

Hemophilic pseudotumor complicated by femur fracture: a case report

Pseudotumor hemofílico complicado con fractura de fémur: a propósito de un caso

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Introduction

Hemophilic pseudotumor is an encapsulated blood accumulation caused by repeated bleeding into soft tissues that did not receive appropriate treatment¹. Its incidence rate in patients with hemophilia is approximately 1%-2%^{2,3}. Due to its rarity, there are no standardized clinical practice guidelines for its treatment.

Available treatments include clotting factors, surgical resection, radiotherapy, and others such as endovascular embolization⁴.

Below, we present a descriptive observational study in the form of a clinical case report.

Case report

This is the case of a 42-year-old male diagnosed with severe hemophilia A (basal factor VIII level of 0.48%) on tertiary prophylaxis with plasma-derived factor VIII at a dose of 20 IU/kg biweekly. The patient presents with hemophilic arthropathy and 2 long-standing pseudotumors: one in the upper third of his left leg—16 cm in diameter—and the other on the anterior aspect of his right thigh (25 cm in diameter).

He visited the emergency department of a tertiary referral center due to acute pain in his right thigh; upon standing, he perceived a cracking sound.

Physical examination revealed an increase in size and intense localized pain in the pseudotumor of the right thigh.

A plain radiograph of the lower limb showed a non-displaced linear fracture of the distal femur.

Computed tomography (CT) confirmed the presence of a 25 cm × 13 cm × 12 cm polylobulated mass of heterogeneous density, with calcifications and a complete linear fracture line. Conservative treatment with plasma-derived factor VIII was initiated, and a plaster cast was applied. The patient was admitted to the medical ward.

Hemostatic treatment plan with plasma-derived factor VIII: day 1: 50 IU/kg bolus; days 2-6: 25 IU/kg every 12 hours; days 7-10: 25 IU/kg every 24 hours.

On day 10, the patient was discharged due to good clinical progress, with a plaster cast and prophylactic treatment with factor VIII at 25 IU/kg 3 times weekly. Thromboprophylaxis with low-molecular-weight heparin at 1 mg/kg daily was also initiated upon admission.

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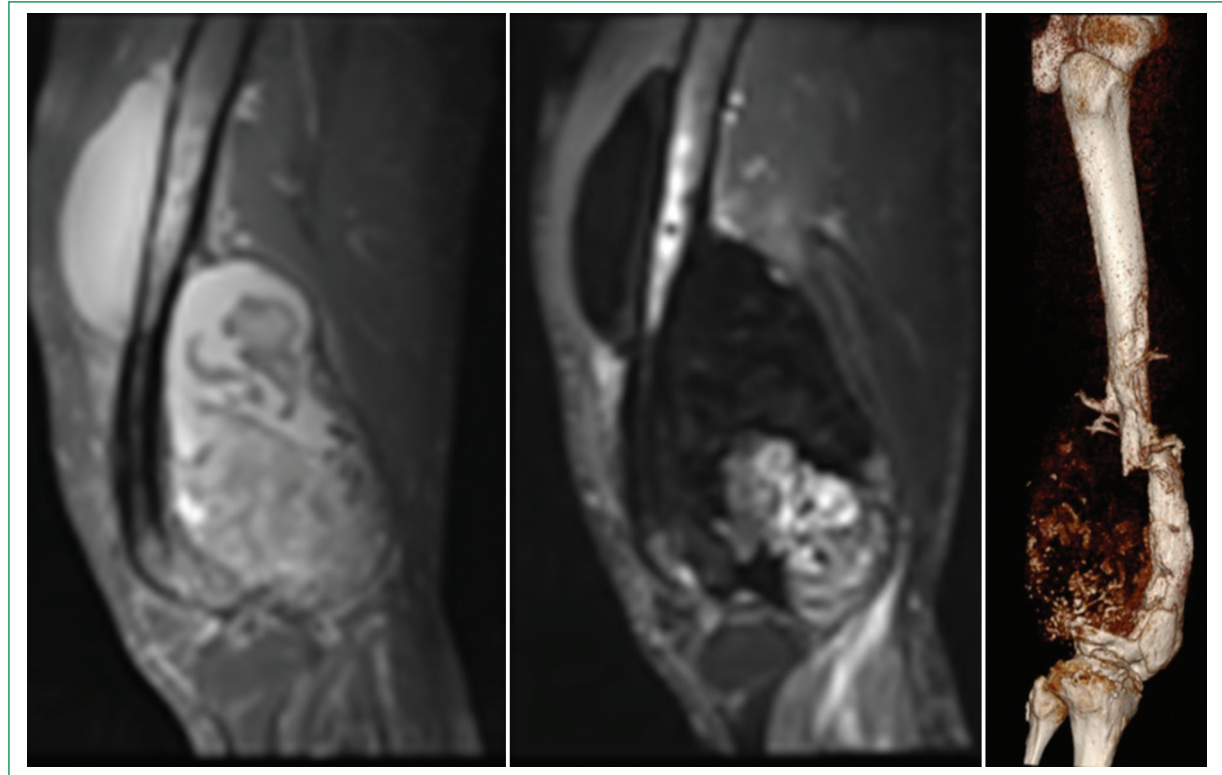


Figure 1. CT scan and 3D reconstruction showing a displaced and overriding fracture associated with a known pseudotumor.

One month later, a follow-up CT scan showed displacement of the fracture with overriding, as well as the persistence of the pseudotumor with the same size (Fig. 1). The patient was readmitted for surgical resolution of the displaced fracture and pseudotumor. The following hemostatic plan was implemented for surgery: immediate preoperative: 50 IU/kg bolus of factor VIII; days 2-6: 25 IU/kg every 8 hours; days 7-14: 25 IU/kg every 12 hours; days 15-17: 15 IU/kg every 12 hours; days 18-20: 20 IU/kg every 24 hours. Serial measurements of factor VIII activity levels were performed (Table 1).

Surgery was divided into 3 surgical acts: opening, lavage, and cavity filling. Under general anesthesia, a wide lateral approach to the thigh was performed, centered on the epicenter of the cavity, extending from the proximal to the distal femur, where a fistulization was present. Upon opening the cavity, the tumor contents drained spontaneously. The hemorrhagic material was removed in one piece, the fracture was corrected, and the cavity was filled with hydroxyapatite. The fracture was stabilized with a plate and screws, and the wound was closed with drainage and a compressive bandage (Fig. 2).

The patient experienced no postoperative complications. Due to good clinical progress, he was discharged after 21 days of hospitalization, with instructions for prophylactic factor VIII at 25 IU/kg 3 times weekly. Physical rehabilitation therapy was initiated, and at 12 months, the patient was able to walk at a normal pace and return to his daily activities (Fig. 3).

The pathology report described the presence of abundant partially organized fibrin mesh, red blood cells, and some amorphous calcifications in the absence of inflammatory infiltrates; all findings consistent with the diagnosis of hemophilic pseudotumor.

Discussion

Hemophilia is a congenital coagulopathy caused by X-linked recessive mutations in the genes encoding factor VIII (hemophilia A) or factor IX (hemophilia B)⁵.

In 1918, Starker first described hemophilic pseudotumor in a 14-year-old adolescent with femoral involvement⁴. Hemophilic pseudotumor is a complication of hemophilia consisting of an encapsulated hematoma with progressive enlargement due to recurrent bleeding,

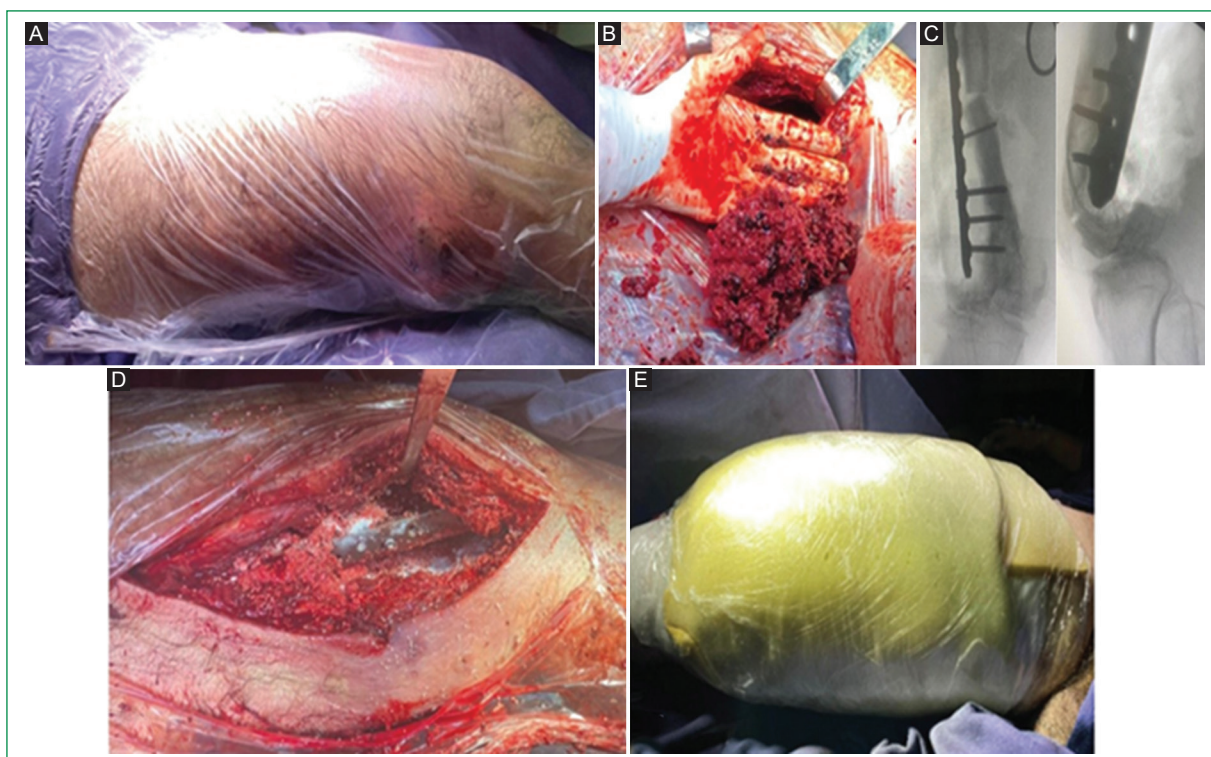


Figure 2. **A:** lateral approach centered on the tumor mass. **B:** evacuation of contents. **C:** revitalization of fracture focus and fixation with osteosynthesis. **D:** appearance of hydroxyapatite. **E:** compressive circumferential bandage with polyfoam and adherent fields.



Figure 3. Patient standing 12 months postoperatively.

Table 1. Plasma factor VIII dosage and coagulation factor VIII activity percentage during and after surgery

Plasma factor VIII dose	Day	Factor VIII activity (%)
50 IU/kg	1	78%
25 IU/kg every 8 hours	2	72%
	3	77%
	4	139%
	5	-
	6	-
25 IU/kg every 12 hours	7	113%
	8	72%
	9	68%
	10	139%
	11	92%
	12	72%
	13	52%
	14	31%
15 IU/kg every 12 hours	15	35%
	16	45%
20 IU/kg every 24 hours	17	-
	18	32%
	19	-
	20	50%

which can endanger limbs or life. Its pathogenesis is related to recurrent bleeding that did not receive adequate treatment⁵.

Pseudotumors most frequently occur in relation to long bones⁶, but small bones of the hands and feet can also be affected.

Clinically, they present as a firm, painless mass that increases in size and adheres to deep structures^{1,7,8}.

In terms of diagnosis, CT is more useful than plain radiography, as it allows for discrimination of size, relationships, and the presence of a fibrous capsule⁵. Magnetic resonance imaging (MRI) is equivalent, but its availability is a limiting factor^{9,10}.

Treatment of a pseudotumor depends on its location, size, growth, and involvement of adjacent structures. Options include replacement therapy with factor and monitoring, radiation, or surgical excision, with no standardized guidelines currently available.

Surgical excision may be necessary in cases of large pseudotumors and complex anatomical relationships, as in our patient. Surgery was planned using factor concentrate treatment and performed at a hemophilia-specialized center, with daily factor VIII dosing.

Evidence for surgical treatment of pseudotumor is based on case reports or case series. In a multicenter study by Magallón et al.³, 8 out of 14 patients with pseudotumor surgery had good outcomes vs conservative treatment with factor, where only 2 out of 15 patients showed good progress.

In a retrospective study by Lin et al.³ spanning 14 years (2006-2017) at the Hemophilia Center of Nanfang Hospital in China, 39 patients were diagnosed with hemophilic pseudotumor, and surgical treatment was decided for 34 of them, with good outcomes. In another retrospective study by Kamal et al.⁸ over a 7-year period in an orthopedic and trauma department in Indonesia, 6 patients with hemophilia A and pseudotumor were described, concluding that surgery was the most effective treatment for these patients.

For major surgery in a patient with severe hemophilia A, the World Federation of Hemophilia guidelines recommend factor VIII levels of 80-100% preoperatively, 60-80% on days 1-3, 40-60% on days 4-6, and 30-50% on days 7-14. This same regimen was used by Lin et al.³ in 34 pseudotumor surgeries.

As seen in [table 1](#), adequate factor VIII levels were achieved for the planned surgery.

Conclusions

We report the case of a 42-year-old man with a hemophilic pseudotumor and a pathological fracture in the femoral diaphysis, treated with radical resection and orthopedic surgery. This case is the first documented in our country to associate a pathological fracture with the surgical treatment of a hemophilic pseudotumor, characterized by a highly favorable outcome.

Funding

None.

Conflicts of interest

None.

Ethical considerations

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The recommendations of the SAGER guidelines have been followed according to the nature of the study.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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