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Role of Imaging in the Diagnosis of Iliac Haemophilic Pseudotumors- A Rare Case Report

Zakia El Yousfi, Kaoutar Imrani, Amal Lahfidi, Nabil Moatassim Billah and Ittimade Nassar
Central Radiology Department, Ibn Sina Hospital, Mohamed V University, BP 10100, Rabat, Morocco.

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ABSTRACT

Haemophilic pseudotumor is a rare complication of haemophilia consisting of chronic, encapsulated hematoma of muscle and or bone due to repeated bleeding. It is estimated to occur in approximately 1-2% of patients with severe disease. Imaging tests (radiography, sonography, CT, and MRI) are fundamental for diagnosis and monitoring the evolution of pseudotumors. In this paper, we report the radiography and computed tomography aspects of a rare case of a patient with large iliac hemophilic pseudotumor, occurring in severe hemophilia.

Introduction

Haemophilic pseudotumors are severe but exceptional complications of haemophilia. They are diagnosed in about 1 to 2% of patients with a severe form of the disease. The pelvis, femurs, and tibias are their most common localisations. Conventional radiography and computed tomography (CT) are essential to diagnosis. Radiologic manifestations are often characteristic [1].

In the following paper, we report a case of iliac haemophilic pseudotumor and describe the different aspects radiologists might encounter.

Case Presentation

A 45-year-old man, with a known history of haemophilia A, is addressed to us. He complains of intense pain in the left hip area beginning some three months ago. He also has trouble walking. Physical examination finds impaired mobility in the left leg.

A conventional radiograph showed an expansile, multilobular osteolytic lesion occupying entire left ilia, with endosteal scalloping and cortical thinning.

A pelvis CT scan revealed a well-circumscribed lesion centred on the left iliac wing, reaching the obturator muscle, with lysis the acetabular roof. There were some gaps in the iliac cortical bone. Areas of increased density suggesting hematic origin were found inside that lesion. The diagnosis of haemophilic pseudotumor was suspected because of the context and imaging findings. However, other bone tumors, could not be excluded. CT-guided biopsy showed that the tumour was made of necrotic debris, fibrin, and blood clots, surrounded by a fibrous capsule.

A conservative therapeutic approach was decided. The leg was immobilized, and the patient received regular doses of factor concentrates. A control CT scan performed three months later showed a lack of significant tumoral growth.

Discussion

Haemophilic pseudotumors are chronic encapsulated hematomas growing with each bleeding episode. Often, they are the result of repetitive minor traumatisms. Their natural history consists of growth, invading of adjacent structures and bone erosion [2]. Common locations include the thigh, the gluteal region, and the iliopsoas muscle. The clinical presentation varies considerably depending on lesion location but is slowly progressive and chronic.

These pseudotumor usually occur in soft tissues (intramuscular), and occasionally in bone or a subperiosteal location, and are classified accordingly [3]. Radiographic aspects are characteristic alongside a known history of haemophilia.

Conventional radiography is the first-line test. In bone tumours, one can often expect expansive, well-defined osteolytic lesions. They can be unilocular or multilocular. Osseous trabeculae frequently traverse the lesions. Occasionally, calcific or ossific foci are present, which are characteristic of older tumours.

Periosteal elevation is a hallmark of subperiosteal pseudotumors, alongside thick radial bone trabeculae and endosteal scalloping of the adjacent bone [4]. Soft tissue pseudotumors evolve from neglected hematomas. Increased soft-tissue density and the presence of calcifications are common signs found in this type of tumour [5].

CT is essential to fully characterize the lesion. In the context of bone pseudotumors, cortical damage, periosteal reaction and detachment can be more thoroughly studied. Assessment of soft tissue pseudotumors also benefits from CT, as their age, density, and limits can be characterized more precisely. Bone lysis is also easier to quantify. The presence of air bubbles inside the lesion can be a sign of infection [5].

Magnetic resonance imaging (MRI) allows for precise evaluation of a lesion's topography and neurovascular relationships. A typical pseudotumor appears as a mass of heterogeneous signal, which have complex signal intensities reflecting the effects of remote and recurrent haemorrhage and clot organization.

Tele:
E-mail address: zakia.ysf@live.fr

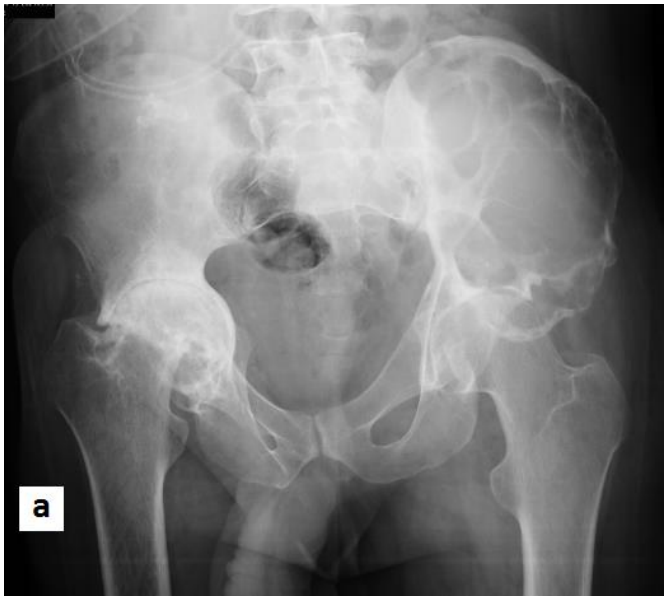

Figure 1. Conventional radiograph (a) reveals well-defined, expansile, multilobular osteolytic lesion involving the left ilia.

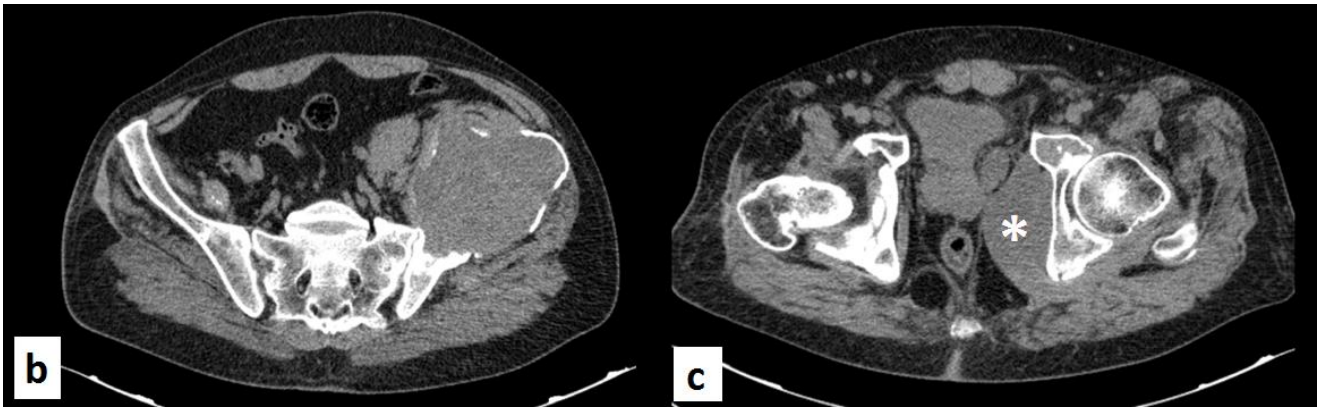

Figure 2. Abdominal unenhanced axial CT scans (B and C) reveal a collection centered on the left ilia, well circumscribed, with increased density suggesting hematic origin, and extension to the homolateral obturator muscle (asterisk)

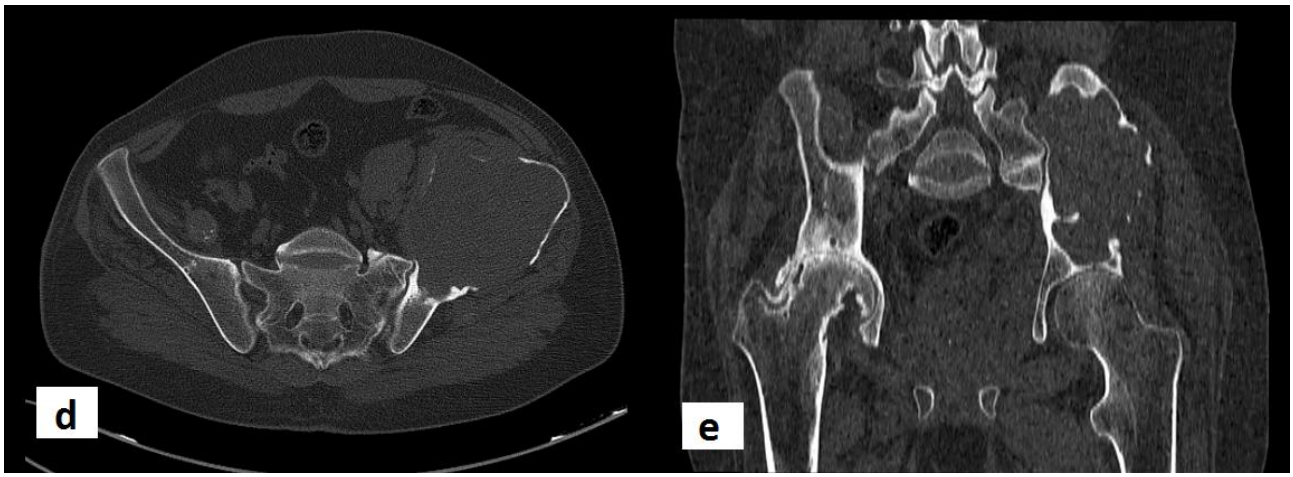

Figure 3. CT bone window scans, axial (d) and coronal plans (e) show extensive bone lysis of the left ilia, with endosteal scalloping. Cortex is focally disrupted.

Acute haemorrhage with intracellular deoxyhemoglobin appears isointense on T1-weighted MR images and hypointense on T2-weighted images. As the T1 relaxation time shortens by methemoglobin, the signal intensity progressively increases, from the periphery to the center, on T1 and T2. The tumour is usually surrounded by dark signal intensity on all sequences, it is consistent with the fibrous capsule or hemosiderin. Sometimes, mural nodules can be found at the capsular wall [5].

Sonography can be a helpful diagnostic tool when anatomical conditions allow it; it's also a valuable follow-up option. It shows a central anechoic area with increased echoes behind the lesion, caused by fluid in the pseudotumor, in the acute and subacute stages [1].

Regardless of the context, polycystic lytic images can be misdiagnosed as an aneurysmal cyst, solitary bone cysts, brown tumors, even bone hydatidosis. There are many malignant bone tumors that may resemble a pseudotumor, including metastatic disease, fibrosarcoma, malignant fibrous histiocytoma, and telangiectatic osteosarcoma. The most difficult differential diagnosis is that of an aneurysmal bone cyst (same signal characteristics, but different history) [6].

Although the puncture is contraindicated because of an added risk of superinfection and fistulisation, it is necessary, if there is any doubt, to perform a biopsy in a fleshy area suspected of tumour.

Treatment is complex and multidisciplinary cooperation is often needed. The conservative approach (limb immobilization) is indicated in young pseudotumors. Surgery is the option of choice when dealing with older or deeper tumours [7].

Conclusion

Haemophilic pseudotumors are rare, often serious complications of haemophilia. Conventional radiography and CT each play an important role in the diagnosing and staging this pathology. They are essential for choosing the adequate therapeutic approach as well as eventual follow-up.

Conflict of interest

The authors do not declare any conflict of interest.

Author's contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

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