Hemophilic Pseudotumors: A Rare Complication of Hemophilia

Abstract

Background:
Hemophilic pseudotumor is a rare but potentially severe complication of hemophilia, seen in 1–2% of patients with severe forms of the disease, particularly in the absence of early prophylactic treatment. These lesions present as slowly expanding, encapsulated hematomas, most commonly located in long bones.

Case Presentation:
We report the case of a 38-year-old man with severe hemophilia A under prophylactic factor VIII therapy, who developed a right iliac bone pseudotumor complicated by an iliopsoas hematoma. CT revealed multiloculated hemorrhagic collections eroding the iliac bone and compressing adjacent organs. Surgical drainage and resection were performed under factor VIII coverage, with favorable postoperative outcomes.

Conclusion:
Although rare, hemophilic pseudotumor should be considered in patients with hemophilia presenting with erosive bone lesions or soft tissue masses. Imaging plays a key role in diagnosis and management planning.

Keywords:
Pseudotumors; Hemophilia; Intraosseous hematoma

Introduction

Hemophilic pseudotumors are rare, occurring in approximately 1–2% of patients with severe hemophilia [1]. They most commonly affect long bones such as the femur, pelvis, and tibia in adult patients and arise from repeated, untreated bleeding episodes that evolve into chronic, encapsulated hematomas. Their incidence has significantly decreased with the widespread implementation of prophylactic factor replacement therapy.

Case Report

A 38-year-old male with severe hemophilia A, receiving prophylactic factor VIII therapy, presented with abdominal distension and tenderness. His history included hemophilic arthropathy of both knees and elbows and a right total hip arthroplasty. He had been under follow-up for a pseudotumor of the right iliac bone associated with a psoas muscle hematoma for the past year.

On examination, the patient was afebrile, with a tender epigastric mass, joint stiffness in both knees and elbows, and walked with assistance. General condition was preserved.

A contrast-enhanced abdominopelvic CT scan revealed:

- Confluent centromedullary osteolytic lesions in the right iliac wing with cortical thinning and breakthrough;
- A large, encapsulated hemorrhagic collection in the iliacus muscle measuring 7.6 × 7.0 × 11.0 cm;

- Endosteal scalloping of the iliac wing with adjacent calcifications;
- A second heterogenous hemorrhagic collection extending from the right psoas muscle into the abdomen, measuring 11.5 × 7.0 × 14.0 cm;
- Mass effect on the right external iliac vessels, bladder, and sigmoid colon without direct invasion.

Imaging Findings

- **Figure 1:** Coronal contrast-enhanced abdominopelvic CT (parenchymal window) – Confluent hemorrhagic collections in the left psoas and iliacus muscles (black arrow).
- **Figure 2:** Axial contrast-enhanced CT (parenchymal window) – Encapsulated hemophilic pseudotumor of the right iliac bone eroding the iliac wing with calcifications (arrow).
- **Figure 3:** Axial contrast-enhanced CT (bone window) – Pseudotumor with cortical thinning, endosteal scalloping, and cortical breakthrough (arrow).

Surgical Management and Outcome

Given the erosive nature and risk of iliac fracture, surgical drainage and resection of the pseudotumor were performed via a right lower abdominal laparotomy. The procedure was done under anesthetic monitoring and perioperative factor VIII coverage.

The postoperative course was uneventful, with no bleeding. Control CT showed significant reduction in the size of both intraperitoneal collections.

Discussion

Hemophilic pseudotumors may develop in intraosseous, subperiosteal, or soft tissue locations. They can remain asymptomatic for long periods, allowing considerable growth. Presenting symptoms may include pain, swelling, fractures, neurovascular compression, superinfection, or hemorrhagic rupture [2,3].

Intraosseous pseudotumors typically involve the femur, pelvis, or tibia. Radiographs show well-defined, unilocular or multilocular expansile osteolytic lesions with peripheral sclerosis and possible soft tissue extension. CT is useful for assessing bone fragility and guiding surgery. MRI reveals multiloculated lesions with signal heterogeneity reflecting hemorrhage stages. Differential diagnoses include aneurysmal bone cysts, unicameral cysts, brown tumors, desmoplastic fibroma, and slow-growing malignancies (e.g., plasmacytoma, metastases, chondrosarcoma).

Subperiosteal pseudotumors elevate the periosteum and cause chronic mass effect, leading to cortical scalloping. CT and MRI effectively visualize periosteal detachment. Thick radial bony trabeculae are suggestive.

Soft tissue pseudotumors are most commonly seen in the thigh, gluteal region, and iliopsoas muscle. On radiography, they appear as dense masses with or without calcifications. Adjacent bone may show erosion or destruction. Ultrasound assists in volume assessment and monitoring. CT and MRI define lesion extent and relationships with adjacent neurovascular structures.

Treatment begins with immobilization and coagulation factor replacement [4]. Surgical excision may be necessary in non-responders and can be preceded by embolization. Radiotherapy is reserved for non-surgical candidates [5]. Biopsy or drainage is contraindicated due to the risk of perforation or infection [2].

Conclusion

Although rare, hemophilic pseudotumors should be considered in hemophilia patients presenting with large, erosive juxta-osseous masses. Timely diagnosis through imaging is essential to avoid complications such as fracture, neurovascular compression, or rupture. Multidisciplinary management is crucial for optimal outcomes.

References

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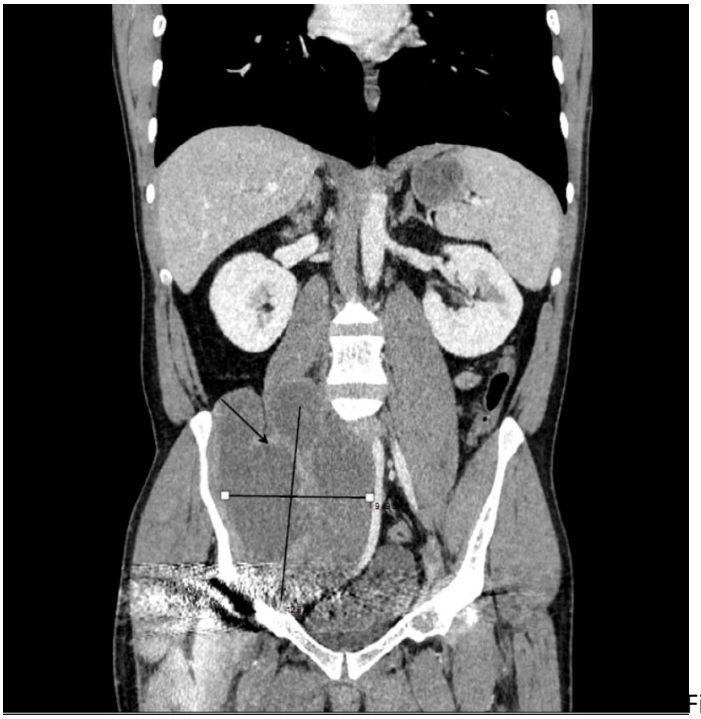

Figure 1

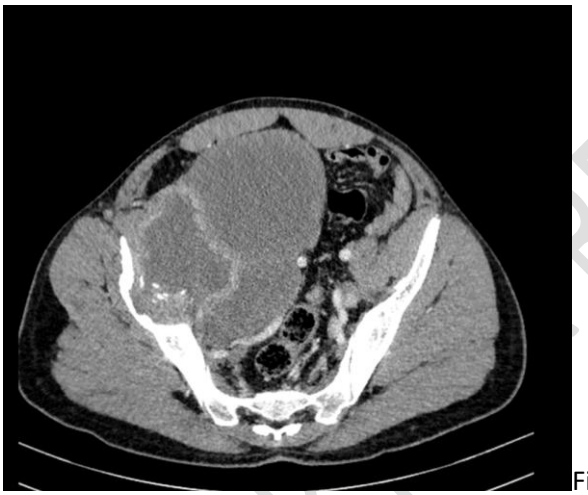

Figure 2

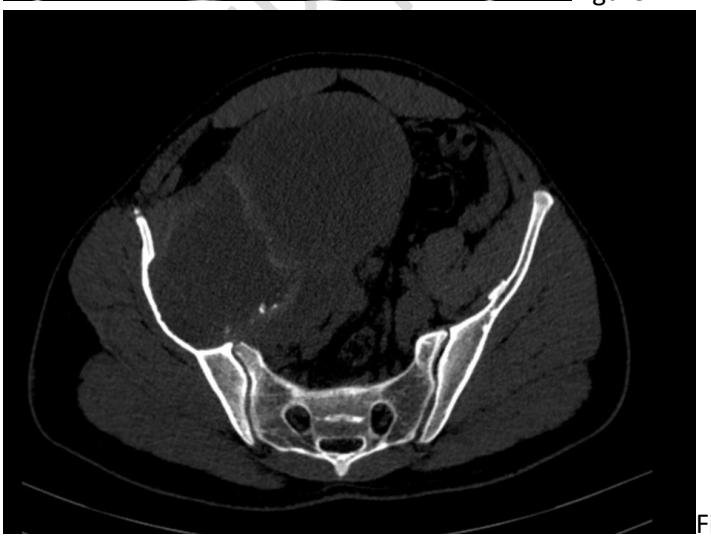

Figure 3