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Case Report

Recurrent High Grade Chondroblastic Osteosarcoma of the Maxilla : A Rare Case Report

Surekha Kanala [1], Sudhakar Gudipalli [2], Manthru Naik Ramavath [3], Santhosh Kumar Venkata Peruri [4], Roger Paul Thota [5]

[1]Professor & HOD, [2]Associate professor, [3,4,5] Post Graduate Student
Department of oral and maxillofacial surgery, Government dental college and hospital, Vijayawada

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ABSTRACT

Osteosarcoma of jaw bones is a rare entity comprising about 6% to 8% of all osteosarcomas, whereas it is reported to be the most common primary malignant tumor of long bones. Osteosarcoma of the maxilla is even rare and is particularly difficult to get wide and clear surgical margins because of its proximity to important structures which may result in recurrence. We report a recurrent case of high grade chondroblastic variant of osteosarcoma of the left maxilla in a 32-year-old male patient. He was treated with subtotal inferior maxillectomy. Unfortunately, recurrence was reported after one month which was treated by total maxillectomy followed by adjuvant chemotherapy and radiotherapy. 2 years clinical and radiographic follow-up revealed no recurrence.

INTRODUCTION

Osteosarcoma (OS) is the most common primary malignant bone tumor with a reported incidence of 1:100,000, in which the neoplastic mesenchymal cells show evidence of osteoid production [3, 15]. It occurs most commonly in the metaphyseal region of long bones of adolescents between 10 and 20 years of age [9]. Conversely, it is rare in the maxillofacial region. Osteosarcoma of Jaw bones (OSJ) represents only about 6% to 8% of all osteosarcomas (OSs) [9]. Approximately, the incidence of the new cases of OSJ per year is 0.07 in 100,000 [6].

OSJ shows different clinical and biologic behaviour than its counterpart in the long bones, though they share histologic features [8]. First, it tends to occur in the third and fourth decades of life, almost a decade after their presentation in the long bones [8]. Second, it has better prognosis than that of OS arising in other sites, which may be due to lower mitotic activity of tumor cells and because cellular anaplasia is found less often in OS of the jaws [15]. Third, it shows fewer tendencies to metastasize than OSs of the long bones [15].

Broadly, OSs can be divided into intramedullary and surface types [15]. The intramedullary or conventional osteosarcoma can be subdivided into the osteoblastic, chondroblastic and the fibroblastic histologic types [8]. OSs arising on the surface of bone are classified into 3 subgroups: parosteal, periosteal, and high-grade surface [15].

In the perspective of rarity of OSJ, case reports are the opportunities to understand and discuss the issues regarding OSJ. So, this article aims to report a case of recurrent high grade chondroblastic osteosarcoma (COS) of maxilla in a 32 years old male patient with

* *Corresponding author:* ***Manthru naik ramavath, Department of oral and maxillofacial surgery, Government dental college and hospital, Vijayawada – 520004, Andhra Pradesh; Telephone number: +91 8790787474; Email id: rmanthrunaik@gmail.com.***

an emphasis on the clinical, histopathological and treatment aspects of the tumor, and to review the literature on OSJ.

CASE REPORT

A 32-year-old male patient presented with the chief complaint of a rapidly growing asymptomatic swelling in the left upper buccal vestibule since 2 months. Swelling was associated with difficulty in opening the mouth and chewing. No history of paresthesia, episodes of bleeding and/or infections. His medical and family histories were insignificant. All the vital signs were within normal limits.

Extraoral inspection revealed facial asymmetry due to a diffuse swelling presented on the left cheek measuring approximately 6 cm X 5 cm in size (Fig. 1). It extended superoinferiorly from the infraorbital margin to 1 cm above inferior border of mandible and mediolaterally from ala of the nose to 2 cm in front of the tragus of the ear. Nasolabial fold was obliterated with elevation of the ala of nose. There was no evidence of nasal obstruction. The skin over the swelling was stretched without any secondary changes. On palpation, all the inspectory findings were confirmed. There was no localized rise in temperature. The swelling was firm, non-tender, smooth and immovable.

On intraoral examination, a solitary localized swelling was seen in left buccal vestibule of maxilla (Fig. 2) measuring approximately 4.5 cm X 4 cm in dimensions extending anteroposteriorly from mesial aspect of first premolar to tuberosity region and superoinferiorly obliterating the buccal vestibule up to 1.5 cm below the occlusal aspect of the molars. Swelling extended medially almost upto the midline of the palate. The surface was irregular, lobulated and covered with slough in some areas. There was no discharge from the swelling. The overlying mucosa appeared to be hypervascular without ulceration. On palpation all the inspectory findings were confirmed. The swelling was firm in consistency, non-tender, non compressible, non pulsatile and fixed to the underlying bone. The premolars and molars associated with swelling were vital with grade II mobility.

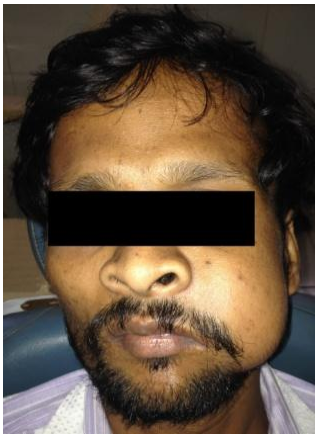

Fig. 1: Preoperative facial view showing swelling with diffuse borders.

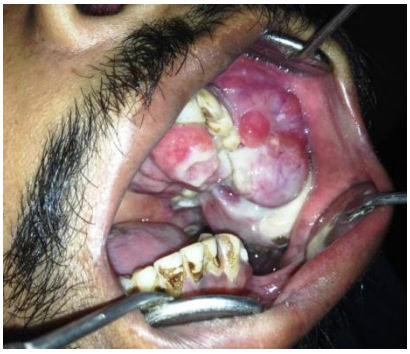

Fig. 2: Intraoral view showing obliteration of left buccal vestibule and palatal aspect of the swelling.

An orthopantomogram (OPG), computerized tomography (CT) scan and 3-dimensional (3D) CT views were taken. OPG and axial cut of the CT revealed a poorly defined radiolucent mass with dispersed areas of radiopacity. Axial view in soft

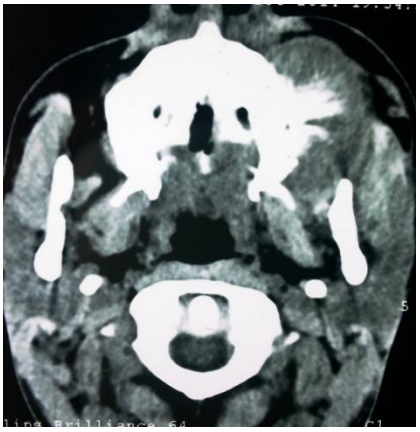

Fig. 3: Axial view showing sunray type of ossification

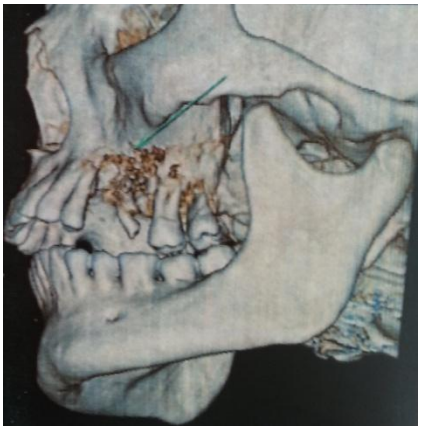

Fig. 4: 3D – CT showing bone destruction in left maxilla.

tissue window revealed 'sunray type of ossification' in the soft tissues (Fig. 3). Coronal view showed tumor encroachment into the maxillary sinus. 3D - CT views showed bone destruction in left maxilla (Fig. 4). Regional pathological lymph nodes were not detected. Aspiration was negative. His hematological and biochemical profile including serum alkaline phosphatase were normal except for leukocytosis.

With a provisional diagnosis of a neoplastic swelling, an incisional biopsy was done to obtain a definitive diagnosis. The microscopic examination revealed predominantly cartilage formation with foci of

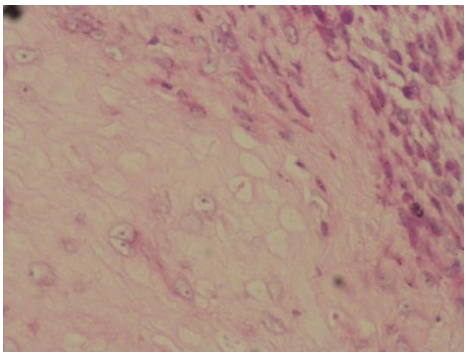

Fig. 5: Showing cartilage formation (H&E, high power, x20)

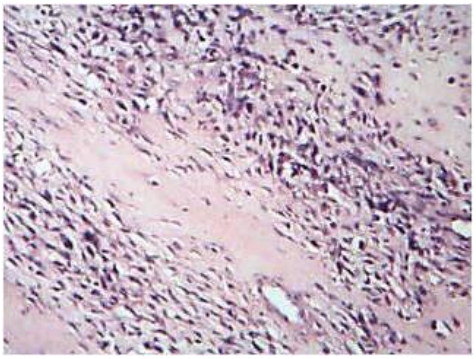

Fig. 6: Showing osteoid formation (H&E, low power, x400)

irregular bony trabeculae in the chondroblastic matrix (Fig. 5, 6). The tumor cells were mostly spindle shaped exhibiting nuclear atypia, hyperchromasia with marked hypercellularity. Immunohistochemical (IHC) studies showed positive reactions for S-100 protein (Fig. 7) and Vimentin (Fig. 8) and negative for cytokeratin-19 and Epithelial Membrane Antigen (EMA). The final diagnosis was high grade chondroblastic variant of osteosarcoma. Subsequently, CT chest, magnetic resonance imaging (MRI) brain and abdominal ultrasonogram were obtained to rule out possibility of metastasis to the lungs, brain and liver.

The patient underwent subtotal inferior maxillectomy on left side through Weber-Ferguson incision. Resected specimen (Fig. 9) was sent for histopathological examination and confirmed the

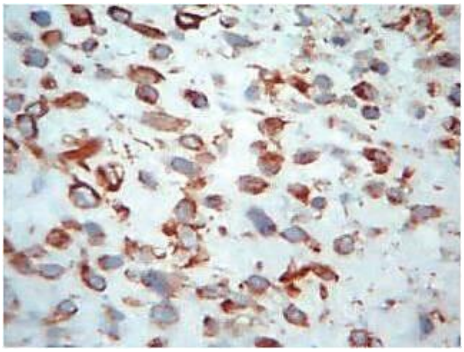

Fig. 7: IHC microphotographs showing S-100 positive (X400)

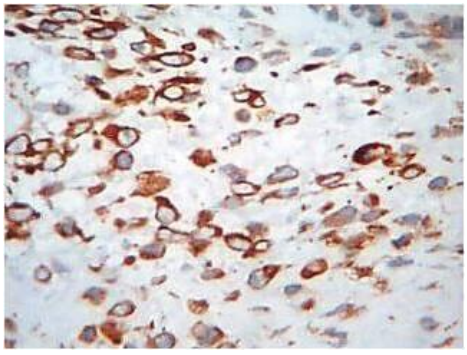

Fig. 8: IHC microphotographs showing Vimentin positive (X400)

preoperative diagnosis. Unfortunately, a recurrent lesion was noted intraorally at the operated site after one month. The patient was planned for second surgery and left total maxillectomy keeping periorbita intact was done. The resultant defect (Fig. 10) was rehabilitated with obturator prosthesis. Owing to high grade nature and recurrence of the tumor, adjuvant external beam radiotherapy by intensity modulated radiotherapy to a total dose of 60Gy; 2Gy per fraction; 30 fractions and chemotherapy (Cisplatin 75 mg and Doxorubicin 40 mg, 6 cycles every 3 weeks) were given. 2 years clinical and radiographic follow-up revealed no recurrence (Fig. 11).

DISCUSSION

Osteosarcoma is a malignancy of mesenchymal cells with the ability to produce osteoid or immature bone [15]. Head and neck OS are rare tumors. OSJ are even

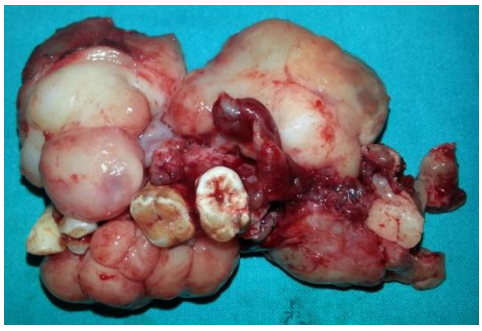

Fig. 9: Resected specimen.

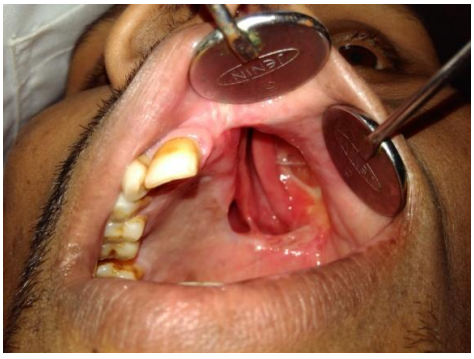

Fig. 10: Introral defect after total maxillectomy.

rarer, representing only about 6% to 8% of all OSs [9]. The etiology is unknown [12]. The average age at onset of OSJ is found in third to fourth decade [2]. A slight male preponderance is reported [2, 8]. Mandible is more affected than the maxilla [7]. The most common presenting symptom is swelling without pain as found in the present case [2, 5]. Other signs and symptoms include displacement and loosening of teeth, paresthesia/anesthesia, epistaxis and nasal obstruction [5, 7].

Mixed radiolucency with radio opaque pattern and poorly defined irregular borders is the most common radiographic presentation as seen in the present case [4]. Symmetric widening of the periodontal ligament space (Garrington sign) [5, 6] and sunray/sunburst

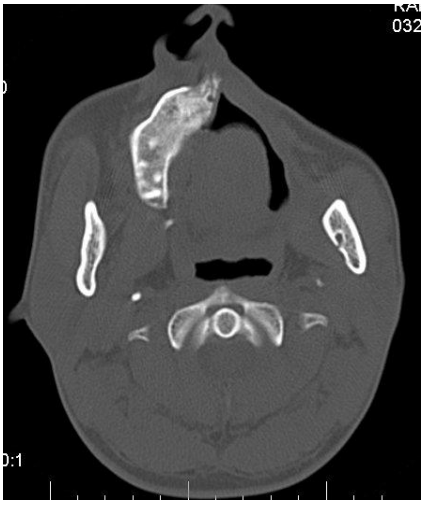

Fig. 11: CT view showing no recurrence after 2 years of followup.

appearance [7] are the other common radiographic findings.

Histologically, OSs are divided into osteoblastic, chondroblastic, fibroblastic variants [8]. The jaw lesions are predominantly chondroblastic while fibroblastic is the least common [6, 8, 15]. High grade OSJ has been reported to be associated with poor prognosis [8]. COS should be differentiated from chondrosarcoma. Osteoid formation within the tumor is the key histomorphologic diagnostic feature of COS [14]. IHC shows CS to be positive for S100 and Vimentin and negative for Cytokeratin and EMA. COS has been found to be positive for S100, Vimentin, EMA, and rarely Cytokeratin [1]. The IHC profile of our patient was S100 and Vimentin positive; EMA and Cytokeratin19 negative. The present case is EMA negative which is in contrast to the reported literature.

Radical surgery with clear margins of 1.5 - 2 cms is recommended as the primary treatment for OSJ [5, 7]. Maxillary lesions are generally approached through Weber - Ferguson's incision or transoral route, depending on extent of the lesion. Generally, subtotal inferior maxillectomy [11] or total maxillectomy with or without orbital exenteration is done based on the clinical situation. The complex anatomy of the head and neck is attributed as the reason for difficulty in achieving clear margins, especially in the maxilla and skull base, where positive margin rates of 31% to 52.4% have been reported [5]. Even though the results of using adjuvant chemotherapy along with surgery for OSJ had been variable, significant improvement in disease free survival rates were reported [13]. In spite of the fact that OS has been regarded as a radioresistant tumor, the present case was treated with radical surgery followed by adjuvant chemotherapy and radiotherapy owing to its high grade nature and recurrence. High grade tumors often recur within the first year [3]. Patel SG et al. [10] and Fernandes R et al. [5] reported the use of postoperative radiation therapy for patients with high-grade tumor or close margins.

SUMMARY

OSJ are uncommon tumors. In the maxillofacial region, the neoplasm manifests as localized disease. Radical surgery with clear margins is the mainstay of treatment. Although distant metastases are rare, adjuvant chemotherapy and radiotherapy have their role in cases of high grade and recurrent tumors. Periodic screening and longer followup is of utmost importance in detecting recurrence and metastasis. Dental professionals are advised to approach such cases with greater concern so as to diagnose them at an early stage leading to better prognosis.

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