Pelvic haemophilic pseudotumour: a case report

Achmad Fauzi Kamal,1 Lugyanti Sukrisman,2 Ismail Hadi Soebroto Dilogo,1 Heka Priyamurti,1 Muhammad Nurul Qomaruzzaman1

1 Department of Orthopaedic and Traumatology, Ciptomangunkusumo Hospital, Faculty of Medicine, University of Indonesia, Indonesia
2 Division of Hematology, Department of Internal Medicine, Ciptomangunkusumo Hospital, Faculty of Medicine, University of Indonesia, Indonesia

ABSTRACT

We report on a 30-year-old haemophiliac man with a pelvic pseudotumour compressing adjacent structures causing pain and swelling and destruction of surrounding soft tissues and bones. He underwent evacuation of the pseudotumour, acetabular reconstruction using the Harrington procedure, and total hip arthroplasty.

Key words: arthroplasty, replacement, hip; hemophilia A

INTRODUCTION

Haemophilic pseudotumour is a progressive cystic swelling caused by repetitive bleeding not treated adequately and involves bone in the form of osteolytic lesions.1,2 Destruction and formation of bone and calcification (ossification) of surrounding soft tissues are noted. This condition is similar to malignant bone tumours such as osteosarcoma and thus known as pseudotumour.1,3,4 Haemophilic pseudotumour is a complication of haemophilia and may cause bone erosion, progressive neurological deficit, shifting of tissue or organ, or loss of normal organ function.5 The overlying skin may undergo necrosis, infection, and rupture.5–7 Mobility may be interfered.5

CASE REPORT

In July 2010, a 30-year-old haemophiliac man presented with a 3-year history of progressive swelling in his left pelvis. He had had recurrent swelling of the joints, especially the knees and elbows for up to 5 times a year. The mass was solid with cystic parts, non-tender, with bluish, smooth overlying skin and ill-defined margin (Fig. 1). Both active and passive range of movements of the hip joint were within normal range.

Blood tests showed a haemoglobin level of 9.3 g/dl, haematocrit of 28%, white blood cell count of 5300/mm³, platelet count of 402 x10⁹/mm³, prothrombin time of 11.3 seconds, activated partial thromboplastin time of 56.4 seconds, and factor VIII
level of 5.6% with negative anti-factor VIII.

Pelvic radiographs 6 months prior to admission revealed expansile lytic lesions with ground glass appearance in the left ilium, and multiple septations accompanied by a soft-tissue mass. Pelvic radiographs before surgery showed an expansile destructive lesion demarcated by calcification from the ilium to the left inferior iliac spine with cortical thinning, with soft-tissue swelling around it (Fig. 2).

Magnetic resonance imaging showed a lobulated homogeneous mass (5.4x6x9 cm) with well-defined margin, hyperintense on T1- and T2-weighted images (Fig. 3). On fat suppression, the mass protruded ventrally reaching the sacrum and destroying the acetabulum, femoral head, and left ilium. The medial border of the pseudotumour pushed the left iliopsoas muscle. Computed tomography showed an expansile mass in the ilium extending into the left ischium and sacrum and infiltrating the fatty tissue. Calcification was noted in the central area, causing thinning of the cortex, and non-homogeneous enhancement on the edge.

The patient was diagnosed with haemophilic pseudotumour of the left ilium and treated with 2500 units of factor VIII concentrate twice a day for 2 weeks, followed by 1000 units 2 times a week as maintenance dose. After consultation with the haematology team, it was decided to perform surgery including evacuation of the haematoma, acetabular reconstruction using a modified Harrington procedure, and total hip arthroplasty.

One hour before surgery, the patient was given factor VIII concentrate to increase the level to 100%. Layer-by-layer incision was made through the iliopsoas approach. The haematoma (3.5 litres in volume) was evacuated, and the cavity was lavaged with normal saline (Fig. 4). The hip joint was then...
opened through the anterolateral approach, and the femoral neck was osteotomised and the femoral head was extracted. The acetabulum was reconstructed with an acetabular cage and cup, followed by insertion of the femoral component. Simultaneously, the left fibular diaphysis was harvested and fixed to the linea inominata as a strut graft (Fig. 5). Histopathological examination of the tissue revealed that it consisted of haemorrhage, reactive osteoid, and fibrous tissue (Fig. 6).

At the one-year follow-up, the patient was in good condition and could work normally. Three years after surgery, the left hip was debrided again because of a new haematoma causing bleeding, fistula, and infection (Fig. 7), which was secondary to inadequate maintenance of factor VIII concentrates and unrestricted daily activities.

**DISCUSSION**

In patients with haemophilia A or B, bleeding in the muscles, joints, and body cavities after trauma may cause chronic disability and require multidisciplinary
management. Haemophilia should be suspected in patients with a history of easy bruising in early childhood, spontaneous bleeding (especially in the joints and soft tissues), excessive bleeding during trauma or surgery, and family history of bleeding. The activated partial thromboplastin time is lengthened.

In patients with mild haemophilia (anti-haemophilic factor >5%), bleeding episodes occur less frequently after trauma or major surgery. In patients with severe haemophilia (anti-haemophilic factor <1%), bleeding episodes occur more frequently and spontaneously, often after very mild trauma or daily activities. More than 90% of bleeding episodes occur in the musculoskeletal system; of them 80% occur in joints.

Pseudotumour as a complication of haemophilia is...
rare, occurring in only 1% to 2% of patients, especially those with severe hemophilia. It is most commonly seen in long bones or the pelvis. The presence of a slowly enlarging mass in a patient with haemophilia should raise the suspicion of a pseudotumour. The pseudotumour arises as a result of inadequate treatment of bleeding, usually in the muscle adjacent to bone; the bone itself may be secondarily involved. The pseudotumour consists of blood products in different evolution levels, and later is surrounded by a fibrous capsule containing macrophages with haemosiderin; it may enlarge and compress adjacent structures and cause pathological fractures, skin and soft-tissue necrosis, fistula, infection, and rupture of the mass causing bleeding that is difficult to stop.

The differential diagnoses of haemophilic pseudotumour include giant cell tumour and aneurismal bone cyst. Based on our patient’s medical history, clinical examination, laboratory, and radiological findings, the most likely diagnosis was haemophilic pseudotumour. Preoperative biopsy was not performed, because in patients with haemophilia, fine needle biopsy or core biopsy may cause further bleeding and expansion of the pseudotumour. In addition, clotting factor coverage for biopsy is expensive and increases the total cost of treatment. After consultation with the haematology team, we proceeded directly to definitive treatment.

Management of haemophilic pseudotumour involves a multidisciplinary approach. Early diagnosis and prevention of haematoma is important. According to the World Federation of Hemophilia, guidelines for the management of haemophilia include prevention of bleeding, management of acute bleeding as soon as possible, hospitalisation for all heavy bleeding, giving clotting factor before invasive procedures, and avoidance of trauma by adjusting lifestyle and avoiding contact sports.

Treatment modalities include conservative treatment, surgery, embolisation, and radiotherapy. Conservative treatment consists of infusion of synthetic clotting factor and immobilisation of the patient, but it fails to prevent progression of the lesion. Although regression is occasionally seen, there are no true cures and recurrence and/or progression is common. Surgery is indicated when the pseudotumour continues to grow despite anti-haemophilic factor replacement therapy. Other indications include haemophilic chronic synovitis, severe soft-tissue contractures, deformity requiring bone osteotomy, severe joint degeneration with disability and severe pain, and non-functioning extremity or the presence of chronic infection.

Percutaneous aspiration is easy, straightforward, and relatively non-invasive. It is also cheap and does not require lengthy hospitalisation. It is indicated for pseudotumours located more peripherally in a well-localised position and for small pseudotumours (<3 cm in diameter). In our patient, the huge pseudotumour had destroyed the ilium and sacrum. Percutaneous aspiration followed by injection of fibrin glue or bone graft is very difficult for a large pseudotumour.

Most pseudotumours require surgery. Surgical options include tumour excision and arthroplasty or amputation, depending on the location and the severity of the destruction. Large pseudotumours should be removed surgically. However, surgery for extensive pseudotumour of the ilium has potential complications. One patient with an extensive haemophilic pseudotumour of the ilium developed a chronic fistula 6 months following en-bloc resection. Another patient required 2 further procedures and the use of a pedicled rectus abdominis flap to eradicate the fistula. If removal of the capsule endangers a vital structure, it is better to leave it intact. In pseudotumour of bone, the cavity should be opened, debrided, and filled with bone grafts and bone substitute. The most effective treatment for large pseudotumours is resection together with factor VIII replacement therapy. In our patient, the haematoma was evacuated, the cavity was debrided and partly filled with bone grafts and bone cement, followed by reconstruction of the acetabulum with Harrington modification procedure and total hip arthroplasty. We did not remove the entire rim of the ilium because it was not malignant and easier to reconstruct. In patients with a large pseudotumour, palliation of symptoms and improving the quality of life are more
realistic goals than total eradication of the lesion. The
strength of the acetabulum in holding the load should
be assessed. The acetabulum should be reconstructed
with anti-protrusion cage to enhance fixation and
distribute the weight-bearing area throughout the
pelvis. Recurrent pseudotumours can be the result
of incomplete resection, failure to identify small
daughter cysts, or postoperative haematomas that
repeat the pathological process.

DISCLOSURE
No conflicts of interest were declared by the authors.

REFERENCES
4. Park JS, Ryu KN. Hemophilic pseudotumor involving the musculoskeletal system: spectrum of radiologic findings. AJR Am
2005.
12. Gilbert MS. Surgical management of the adult haemophilic blood cyst (pseudotumours). In: Rodriguez-Merchan EC,
14. Heeg M, Smit WM, van der Meer J, van Horn JR. Excision of a haemophilic pseudotumour of the ilium, complicated by
17. Faisham WI, Muslim DA, Bhavaraju VM, Nawaz AH, Zulmi W. Modified Harrington procedure for acetabular insufficiency