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Chondroblastic Osteosarcoma – A Case Report and a Review of Literature

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Abstract

Teratoma Osteosarcoma is the most common malignancy of mesenchymal cells mostly originating within long bones, but rarely in the jaws. The World Health Organization (WHO) assorts several variants that differ in locale, clinical behavior and cellular atypia. This report illustrates a case of one of the histological variants of osteosarcoma i.e., chondroblastic osteosarcoma in the region of anterior maxilla in a 58 year old male patient previously treated for ossifying fibroma of the same site

Keywords: Osteosarcoma; Sarcoma; Maxilla; Bone neoplasms

Introduction

Chondroblastic osteosarcoma as defined by WHO is a histological entity characterized by predominant presence of chondroid matrix, which tends to exhibit a high degree of hyaline cartilage and is intimately associated with the non-chondroid element (osteoid or bone matrix) [1]. It is the most frequent histologic type of osteosarcoma (OS) [2,3] OS of jaw bones is rare and comprises 6-9% of all osteosarcomas and less than 1% of all head and neck malignancies [4,5].

Due to the rare occurrence of OS in jaw bones, herein we report a case of chondroblastic osteosarcoma in anterior maxilla with emphasis on the clinical and radiological aspects of the tumor.

Case Report

A 58-year old man, reported to the Department of Oral Medicine and Radiology, Manav Rachna Dental College, Faridabad, Haryana, India complaining of a swelling in the left anterior region of upper jaw since 1 year. The swelling, as reported, started within the oral cavity and gradually increased to its present size i.e., that of a large walnut. He reported no associated symptoms of pain, numbness, difficulty in breathing or swallowing, fever, weight loss, or any other swelling elsewhere on body.

Patient reported a similar swelling in the same region 18 years earlier which was operated along with extraction w.r.t. 11, 12, 13, 14, 21, 22, 23, 24 and 25 and was histologically diagnosed as an ossifying fibroma. A removable partial denture was then fabricated which he efficiently used till 1 year ago. For the last 17 years, he was asymptomatic with no recurrence of swelling (Figure S1).

The patient's medical history was non-contributory. He was a smokeless tobacco user for the past 20 years. On general examination, he was moderately built and nourished with all vital signs being within the normal limit.

On extra-oral examination, a slight bulge raising the ala of nose (left) was observed on the left middle third of face. The lymph nodes of head and neck region were not palpable.

Intraoral examination revealed a solitary, well-defined, oval shaped swelling in the premaxillary region extending from midline till the mesial aspect w.r.t. 26. Its anterior margin obliterated the labial vestibule and posteriorly it extended till the mid of hard palate. The swelling was lobulated and pink in color. It was non-tender and bony hard. A well-defined grayish brown mucosal patch (Figures 1-3).

The history and clinical features of the lesion suggested a central, benign, osseous neoplasm possibly a recurrant ossifying fibroma. Literature states a recurrence rate of 20% in ossifying fibroma of jaws.

The clinical differential diagnosis included desmoplastic variant of ameloblastoma which occurs predominantly in anterior maxilla and presents as a slow growing asymptomatic swelling. Another odontogenic tumor which is slow growing, asymptomatic and affecting middle-aged males is Calcifying epithelial odontogenic tumor. Among the malignancies, low grade chondrosarcoma was considered as it shows similar features as noted in our case. Clinically, osteosarcoma was not considered as a differential diagnosis because the patient didn't show obvious signs strongly suggestive of OS.

Radiological tests performed included intraoral periapical and occlusal radiographs, Digital Panoramic Radiograph (DPR) and Cone Beam Computed Tomography (CBCT) (Figure 3 and Figures 4a-4d).

Radiographic examination revealed a mixed radiopaque-radiolucent lesion at the edentulous premaxillary region. The maximum dimension of the mass was 46.1 × 31.9 × 19.5 mm. The lesion appeared roughly ovoid in shape. In some areas, the borders showed a wide zone of transition thereby blending with the surrounding normal bone. At other areas, the borders were relatively well defined with a narrow zone of transition and surrounded predominantly by a thin radiolucent halo separating the lesion from normal bone as an encapsulation. Internal structure consisted of numerous, ill-defined, irregular radiopaque areas of varying size and density scattered among lytic areas resembling cotton-wool or wisp-like appearance. Peripheral periosteal bone was seen as radiating lines perpendicular to the expanded cortex showing a 'sunray' or 'hair on end appearance'. Thickening of the sinus membrane could be seen. The lobulated appearance of the mass could be appreciated. Nasopalatine canal was displaced anteriorly. In the coronal sections, the canal was seen to be displaced also to the right side. Floor of the nasal cavity was breached and irregular on the left side. Thickening of nasal mucosa as well as the antrum membrane could be

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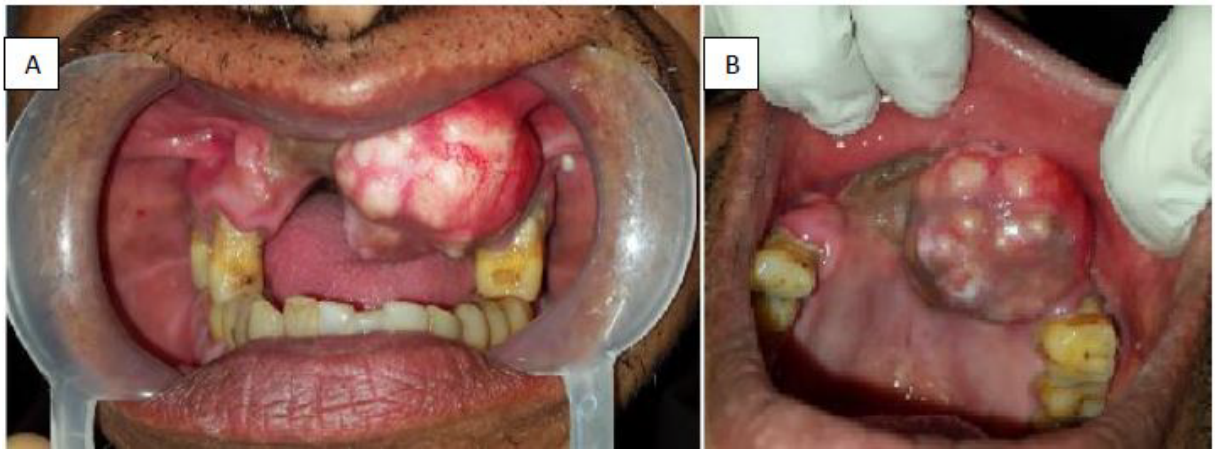

Figure 1: Intraoral examination showed a swelling in the anterior maxillary region obliterating the labial vestibule.

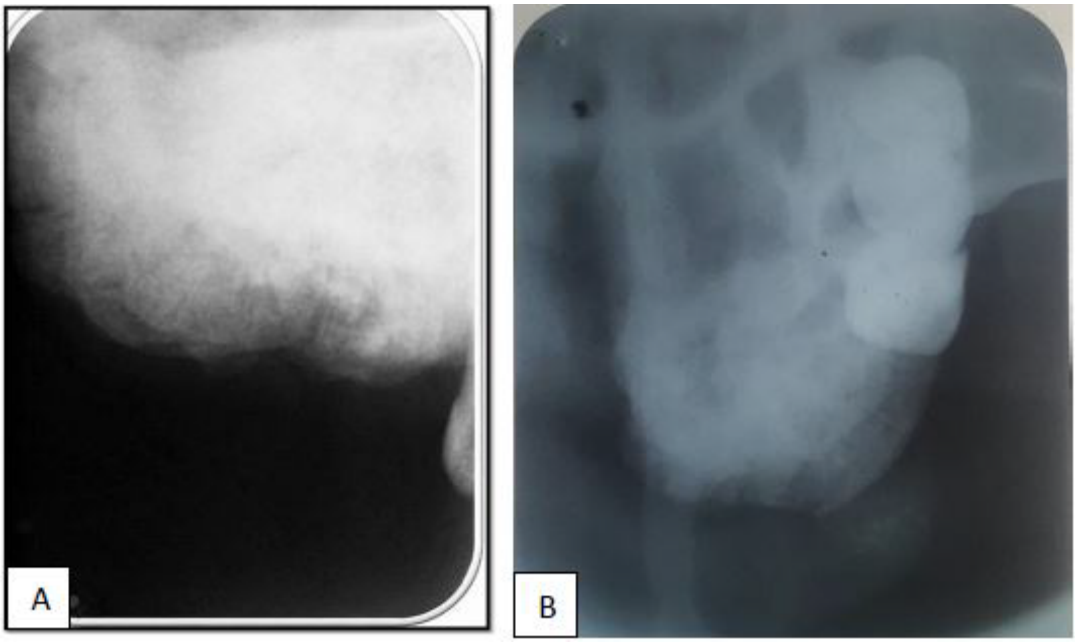

Figure 2: IOPAR revealed a mixed radiopaque-radiolucent lesion at the edentulous premaxillary region. In the maxillary lateral occlusal radiograph, peripheral periosteal bone was seen as radiating lines perpendicular to the expanded cortex showing a 'sunray' or 'hair on end appearance'.

seen indicating the lesion to be infiltrating both the nasal cavity and the antrum. The adjacent tooth 26 showed widening of periodontal ligament (PDL) space. Generalized periodontal bone loss was present. Dental caries involving pulp w.r.t. tooth 17 was present (Figures 2a and 2b).

In liew of the additional information obtained from radiographic examination the provisional diagnosis was modified as low grade osteosarcoma due to the sunburst appearance and lobulation. Incisional biopsy was performed from the labial aspect of swelling. A hard bony tissue was removed and decalcified. Histopathological examination confirmed it as a case of chondroblastic variety of osteosarcoma.

PET-CT scan was obtained from the level of the vertex of skull till mid thighs in arms down position. 185 MBq of Radiotracer agent F18 fluorodeoxyglucose (FDG) was injected. An I.V. injection of non-ionic contrast was also given. An abnormal focal hypermetabolic mass at anterior maxilla was noted measuring 31 (AP) * 34 (TR) * 18(CC) mm. The largest node measured 10 mm in the paratracheal region with standardized uptake value (SUV) max 2.7. No adenopathy was noted in the supraclavicular, axilla, mediastinum or hilum. No pulmonary nodules or masses were identified. No definite evidence of skeletal metastasis was noted. PET-CT scan thus ruled out possible metastasis and signs of secondary tumors (Figure 5).

In accordance with the Enneking System of staging and grading, the tumor was staged as II A that involved G2, T1 and M0 stages.

Since there was no metastatic involvement, complete resection was planned with clearance margins of 0.5-1cm. The patient was then

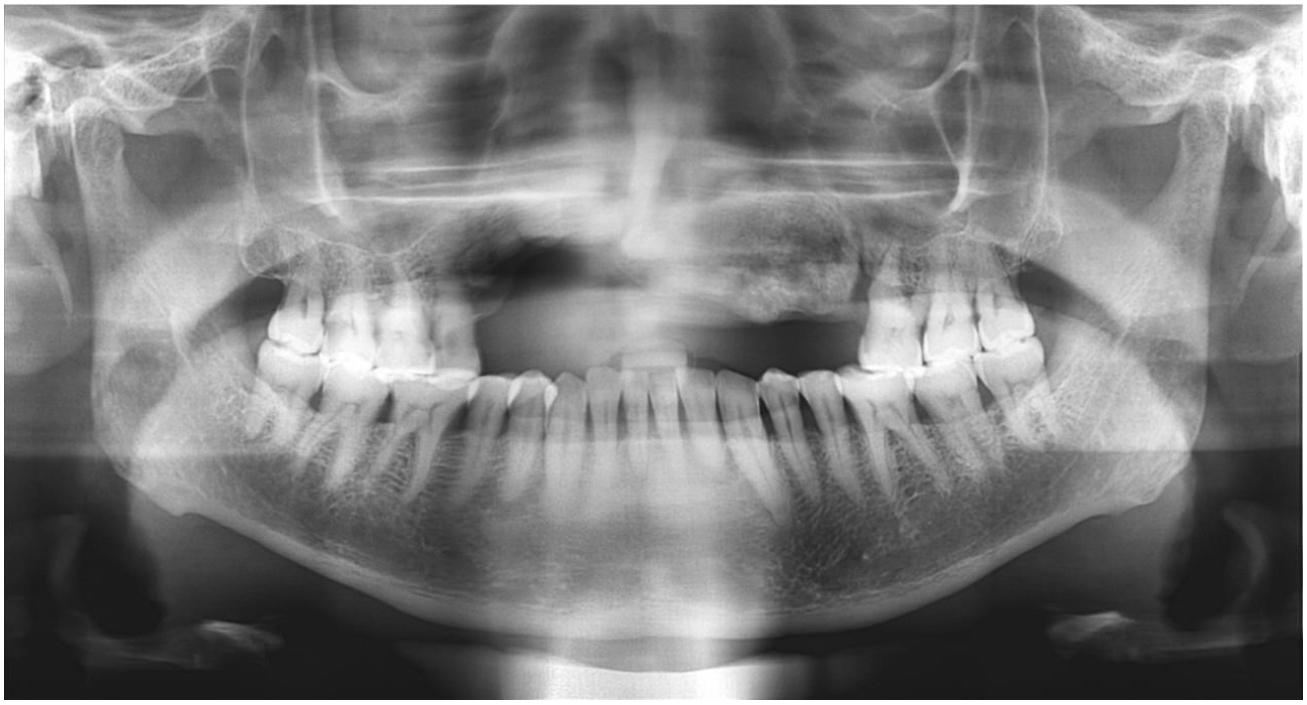

Figure 3: Digital panoramic radiograph showing the lesion at anterior maxilla with a well-defined distal margin with a narrow zone of transition and surrounded by a radiolucent halo. The adjacent tooth 26 showed widening of periodontal ligament space.

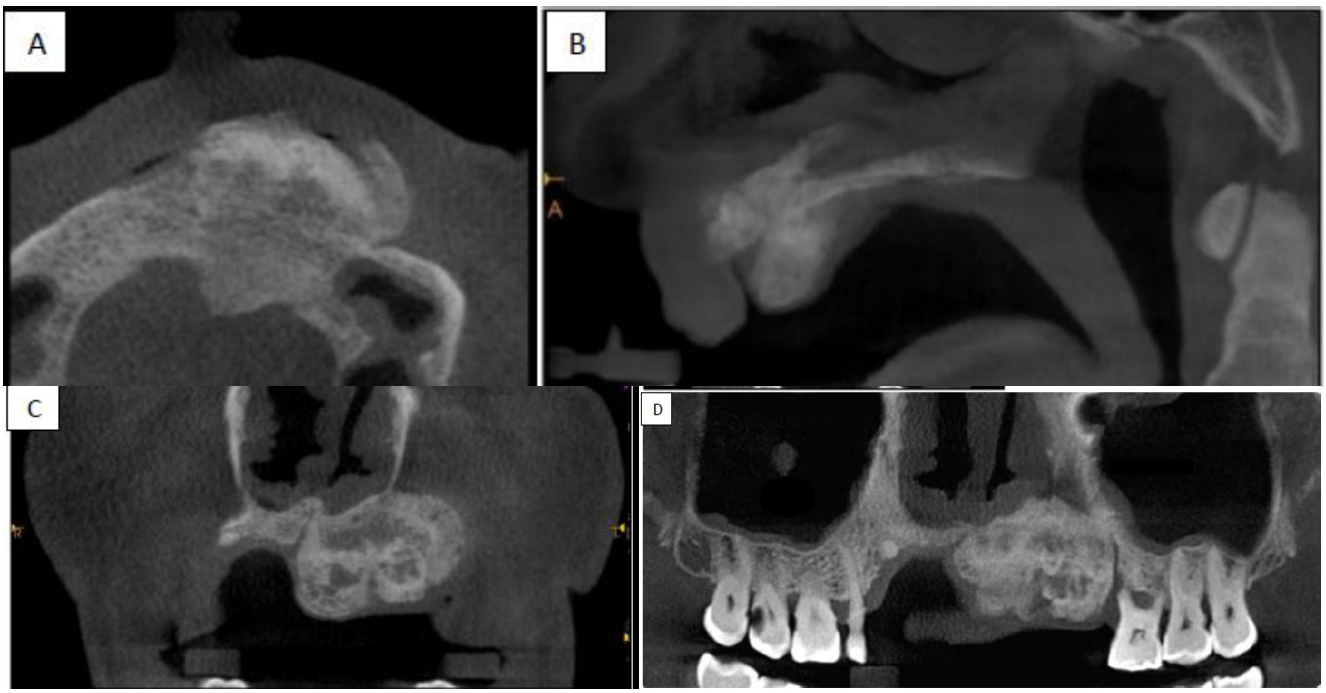

Figure 4: (A) CBCT scan of the maxilla showing poorly defined margins (B) displacement of nasopalatine canal anteriorly and (C) to the left side and (D) Infiltration into the maxillary sinus and nasal cavity.

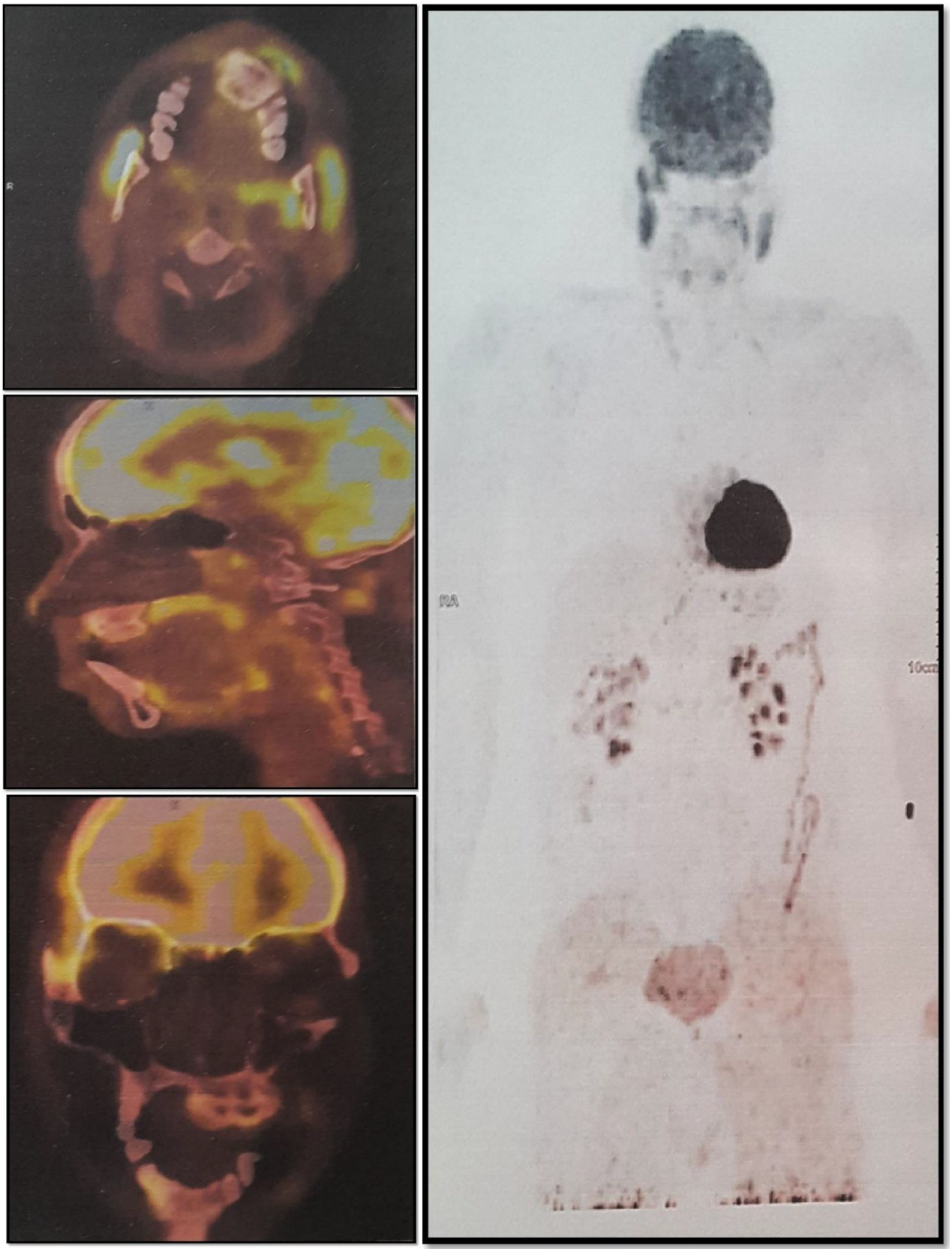

Figure 5: PET-CT showing an abnormal focal hypermetabolic mass at anterior maxilla and no metastasis and signs of secondary tumors.

referred to an Oncology Service. Partial maxillectomy was performed under general anesthesia. Post-surgical obturator was then placed. The patient 1 year after surgical excision reports to be asymptomatic.

Results and Discussion

Chondroblastic osteosarcoma is a subtype of osteosarcoma, characterized by the production of chondroid matrix of variable cellularity, most commonly high-grade hyaline cartilage. OS is the second most common malignant bone tumor after multiple myeloma accounting for 15-35% of all primary malignant bone tumors followed by chondrosarcoma and Ewing's sarcoma [6].

OS of jaws is seen mostly in third-fourth decade (a decade later than mean age of OS incidence in long bones) [7-9] with male predominance and although literature mentions the mandible as a more likely location than the maxilla [7]. Swelling is the dominant complaint in OS of jaws whereas pain is common in OS of long bones [9]. Other features such as swelling, tooth mobility and paresthesia may be present. Pain, fever or weight loss is rare [1].

Only one reported case of transformation of cemento-ossifying fibroma into osteosarcoma has been reported till date [10]. Incidence of osteosarcoma secondary to paget's disease and fibrous dysplasia is 0.95% and 0.7% respectively. Incidence of radiation Induced Sarcoma is 0.03% to 0.3%. Incidence of radiation induced osteosarcoma is 0.03%. This present case however seemed to occur *de novo* due to absence of histopathological evidence suggesting otherwise.

Radiographic findings vary from radiopaque to mixed to radiolucent. Cotton balls, wisps or honeycomb pattern is seen. "Classic" sunray or sunburst appearance due to osteophytic bone production is an important feature [11]. Garrington's sign (widening of PDL space around affected teeth) with tapered resorption of tooth roots maybe present [9]. There may be cortical bone destruction and adjacent soft tissue involvement [1]. The antral or nasal wall cortices may be affected in maxillary lesions. Our case showed a lobulated mass with cotton wool internal structure with peripheral periosteal sun ray pattern. In mandible neurovascular canal cortex may also be affected.

According to WHO, patterns of bone destruction are indicative of the aggressiveness of the lesion [12] Lodwick et al. [13] proposed the radiologic signs necessary to establish survival rates Pattern of tumor margins only implies the progression rate and not directly its malignancy. The geographic pattern is considered least aggressive while the permeative pattern is most aggressive. However, the present case showed radiographic evidence of type 1C pattern which exhibits a less sharp limit.

Nakayama et al. [14] proposed a classification of CT pattern found in OS of jaws based on osteogenesis and signs of bone destruction and reported its significant association with survival outcome (prognosis). The present case falls into the group of osteogenic type without bone destruction.

Immunohistochemistry helps in differentiating chondroblastic osteosarcoma from chondrosarcoma as it is positive for vimentin, epithelial membrane antigen, S100 and rarely positive for cytokeratin whereas chondrosarcoma is positive for vimentin and S100 [15]. In chondroblastic osteosarcoma, the presence of osteoid distinguishes it from chondrosarcoma. Raised serum alkaline phosphatase in osteosarcoma also distinguishes it from chondrosarcoma [16].

OS of jaws have a better prognosis than OS of long bones and most commonly metastasizes to lung [17]. Staging of the tumor is crucial for estimating prognosis. The Enneking Staging System for bone sarcomas is based on grade (G), local extent of the primary tumor (T), and metastasis (M) [18]. The present tumor was staged as II A that involved G2, T1 and M0 stages.

The treatment of choice in oral osteosarcomas is surgical resection [7,19,20]. Complete surgical excision with negative margins continues to be the mainstay of treatment but osteosarcomas of maxillofacial region pose difficulties in obtaining tumor-free margins because of their complex anatomy and close proximity to the cranium [20].

Surgery may be complemented by radiotherapy and/or chemotherapy. The use of chemotherapy before and after surgery promotes local control by size reduction [21]. Currently, doxorubicin, cisplatin, methotrexate with leukovorin and ifosfamide are considered the most active agents against osteosarcoma. The optimum time for commencing chemotherapy is within 21 days of surgery. Overall prognosis in OS is 25-50% with 5-year survival rate [11]. The present case showed absence of metastasis thereby chemotherapy was not done, and radiotherapy was avoided so only surgical excision was done.

Conclusion

OS of maxillofacial region have variable appearances clinically as well as radiologically which poses a diagnostic challenge for clinicians especially in its imaging diagnosis. This case also presents an unsolved mystery about whether it occurred *de novo* or secondary to the central ossifying fibroma. OS of jaws being rare in the oral cavity supplemented by its life-threatening nature accompanied by limited knowledge about its features necessitates further studies on more cases to determine the behavior of this neoplasia with greater precision.

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