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CASE REPORT

Chondroblastic osteosarcoma—A case report and review of literature

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Key Clinical Message

Osteosarcoma is the most common malignancy of mesenchymal cells mostly originating within long bones, but rarely in the jaws. This report illustrates a case of 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

bone neoplasms, maxilla, osteosarcoma, sarcoma

1 | 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 nonchondroid element (osteoid or bone matrix).[1] It is the most frequent histologic type of osteosarcoma (OS).[2,3] Osteosarcoma of jaw bones is rare and comprises 6%-9% of all osteosarcomas and <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.

2 | 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, that is, 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 of 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 until 1 year ago. For the last 17 years, he was asymptomatic with no recurrence of swelling.

The patient's medical history was noncontributory. 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 extraoral 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 to the mesial aspect of tooth 26. Its anterior margin obliterated the labial vestibule and posteriorly it extended to the mid of hard palate. The swelling was lobulated and pink in color. It was nontender and bony hard. A well-defined grayish brown mucosal patch was present on the labial vestibule adjacent to the medial aspect of swelling (Figure 1).

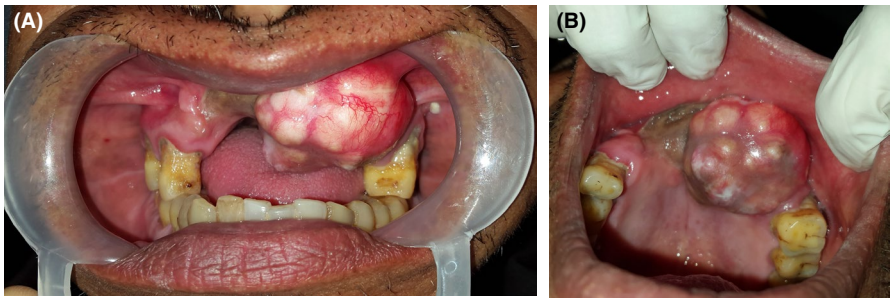

FIGURE 1 Intraoral examination showing a swelling in the anterior maxillary region obliterating the labial vestibule

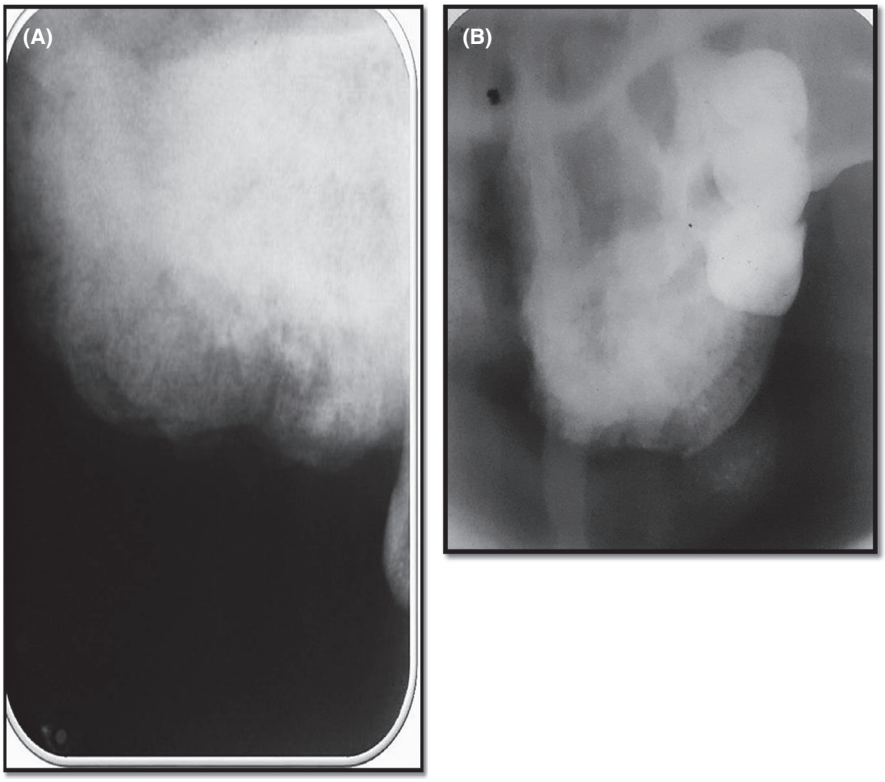

FIGURE 2 IOPAR revealed a mixed radiopaque-radiolucent lesion at the edentulous anterior maxillary region (A). In the maxillary lateral occlusal radiograph, peripheral periosteal bone was seen as radiating lines perpendicular to the expanded cortex showing a "sunray" or "hair on end appearance" (B)

The history and clinical features of the lesion suggested a central, benign, osseous neoplasm possibly a recurrent 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 did not 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) (Figures 2-5).

Conventional 2D imaging 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."

The 3D CBCT examination showed thickening of the sinus membrane. The lobulated appearance of the mass could be appreciated. The nasopalatine canal was displaced

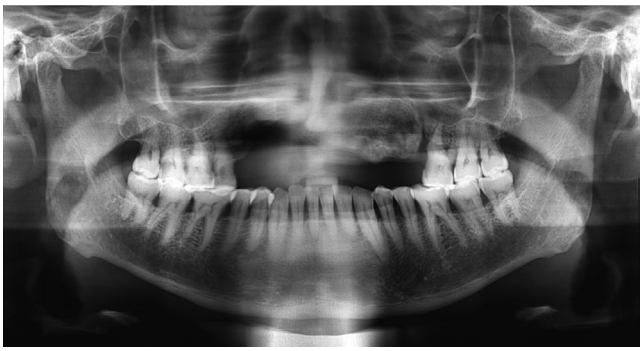

FIGURE 3 Digital panoramic radiograph showing the lesion at anterior maxilla with a well-defined distal margin with a narrow zone of transition and surrounded by a radiolucent halo. The adjacent tooth 26 showed widening of periodontal ligament space

anteriorly and to the right side. Floor of the nasal cavity on left side was breached and irregular. Thickening of nasal mucosa as well as the antrum membrane could be 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 of tooth 17 was present.

In lieu 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 to mid thighs in arms down position. 185 MBq of radiotracer agent F18-fluorodeoxyglucose (FDG) was injected. An intravenous injection of nonionic 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.

In accordance with the Enneking System of staging and grading,[6] the tumor was staged as II A that involved G2, T1, and M0 stages.

As there was no metastatic involvement, complete resection was planned with clearance margins of 0.5-1 cm. The patient was then referred to an oncology service. Partial maxillectomy was performed under general anesthesia. A postsurgical obturator was then placed. The patient 1 year after surgical excision reports to be asymptomatic.

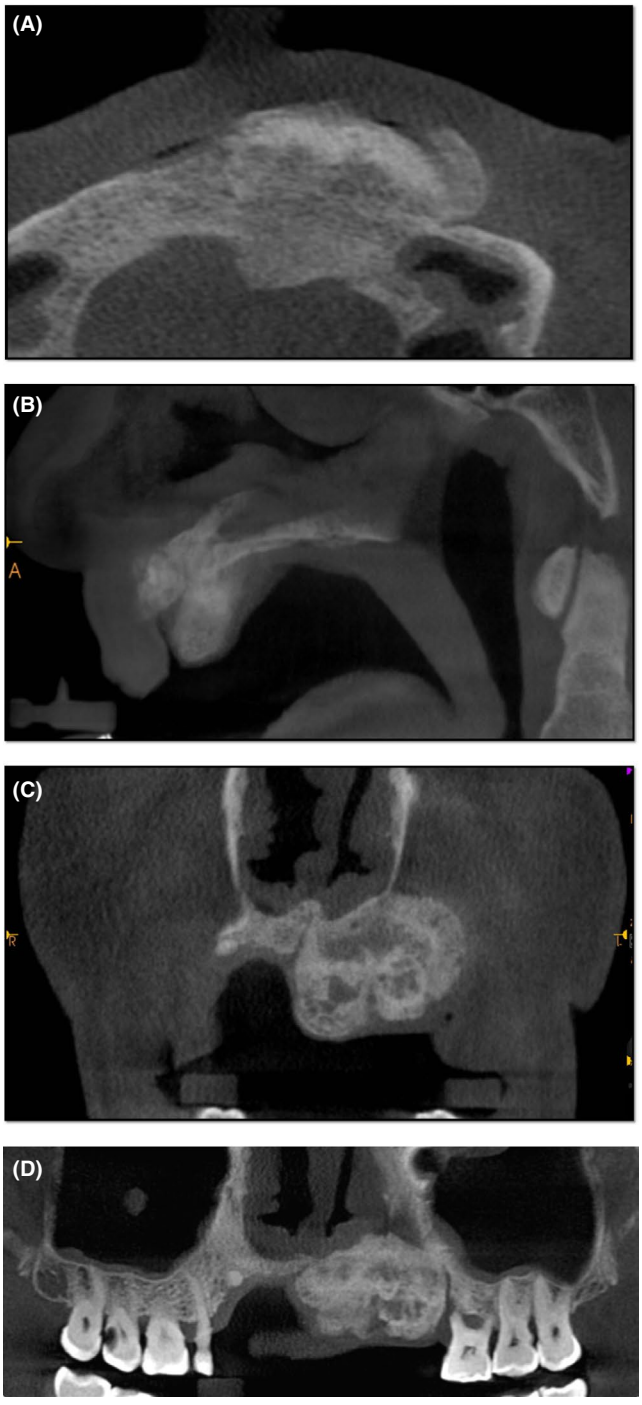

FIGURE 4 CBCT scan of the maxilla showing poorly defined margins (A), displacement of nasopalatine canal anteriorly (B) and to the left side (C) and Infiltration into the maxillary sinus and nasal cavity (D)

3 | DISCUSSION

Chondroblastic osteosarcoma is a subtype of osteosarcoma, characterized by the production of chondroid matrix of variable cellularity, most commonly high-grade hyaline cartilage. Osteosarcoma is the second most common malignant bone

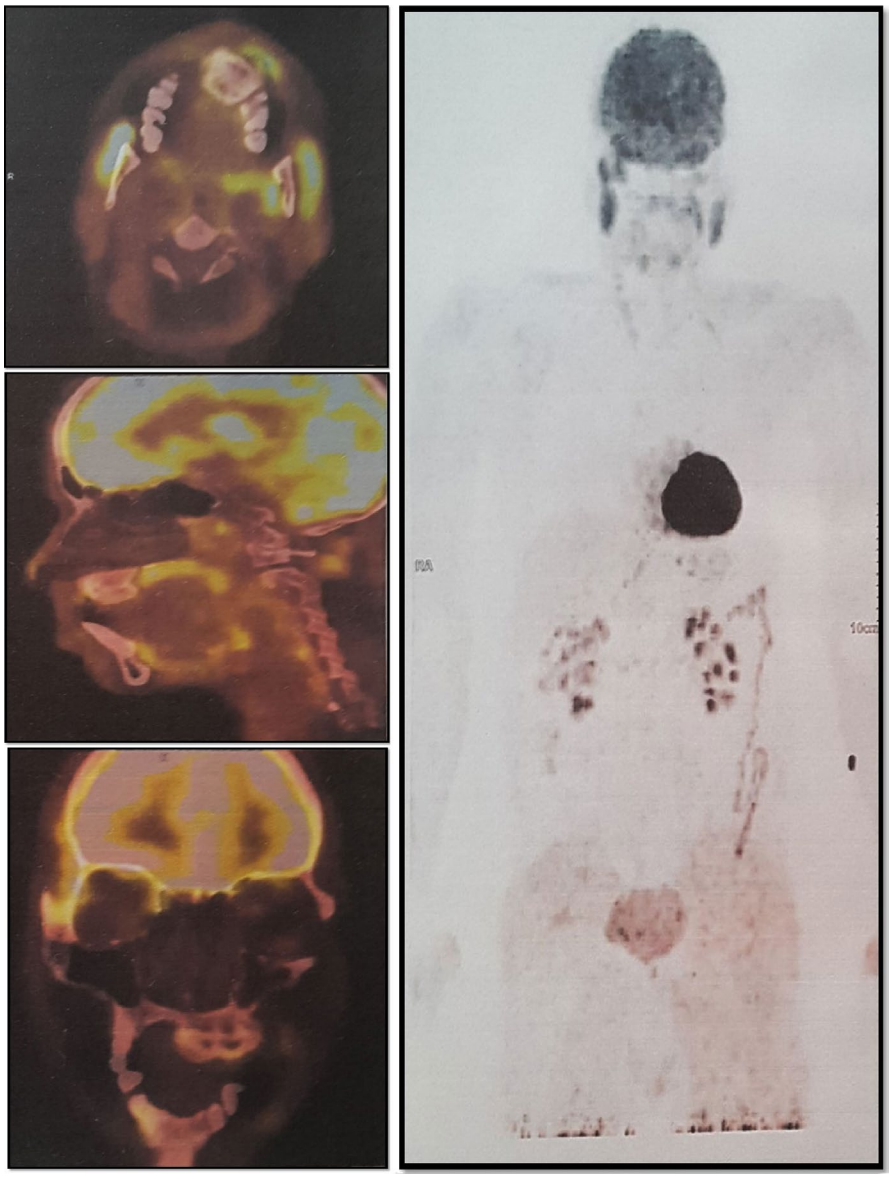

FIGURE 5 PET-CT showing an abnormal focal hypermetabolic mass at anterior maxilla and no metastasis and signs of secondary tumors

tumor after multiple myeloma accounting for 15%-35% of all primary malignant bone tumors followed by chondrosarcoma and Ewing's sarcoma.[7]

Osteosarcoma of jaws is seen mostly in third-fourth decade (a decade later than mean age of OS incidence in long bones)[8–10] with male predominance. The literature mentions the mandible as a more likely location than the maxilla.[8] Swelling is the dominant complaint in OS of jaws whereas pain is common in OS of long bones.[10] Other features such as swelling, tooth mobility, and paresthesia may be present. Pain, fever, or weight loss are rare.[1]

Only one reported case of transformation of cemento-ossifying fibroma into osteosarcoma has been reported to date.[11] 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%-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.[12] Garrington's sign (widening of PDL space around affected teeth) with tapered resorption of tooth roots maybe present.[10] In lesions involving mandible neurovascular canal cortex may also be affected. 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.

According to WHO, patterns of bone destruction are indicative of the aggressiveness of the lesion.[13] Lodwick et al[14] proposed the radiologic signs necessary to establish growth

rates of bone lesions. 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[15] 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.[16] In chondroblastic osteosarcoma, the presence of osteoid distinguishes it from chondrosarcoma. Raised serum alkaline phosphatase in osteosarcoma also distinguishes it from chondrosarcoma.[17]

Osteosarcoma of jaws has a better prognosis than OS of long bones and most commonly metastasizes to lung.[18] 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).[6] 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.[8,19,20] 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.[12] The present case showed absence of metastasis thereby chemotherapy was not performed and radiotherapy was avoided. Complete surgical excision was performed under general anesthesia.

Osteosarcoma of maxillofacial region has 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. Osteosarcoma of jaws being rare in oral cavity, its life-threatening nature and the limited knowledge about its features necessitates further studies on more cases to determine the behavior of this neoplasia with greater precision.

CONFLICT OF INTEREST

None declared.

AUTHORSHIP

PM: Drafted and wrote the article. VD: Edited and reviewed the article. NSB: Provided guidance and helpful feedback on the draft. ND: Revised it critically for important intellectual content. PK: Revised it critically and provided suggestions for final preparation of the manuscript.

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