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Percutaneous treatment of haemophilic digital pseudo tumours

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Abstract In young patients with haemophilia, blood cysts in the finger bones may grow rapidly and imitate a malignant tumour (pseudo tumour). The condition must initially be treated with administration of factor VIII-IX. If this does not control the growth, surgical treatment is indicated. We have advised a percutaneous treatment with evacuation of the blood and injection of the cavity with fibrin seal.

Résumé Chez les jeunes patients hémophiles les kystes sanguins des os digitaux peuvent se développer rapidement et évoluer comme une tumeur maligne (pseudotumeur). Le traitement initial doit être l'administration de Facteur VIII-IX. Si cela ne contrôle pas l'augmentation de volume, le traitement chirurgical est indiqué. Nous recommandons un traitement percutané avec évacuation du sang et injection de la cavité de dérivé de fibrine.

Introduction

Haemophilic cysts (blood cysts) appear in 1–2% of patients with severe factor VIII or IX deficiency and are frequently seen in the second or third decade [3–5]. They may invade the surrounding tissue and imitate a malignant tumour (pseudo tumour) [3]. Haemophilic cysts are most frequently found in the femur, followed by the pelvis and tibia [1], but may also be found in the small bones of hands and feet. Generally, the growth is slow, but when they occur in the hands and feet of young patients with epiphyses still open, they are fast growing.

Material and method

We recently treated three young patients. The first was a 15-year-old male Caucasian with antecedents of haemophilia A, mild (more than 5% factor VIII activity), where a tumour-like swelling of a finger was seen at the proximal phalangeal level and re-absorption of this phalanx was evident in X-rays (Fig. 1). The second patient was a 14-year-old male Caucasian with antecedents of haemophilia A. The disease was moderate (<3.3% of factor VIII activity) and diagnosed when the patient was 6 years old. He did not attend the periodical outpatient checks. He was referred with a fusiform tumour in the first phalanx of the left index finger of 8 months' duration. It had progressively increased in size and was now accompanied by pain and changes in skin colour.

The third patient was a 12-year-old male Caucasian also with antecedents of haemophilia A. The disease was moderate (<3.3% of factor VIII activity) and had been diagnosed when he was 3 years old. This patient did not attend the periodical outpatient checks. He was referred for a tumour in the second phalanx of the left ring finger. The tumour had developed over the previous 5 months and was accompanied by pain and limitation of flexion.

Under factor VIII cover all patients were treated with percutaneous evacuation of the clot, curettage of the cavity and filling with fibrin prepared in our own facilities. We used a dose of 1 ml fibrin seal for each 4 ml of blood extracted.

Discussion

The haemophilic pseudo tumour is a severe complication of bleeding into the bone or muscle. It should be treated early before it grows to become a tumour of giant size that compromises the limb. As soon as a haematoma does not respond to the appropriate treatment with clotting factor, or if osseous bleeding progresses, the possibility of the development of a blood cyst leading to a pseudo tumour should be born in mind [6].

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Fig. 1 AP and lateral X-rays of haemophilic cyst proximal phalanx, middle finger. Observe cortical involvement, great osteolysis, septal alveolar aspect from epiphysis to epiphysis and enlargement of the bone [2]

When the localisation is in the fingers, the treatment should initially be conservative, with the administration of factor VIII or IX in order to maintain a level of 25–30% above coagulation level for 8–10 weeks. It is also important to immobilise the hand to avoid pathological fractures. The pseudo tumours with uncontrollable growth need to be treated surgically and must be extirpated if the integrity of the affected extremity is in danger or if pathological fractures occur.

We have previously proposed a percutaneous treatment [4], which is indicated when the cyst continues to grow in spite of the administration of factor VIII or IX; when rupture of the haematoma is imminent. This action prevents skin necrosis and is also needed when there is evidence of neurovascular compromise [4, 7]. We have suggested a percutaneous evacuation of the clots and curettage of the cavities, followed by the injection of fibrin glue. Fibrin glue consists of two separate solutions of fibrinogen and thrombin. When mixed together, these two solutions mimic the final stages of the clotting cascade to form a fibrin clot. This clot may produce haemostasis and thus promote wound healing.

In the treatment efforts should be focused on prevention of pseudo tumour by ensuring adequate education of the patient and relatives; training of health personnel and elimination of social barriers to ensure early access to adequate treatment in order to avoid progression to a giant pseudo tumour. In developed countries, this pathology is not seen

frequently as there are better facilities for follow-up of the haemophilic patients and better resources for treatment.

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