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Research Article

AGGRESSIVE CENTRAL GIANT CELL GRANULOMA OF MANDIBLE- A CASE REPORT

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Central giant cell granuloma (CGCG) is a benign intraosseous osteolytic tumor of the jaws with an unknown etiology. Clinically and radiologically, there are two variants, aggressive and non-aggressive. The lesion usually occurs in patients younger than 30 years. Histologically identical lesions occur in patients with known genetic lesions such as cherubism, Noonan syndrome, or neurofibromatosis type 1. Surgical curettage or in aggressive lesions, resection, is the most common therapy. Histologically this lesion consists of cellular fibrotic tissue with multinucleated giant cells, multiple foci of hemorrhage and trabeculea of woven bone.

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INTRODUCTION

Central Giant Cell Granuloma is a benign, rare, non-neoplastic and proliferative lesion of jaw bones.¹ It was first introduced by Jaffe in the year 1953 as a Reparative Granuloma.¹ The term reparative is not in use nowadays because this lesion has been seen to cause destruction of involved bones. The WHO has defined CGCG as an intraosseous lesion which consist of cellular fibrous tissue containing multiple foci of hemorrhage, aggregations of multinucleated giant cells and sometimes trabeculae of woven bone".²

It presents itself as a solitary lesion which radiographically can be seen as a multilocular radiolucency with scalloped margins with a honeycomb or soap bubble-like appearance. It constitute for less than 7% of all the benign tumors of the jaws. The mandible is more commonly affected than maxilla ranging in 2:9-11:9.³ The predilection for female is more than males with ratio of 2:1.⁴

Clinical behavior of CGCG is not constant/variable. It is asymptomatic in most cases with slow growth or may show aggressive nature with rapid hollowing out of bone with cortical expansion, thinning and perforation. It may exhibits root resorption, displacement of adjacent structures including teeth and nerves supplemented with pain. Rate of recurrence is high which is about 15%-20%.³

CASE REPORT

A18 year old female reported to Department of Oral and Maxillofacial Surgery, King George's Medical University with the chief complaints of swelling in her lower left jaw since 3 months. The swelling was small initially and gradually reached to its present size (refer to Figure 7). Her medical and family histories were unremarkable. Patient gave a history of extraction of lower left back tooth region. Extra-orally swelling was evident on left side of mandible measuring 4x5 cm in diameter which was tender and bony hard on palpation .Intraorally the size of lesion was approximately around 1x 2 cm present over lower left region of mandible with overlying mucosa ulcerated and inflamed. (Refer to Figure 1).



Fig 1 Pre-operative clinical photograph

Orthopantogram (OPG) (refer to Figure 2) showed mulitilocular radiolucent lesion in left mandibular body-ramus region, with resorption of dental roots in relation 36 and 37. CBCT (refer to Figure 3) showed relatively well defined osteo-expansile and osteolytic lesion measuring approximately 35.5x39.6x68mm involving ramus and body of left hemimandible. Incisional biopsy was performed and it confirmed with the diagnosis of central giant cell granuloma.

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Fig 2 OPG of the patient



Fig 3 CBCT of the patient

Based on clinical, radiographic and histopathological features the patient was diagnosed as a case of central giant cell granuloma of mandible and was planned for surgical resection under general anesthesia. The surgical procedure was done through an extra oral approach an incision was given on left submandibular region extending 2cm away from midline to left angle of mandible and flap was raised and the lesion was exposed using chisel and mallet and straight fissure bur. The bone was osteotomised and segmental resection was done extending from distal of 34 to left condyle of mandible and reconstruction was done with 2.4mm Titanium angle-condyle reconstruction plate of 16 holes with 4 2.7mm screws (Refer to Figure 4 and Figure 5) and the resected specimen was sent for histopathological examination which confirmed the diagnosis of Central Giant Cell Granuloma.



Fig 4 Exposure of lesion intraoperatively



Fig 5 Reconstruction with Reconstruction plate

Follow-up of the same patient showed restored symmetry of the face with good contour of the mandible on the resected half (refer to Figure 6 and 7)



Fig 6 Post operative clinical photograph



Fig 7 OPG during the post-operative period

DISCUSSION

The Central Giant Cell Granuloma of the jaw is a tumor which rare, benign and have an etiology which is unknown.⁵ The estimated incidence is 0.0001% with 60% of cases which is occurring below the age of 30 with slight female predilection. CGCG is mostly seen in the jaws (anterior region) and often crosses the midline. The mandible is affected more as compared to maxilla and confined to the tooth-bearing areas of the jaws.⁶ The etiology of CGCG is not assured and put is forwarded with many theories for its pathogenesis. It was thought to be a hyperplastic reparative reaction to the trauma induced intraosseous hemorrhage but a proper history of trauma may not be consistently found. Pathogenesis theories includes infectious and repair process, developmental disturbance, or even inflammatory causes but none of them has been accepted. The genetic cause has also been considered, however there is no evidence to support this hypothesis.⁷ In the etiology of giant cell granuloma the role of t(X; 4) (q22; q31.3) has also been stated ⁸

Clinically CGCG can be classified as aggressive or nonaggressive in nature. Aggressive lesions are those exhibiting size equal to or greater than 5cm with rapid growth. They might show tooth displacement, root resorption, cortical bone thinning and recurrence after curettage (Chuong et al). When compared to non-aggressive lesions, the aggressive lesions show high recurrence rate, however no histological difference has been noticed9. Nearly 70% cases of CGCG are of a nonaggressive nature which are asymptomatic, slow-growing and the remaining 30% displays aggressive and gradually destructive behavior.¹⁰ The clinical behavior as already mentioned is variable in case of CGCG. It ranges from slow growing asymptomatic swelling to an aggressive lesion. Generally the swelling is painless.⁸ However the swelling may be associated with facial asymmetry and can cause difficulty in mastication.¹ Only in about 25% cases the lesion may be painful. Sometimes it may be a by chance finding seen in the radiograph that has been taken for any other purpose. The

teeth associated with the lesion may show mobility however uphold their vitality.⁸

Radiographically the lesion of CGCG shows variability in its appearance. It may present as a Unilocular or a multilocular lesion with solitary radiolucency. Multilocular appearance is more commonly seen with the formation of septa in locules by calcifications.¹¹ Most of the cases of CGCG are seen in mandible with their epicenter lying in the region of first molar in young patients however, after first two decades of life the epicenter may be seen in posterior region. Anterior to canine is most commonly affected region in maxilla. Borders may be Well-defined or ill-defined along with expansion and destruction of cortical plates. The lesion may show fine granular bone pattern internally accompanied with straight, coarse and wispy septa. In some cases resorption and displacement of teeth may also be seen ¹²

CGCG histologically consist of two different types of cell population that are multinucleated giant cells and spindle shaped stromal cells irregularly distributed in collagenous stroma.¹⁴WHO has histologically defined CGCG as an intraosseous lesion which consist consist of fibrous tissue with multiple foci of hemorrhage, aggregations of multinucleated giant cells and occasionally trabeculae of bone.10In CGCG multinucleated giant cells that are found to be of a foreign body type/osteoclast-like which has approximately 30 nuclei and equally scattered around the lesion¹¹³The connective tissue stroma of CGCG shows some areas of hemorrhage with hemosiderin deposits, aggregates of inflammatory cell and fibrosis can be demonstrated. Blood vessels which are interspersed are also seen⁴

The CGCG differential diagnosis includes aneurysmal bone cyst, benign chondroblastoma, and brown tumor of hyperparathyroidism, cherubim, fibrous dysplasia, non-osteogenic fibroma, osteosarcoma and true giant cell tumor.⁷ Above mentioned lesions should be distinguished from CGCG before confirming the diagnosis.¹⁴

Both surgical as well as non-surgical treatment has been included in the treatment of CGCG ¹³ The treatment which is included in the surgery comprises of simple curettage, curettage with peripheral ostectomy, enucleation, and en bloc resection. The management of Aggressive lesions is by curettage. Successful results have been reported with the use of intralesional injection of corticosteroids.¹⁴ Calcitonin therapy has been seen to act as important role in the management of CGCG. The recurrence rate of CGCG is between 11% and 49% in case of curettage alone but it has been reported that aggressive subtype of this lesion has a high recurrence rate of nearly 72%.

CONCLUSION

CGCG is a rare disease of jaw with a locally aggressive behavior. Hence, correct diagnosis is established by correlating clinic-radiographic and histological features. Proper biochemical investigations must be done to rule out hyperparathyroidism. Surgery is the conventional and most accepted treatment but can be combined with local injection of steroids and calcitonin in order to avoid recurrence.

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