INTRODUCTION

Giant cell tumor (GCT) is an aggressive but benign neoplasm containing spindle-shaped stromal cells, mononuclear round to oval cells resembling histiocytes and abundant evenly distributed osteoclastic giant cells. This lesion 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. We report a rare case of GCT of the mandible in a young patient causing massive resorption.

CASE REPORT

A 12-year-old female patient was referred to the Department of Oral and Maxillofacial Surgery with a complaint of painless swelling of the lower jaw. The patient had noticed the swelling 4 years back which gradually increased to the present size. Extraoral examination revealed the presence of an asymmetric swelling of the mandible causing deviation of the chin to the right side. The skin over the swelling was normal. The jaw movements and the temporomandibular joint (TMJ) movements were normal [Figure 1]. On intraoral examination, the swelling was seen to extend from the right angle to the ramus on the left side. The labial and buccal mandibular sulci were obliterated and buccolingual expansion was evident.

The teeth present were 33, 36, 41, 42, 43, 45 and 46 with multiple missing teeth and displacement of 33, 41, 42 and 43 [Figure 2].

Orthopantomograph (OPG) revealed a multilocular radiolucency extending from the right ramus to the left ramus region with an expansile swelling in the lower border of the mandible [Figure 3]. Apart from the teeth present clinically, impacted second permanent molars on both sides as well as an impacted paramolar on the right side were discerned. Expansion of the cortical plates buccally and lingually was appreciated. Displacement of 33, 41, 42 and 43 and resorption of roots of 41, 42, 43, 36 and 46 could be appreciated.

Visor incision was placed, layer by layer dissection was performed at the subplatysmal layer and flap was elevated to explore the mandible from the right to left condylar region. Facial artery and vein were preserved for the reconstruction procedure. Genioglossus and geniohyoid muscles were resected. The affected portion of the body of the mandible was resected from right to left subcondylar region after making bilateral osteotomy cuts. Right fibular graft harvested by the plastic surgery team was osteotomized at the bilateral canine region, adapted to the titanium reconstruction plate and fixed with screws. Microvascular anastomosis was done. Glove drain and Ryles tube were placed. It was closed with 3-0 vicryl suture and 4-0 ethilon sutures. The resected tumor mass was sent for histopathological examination.

A specimen x-ray was taken which revealed similar radiologic findings as the preoperative OPG [Figure 4]. Multiple sections were grossed from different areas of the specimen [Figure 5]. The cut section shows areas of yellowish white coloration.
along with pale hemorrhagic areas with rough borders. It was friable and soft in consistency measuring 4.5 × 5.5 cm in dimension. Brownish fluid oozed out while grossing (suggestive of cystic degeneration). Fibrous areas intermixed with fatty areas were seen. Right posterior border of the specimen was grossed. It was creamy white in color, hard in consistency, measured 2.7 × 2.3 cm in dimension and contained the second permanent molar.

The histopathological examination of the soft tissue revealed a richly cellular stroma comprising of plenty of evenly distributed multinucleated giant cells [Figure 6]. Few areas of pathologic resorption of bone were also evident [Figure 7].

Figure 1: Extraoral swelling exhibiting asymmetry and deviation of the chin

Figure 2: Intraoral view of the swelling

Figure 3: Preoperative orthopantomograph exhibiting an expansile swelling of the lower border of the mandible

Figure 4: Radiograph of the resected specimen

Figure 5: Grossing of the specimen

Figure 6: A richly cellular stroma with evenly dispersed giant cells (H&E, x40)
High power view showed multinucleated giant cells with agglomeration of around 25–50 nuclei in center surrounded by clear cytoplasm, the proliferating stromal cells were hyperchromatic and pleomorphic [Figures 8 and 9]. Decalcified section of the specimen with impacted teeth showed dentin, bony trabeculae and myeloid tissue. The serum calcium levels and alkaline phosphatase levels were evaluated to rule out hyperparathyroidism.

Based on the characteristic appearance of giant cells, stromal cells findings and lab investigations; a diagnosis of GCT was given.

**DISCUSSION**

GCT makes up 4.9–5% of all osseous neoplasms, 21.87% of the benign tumors of bone and head and neck region accounts for 2% of all GCTs.; Most of the lesions occur in the ethmoid, sphenoid and temporal bones; the occurrence in the mandible is less than 2%. The case being discussed involves the mandible.

The age distribution of GCT is around 20–40 years. The sex distribution is 1.3:1:female:male. The case being discussed occurred in a 12-year-old female. This is in concurrence with the cases reported to have occurred in jaw bones in the literature (based on PubMed search engine). The symptoms were pain, swelling of the affected region, weakness and limitation of the jaw movement associated with elicitation of moderate tenderness and eggshell crackling. It may cause pathologic fracture or atrophy of the muscles. The case was painless with swelling of the affected region without limitation of the jaw movements. Eggshell crackling was elicited, but the case did not exhibit tenderness, pathologic fracture or atrophy of muscles.

The typical radiologic features are an expanding zone of radiolucency either eccentric or central with either cortical thinning or involvement of the soft tissue without a sclerotic margin. It may be either well or poorly marginated. Similar radiologic features are observed in our case which had a multilocular radiolucency with poor margination evincing a stretching of the lower border of the mandible.

The gross appearance exhibits chocolate brown or greyish white foci with yellowish discoloration interspersed with hemorrhagic or cystic areas with a soft and friable consistency. The characteristic histopathology of GCT is well-appreciated in the histopathology of the case being discussed.

The differential diagnoses include central giant cell granuloma (CGCG), aneurysmal bone cyst, hyperparathyroidism and other giant cell lesions.

The frequently described histological differences between the GCT and CGCG are: (i) the rounded, larger giant cells which are uniformly dispersed with an increased number of nuclei which tends to aggregate centrally in the GCT, (ii) fresh hemorrhage and hemosiderin deposits and inflammatory component being found more commonly in the CGCG than in the GCT, (iii) osteoid or new bone formation and increased number of spindled fibroblasts with areas of fibrosis in the CGCG and (iv) the presence of necrotic areas in the GCT, but not in the CGCG. This is summarized in Table 1.

The stroma of the case was very cellular with an almost even distribution of giant cells. The stromal cells were pleomorphic and hyperchromatic. Osteoid or new bone formation, fresh hemorrhage and hemosiderin deposits were not observed. The striking feature was that the giant cells exhibited a general tendency towards central aggregation of nuclei with a peripheral zone of eosinophilic cytoplasm. The histopathological features observed were in favor of GCT.

The normal serum calcium and alkaline phosphatase levels found in our case ruled out hyperparathyroidism. In aneurysmal bone cyst and other such lesions, giant cells are found close to areas of hemorrhage; whereas in our case,
Giant cell tumor of the jaw

Figure 9: A composite photomicrograph of four different fields exhibiting cellular stroma with giant cells (H&E stain, x100). Inset of each image shows high power view of the giant cell in the particular field (H&E stain, x400)

Table 1: Characteristic differences between giant cell tumor and central giant cell granuloma

| Giant cell tumor                                           | Central giant cell granuloma                  |
|------------------------------------------------------------|-----------------------------------------------|
| Giant cells with an agglomeration of nuclei in the center with a peripheral band of cytoplasm | Giant cells with scattered nuclei              |
| Stromal cells are pleomorphic and hyperchromatic            | Nonreactive stromal cells                      |
| Vasculature is evident                                     | Vasculature is more with evidence of fresh hemorrhage and hemosiderin deposits |
| Bone destruction                                           | Reactive bone formation                        |

the giant cells were evenly distributed. The treatment of choice is wide resection of the tumor mass.[4] Other treatment modalities are cryotherapy, chemotherapy and curettage with adjuvant agents. The case underwent resection from right to left subcondylar region. The documented recurrence in the literature after resection of the tumor mass is 7%. The patient is being reviewed regularly and is normal. The documented malignant transformation rate is 1–5%.[6]

CONCLUSION

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.[7-10]

Careful histopathological examination is emphasized on with exclusion of other possible lesions to arrive at the appropriate diagnosis. The normal serum calcium and alkaline phosphatase levels ruled out hyperparathyroidism. The case being discussed was in a 12-year-old female, the age being concurrent with cases occurring in the jaw and in accordance with the female predilection usually observed. Eggshell crackling was elicited, but there was no associated tenderness. Radiologic features were also concurrent with a multilocular radiolucency with poor margination. Histopathologically, the even distribution of giant cells throughout the cellular stroma composed of hyperchromatic and pleomorphic cells, the giant cells with central aggregation of nuclei close to 25–50 all led to the diagnosis being made in favor of GCT of the mandible.

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A consent form has been received from the patient to reveal the identity of the patient.