Pseudotumour of the Mandible Associated with von Willebrand's Disease

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Abstract Patients with bleeding disorders may occasionally present with pseudotumours. Most commonly these occur in the soft tissues and long bones, and are very rare in the maxillofacial region. We present the clinical details and management of a pseudotumour of the mandible in a 12-year-old girl with von Willebrand's disease.

Keywords von Willebrand's disease · Pseudotumour · Mandible

Introduction

Patients with bleeding disorders infrequently present with pseudotumours secondary to bleeding into soft tissues, bones less frequently and from joints. This phenomenon was first reported by Starker in 1918 [1]. The common sites of occurrence are the soft tissues, pelvis, femur, tibia and other long bones [2]. Pseudotumours of the maxillofacial region are rare [3]. While pseudotumours of the maxillary sinus have been described in a patient with von Willebrand's disease (vWD), mandibular pseudotumour has not been reported in vWD.

We describe here a case of pseudotumour of the mandible in a child with vWD, type III.

Case History

A 12 year old girl presented to the Dental and Oral Surgery Department, Christian Medical College, Vellore, with a history of swelling in the right side of the jaw for the past 5 months with associated intermittent pain and intraoral oozing of blood. On physical examination there was a diffuse swelling in the right side of the mandible. The swelling was firm and mildly tender. There was cortical expansion on the buccal side and mild continuous ooze from the inflamed gum line. Intraoral examination revealed obliteration of the vestibule over the region of the 1st and 2nd right molar area with clotted blood surrounding the area. On palpation there was no mobility of the permanent teeth whereas retained deciduous canine and molar demonstrated grade I mobility. A panoramic radiograph revealed a multilocular radiolucency in the apices of the first and second molars on the right side along with lower border expansion and no obvious areas with perforation. Root resorption was also noted in the second molar (Fig. 1).

The family gave a history that since she was a year old, the patient has had intermittent bleeding from the gums on brushing the teeth and prolonged bleeding after injury. A similar history was present in her elder brother and a maternal uncle.

She was evaluated by the haematologist and was found to have normal platelet count and morphology, prolonged bleeding time, normal prothrombin time, prolonged activated partial thromboplastin time with decreased levels of Factor VIII C, von Willebrand factor (vWF): Ristocetin Cofactor (RCo) and undetectable levels of vWF: Antigen (Ag). Based on these findings she was diagnosed to have vWD Type III.

A differential diagnosis of giant cell lesion and odontogenic tumour was considered. Intra-bony biopsy was

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Fig. 1 OPG at presentation

Fig. 2 Pre OP OPG second visit

Fig. 3 Pre OP lateral oblique view of mandible

planned. Under local anaesthesia and with factor support during and after the procedure, the right lower first molar was extracted and the cystic lining of the cavity was curetted out. Post operatively haemostasis was achieved. Histopathologic examination of the lesion revealed parts of a cyst wall composed of fibrocollagenous tissue with foci of recent haemorrhage, fibrinous deposits and scattered

Fig. 4 Slide showing hemorrhage and bony trabeculae

Fig. 5 Post OP PNS view of mandible

osteoclast type multinucleate giant cells. There was focal evidence of new bone formation and no evidence of malignancy.

The patient presented 13 months later with similar complaints of a swelling in the right side of the jaw and bleeding inside the oral cavity of 10 days duration. Facial asymmetry was present due to the swelling. A lateral oblique view showed multilocular lytic lesions with thin cortices of the inferior border of the mandible with further anteromedial extension of the lesion, involving the third molar toothbud posteriorly and anteriorly involving the second premolar tooth (Figs. 2, 3).

With factor support, segmental resection of the right body of mandible was done and reconstruction was carried out using a titanium plate under general anaesthesia. Despite other haematological factors being normal, mild

Fig. 6 Post-operative lateral oblique view

ooze was noted postoperatively from the extra oral surgical site. On the second post-operative day the extra oral site was re-explored, a bleeder was identified and homeostasis achieved. Thereafter, there was no further ooze or bleeding and she was discharged from the hospital on the tenth post-operative day. Histopathology revealed fibrocollagenous tissue with inflammatory granulation tissue, extensive areas of recent and old hemorrhage, necrosis and mild chronic inflammation composed of lymphocytes, histiocytes and few plasma cells. Bony trabeculae were also present (Figs. 4, 5, 6).

She was reviewed a month after her last surgery at which time she had no complaints and both the intra and extra oral surgical sites had healed satisfactorily.

Discussion

von Willebrand disease (vWD), first described by Dr. Erik von Willebrand is the most common inherited bleeding disorder [4]. It is caused by deficient or defective plasma von Willebrand factor (vWF), which mediates the adhesion of platelets to sites of vascular injury and binds and stabilizes the procoagulant protein factor VIII. Defective vWF interaction between platelets and the vessel wall impairs primary haemostasis leading to heavy and continuous bleeding after an injury. vWD is divided into three major categories with type III (total deficiency) being the least common and manifesting with severe mucocutaneous and musculoskeletal bleeding.

Pseudotumours, also called “blood cysts”, are very rare complications occurring in about 1–2 % of patients with

severe bleeding disorders [5]. They occur in affected individuals who have experienced multiple episodes of bleeding over several years. The classic presentation is painless swelling with subsequent bony destruction. The ‘tumour’ consists of extravasated clotted blood surrounded by a fibrous capsule [5, 6]. Histological examination of the specimen usually reveals organised fibrous tissue, debris, blood clots, bone fragments, neovascularisation, osteoid neoformation, multinucleated giant cells and foreign body type cells [5].

Pseudotumours usually occur in the long bones where trauma is most likely to occur [5], and may be as a result of subperiosteal or intra osseous bleeding. Intra muscular pseudotumours may be seen in areas of greater mobility and more prone to minor trauma such as the major flexors of the limbs, the calf, and forearm flexors. Our review of literature suggested that pseudotumours of the mandible or maxilla, occurred in young patients rather than in adults [2, 3, 7–10]. It has only been described in the dentate region of the mandible and not elsewhere. The possible reasons for pseudotumours arising in the mandible are not clear. Though trauma has been established to be an important etiological factor, sometimes these may occur in the absence of a history of trauma [8]. Tooth eruption is known to cause bleeding [11]; we postulate that the physiologic process of tooth eruption of either the premolar or the second molar itself may have been the cause for pseudotumour developing in the mandible in this case. Fig 1 shows erupting pre molar and resorbed roots of second molar. Repeated bleeding during the eruption of any of these teeth could have led to pressure necrosis and resorption of the second molar roots.

Various conservative modalities of treatment of pseudotumours have been advocated such as embolization of the vessel [12, 13] and radiotherapy [14]. Surgical removal has been shown to be the best option especially in the case of recurrence [2, 5, 15], as in this case where incomplete removal of the lesion during the first attempt resulted in the recurrence of the lesion. But this must be undertaken only with adequate support from a haematology unit [16].

Conclusion

Though rare, a diagnosis of pseudotumour must be considered a possible differential diagnosis in patients with rapidly growing jaw swellings in the context of a bleeding disorder.

It is possible that the eruption of teeth may result in haemophilic pseudotumours, therefore young patients in the teeth erupting stage with a comorbid bleeding disorder should be closely monitored.

When a pseudotumour is diagnosed, surgical treatment, with adequate haematological cover, is considered the optimal treatment choice.

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