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Giant cell tumor of maxilla – A clinical rarity

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Abstract

Background: Giant cell tumor (GCT) is an aggressive but benign neoplasm that constitutes 5% of all primary bone Tumors. The frequency of occurrence is 25% in the epiphyses of long bones and 2% in the craniofacial bones commonly in the sphenoid, ethmoid and temporal bones. Their occurrence in the maxilla is notably uncommon in the craniofacial skeleton

Case presentation: In this case report, we present a rare case of Giant cell tumor of maxilla in a 28-year-old, young man who reported to our department with a painless swelling of right anterior maxilla region. An osteolytic radiolucent lesion with poorly defined margin was evident in the occlusal radiograph and CBCT scan. An incisional biopsy confirmed it to be a case of Giant cell tumor

Conclusion: The present case needs emphasis because the clinical and radiographic presentation resembles a malignant tumor and definitive diagnosis should be always based on histopathological findings. Giant cell Tumors may undergo malignant trans formation and metastasis

Keywords: Giant cell tumour, maxilla, osteolytic, multilocular

Introduction

Giant cell tumor (GCT) is one of the most common benign bone Tumors, which occurs in young adults 20-40 years old with a high recurrence rate and a potential for aggressive behaviour. Approximately half of the Tumors occur in adults in their third and fourth decades of life. It is rarely seen in patients older than 50 years.

The GCT presents as slow growing tumor, with diverse symptoms depending on the location of primary lesion; symptoms include swelling, pain, epistaxis, neurological deficits, proptosis, visual defects, tinnitus, and malocclusion. Here we present a case of Giant cell tumor of maxilla in a 28-year-old male patient who reported to our OPD.

Case Report

A 28-year-old male patient reported to our OPD with the chief complaint of a painless swelling in the right cheek region since 3 months. The swelling was of insidious onset and increased gradually to the present size. There was a history of trauma to the face 15years ago. There was no history of pain, paresthesia, nasal discharge, epiphora, loosening of tooth or systemic symptoms associated with the swelling. He had the habit of chewing tobacco since 7 years. Extraoral examination revealed a diffuse swelling lateral to the ala of nose on the right side with obliteration of right nasolabial fold. (Figure:1) It was bony hard in consistency and nontender on palpation. The overlying skin was normal and no local rise in temperature was noted. On Intraoral examination, a well-defined solitary swelling of size 2×2 cm noted in the labiogingival sulcus of 12,13,14 region (figure 2). It extends anteriorly from the distal aspect of 12 to 14 posteriorly. The mucosa over the swelling was intact, smooth with normal color and texture. On palpation, the swelling was non tender, non-fluctuant and bony hard in consistency. Ellis class II fracture was noted in 13 and was non responsive on electric pulp vitality test. Based on this clinical finding, a provisional diagnosis of periapical cyst was made. Occlusal radiograph revealed a ill-defined radiolucent lesion with altered trabecular pattern extending from the distal aspect of 12 to the mesial aspect of 14 region (figure 3). The root of 12 is displaced mesially and 13 distally by the lesion. CBCT showed an expansile multilocular radiolucent lesion noted in the right anterior maxilla region (figure 4a,4b,4c). It extends anteriorly from the distal aspect of 11 to distal aspect of 14 posteriorly. Superiorly there is destruction of floor and lateral wall of right nasal cavity and involves the anterior aspect of the right maxillary sinus. Inferiorly it extends to involve the alveolar crest of 12,13,14. Root of 12 is displaced mesially by the lesion. There was loss of lamina dura on the mesial aspect of 14, mesial and distal aspect of 13 and distal aspect of 12. Bi cortical expansion with perforation noted in 13 regions. On the basis of clinical and radio logical findings, a provisional diagnosis of adenomatoid odontogenic tumor was made. Incisional biopsy of the lesion was done and the tissue was sent for histo pathological examination. Histo pathological report showed a giant cell lesion permeating through the bone. It composed of clusters of osteoclasts like giant cells set in a mononuclear stroma. Nuclei of both the giant cells and stromal cells were identical suggestive of Giant cell

tumor. The patient was managed with wide enbloc resection and currently under follow up.

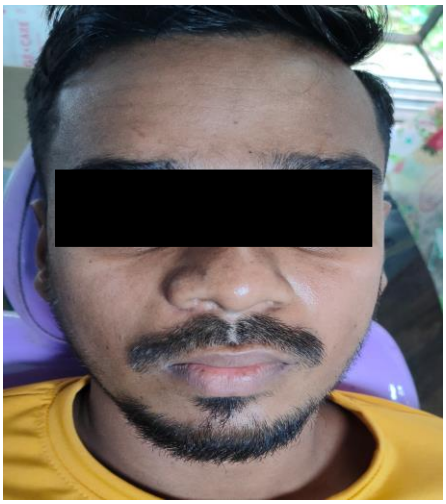

Fig 1: Diffuse swelling noted lateral to ala of nose on the right side obliterating the nasolabial

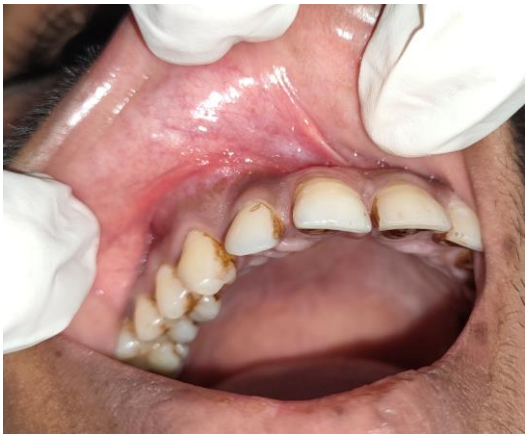

Fig 2: swelling noted in the labiogingival sulcus of 12,13,14

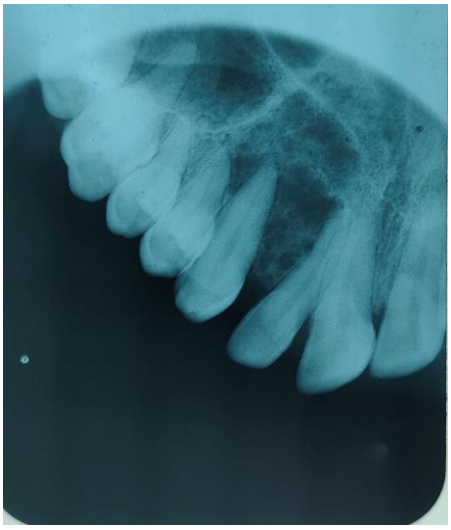

Fig 3: ill-defined radiolucent lesion with altered trabecular pattern extending from the 12 to 14 with displacement of roots of 12 and 13

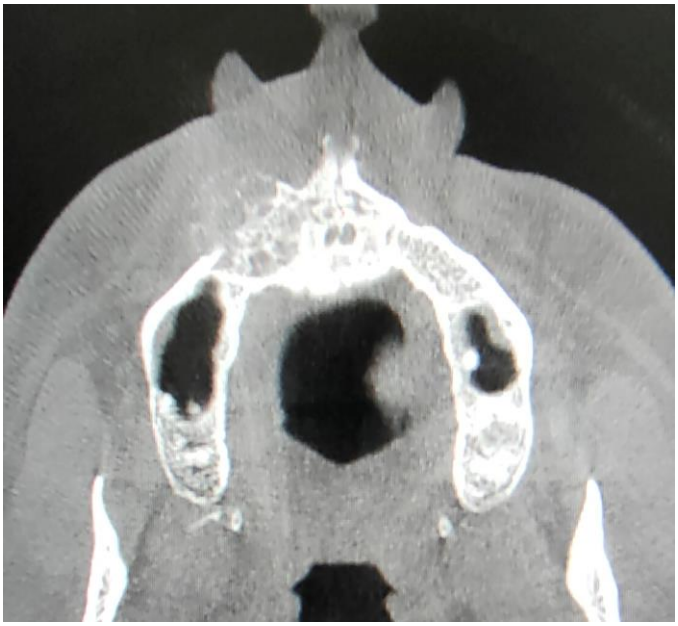

Figure 4a: CBCT axial view

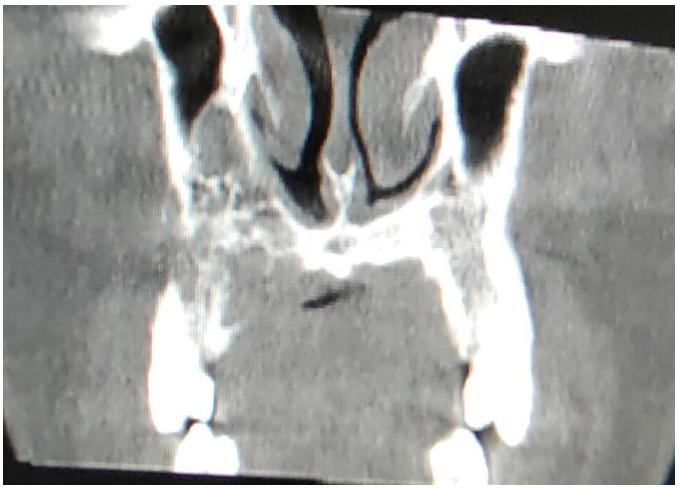

Figure 4b: CBCT coronal view

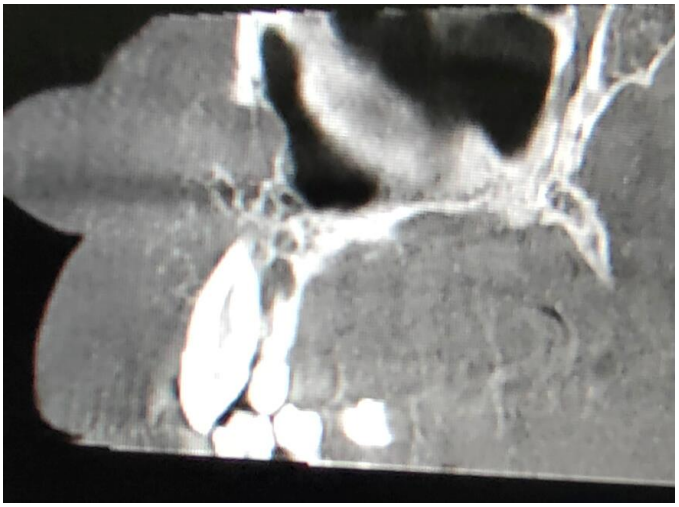

Figure 4c: CBCT sagittal view

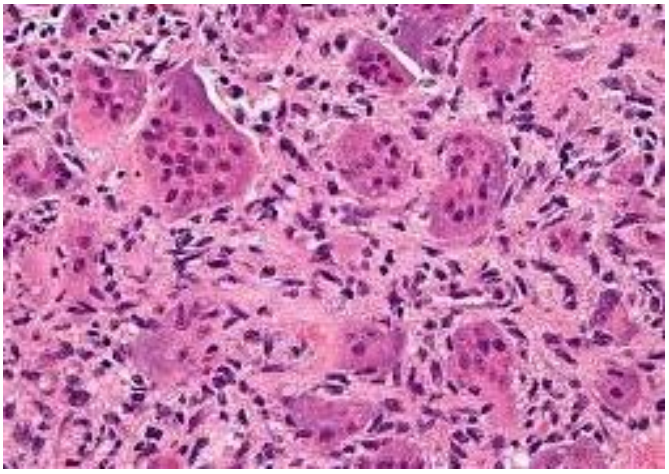

Figure 5: photomicrograph showing areas of clusters of osteoclasts like giant cells set in a mononuclear stroma.

Discussion

Giant cell tumor of bone (GCT) was described by Cooper and Travers in 1818. It is a true neoplastic process originating from the undifferentiated mesenchymal cells of the bone marrow, characterized by a profuse multinucleate giant cell scattered throughout the stroma of mononuclear cells. These giant cells have some similarity with osteoclasts, and so are called osteoblastoma. It is generally considered as benign but severe bony destruction may result occasionally depending on the location and clinical presentation of the tumor, making tumor management very challenging. Although benign, these Tumors exhibit locally aggressive behaviour and own the potential to metastasize. Metastases most frequently occur in the lungs. Varying degrees of local aggressiveness, like a simple cortical breakthrough, extension into surrounding soft tissues, and articular structures can cause severe and debilitating local complications. The risk of recurrence is approximately 35%. The incidence of the GCT is 5% of all primary bone neoplasms. Although only 2 to 4% of all GCTs occur in the head and neck. The case report described here has its presentation in the right side of maxilla. There are few reports found in the literature describing involvement of jaw bone by this tumor. This adds one more aspect in diversity of clinical and biological behaviour of GCT in maxillofacial region. The exact etiology of GCT is not fully understood yet. It remains uncertain whether it is a true neoplasm or just a reactive condition. A 20q11 amplification is seen in 54% of GCTs, over-expression of p53 in 20% of them. Centrosome amplification, and boosted telomerase activity with the prevention of telomeres shortening support a neoplastic etiology. Clinically, the lesion usually occurs in young adults. The above description of the clinical age is seen in our case report. The GCT presents as slow growing tumor, with diverse symptoms depending on the location of primary lesion; symptoms include swelling, pain, epistaxis, neurological deficits, proptosis, visual defects, tinnitus, and malocclusion. In this case, GCT arising from the maxilla caused no symptoms. Radiological findings vary from small unilocular lesion to large multilocular lesion with well or ill-defined borders. It may also be associated with cortical bone perforation and root resorption.

The final diagnosis is established only on the basis of a biopsy. Histologically, GCTs are composed of multinucleated giant cells in a vascular stroma of epitheloid or spindle-shaped mononuclear cells, with

peripheral osteoid formation. Various treatment modalities have been used in management of GCT. The treatment of choice is surgical excision. Radiation has been used as a therapeutic modality, but the subsequent development of sarcoma is possible and has been reported Other treatment modalities including cryotherapy, chemo therapy, intralesional steroids, calcitonin, interferon alfa and curettage with adjuvant agents (phenol and methyl methacrylate) have been tried, but have yielded less effective results. Adjuvant treatments (liquid nitrogen, phenol, or HO and argon beam coagulation), topical or systemic bisphosphonates like zoledronate or pamidronate can be used as a novel adjuvant therapy for GCT. Denosumab, a monoclonal antibody, is widely used to treat unresectable GCTs of bone in adults and skeletally-matured adolescents, and acts by specifically binding to RANKL.

Conclusion

Giant cell tumor of maxilla is a rare clinical entity. Numerous bone Tumors have multinucleated giant cells that must be distinguished from conventional GCT. These range from benign lesions such as ossifying fibroma to locally aggressive lesions like CGCG, aneurysmal bone cyst, high-grade sarcomas and also metabolic disorders such as hyperparathyroidism which is disguised by masses of reactive osteoclast-like giant cells. Careful histopathological examination is emphasized on with exclusion of other possible lesions to arrive at the appropriate diagnosis.

Declaration of patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images

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