Case 8055

Hemophilic pseudotumor
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Section: Musculoskeletal system
Case Type: Clinical Cases
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Patient: 50 years, male

Clinical History:

50-year-old Vietnamese male, with history of factor VIII deficiency haemophilia. The patient presents with multiple palpable masses within the right buttock region.

In the remote past, the patient had a left leg above knee amputation, secondary to extensive bleeding.

Imaging Findings:

Oblique radiograph of the pelvis demonstrates a soft tissue convexity with multiple scattered calcifications.

Ultrasound image of the palpable mass in the right buttock demonstrates heterogeneous mass, with mixed isoechoic and hypoechoic regions.

CT images demonstrate multiple masses within the right gluteus maximus and medius, with multiple foci of coarse calcification, which are near the periphery of the masses. The masses are hyperdense at their periphery, with several nodular soft tissue densities centrally. There is lytic change involving the posterior right ilium adjacent to the right sacro-iliac joint, as well as the right sacrum. There is no associated periosteal reaction.

MR images again show the multiple lesions. All of the lesions demonstrate very low signal at their periphery on all imaging sequences, with susceptibility artifact on the gradient recalled echo images, consistent with haemosiderin deposition.

One of these lesions was aspirated under ultrasound guidance, returning a total of 450 ml of old blood.

Discussion:

Haemophilic pseudotumour is a rare complication of haemophilia, estimated to occur in approximately 1-2% of patients with severe disease [1]. It is seen almost exclusively in men, as the predisposing diseases of haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency) are both X-linked recessive diseases.

A pseudotumour is a chronic, encapsulated haematoma, and may initially occur either due to spontaneous or post-traumatic haemorrhage. The fibrous wall often maintains a blood supply, although the central portions of the lesion are avascular [2]. Repetitive bleeding at the level of the wall into the haematoma due to derangement of the clotting cascade leads to expansion and enlargement of this mass. This may result in erosion of the adjacent bone, or necrosis or denervation of adjacent muscle.

On radiography, pseudotumours appear as an area of increased soft tissue density, which may contain internal calcifications. There may be remodelling or frank destructive change of the adjacent bones.

On both ultrasound and CT, the appearance is widely variable, due to the possible different stages of haemorrhage.
within the lesion. Both anechoic and heterogeneously echoic collections have been noted on ultrasound. Similarly, pseudotumours of low, intermediate, and high density have all been reported [3].

On MR, the signal internally will vary on both the T1 and T2 weighted images due to the evolution of the blood products. However, pseudotumours have a characteristic rim of dark signal on all imaging sequences, due to the presence of the fibrous and haemosiderin-laden wall. On MR imaging, the differential diagnosis is relatively limited, due to the characteristic low signal rim. A rare mimic of this lesion is the aneurysmal bone cyst of soft tissue. [6]

The management of these lesions is controversial. Prevention of these lesions by factor replacement is effective [4]. Once the lesion is present, many forms of therapy have been used, to variable success, including: irradiation, embolisation, percutaneous evacuation with instillation of fibrin seal, surgical resection, and as a last resort, amputation [5].

**Differential Diagnosis List:** Hemophilic pseudotumour.

**Final Diagnosis:** Hemophilic pseudotumour.

**References:**


Figure 1

Description: Oblique pelvis radiograph demonstrating a soft tissue mass in the right gluteal region with multiple foci of calcification. Origin:
Figure 2

Description: Ultrasound image at the right gluteal region demonstrating a heterogeneous hypo-echoic to iso-echoic solid mass. Origin:
**Description:** CT on soft tissue windows demonstrating a thick-walled solid mass in the right gluteal region. Centrally, the mass is of low density, with multiple higher density internal nodules. A few foci of calcification are present at the medial aspect of the wall. **Origin:**
Description: CT on soft tissue windows demonstrating communication of the mass with a larger component more posteriorly. Origin:
**Description:** CT on bone windows demonstrating the mass abutting the right ilium. **Origin:**
Description: CT on bone windows demonstrating the mass abutting the right ilium, with associated erosive changes. Origin:
Description: Coronal CT shows additional lesions within the right thigh. Origin:
Description: Ax T1 weighted MR image of pseudotumour. (1/2) Origin:
Description: Ax T1 weighted MR image of pseudotumour. (2/2) Origin:
Description: Ax T2 weighted MR image of pseudotumour. (1/2) Origin:
Description: Ax T2 weighted MR image of pseudotumour. (2/2) Origin:
Description: Cor STIR MR image of pseudotumour. (1/2) Origin:
Description: Cor STIR MR image of pseudotumour. (2/2) Origin:
Description: Sag GRE MR image of pseudotumour. (1/2) Origin:
Description: Sag GRE MR image of pseudotumour. (2/2) Origin: