Pelvic haemophilic pseudotumour in a patient with haemophilia

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We report a 20-year-old male patient, with a history of haemophilia presented with a sudden onset of pain in the right gluteal region of 5 days duration. He is afebrile; there is no history of trauma. He developed painful swelling in his right gluteal region for nearly 1 week with difficulty in walking. Physical examination revealed tenderness at the same site.

CT examination confirmed the presence of a large heterogeneously enhancing lobulated pseudotumour (12×16 cm) and a major lytic defect in the right iliac bone (figure 1).

It is important to investigate primary malignancies, as mass lesions could be mistaken for pseudotumours of haemophilia, as there are reports for malignancies such as sarcomas and non-Hodgkin's lymphoma may mimic a pseudotumour in haemophilia patients. Preoperative biopsy is contraindicated so surgical excision was performed for final diagnosis and treatment.

Haemophilic pseudotumours occur in 1–2% of patients with severe haemophilia. Clinically, haemophilic pseudotumours usually present as a painless expanding mass growing slowly, but in our case, the patient was presented with a sudden onset of pain and swelling of nearly 1 week duration. Their most serious sequel is pathological bone fracture and uncontrollable bleeding. Two types of pseudotumours have been described by Gilbert: a proximal (occurs in the long bones and the pelvis) and a distal type (affects the small bones).

Conservative treatment with replacement therapy and immobilisation may resolve the distal type but the proximal type of haemophilic pseudotumour like our case does not respond to conservative treatment.

Learning points

▸ The haemophilic pseudotumour is a rare but severe complication of haemophilia.
▸ Chronic haematomas may mimic soft tissue sarcomas.
▸ Treatment of haemophilic pseudotumour is currently based on conservative measures or surgical resections.

Figure 1  Contrast-enhanced CT scan of the pelvis in the axial plane showing a large lobulated heterogeneous mass (12×16 cm) and a major lytic defect in the right iliac bone: (A) soft tissue window and (B) bone window.

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REFERENCES