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Giant Cell Tumour of Maxilla - A Rare Occurrence in Paediatric Age Group

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Abstract: ***Giant cell tumour (GCT) of the bone conventionally occurs in the epiphyses of long bones. Their incidence in craniofacial skeleton is atypical, constituting only about only 2% in the head and neck region. Their occurrence in paediatric age groups has been seldomly described. Here, we report a case of GCT maxilla in a 6 year old child who presented with oral exophytic lesion. The mass was excised via sub labial approach under general anaesthesia with no recurrence in the follow up period. GCT of head and neck region in paediatric age group is not aggressive and simple excision with follow up is only required.***

Keywords: Giant Cell Tumour, GCT, Giant Cell Reparative Granuloma, GCRG

1. Case History

A 6-year-old female patient presented to the Department of Otorhinolaryngology with swelling over the right side of cheek for 3 months. The swelling was of insidious onset and progressed gradually to the present size (fig 1A-B). The swelling was painless but was causing difficulty in chewing food. No other systemic symptoms were reported. Clinically, the pinkish swelling was visible per orally as it was causing lip approximation not possible. It measured approximately 4x4 cm in size and was covered with smooth mucosa. On intra-oral examination, the swelling was found to be arising from the anterior part of the maxilla with involvement of the upper alveolar margin with grossly malaligned teeth. On palpation, the swelling had a smooth surface which readily bled on touch; it was firm in consistency, non tender and adherent to the underlying maxillary bone. Rest of the oral cavity, neck and systemic examination was clinically normal.

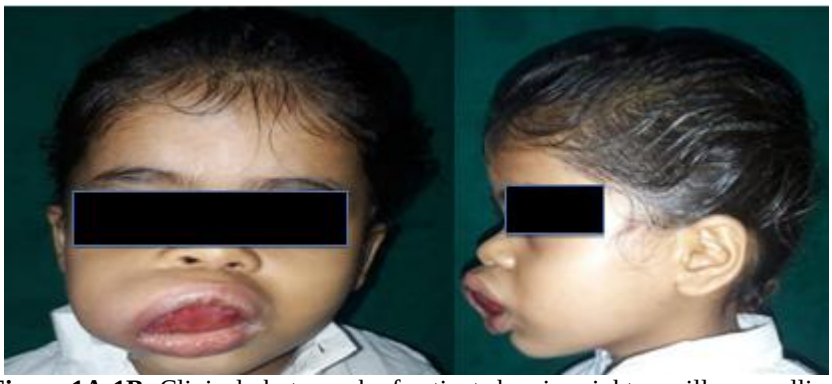

Figure 1A-1B: Clinical photograph of patient showing right maxillary swelling

Contrast enhanced computed tomography (CECT) of face revealed (figure 2A-2B) a well defined heterogeneous expansile lytic lesion arising from the right anterior maxilla causing bony scalloping and destruction of maxilla extending into the adjacent gingivo buccal sulcus. It was avidly enhancing, lytic expansile soft tissue mass measuring 2.4 x 3.5 x 4.3 cm centered in the right maxillary alveolus showing buccolingual expansion and destruction of maxillary alveolus with areas of cortical breach. Anteriorly, it extended into the gingiva-labial sulcus reaching upto the hard palate postero-superiorly causing expansion of the alveolar arch with wide separation of right upper lateral incisor and canine teeth.

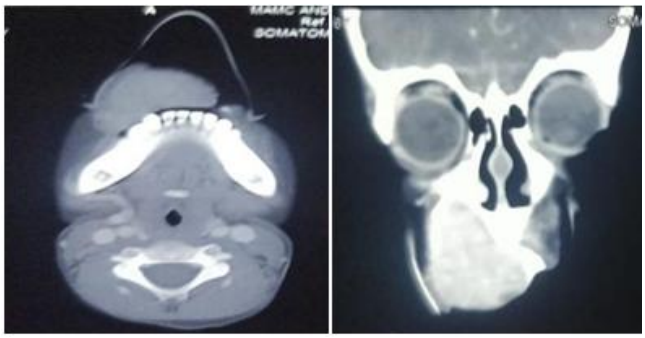

Figure 2A-2B: CECT of face revealing awell defined heterogenous expansile lytic lesion arising from the right maxilla

A biopsy revealed Giant cell reparative granuloma. The hematological and biochemical investigations were unremarkable. Specifically, calcium, phosphate, alkaline phosphatase and PTH assay was done and all were within normal ranges.

Excision of tumour was done under general anesthesia via a right sublabial incision extending from the right upper canine to the right upper premolar was done (Figure 3A-3B). Tumour was excised completely by following a subperiosteal plane from its surrounding bone and sent for histopathological examination (Figure 4). Post operative recovery was uneventful.

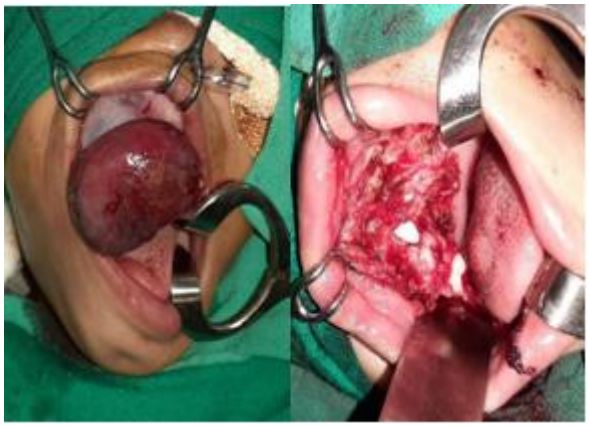

Figure 3A-B: Excision of tumour via right sublabial approach

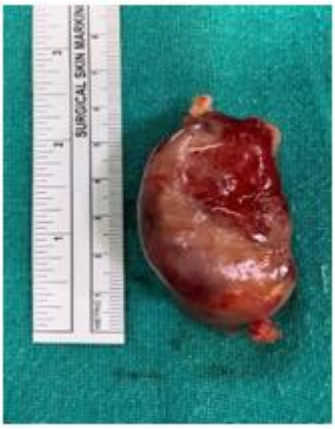

Figure 4: Resected specimen of tumour measuring 2-5 x1 cm

The final histopathology revealed even distribution of giant cells interspersed in cellular stroma consisting of mononuclear cells with prominent spindling which were consistent with GCT (Grade II). The patient was being followed up and there is clinically no evidence of recurrence.

2. Discussion

GCT is a locally aggressive benign neoplasm that is associated with a large biological spectrum ranging from latent benign to highly recurrent and occasionally metastatic malignant bone tumour.[1, 2] It accounts for 4–10% of all bone tumours and typically presents as a lytic lesion in the epiphyseal/metaphyseal part of long bones. It is seen to affect age groups between 15 and 40 years, with a peak incidence in the third decade of life.[3-5] The most common site involved is the distal femur, followed by the distal radius, sacrum, and proximal humerus.

The occurrence of GCT in head and neck region is seldom reported.6Predisposing sites within this region include mandible, maxilla, ethmoid, sphenoid, temporal bone, zygoma and temporal bone. The incidence in children is exceptionally rare and hence its course in the immature skeleton is sparsely described.7

Within the head and neck region, It presents as a swelling which is gradually progressive in nature, often painless with no reported evidence of other systemic manifestations.

Considering the rarity of its occurrence in the head and neck region, it is vital to bring it to light the differential lesions that can occur within this region which could be clinically masquerading as a GCT. Few worth mentioning include Giant cell reparative granuloma (GCRG), Osteosarcoma and Rhabdomyosarcoma, Brown tumour of hyperparathyroidism, Aneurysmal bone cysts and Cherubism. Hence, it becomes imperative to distinguish between these entities clinically, biochemically, radiologically and histologically.

Radiologically, a typical GCT will reveal extensive bone destruction, lytic expansile cortical breach with soft tissue invasion and histologically it shows uniform distribution of giant cells interspersed in cellular stroma consisting of mononuclear cells with prominent spindling in contrast to lesions simulating GCT show foci which contains numerous clumped giant cells alternating with large areas devoid of giant cells.

There is a remarkable resemblance of GCT and GCRG to each other, which were initially considered to be spectrum of the same disease but now widely accepted as they are two distinct lesions. GCRG occurs in the first and second decade and in the jaw it is seen peripherally involving the gingiva and alveolar mucosa or centrally as an endosteal lesion within the jaw bone.8It can be histologically differentiated from GCT by its hemorrhagic fibroblastic background with innumerable giant cells.9 In our case it was initially diagnosed on histopathology as GCRG and later it was on post operative histopathological examination that the tumour mass was confirmed to be a GCT.

Likewise other lesions like brown tumour which typically involves the maxilla or the mandible should be clinically differentiated from GCT. Biochemical laboratory investigation which include serum calcium, phosphate, ALK phosphatase and PTH assay aid in its differentiation.

In our case report as well, the fore mentioned biochemical tests were subjected and yielded an exclusion diagnosis of Brown's tumour.

Tumour resection is the accepted treatment modality which will also depend on tumour location. Marrioni et al. reported performing a maxillectomy with reconstruction using a calvaria and temporalis muscle pedicled flap in an advanced GCT lesion of maxilla.1 Radiotherapy has been used collaboratively for lesions that impede surgical approaches. Surgical resection was found to be curative without the need for an extensive approach which also yielded more cosmetically acceptable results.

The recurrence rate for GCTs occurring within the head and neck region is very minimal but owing to its nature of aggressiveness and malignant potential in other regions of the body, long term follow up is often advised.

3. Conclusion

In conclusion, despite being a rare condition in the immature skeleton, GCT should be considered as a differential diagnosis in children and in all cases of exophytic maxillary swelling. The biological behaviour of tumour in children does not have any significant difference from that which is reported in adults. Surgical resection is found to be sufficiently curative for GCTs occurring within the head and neck region, thereby circumventing the need for more radical approaches which would lead to a cosmetically disfiguring scar. Minimal recurrence is reported for GCTs occurring within the head and neck region.

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