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Bilateral Haemophilic Pseudo-Tumour of the Mandible – A Rare Case Report

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Abstract

Haemophilic pseudo-tumour is an exceedingly rare but characteristic manifestation of haemophilia. It is an extra-articular osteolytic lesion, mostly affecting the long bones and rarely in the mandible. These lesions develop due to repeated and unresolved haemorrhage in soft tissue and bone, remote from joints. We report a case of a 5-year-old male patient brought to our department with a painless gradually increasing swelling of both sides of the mandible since 4 months. The child was a known case of severe Haemophilia A (FVIII < 1%) requiring Factor VIII transfusions based on need. He was treated with twice weekly Factor VIII for 4 weeks followed by weekly transfusions for the next 4 weeks and as per need thereafter. He reported 3 months later and we observed a significant decrease in the size of the swelling on both sides. With such rare incidence and non-specific radiographic picture, we aim to make clinicians aware of the possibility of haemophilic pseudo-tumours occurring in the jaws. The first clue to diagnosis is the presence of haemophilia itself. In the event of no prior or familial history of haemophilia and non-specific histopathological findings in children, intra-bony lesions of the jaws, particularly malignant neoplasms must be considered. Tailoring the treatment of choice for each patient can have beneficial results, provided that adequate factor levels are maintained.

Keywords: Haemophilia A, Haemophilic Pseudo-tumour, Factor VIII concentrate, transfusions

Introduction

Haemophilia is the most common congenital coagulation factor deficiency encountered worldwide. It challenges the Oral and Maxillofacial Surgeons by inducing excessive bleeding during treatment which can even lead to life threatening situations. Haemophilia is broadly classified into Haemophilia A and Haemophilia B. Both are X-linked recessive disorders resulting from a deficiency of either factor VIII (Haemophilia A) or factor IX (Haemophilia B). Classic symptoms include soft tissue bleeding and hemarthroses, which can result in

debilitating arthropathy.[1]Hemarthrosis, or bleeding into a joint, affecting particularly the knees, ankles, and elbows in children, is one of the most common complications of haemophilia.[2] Haemophilic pseudo-tumour also known as haemophilic cyst of bone is an exceedingly rare (occurring in 1–2% of severely haemophilic patients) but characteristic manifestation of haemophilia. It is an extra-articular osteolytic lesion, first reported in 1918 by Starker in the femur of a 14 year old boy.[3] They mostly affect the long bones, occurrence in the mandible is rare. These lesions develop due to repeated and unresolved haemorrhage in soft tissue and bone, remote from joints. The classic presentation is a painless swelling with subsequent bony destruction.[4] The presentation of a rapidly growing swelling with bony destruction occurring at a young age may be indistinguishable from those of malignant osteolytic tumours and should always be considered as a potential differential diagnosis.

Case Report

A 5-year-old male patient was brought to the Department of Oral and Maxillofacial Surgery of our institution with a painless gradually increasing swelling of both sides of the mandible since 4 months, the left side larger in size than that of the right(Fig. 1). There was no history of previous injury over the area. The child was a known case of severe Haemophilia A (FVIII < 1%) requiring Factor VIII transfusions based on need. He was hospitalized two times in the past for episodes of acute oral bleeding which was successfully treated with Factor VIII concentrate. He also presented with hemarthroses of both the knees, his left knee showing more swelling than the right (Fig. 2)

On examination, his face was asymmetrical with the swelling on the left side of an increased dimension than that of the right. Both the swellings were bony hard, non-tender, non-fluctuant with bucco-lingual expansion. No palpable thrills or pulsations were present. On intra-oral examination, there was grade 1 mobility of the left deciduous second molar. On application of pressure over that tooth, mild oozing of blood from the gingival crevice was noted. Orthopantomogram (Fig. 3) revealed a multilocular honeycombed radiolucent lesion with scalloped borders in the mid ramus and angle region on the right side. On the left side, a unilocular radiolucent lesion was present with a definite scalloped margin anteriorly, extending from the posterior border to the anterior 1/3rd of the ramus. Based on the clinical and radiographical features and from the history of presence of severe haemophilia, a provisional diagnosis of haemophilic pseudo-tumour was made.

Topical application of tranexamic acid stopped the mild oozing from the gingival crevice. Given the risk of bleeding, the decision to forgo surgery of the mandibular lesions was made and he was referred to the haematology department for further transfusions. He was treated with twice weekly Factor VIII for 4 weeks followed by weekly transfusions for the next 4 weeks and as per need thereafter. He reported 3 months later and we observed a significant decrease in the size of the swelling on both sides (Fig. 4). However, he did not turn up for further follow-up.

Discussion

Haemophilia is an X-linked recessive disorder caused by deficiency of factor VIII (type A) or factor IX (type B) deficiency. Haemophilia A can also be classified according to its severity as severe, moderate, and mild.[5] In severe cases, there is <1% of factor VIII normal plasma level and patients tends to bleed spontaneously in cases of mouth abrasions, during eruption or shedding of deciduous teeth and due to minor trauma. In these kind of situations, where severe bleeding is anticipated, the patients should be managed with a multidisciplinary

approach in connivance with a medical team, preferably in a hospital setup which has a haemophilia treatment center.[6]

Haemophilic pseudo-tumours are believed to occur in these patients following spontaneous or traumatic haemorrhages into the bone or soft tissue.[7] Majority of the patients developing these tumours are less than 14 years of age with trauma as the initiating factor. However, in our case, the tumour developed spontaneously without any prior inciting event such as trauma.[8] In 1918, Starker[3] described the first case of ‘‘haemophilic pseudotumor’’ in a 14-year-old boy with haemophilia who presented with a swelling of the right thigh and cortical destruction of the femur that had been thought to be osteogenic sarcoma clinically, but was not confirmed as such on histopathological evaluation. The pathogenesis of haemophilic pseudo-tumour is thought to be a pressure necrosis phenomenon where unresolved bleeding into a closed subperiosteal space or interosseous bleeding may cause enough pressure to induce necrosis of bone. Haemorrhage associated with haemophilia usually occur in three distinctive anatomic sites: a) haemarthosis in joints which leads to disabling arthropathy; b) bleeding in the soft tissues, which may compress the surrounding nerves leading to neuropathy; and c) in the proximity of bone, which leads to formation of pseudotumor.[9]

A pseudotumor may remain asymptomatic and unchanged and might be an accidental discovery on radiographs. Sometimes, in cases of severe FVIII deficiency, there can be sudden exacerbations leading them to become the source of active bleeding and bony perforation. Repeated haemorrhage into these closed spaces is thought to cause the bony changes associated with pseudo-tumour.

The radiological picture is non- specific and the skeletal changes consist of areas of bone destruction and new bone formation. It can present as a well-defined unilocular or multilocular lesion with scalloped border and trabeculations . Cortical thickening or sclerosis may be present due to subperiosteal reaction. Important differential diagnosis include malignant or metastatic osseous tumours, giant cell reparative granuloma, Langerhans’ cell histiocytosis, aneurysmal bone cyst, solitary bone cyst, Brown’s tumour and osteomyelitis.[8]

Treatment of haemophilic pseudo-tumours varies from conservative approach and supportive therapy to surgery and radiation depending on the presenting features. Supportive care consists of immobilization and factor replacement. This management seems to be successful in lesions of recent onset. Ahlberg in 1975 studied seven cases of haemophilic pseudo-tumour for 10 years.[10] He showed that adequate and early factor replacement therapy will prevent occurrence of the pseudo-tumours, with replacement advocated when factor VIII or IX was reduced to 50% of normal levels.

Surgical treatment has been recommended only when conservative treatment methods have failed. Before the availability of factor VIII transfusions, surgical intervention was usually fatal. Coordination with the haematologist at all times is essential and factor levels should be closely monitored during and after the surgical procedure to maintain factor VII/IX levels at least above 50%.

Radiation therapy is reportedly useful in cases in which an inhibitor to factor VIII has developed and thus, achieving haemostasis is difficult even in the presence of replacement therapy. Inhibitor to factor VIII has been reported to develop in 15%of patients with factor VIII deficiency.[11]

Recurrent lesions can be the result of incomplete excision, or postoperative hematomas that repeat the pathologic process.

Conclusion

With such rare incidence and non-specific radiographic picture, we aim to make clinicians aware of the possibility of haemophilic pseudo-tumours occurring in the jaws. The first clue to diagnosis is the presence of haemophilia. In the event of no prior or familial history of haemophilia and non-specific histopathological findings in children, intra-bony lesions of the jaws, particularly malignant neoplasms must be considered. Tailoring the treatment of choice for each patient can have beneficial results, provided that adequate factor levels are maintained.

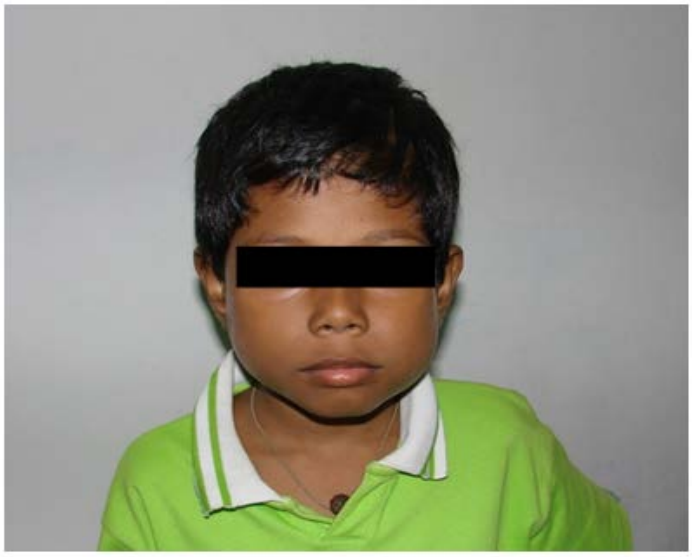

Fig. 1: Bilateral swelling of mandible

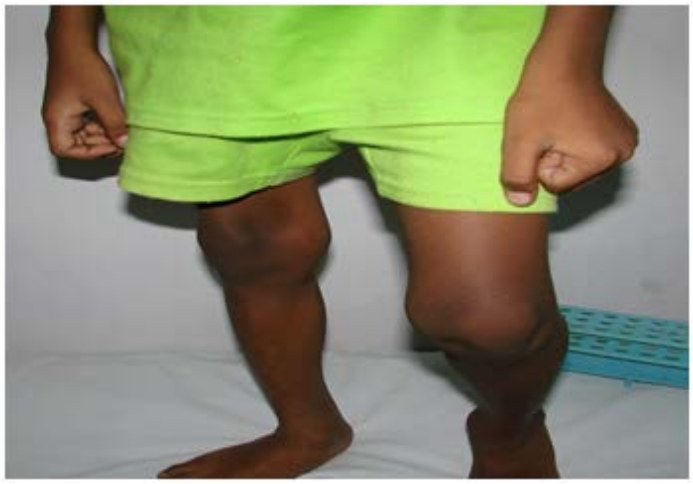

Fig. 2: Haemarthrosis of knee joint

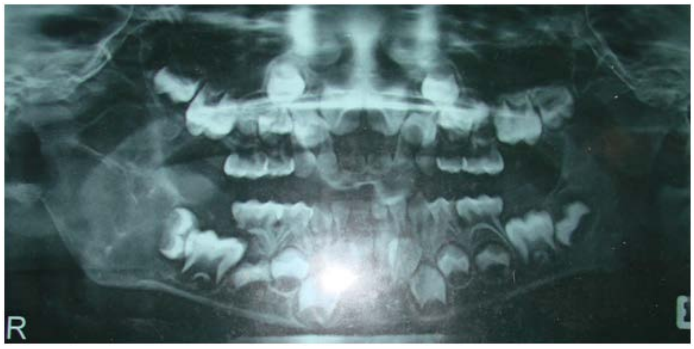

Fig. 3: Orthopantomogram showing the lesions

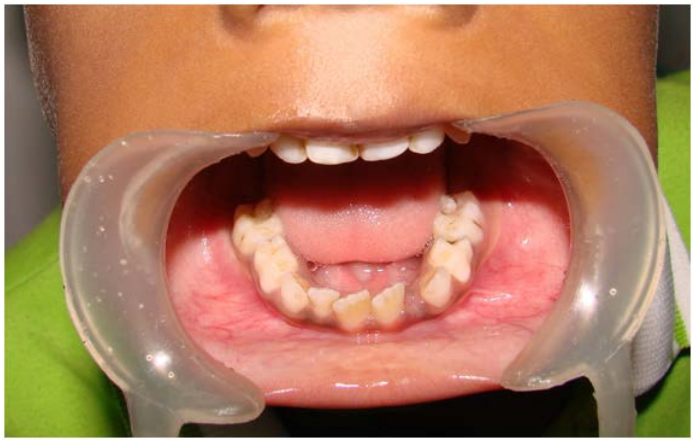

Fig. 4: No intra-oral bleeding observed in the follow up after 3 months

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