Central Giant Cell Granuloma of the Maxilla: A Rare Presentation

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Abstract: Giant Cell Granulomas (GCG) are rare tumors affecting the Head and Neck region and other sites of the body. It occurs most commonly in the mandible. The case reported here resembled a wide variety of conditions that led to a misdiagnosis both clinical and radiographic examination but was histopathologically diagnosed as CGCG. We describe a case of central giant cell granuloma arising from the maxilla to highlight to the general dental practitioner the importance of histopathology in the diagnosis of this enigmatic lesion.[S Shah, Natl J Integr Res Med, 2018; 9(2):102-105]

Key Words: Central giant cell granuloma, swelling of maxilla, giant cells

Introduction: Giant cell granulomas (GCG) are rare benign tumors that can affect the head and neck region. GCG comprise 0.1% of all tumors of facial bones. It was first described by Jaffè in 1953. Central giant cell granuloma (CGCG) are defined by the World Health Organization as “an intraosseous lesion consisting of cellular fibrous tissue that contains multiple foci of hemorrhage, aggregations of multinucleated giant cells and occasionally trabeculae of woven bone”. CGCG of the jaws mainly occurs in children or in young adults, with a predilection for females. It is more common in the mandible than in the maxilla, and can be confined to the tooth-bearing areas of the jaws. Surgery is the traditional and still most accepted treatment for GCGs. The case history reported here presented with a giant cell lesion that involved the right side of the palate.

Case Report: A 41 year old male patient reported to our out-patient department of Oral and Maxillofacial surgery with a chief complain of swelling and difficulty in chewing food from upper right back tooth region since 2 months. Complete case history revealed that the swelling started as small size and progressively increased to the present size over a period of two months. Before one month patient had sought treatment from his family dentist who extracted his 16 because of severe mobility but the swelling persisted even after extraction. There was no history of trauma, neurological deficit, and fever, no loss of appetite, nasal discharge or difficulty in swallowing. Patient was systemically healthy. Medical and family histories also not reveal any abnormalities.

Extra oral examination revealed a diffuse oval shaped swelling on the right side of face with ill-defined margins causing slight obliteration of nasolabial fold which results in facial asymmetry. Swelling was about 3*4 cm in size with smooth surface and no change in the color. The overlying skin appears free and normal with no evidence of involvement of facial nerve. Regional lymphadenopathy was absent on palpation. The swelling was firm in consistency and was slight tender on palpation (Figure 1). Mild compressibility can be seen with no reducibility. The swelling had no localized elevation oftometer. No pus discharge with absence of sinus tract or fistulae.

Intraoral examination shows slightly red expansile mass in the region extending anteroposteriorly from distal side of 13 upto mesial side of 18 and mediolaterally from median raphe and extending on buccal side upto the buccal vestibule of 4*5 cm in size. Swelling was not crossing the midline. The swelling had smooth surface which was firm and slightly tender on palpation (Figure 2). The extracted socket 16 was healed with reddened inflamed mucosa with no pus discharge or sinus tract opening. Grade 2 mobility was present with 12, 13 and 18 tooth number. The swelling also caused slight displacement of the upper right anterior teeth. Patient was not having nasal complain.

Cone Beam Computed tomography (CBCT) scan revealed well defined, well circumscribed hypodense area. The mass extended across the arch from teeth 11 to 17 region. Supero-inferiorty from within right maxillary sinus to alveolar crest and medio-laterally perforating both cortical plates (Figure 3). Displacement of roots of lateral incisor and canine was observed with no resorption of teeth was seen in this case.
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Routine haematological and urine examination were normal. On the basis of clinical and radiological examination a provisional differential diagnosis of Periapical Radicular Cyst, Residual Cyst Adenomatoid Odontogenic Tumor (AOT), Desmoplastic Variant of Ameloblastoma Odontogenic Myxoma, Calcifying Epithelial Odontogenic Cyst (CEOC), Fibrous Dysplasia, Browns Tumor of Hyperparathyroidism and Central Giant Cell Granuloma was made.

Intraoral aspiration of the lesion was attempted with a 20-gauge needle under local anesthesia but it did not yield any aspirate. After the investigations were carried out, we ruled out radicular cyst, residual cyst, CEOC and fibrous dysplasia. An unproductive aspirate ruled out cystic lesions like radicular cyst and CEOC. The absence of characteristic ground glass appearance in the radiograph and presence of a radiolucent lesion, eliminated fibrous dysplasia from the diagnosis. Desmoplastic variant of ameloblastoma was also ruled out because it typically presents with a mixed radiolucent and radiopaque appearance within the dense fibrous septa. Negative aspiration did not rule out AOT because this lesion may or may not be associated with aspirate. This lesion was still considered in the differential diagnosis based on the fact that the lesion may appear completely radiolucent or a mixed radiolucent radiographically. Hence based on history, clinical features and investigations we considered AOT and CGCG in our diagnosis.

The case was planned for surgery under general anaesthesia. Through the intraoral approach the lesion was exposed labially from 11 to 17 region and palatally from 23 area to 17 region. Enucleation with extensive curettage + peripheral ostectomy was done. During surgical procedure, the lesion was found to be communicating with maxillary sinus and involving the right nasal floor. Cystic fluid was not encountered during surgery. Good amount of bleeding was encountered during this procedure. Maxillary sinus and palatal region was extensively curetted taking care not to injure the greater palatine vessels while curetting the palatal region. Haemostasis was achieved and the entire curetted area was toileted with betadine. Nasal antrostomy was done and the pack was removed from right nostril after 48 hours. The specimen was brown in color and firm in consistency (Figure 4). No signs of recurrence was found in 18 months period of follow up.

Histopathological examination of the surgical specimen confirmed the CGCG diagnosis. In a background rich in multinucleated osteoclastic-like giant cells within a vascular stroma with thin walled capillaries adjacent to the giant cells (5 to 10 nuclei /
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Histologically, the features of CGCG are indistinguishable from brown tumor of hyperparathyroidism and giant cell lesions, but biochemical tests such as serum calcium, phosphorus, and alkaline phosphatase can be taken into consideration to rule out these lesions. Brown tumor of hyperparathyroidism was excluded by demonstrating normal levels of serum calcium, phosphorus and alkaline phosphatase levels, thus establishing the diagnosis of Central Giant Cell Granuloma of the maxilla.

Discussion: Giant cell granulomas are benign lesions of the craniofacial skeleton. Giant cell granulomas of the jaw bones may be peripheral or central. Peripheral lesions present as pedunculated or sessile lesions on the gingiva while central lesions are endosteal. CGCGs are endosteal and arise from within the cortex, frequently it is only a painless swelling, but growth in some cases is so rapid and the mass can also rarely erode through bone particularly of the alveolar ridge to produce a soft tissue swelling. It can occur at any age but presents most frequently in the 2nd and 3rd decades. It is twice as frequent in females. Trauma has been considered as an important etiologic factor in the initiation of this lesion. The lesions increase by accumulation of tissue which is produced by slow, minute, continuous hemorrhages of multicentric nature due to trauma and somedefect in the capillaries. Despite the fact that the course of the disease is considered benign, there still exist some reports in literature where metastasis has been observed.

Histologically, these lesions are characterized by the presence of numerous multinucleated giant cells embedded in a fibro-cellular stroma oftenfound adjacent to blood vessel walls. Central giant cell granuloma is made up of a loose fibrillar connective tissue stroma with many interspersed proliferating fibroblasts and small capillaries.

Central giant cell granuloma of the jaw usually presents as a painless solitary radiolucent expansion in most of the cases. Some lesions are more destructive with a marked tendency to recur. A more aggressive type of such lesion will require more radical treatment. The recurrence rate is reported to be 13–22%, with most treatment failures manifesting within the first two years of the therapy. The management of CGCG will depend on the clinical and radiographic findings. Generally, curettage of well-defined localized lesions is associated with a low rate of recurrence. In extensive lesions with radiographic evidence of
perforation of cortex, a moreradical excision is mandatory. The medical management of CGCG as an adjunct to surgery includes treatment with steroids or calcitonin which inhibits osteoclastic activity.\(^2\) Harris was the first to report on the use of synthetic human calcitonin as a therapy for CGCG. In a study on 4 patients, a total remission of the lesions was obtained.\(^3\) Interferon-alpha appears useful in the management of aggressive CGCG, presumably due to its anti-angiogenic effects. Bisphosphonates have been administered intravenously in CGCG with promising results.\(^2\)

The clinical behavior of this lesion is quite variable and difficult to predict. Hence we suggest that CGCG should also be considered in the differential diagnosis of the swellings in maxillary posterior area even though it has a marked propensity to occur in the mandibular posterior area.\(^8\)

**Conclusion:** CGCG of maxilla is very rare and constitutes a diagnostic challenge. This site seems to be associated with an aggressive behavior based on the high incidence of bone erosion and recurrence. Radical surgery appears to be the gold standard for treatment with no evidence related to adjuvant treatment or other treatment modalities.

**References:**