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FAMILIAL CENTRAL GIANT CELL GRANULOMA INVOLVING BOTH JAWS BILATERALLY: A RARE CASE REPORT

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ABSTRACT

BACKGROUND: Central giant cell granuloma is a benign, idiopathic proliferative lesion of the jaw. This lesion may be of two types: Aggressive or Non-Aggressive based on the pain, paraesthesia, root resorption and recurrence. **AIMS:** We have reported a rare case report of a 12 year old child with multiple CGCG of jaw involving posterior part of both the jaws bilaterally and showing familial genetic predisposition, as patient's whole maternal side of his family including his mother, 5 of his uncles and 3 of his aunts were involved with similar type of jaw lesions. **RESULTS:** Good bone formation was noted in postoperative OPG after 6 months of follow-up and no recurrence was noted. Mild paresthesia on both sides of the mandible at the angle region was observed for 3 months post-op but it gradually improved on successive follow-ups. **CONCLUSION:** In the literature, there are few studies showing genetic relationship of CGCG, so further study is needed to know familial genetic relationship of CGCG. In our case, we can clearly notice looking at the family history, that a definite genetic inheritance of CGCG is present.

INTRODUCTION

Central giant cell granuloma is a benign, idiopathic proliferative lesion of the jaw which occurs most often in the mandible.[1] It occurs mainly in the younger age group below 30 years and mainly occurs in anterior mandible which can cross the midline.[1, 2] Its etiology is mainly unknown, but has been reported to be associated with trauma, inflammatory cause or genetic predisposition.[3] CGCG is defined by WHO as an intraosseous lesion consisting of cellular fibrous tissue that contains multiple foci of haemorrhage, aggregations of multiple nucleated giant cells, and occasionally trabeculae of woven bone.[1-4] It contributes to 7% of all the benign jaw lesions, having two types of biological behaviours, either nNon-aggressive or Aggressive based on severity of pain, root resorption, paresthesia and tendency to recur spontaneously after excision.[4, 5] In the literature, we have noted that a very few cases are reported with multiple CGCG of jaws with familial involvement.[6] Here we are reporting a case report of a 12 year old child with multiple CGCG of jaws involving posterior part of both the jaws bilaterally and showing familial genetic predisposition, as the patient's whole maternal side of his family including his mother, 5 of his uncles and 3 of his aunts were involved with similar type of jaw lesions.

Case history

A 12 year old young boy presented to our department OPD, with a painless bilateral jaw swelling since 4 months. He was apparently asymptomatic 4 months back until his parents noticed a small swelling over his right side of gums and palate. He also complained of bleeding from the lesion due to repeated self-induced trauma when patient used to close his jaws in occlusion. The lesion expanded so much that it came at the occlusal level. Patient visited some nearby private dental clinic, where he was managed by some medication for 15 days but didn't get any relief. After that, the patient was immediately referred to our centre. Patient was then examined, evaluated and adviced for an orthopantamogram (OPG) where we noticed a bilateral multilocular radiolucent lesion involving posterior part of maxilla and mandible. Patient gave family history of similar type of jaw swelling on his maternal side including his mother, 5 of his uncles and 3 of his aunts affected with a similar type of jaw lesion. They didn't seek any treatment for the same, they are still having the lesion which is growing gradually. Patient father was free from any lesion. All of his uncles, and aunts were above 30 years of age. (fig. 1) (fig. 2)

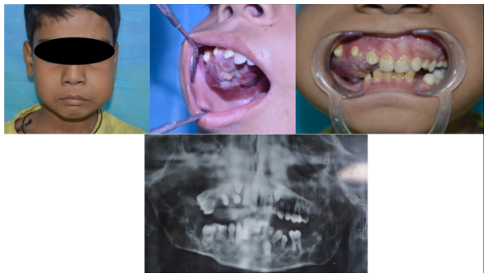

Fig. 1- Pre-op clinical and radiographic pictures

Fig. 2- Patient with his family photographs, which shows involvement of similar jaw lesion in his mother, and maternal side relatives but absent in his father.

Patient is having low socioeconomic status and doesn't wants to undergone genetic study of the lesion. All blood parameters were within normal limit. Based on these features, differential diagnosis may include, Central giant cell granuloma, Odontogenic keratocyst, Gorlin-Goltz syndrome, Cherubism, Neurofibromatosis I, and Noonan's syndrome. After proper consent taken from the patient, incisional biopsy was done, which came with histopathological report of Giant cell reparative granuloma. Patient was then given intralesional corticosteroid injections once weekly for 2 month. But lesion growth did not subside and no changes were found in radiographs. The patient underwent surgical curettage of the lesion with peripheral ostectomy under GA. The lesions on the right side of both the jaws were treated first surgically and then lesion present in the left side were treated after 2 months under General Anaesthesia. Specimen collected after curettage were also sent for histopathology, which also was diagnosed histopathologically as central giant cell granuloma. After curettage maxillary bony defects were filled with iodoform dressing and mandibular site was closed primarily. After surgery, post-operative OPG was done at regular interval of 1 day, 2 month, and 4 month and 6 month, we have found good bone healing with good bone formation during successive follow-up. (fig. 3)

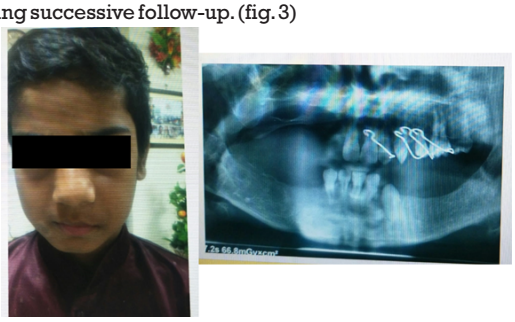

Fig. 3- Post-op clinical and radiographic pictures (OPG) showing good bone formation 6 months post-op and no recurrence noted.

RESULTS

Postoperative recovery of the patient was uneventful and smooth. Good bone formation was noted on postoperative OPG after 6 months of follow-up and no recurrence was noted. Mild paresthesia in both sides of the mandible at angle region was observed for 3 months post-op but it gradually improved on successive follow-ups.

DISCUSSION

In 1953, Jaffe was the first who described central giant cell granuloma.[1, 2] Jaffe described this lesion as giant cell reparative granuloma of the jaw bones and he postulated that it is not a true neoplasm instead it results from a local reparative reaction.[1,2] In most cases, it presents as a painless, slow growing swelling of the jaw.[3] Aggressive CGCGs are usually painful, show root resorption, cortical destruction of involved bone, and further involvement of soft tissues.[1] CGCG occurs more commonly in females as compared to males.[4, 5] Central giant cell granuloma (CGCG) exhibits variable clinical and histological features.[4-8] Radiological features of this lesion are also not unique and could be easily confused with other jaw lesions like fibrous dysplasia, brown tumour of hyperparathyroidism, Aneurysmal bone cyst and other fibro-osseous lesion.[2, 3] Several studies reports that in 60% of the cases of CGCG it is multilocular.[2,3]

It is believed that occurrence of CGCG with a known genetic origin are mainly neurofibromatosis type I, cherubism, and Noonan's syndrome.[5-7] But these lesions can be differentiated from CGCG by clinical features, histological features and involvement of other parts of the body.[7] Cherubism is found to be due to genetic alteration in the SH3BP2 gene on the short arm of chromosome 4.[7] VM et al in 2009 reported a case of CGCG with mutation of this gene while some studies reported that no gene mutation was found in CGCG cases.[6, 7] Histologically this lesion is characterized by abundant evenly distributed multinucleated giant cells within the ocean of mesenchymal spindle shaped stromal cells, distributed throughout the fibro-vascular connective tissue stroma with containing areas of haemorrhage.[4,8] Management of CGCG is variable depends on the severity and extent of the lesion.[5, 7] CGCG can be managed by pharmacological therapy or surgical therapy.[5, 7] Aggressive lesions, recurrent lesion and the lesion which show no response using pharmacological therapy should be treated by surgical therapy.[5] Pharmacological therapy includes Intralesional corticosteroids, calcitonin, interferon alpha, Imatinib, and Denosumab, etc.[5, 7] Surgical management includes curettage with or without peripheral ostectomy, enucleation, and enbloc resection of the jaw along with 0.5 cm safe healthy bone margins.[5] According to various reports surgical treatment is the best option for CGCG at present, similarly in our case, initially we have tried with intralesional corticosteroids injection weekly for 2 months but the lesion did not respond and it gradually kept on increasing in size and no changes were seen in radiographs as well.[5, 7] So, we did surgical curettage of the lesion with peripheral ostectomy which showed great results during post-op period. Various previous study reported that there is known genetic origin of CGCG present. In our case as well, there is definite autosomal genetic relationship present as the patients whole maternal side was affected with similar type of lesions, but our patient refused for any genetic study.[5,7]

CONCLUSION:

A definite genetic study is needed to rule out cherubism, and other genetic syndromes. It is also important to correlate the relationship of CGCG with these syndromes. Central giant cell granuloma involving both the jaws bilaterally is usually a rare case and its familial genetic relationship definitely needs to be found out. In literature, there are some studies showing genetic relationship of CGCG. In our case, we can clearly notice that looking at the family history, a definite genetic inheritance of CGCG is present.

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