Images: Central hemangioma of the maxilla

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Hemangiomas are common benign vascular tumors. They can occur in skin or soft tissues like muscles, tendons, connective tissue, fatty tissue, synovium or bone.[1,2] Of these, central (intraosseous) hemangioma is most often found in the vertebrae and skull. It rarely develops in the jaw and fewer than 50 cases of mandibular hemangiomas and a still smaller number of maxillary lesions have been reported.[3] Our patient showed extremely typical images of a maxillary haemangioma.

Case Report

A 21-years-old lady came with a slow and gradually increasing swelling on the right upper jaw for one and a half years. There was a history of trauma one and a half years ago when the swelling was noticed. There was tipping followed by exfoliation of one of the upper right teeth, following which there was an increase in the size of the swelling. On clinical examination, there was facial asymmetry in the upper anterior region, loss of the nasolabial fold on the right side and incompetence of the lips [Figure 1]. On intraoral examination, a single soft tissue mass, measuring approximately 4.0 x 3.0 cm was seen labiopalatally extending from the right midline to the mesial aspect of the right upper second premolar.

There was extrusion and palatal tipping of the right upper central incisor and pathological migration of the right upper canine and the first and second premolars with a missing right upper lateral incisor, due to exfoliation [Figure 2]. The swelling was bony hard and non-tender on palpation.

The occlusal radiograph and orthopantomogram (OPG) showed a mass with a ground-glass appearance, with a coarse trabecular pattern, extending from the right upper central incisor to the right upper first molar. The ground-glass appearance extended up to the inferior border of the maxillary sinus, as seen on the OPG [Figure 3]. There was expansion of the external cortical plate and palatal extension bilaterally from the right upper first molar to the left upper first molar [Figure 4]. There was pathological displacement of the upper right central incisor, canine, first and second premolars with a missing upper right lateral incisor.
A provisional diagnosis of fibrous dysplasia or adenomatoid odontogenic tumor was made. On fine needle aspiration cytology (FNAC), there was a spurt of blood from the swelling. A CT scan was then performed to further study the lesion, using a spiral CT scanner (Somatom, Siemens, Erlangen), in the axial and coronal planes. It showed an expansile, multiseptate mass with a honeycomb appearance, arising from the right side of the maxilla, involving the alveolar margin, the hard palate and the floor of the right maxillary sinus with extension across the midline, though no extension was seen into the maxillary sinus. The cortical margins were well-defined, with intact but thinned inner and outer cortices. No obvious destruction of the bone was seen [Figures 5, 6]. There was no associated soft tissue mass. No periosteal reaction was seen. On the basis of the radiological and FNAC findings, a provisional diagnosis of hemangioma or aneurysmal bone cyst was made.

An incisional biopsy was then performed and histopathology showed large blood sinuses with thin walls, each showing endothelial lining with and sinusoidal spaces filled with blood [Figure 7]. The diagnosis of an intra-osseous cavernous hemangioma of the maxilla was thus made.

Discussion

Hemangioma, a tumor-like malformation is composed of seemingly disorganized masses of endothelial-lined vessels that are filled with blood. Both cavernous and capillary hemangiomas have been described, the former
consisting of large blood-filled lakes, the latter of masses of proliferating vessels of capillary dimension.[4] In our patient, the hemangioma was of the cavernous type.

Hemangiomas have been described in almost all locations in and around the oral cavity and face[4] and may be single or multiple. Central hemangiomas which are more common in the vertebrae and skull, rarely develop in the jaws where they are more commonly seen in the mandible than the maxilla.[3] The lesion may be developmental or traumatic and the developmental lesion may enlarge and can become obvious due to trauma as perhaps must have happened in our patient.[3] These hemangiomas are more prevalent in women than men, the ratio being 2:1, occurring, most often in the first decade.[3] They present as bony hard, non-tender swellings of the jaw.[3] The lesion may cause loosening and migration of the teeth in the affected area. Bleeding may occur from gingival margins around the neck of the teeth.[3]

Both plain radiographs and CT are being used to diagnose and to stage these masses.[6] CT is the best modality to see the exact extension of the tumor.[7] On plain radiographs, the periphery of the lesion may be well-defined and corticated or ill-defined.[8] The lesion often has a multi-loculated appearance because of residual bone trapped around the vessels. Small radiolucent locules may resemble enlarged marrow spaces surrounded by coarse, dense and well-defined trabeculae.[3] These internal trabeculae may form a honeycomb pattern.[9] The formation of linear spicules with a sun-ray appearance can occur and simulate an osteosarcoma.[5] The roots of the teeth in the region of the vascular lesion often are resorbed or displaced.[3]

The CT appearance is variable and in the calvaria most commonly shows a characteristic sharply margined, expansile lesion, with intact inner and outer tables, suggesting a benign lesion with a sunburst pattern of radiating trabeculae.[7] “Soap-bubble” and “honeycomb” configurations may also occur, because of radiolucent enlarged marrow spaces surrounded by coarse, dense and well-defined trabeculae as seen in our patient.[7]

Hemangioma should always be part of the differential diagnosis of multilocular lesions of the jaw. Other conditions include giant cell reparative granuloma, ameloblastoma, adenomatoid odontogenic tumor, aneurysmal bone cyst and fibrous dysplasia. Fine needle aspiration helps before a formal biopsy, as biopsy can lead to torrential hemorrhage in the case of a hemangioma.[6,8] Central hemangioma should be treated without delay, because trauma that disrupts the integrity of the affected jaw may result in lethal exsanguinations.[6] Treatment of hemangiomas continues to be a big problem fraught with the danger of uncontrollable hemorrhage.[4] Besides conventional surgical excision, cryosurgery, laser surgery after injection of sclerosing solution or intravascular embolization with plastic spheres, are some of the commonly used and successful approaches.[6,8]

References


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