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CASE REPORT

Extensive Giant Cell Tumour of the Mandible: A Case Report with Review

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Abstract Giant Cell Tumour (GCT) is a rare benign, osteolytic, pseudocystic solitary localized lesion. The lesion is common in skeletal structure but not as common in craniofacial skeleton. They are composed of sinusoidal and vascular spaces filled with blood and surrounded by fibrous tissue septa. There is a controversy as to whether it is a distinct radiological and pathological entity or a pathological change superimposed on a preexisting lesion. We present a case of a 19 year old female patient who reported with swelling and pain in the right mandible associated with pain and gradual increase in size since 4 years. On the radiographic study expansive, multilocular lesion extending to right coronoid process was observed. Incisional biopsy showed the lesion to be a dental cyst, however, enucleation with curettage of the cyst confirmed it to be GCT. GCT are non neoplastic but locally aggressive tumors with occasional rapid growth that may be differentiated from other multilocular lesions like ameloblastoma, giant cell granuloma and sarcomas. There have been reports which have appeared regarding its pathogenesis, response to treatment. However many questions remain regarding its treatment and prognosis.

Keywords Giant Cell Tumour (GCT) · Osteolytic · Pseudocystic · Multilocular

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Introduction

According to Robert Marx, the name of a disease undergoes perpetual change, with good reason due to up-to-date knowledge and information.

The preponderance of evidence shows that aneurysmal bone cyst can be conceptualized as a rapidly proliferative variant of central giant cell tumour. Hence in the following case discussion, Aneurysmal Bone Cyst (ABC) will be referred as Giant Cell Tumour (GCT) [1].

Jaffe and Lichetenstein first described the lesion in the early 1940 in studies of unicameral bone cysts. They are considered as pseudocyst because of lack of epithelial lining [2]. The GCT's had been described under a variety of names viz: haemorrhagic osteomyelitis, ossifying haematoma, osteitis fibrosa cystica, atypical subperiosteal giant cell tumor, aneurysmal giant cell tumor, hemangiomatous bone cyst, subperiosteal bone aneurysm, expansile haemangioma and pulsating giant cell tumor [3]. They are principally located in long metaphysis like the femur and tibia (more than 50% of giant cell tumour) and spine (12–30%) [4]. The incidence of these tumours in facial bone is infrequent, with a 2–12% of all giant cell tumours of the body [5]. In case of craniofacial location, the mandible is more frequently affected than the maxilla with a proportion from 2:1 to 11:9 [6, 7]. The body and the mandibular ramus are the main location with rare case reports in the coronoid process and the mandibular condyle [7].

The age of presentation is the second or third decades of life. [8] The median age of diagnosis is 13 years. 80% of the patients are under the age of 20, with greater sex predilection in female (62%) [9]. Although GCT is a benign lesion, it can behave locally in an aggressive manner because of its rapid growth and osteolytic capacity.

The lesion represents less than 1% of all the bone cysts biopsied. There are many theories about its etiology, mostly referring to alterations of the hemostatic-vascular equilibrium of bone [10]. The best and the most accepted theory of relationship between GCT, Central giant cell granuloma (CGCG), and traumatic bone cyst (TBC) is presented by Hillerup and Hjorrting-Hansen [11], who proposed that these lesions are different manifestations of the same general process, the cause of which is a "vascular mishap" resulting from trauma, primary bone disease or malformation. The rapid growth may result in the erosion of the cortical plates of an asymptomatic slow growth lesion that then becomes symptomatic [2, 5]. The multilocularity with soap bubble or honey comb appearance radiographically, should be differentiated with ameloblastoma, giant cell granuloma, ossifying fibroma, sarcomas. The histologic features consist of a fibrous connective tissue stroma containing many cavernous or sinusoidal blood filled spaces. Surgical curettage and excision have been the treatment of choice. Various modalities have been used in the treatment of GCT including enucleation, curettage, cryotherapy, radiation, resection, amputation [12].

Case Report

A 19 year old female reported to the Department of Oral and Maxillofacial Surgery with a complaint of pain and swelling in the right mandible. The patient first noticed the swelling 4 years ago which gradually increased in size to the present proportions. There was rapid size increase in the last 3 months (Fig. 1). There was a dull aching pain on pressure with no history of trauma, pus discharge, bleeding or regression of the lesion. The extra oral examination revealed a single diffuse swelling of size 10 × 3 cm on right side of the mandible extending anteroposteriorly from symphysis to right angle of the mandible and extending up to the inferior border of mandible. The overlying skin was normal with no visible pulsation. On palpation the swelling was hard in consistency, non-tender, non-compressible, non-pulsatile. No paresthesia was recorded. The expansion on the inferior border was significant. The intra oral inspection and palpation revealed a single well defined swelling from midline to right distal margin of 47 from gingival margin to depth of the buccal sulcus, with no intraoral draining sinus or fistula. There was bicortical expansion, except between 46 and 47. There was lingual tipping of 44, 45, 46, 47 with a dull note on percussion, distal movement of 43. The pulp vitality test revealed non-vital 44. The 48 was not visible in the oral cavity. The provisional diagnosis of benign odontogenic tumor was made. The panoramic radiography revealed multilocular, radiolucency extending from lower left canine to mid ramus region. In the 3D CT (Fig. 2) scan the postero-anterior and lateral view presented a large, well-defined, expansile, multilocular lesion measuring 5.1 × 2.2 cm in the right body of the mandible extending from 33 to mid ramal region, right coronoid process. The incisional biopsy in the region of 45 and 46 was reported as a dental cyst. The patient was referred to Dept of Conservative dentistry and Endodontics for opinion on 44. Considering age of the patient endodontic treatment of 41, 42, 43, 44, and 45 was done. Routine laboratory investigations were within normal limits. The surgical management involved enucleation and curettage with surgical removal of 48 under general anaesthesia (Fig. 3). The welling of blood was noticed, with multilocularity in the bone of the mandible extending to right coronoid process. The upper and lower jaws were stabilized with arch bars and elastics to prevent pathologic fracture, due to extensive nature of the lesion. The post operative histopathological report showed large irregular sinusoidal spaces devoid of endothelial cells and filled with

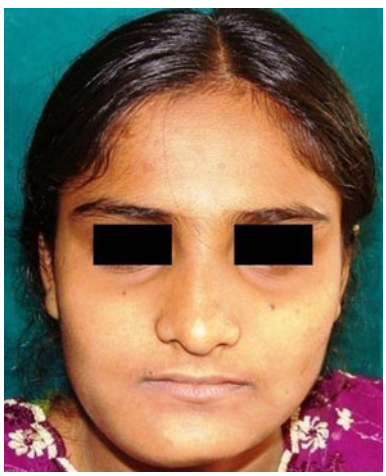

Fig. 1 Preoperative frontal view

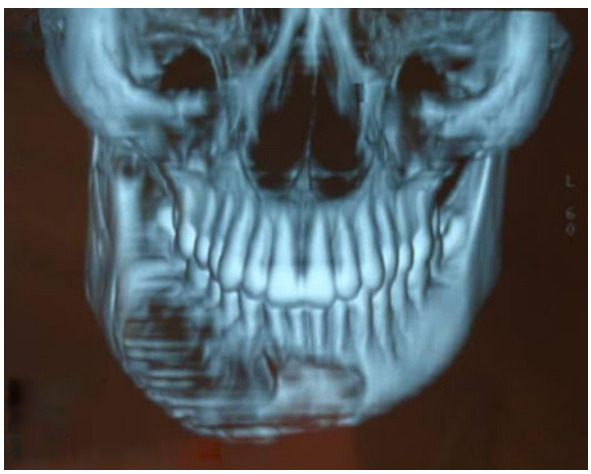

Fig. 2 Preoperative 3D-CT scan

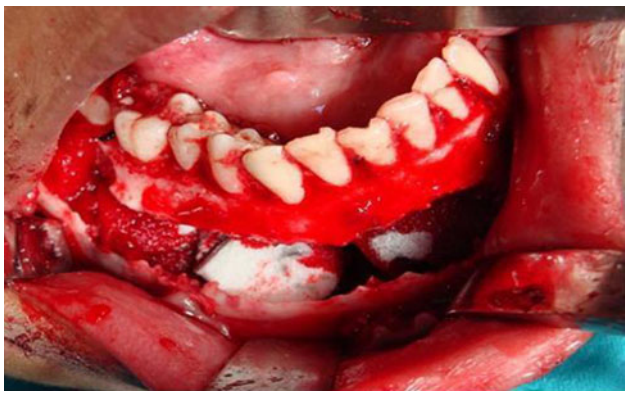

Fig. 3 Intraoperative view

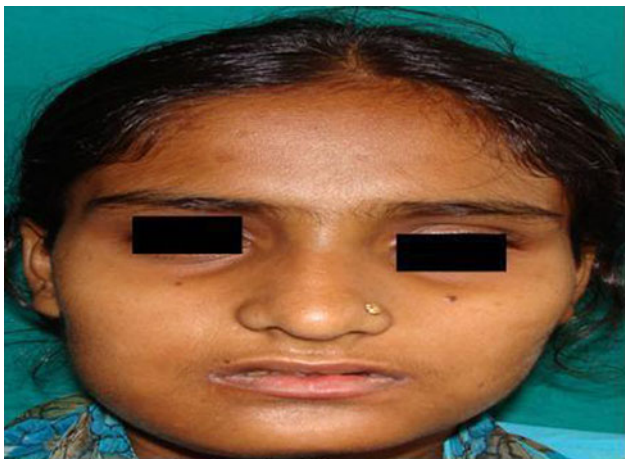

Fig. 4 Postoperative 3 months follow up

blood which were separated by septae. Periphery of the lesion showed osteoid formation. Giant cells are also evident in focal areas surrounding the bone and vascular spaces. The findings correlated with the clinical presentation, thus suggestive of Giant Cell Tumour (GCT). The patient hence has reported to department on regular follow-up. Three month follow up shows adequate bone formation on the right side of the mandible with no recurrence and the patient in good health (Figs. 4, 5)

Discussion

Giant Cell Tumour (GCT) is an uncommon, osteolytic, benign, pseudocystic, solitary localized lesion. The term 'Aneurysmal Bone Cyst' was coined in the year 1950 to describe the characteristic "blow out" or "balloons out" of the cortex, however Robert Marx refers the lesion as Giant Cell Tumour (GCT). The incidence of the GCT is 2–12% in head and neck region. 90% of the cases affect the posterior mandible i.e. body of mandible 40%, ramus 30%,

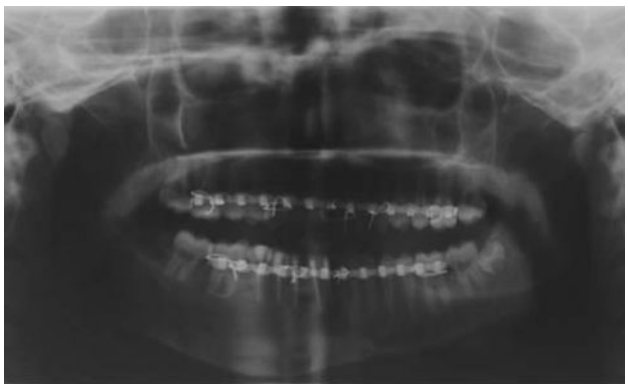

Fig. 5 Postoperative OPG

angle 19%, symphysis 9%, condyles 2%. [9] The case report described here has its presentation in the right side of mandible involving ramus, symphysis, angle and coronoid process. There are few reports found in the literature describing involvement of coronoid process by GCT [5, 13]. This adds one more aspect in diversity of clinical and biological behaviour of GCT in maxillofacial region.

Clinically, the lesion usually occurs in young adults below 20 years of age. There seems to be slight preponderance towards females [8]. The above description of the clinical age and sex predilection is seen in our case report.

The pathogenesis of GCT has added to diagnostic dilemma. Lichtenstein in 1950 postulated that GCT is a manifestation of altered haemodynamics. The pressure secondary to circulatory disturbance leading to congestion leading to bone resorption, with deposition of fibrous connective tissue, osteoid and new bone. A second hypothesis was given by Biesecker et al. [12] where they proposed that a primary lesion initiates an osseous, arteriovenous malformation and therby creates, via its haemodynamic forces, a secondary reactive lesion of bone. The most commonly accepted theory of Hillerup and Hjorting-Hansen [11] suggested that GCT, Central Giant Cell Granuloma, Traumatic Bone Cyst were all related lesions. Minute trauma or the presence of unidentified small aneurysmal enlargements may result in intramedullary bleeding leading to haematoma. If the blood supply is lost TBC may develop. If only small vessels or low pressure is present then capillary and endothelial proliferation occurs resulting in CGCG. If circulation is maintained, creating high pressure, large pools of blood are formed and GCT results. The clinical case did not give any history of trauma and hence this cannot be assigned as the cause of pathogenesis. Among the lesions that have been associated with GCT are fibrous dysplasia, ossifying fibroma, cementifying fibroma, giant cell granuloma. Fibrous dysplasia may occur in association with GCT especially with monostotic type, this can be ruled out in our case since she

did not present with clinical features of fibrous dysplasia where giant cells have limited foci. A differentiating feature of presence of blood filled multiple vascular spaces with giant cells differentiates it from CGCG. The cemento-ossifying fibroma histopathologically represents itself with calcifications which was not seen in our case. The lack of pulsatality in the lesion aids in our diagnosis to rule out arteriovenous malformations causing GCT.

The GCT presents as an enlarging well circumscribed firm mass associated with swelling [5] and malocclusion, may or may not be tender on palpation. The enlargement may be slow or rapid with blow out distension of part of the affected bone causing bony expansion. There is usually an intact periosteum and very thin shell of bone covering the cyst. Pain is an infrequent symptom except with rapid growth [6].The teeth may be missing or displaced but root resorption is rarely seen. Also no paresthesia is recorded. These cardinal clinical presentation were also seen in our case report. Histologically, these cysts are described as blood-filled cavities and sinusoidal spaces separated by fibrous connective tissue septa with osteoid trabeculae. Variable amounts of hemosiderin and giant cells can be found [2, 4]. The similar histopathological report in our case report supports its final diagnosis as GCT.

Panoromic radiography frequently shows the presence of a cystic radioluscent imaging, usually multilocular with a cystic meshwork divided by coarse septa. Bony cortex can also be expanded. The multilocular effect gives this cyst the characteristical but no pathognomical "honey comb" and "soap bubble appearance" seen in other lesions such as giant cell granuloma, myxoma, desmoblastic fibroma, haemangioma, ameloblastoma, and other tumors. Occassionally, destruction of bony cortex may be identified, displaying a periosteal reaction imaging or "sun-ray" effect that is characteristic of osteosarcoma [14].

Asaumi et al. [14] concluded the most differential radiological characteristics as: multiple anomalous branches in angiogram, multilocular cystic structure in the Computed Tomography (CT) with bone window, presence of liquid both in the CT or Magnetic Resonance Imaging (MRI), accumulative pattern in the scintigraphy and in the angiography with radionucleids, bubble like structure in MRI T2W1. MRI is mandatory in complex cases in order to improve plain radiographic examination and CT scan in soft tissue contrast. Angiography is occasionally used in the diagnosis of pseudocysts, however it may be necessary if any haemangioma or high grade neoplasms is suspected when MRI shows hypervascularization [6]. Fine needle aspiration and incisional biopsy may also be performed when high grade tumors are suspected [5].

Concerning the treatment, although many options have been performed, the gold standard is still the surgical excision and curettage of the cavity [15]. There have been various treatment modalities used in the treatment of GCT, including enucleation, curettage, cryotherapy, radiation, resection [12]. The gold standard of surgical enucleation and curettage was used in our case. Curettage used in combination with cryotherapy may reduce the rate of recurrence in nonfacial bones, which has been reported to be greater than 50% when curettage alone is used [12]. Radiation has been used as a therapeutic modality, but the subsequent development of sarcoma is possible and has been reported. Segmental resection must be done only in case of multiple recurrences or extension to overlying tissues [2, 5, 6]. The effort should be expended preoperatively to ensure that the lesion treated is GCT. Bleeding is expected and may be severe, therefore wide exposure for access to afferent vessels, rapid curettage of the lesion, and the liberal use of gauze packing and hemostatics should be employed. Recurrence rates range from 20 to 30% according to different sizes and seems to occur most frequently within the first year of surgery [5, 6].

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