by a fibrous capsule [7]. While CT can visualize the pseudocapsule, it cannot differentiate a hematoma from a chronic abscess. It is valuable for assessing the extent of involvement of bones, soft tissues, and vascular or nervous structures [6,9].

Magnetic Resonance Imaging (MRI) is not specific for HPT but typically shows soft tissue masses with variable signal intensities on T1- and T2-weighted sequences, reflecting clots at different stages of evolution, with a hypointense capsule observed more clearly on T1- and T2-weighted images. Mural nodules may be visible in soft tissue pseudotumors [4,10]. However, the presence of fat, myxoid stroma, or extensive necrosis in the lesion should prompt consideration of well-differentiated liposarcoma, myxoid tumor, or necrotic leiomyosarcoma, respectively [7].

There is no consensus on the management of HPT [2,4,6,7,10]. Multiple authors emphasize the importance of a multidisciplinary approach involving surgeons, hematologists, radiologists, traumatologists, urologists, and more recently, vascular-interventional radiologists, to achieve optimal outcomes [6,8]. Fine needle aspiration is typically contraindicated due to the risk of infection, fistulization, or hemorrhage [2,7,9]. In many cases, long-term replacement therapy or local radiotherapy have been chosen [3]. Surgical excision is the preferred treatment approach, although it is not always feasible [2,5]. Technical difficulties may arise due to tumor size, adhesions, or capsule rupture. Bleeding complications, such as those observed in our patient despite proper FVIII treatment, are of particular concern [3]. In such cases, radiotherapy and arterial embolization should be considered as treatment options prior to surgery, either alone or in combination [2].

Although only a few cases have been reported, pre-surgical embolization aims to reduce tumor blood supply, minimizing the risk of bleeding during surgery [1,4,9,10]. Cone-beam CT imaging has become an essential tool for guiding interventions in the angiography suite, particularly for hepatic vascular procedures in conjunction with digital subtraction angiography. In our case, we found it to be a fundamental tool for accurately identifying the relevant angles and branches for intervention, significantly reducing procedure time and radiation exposure for both the patient and the operator. We performed embolization using polyvinyl alcohol microparticles and coils, consistent with the limited cases reported in the literature [3,11]. Mingming, et al. successfully described the embolization of a pseudotumor in the jawbone of a child using ethanol, although we acknowledge that bony pseudotumors may differ in characteristics from intra-abdominal HPT. The use of liquid agents, such as Onyx, could be considered. Klamroth et al. achieved successful embolization of elbows and knees in hemophilia patients using Onyx, although the cases were not HPT and the embolization was not performed pre-surgically [12].

If embolization is performed two weeks before surgery, the lesion contracts without sufficient time for re-establishing new vascularization [2]. However, embolization alone cannot reduce the size of the capsule, and it should be considered as an adjunctive treatment to surgery [1]. Nonetheless, the choice between conservative treatment and surgery must be evaluated on an individual basis for each patient [2,7].

Conclusion

In individuals with severe hemophilia, the identification of an intra-abdominal lesion exhibiting characteristics such as a hematoma at different stages of evolution, enclosed within a fibrous capsule, should raise suspicion of a hemophilic pseudotumor. Preoperative arterial embolization has been shown to be beneficial in reducing the size of the pseudotumor and minimizing the risk of bleeding both during the surgical intervention and in the postoperative period.

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