Haemophilic Pseudotumour: Surgical Management of a Rare Case

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Abstract Haemophilia is a common cause of genetically inherited bleeding disorders. Pseudotumours occur in 1–2 % of persons with severe forms of haemophilia. These are a result of repeated haemorrhage into soft tissues, subperiosteum or a site of bone fracture with inadequate resorption of the extravasated blood. There are a number of therapeutic alternatives for this dangerous condition: surgical removal, percutaneous management, irradiation, embolization etc. In this case report, we describe the natural history, clinical course and successful surgical management of a patient with haemophilia who presented with a massive pseudotumour. We also briefly review the relevant literature on the various therapeutic modalities that have been implemented in the management of this rare complication. Though surgeons may be averse to operate on haemophiliacs, primary surgical management as done in our case may prove to be the definitive treatment option for such patients.

Keywords Haemophilia · Pseudotumor · Surgery · Management

Introduction

Haemophilic pseudotumour is an encapsulated, slowly expanding haematoma occurring in 1–2 % of patients with severe haemophilia (functional plasma levels of coagulation factor <1 %). It is the result of repeated episodes of bleeding into soft tissues, a bone fracture site or a result of subperiosteal haemorrhage. Inadequate resorption of extravasated blood results in an encapsulated area of clotted blood and necrosed tissue. With successive haemorrhagic episodes, these lesions expand over time causing symptoms by mass effect [1].

Herein, we report on the rare case of a massive pseudotumour affecting the right thigh of a 40-year haemophiliac patient and its successful surgical management.

Case Report

Our patient was a 40-year-old man, a known case of haemophilia A with complaint of a swelling on his right thigh for the last 6 years. The swelling was insidious in onset and gradually progressive. It was accompanied by dull-aching pain, localised to the thigh, aggravated on walking around and relieved by rest. There was no history of significant trauma to the site.

In 2006 and then again in August 2012, the patient presented at another hospital where the swelling was diagnosed as a haematoma, and simple aspiration was done. Though the swelling had resolved the first time, in the second time, in spite of aspiration of approximately 4 L of fluid as documented, there was no appreciable change in the size of the swelling. In October 2012, the patient presented to our outpatient department for treatment.

On examination, a massive 20×20-cm mass was found over the anteromedial aspect of the right thigh approximately 5 cm below the inguinal region (Fig. 1). Its margins were well

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Fig. 1 Clinical photograph showing massive pseudotumour occupying the anteromedial regions of the right thigh of the patient

defined and its surface, smooth. The overlying skin was stretched, and the local temperature was not elevated. Fluctuation test was positive. Mobility was restricted in both longitudinal and transverse planes. Hip joint had a normal range of motion, and all pulses in the leg were palpable and of normal volume.

Investigations included a normal haemogram with bleeding time of 2 min, clotting time of 8 min and 30 s, prothrombin time of 14/13 and APTT 49.6 with a factor VIII level of 5 %. An MRI obtained at this stage showed in the right thigh a collection with internal debris; compressing the adjacent muscles but with clear fat planes. A CT angiogram was done to rule out a significant feeder vessel suitable for embolization.

A decision to excise the pseudotumour was made.

In conjunction with internists, factor VIII infusion was instituted 1 h preoperatively and confirmed to be raised to the recommended 100 % level.

Formula : Dose (IU) = Target Factor VIII levels–baseline level $\times$ body weight $\times$ 0.5 unit/kg

Intraoperative/peroperative findings revealed a thick-walled cyst containing fibrosed tissue and clotted blood. The cyst wall was densely adherent to muscles of the adductor compartment. Muscles were thinned out and flabby, and those of the extensor and posterior compartment were pushed laterally by the mass. The entire mass was carefully dissected out with use of harmonic scalpel to simultaneously achieve good haemostatic control (Fig. 2a, b). In addition, a 5 % solution of

Fig. 2 **a** Clinical photograph showing the cyst being dissected free of surrounding muscles. **b** Clinical photograph after removal of the cyst; surrounding muscles are stretched and flabby

tranexamic acid was poured into and kept in contact with the cavity for about 5 min to aid haemostasis.

Postoperatively, therapeutic factor VIII levels were maintained till suture removal on the tenth postoperative day. At follow-up, the patient was doing well with the resolution of all symptoms.

Discussion

Pseudotumours or “blood cysts” are a rare complication of severe haemophilia which was first described by Starker in the femur of a 14-year-old boy. Patients present with painless palpable masses or with painful crises due to episodic acute bleeding into the tumour. Most of the morbidity from pseudotumours is due to their compressive effect on the surrounding structures including bone destruction, muscle and skin necrosis [2].

Given the rarity of haemophilic pseudotumours, there is no consensus among authors about the best management strategy.

A conservative approach includes factor replacement and immobilisation of the affected area. This method has been suggested as the preferred management of pseudotumours caused by recent haemorrhage [3]. Aspiration of the fluid within the cysts for either diagnostic or therapeutic purposes is absolutely contraindicated [4].

Surgery has been recommended early in the management of pseudotumours. Indications for surgery include clinical or radiographic evidence of progressive enlargement of the haematoma, haemodynamic deterioration or occurrence of a complication [4]. Therapeutic arterial embolization may be considered especially for tumours of large size which may result in reduction of the size of the tumour as well as reduction of bleeding complications during surgery. Radiotherapy is a viable alternative where surgery is not feasible and conservative management is not effective. Around 6–23.5 Gy in fractions of 2 Gy or less has been found sufficient [5].

Conclusion

Though most surgeons are reluctant to operate on haemophiliacs, in the case of a pseudotumour, surgical

management can provide the best outcome especially when selected as the primary line of treatment.

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