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Osteosarcoma of Maxilla – An Unusual Entity: Case Report and Review of Literature

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Case Report

History

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ABSTRACT

Osteosarcoma of the jaw is a rare malignancy. It affects the mandible more often than the maxilla. There have been rare cases of osteosarcomas reported in the maxilla; however, given the lack of specific signs and a diverse range of radiographic features, the diagnosis is often difficult and requires a multidisciplinary approach. This is a case report of a 56-year-old female presenting with a well-defined, dome-shaped firm swelling having radiographic features of extensive destruction of the buccal cortical bone in the maxillary right posterior region. Osteosarcoma of the jaw is difficult to diagnose and manage due to the high frequency of errors in biopsy results, the few specific radiological characteristics, and difficulties in appropriate resection because of the proximity to vital structures. Over the years, the survival rate of patients has greatly improved, due to systematic approach and refined surgical and reconstructive techniques. Due to the aggressiveness of this disease, early diagnosis of the lesion is required.

Key words: Diagnostic Imaging, Maxillary Neoplasms, Osteosarcoma, Osteogenic Sarcoma, Bone Neoplasm.

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Introduction

Osteosarcoma (OS) or osteogenic sarcoma is a rare entity that accounts for approximately 6-10% of jaw bone lesions.[1] In the human skeleton excluding the jaw bones, OS is the second most common malignant bone tumor with tumors of the bone marrow being first. It follows an aggressive course and can cause considerable morbidity and mortality as local recurrence is common.[2] OS displays a bimodal distribution with two peaks, one in the first or second decade and the other in the fourth decade.[1,3]

OS is classified into two types – primary and secondary. The etiology of the primary type is not clearly known and may be due to genetic influences or other environmental factors. In the secondary type, craniofacial osteogenic sarcomas occur in older patients with skeletal Paget's disease, fibrous dysplasia of bone, Li-Fraumeni syndrome, and as a late sequela to cranial irradiation. Environmental factors such as ionizing radiation and chromic oxide (a radioactive scanning agent), have been incriminated as possible causes. Genetic mutations in tumor suppressor gene p53 and mutated retinoblastoma gene have been claimed to be amongst other etiologic factors.[4]

In a study by Nissanka *et al.,* most patients related the occurrence of the tumor to previous dental treatment, most commonly dental extractions. The reason for this is most likely the rapid growth of tumors immediately following trauma, a phenomenon usually seen in skeletal OS.[5]

OS has no specific clinical signs. Given the lack of specific signs and the diverse nature of the radiographic features, like sunray spicules, "hair-on-end" trabeculae or Codman's triangle, internal osseous structure taking up the appearance of granular or sclerotic-appearing bone, cotton balls, wisps, or honeycombed internal structures in areas with adjacent destruction of the preexisting osseous architecture, it can lead to misleading interpretation.[6] This lesion presents itself with the features seen in most malignant tumors, such as bone destruction with no periosteal reaction and irregular subperiosteal new bone formation mass.[7] The diagnosis is often difficult and requires a multidisciplinary team including a molecular biologist. Nearly 20% of all metastases occur in the lung, making it the most frequent site.

In the orofacial region, the mandible is more commonly affected than the maxilla with a ratio of 1.5:1, with males showing a predilection for occurrence in the mandible and females in the maxilla.[8,9] In mandible, the body is most commonly involved followed by angle, symphysis, ascending ramus, and the antrum.[4] This article reports a case of OS in the posterior maxilla.

Case report

A 58-year-old female developed a painless swelling on the maxillary right posterior region since one month for which she had visited a local dentist, who thought it to be associated with the maxillary right first molar, which was subsequently removed. However, not only did the swelling not subside, but instead there was a rapid increase in size, following which she was referred to our facility. Her past medical history revealed that she had undergone anti-tubercular therapy for extra-pulmonary tuberculosis, which lasted 2 years. On examination, a single, well-defined, dome-shaped, firm swelling with a sessile base of size 5 x 4 cm was noted attached to the alveolus. Its superior margin was located 2 cm below the inferior orbital margin and its inferior margin was at the level of the alveolar ridge. Antero-posteriorly, the extent ranged from the distal aspect of tooth 13 to the mesial aspect of tooth 16 (Figure 1) displaying a surface appearance that was partly erythematous and partly white with the surrounding tissues appearing normal. There was no bleeding or pus discharge seen (Figure 2).

Considering the location, the absence of tenderness, and its rapid growth, a provisional diagnosis of "Aggressive, non-odontogenic neoplasm of the maxillary right posterior region" was given.

A cone beam computed tomography (CBCT) was advised which revealed a single, unilateral, radiolucent area present in the region pertaining to teeth numbers 15 and 16 with extensive bone destruction of the buccal cortex and minimal perforation of the palatal cortex. Alteration in the trabecular architecture and sclerotic bone formation was noted posterior to the lesion and extended to the area of the maxillary tuberosity and appeared to blend into the surrounding cortical plate with radiopaque but irregular margins confirming the presence of an aggressive bony lesion (Figure 3).

An incisional biopsy was taken which showed the presence of spindle-shaped cells with pleomorphic vesicular nuclei, coarse chromatin, distinct nucleoli, and moderate cytoplasm arranged in sheets. Besides, malignant osteoid formation was also noted (Figure 4) leading to the final diagnosis of "OS of the maxillary right posterior region". Further investigations with Positron Emission Tomography (PET) were performed which showed a metabolically active primary malignant mass lesion in the right maxillary region and no evidence of other metastasis (Figure 5). It was decided that surgical intervention was the best available treatment option and she had undergone subtotal right maxillectomy and the histopathologic report came out with the pathologic stage classification of pT_1N_0 OS. A PET scan was taken post-surgically which showed the area was not metabolically active and there was no evidence of metastasis (Figure 5).

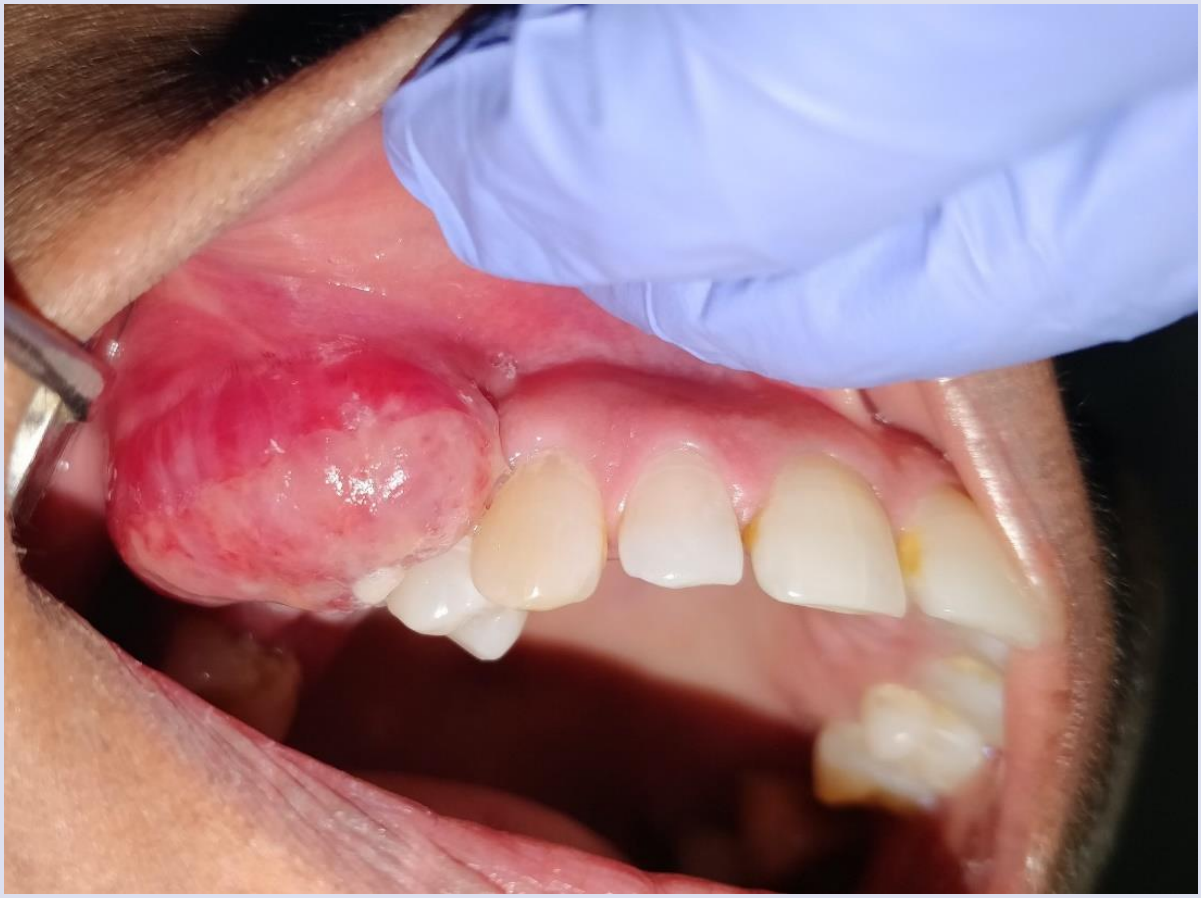

Figure 1. Swelling was seen on the maxillary right posterior region.

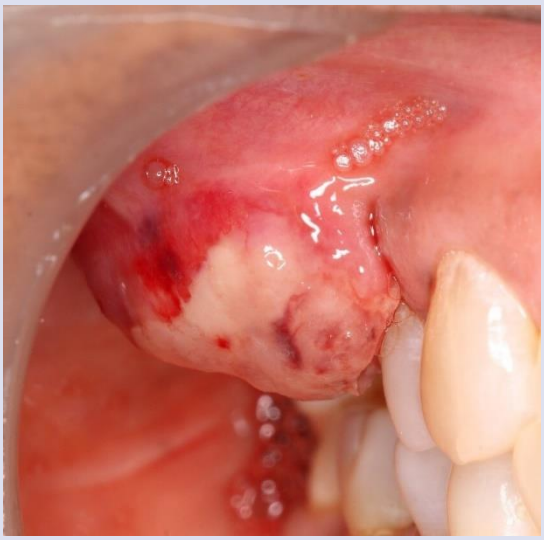

Figure 2. Swelling was presented with surface appearance that was partly erythematous.

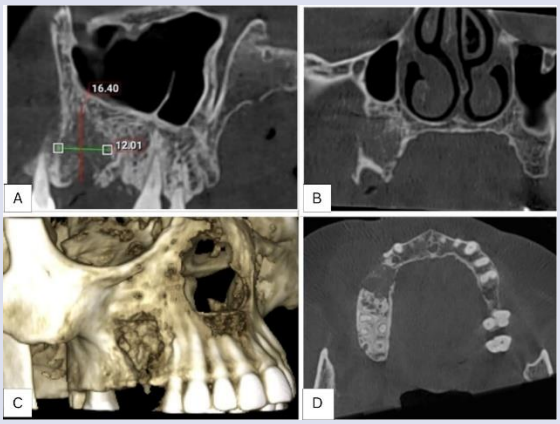

Figure 3. CBCT images showing osteolytic areas on the maxillary right posterior region
***A:** Sagittal view **B:** Coronal view **C:** Three-dimensional view **D:** Axial view*

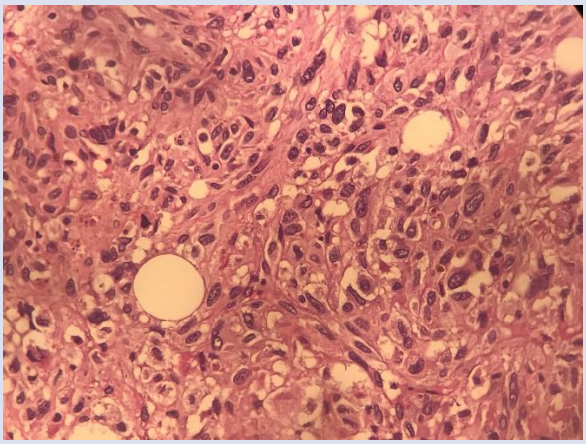

Figure 4. Histopathological picture showing the presence of malignant osteoid formation.

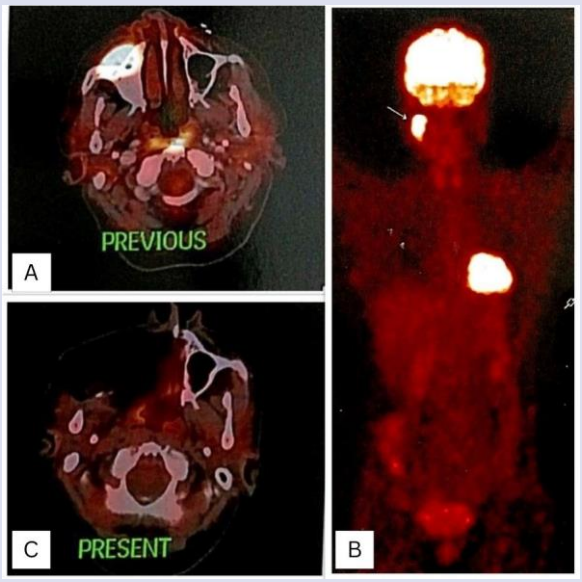

*Figure 5. PET images **A, B:** Preoperative, **C:** Postoperative*

Discussion

OS is a heterogeneous group of primary malignant neoplasms in which mesenchymal cells produce osteoid or immature bone. More than half of all OS arise in the long bones of the limbs, particularly in the region of femur, tibia, and pelvis.[10] Craniofacial OS accounts for only 1% of all head and neck malignancies.[11] Jaws are the fourth most commonly afflicted site but constitute only approximately 6 to 7% of all skeletal cases of OS.[12,13] OS of jaw bones have some distinct features from those of long bones such as older age at presentation with local recurrences being difficult to control, which leads to death of the patients.[14]

OS usually affects the long bones in growing children and young adults due to the rapid growth in their bones.[14] Jaw lesions typically occur with a peak in the fourth decade, about 10 years later on average than the occurrence in long bones which shows bimodal distribution. Mean age according to Garrington *et al.,* ranges from 34 to 36 years.[15] However, this case represents the lesion in a patient of age 58 years.

The occurrence of the disease in males is twice as frequent as in females. This has been attributed to a longer period of skeletal growth and an additional volume of bone in men, though neither has been confirmed.[4] Some authors have reported an equal predilection.[12,13] This article represents the lesion in a female patient.

Clinically, OS presents with myriad symptoms which include bony swelling, facial deformity, loosening, and/or separation of teeth. Paresthesia, toothache, and regional pain are mostly related to regional compression by the growing mass.[5,15] These tumors rarely present with symptoms of ulceration, epistaxis, or visual problems.[6] They grow rapidly causing expansion of the cortical plates with displacement and resorption of roots which were also observed in this case. They easily invade adjacent structures due to their invasive growth pattern with mucosal ulceration and pathologic fractures being common. Sensory abnormalities are encountered when the peripheral nerve is involved.[16] However, in our case, no sensory deficits or pathologic fractures were noted and the only clinical presentation was an intraoral hard tissue swelling with expansion of the buccal cortical plate.

Radiographic features show sunray spicules (ossific laminae radiating in sun-burst pattern from the affected bone surface) or “hair-on-end” trabeculae may be seen because the tumor grows very rapidly, new bone formation tends to occur in a straight line, at an angle of 90º to the bone surface. The lesion involves the periosteum directly or by extension if the periosteum is elevated due to rapid expansion and maintains its osteogenic potential only at the periphery and a Codman’s triangle at the edges is formed, which is rare in the maxilla.[17]

OS may be entirely radiolucent, mixed radiolucent-radiopaque, or quite radiopaque.[6] The Garrington sign, which is the symmetric widening of the periodontal ligament of one or more teeth on a periapical radiograph, is also an early radiologic feature and should raise suspicion for OS.[10] Garrington *et al*., in their analysis of 56 OS cases, reported the presence of a “sun ray” effect in about 25% of cases. However, these features are not specific to OS; hence, radiologic impressions can often be misleading and histopathology is considered the gold standard diagnostic modality. In addition, Magnetic resonance imaging (MRI) can demonstrate invasion of the surrounding soft tissue and peripheral mineralization.[3,10,15] In this case, CBCT images showed the lesion was radiolucent with ill-defined margins representing extensive bone loss on maxillary right posterior regions.

A retrospective analysis of 74 OS cases by Paparella *et al.* revealed varied findings in 25 cases with available

radiologic images. The presumptive clinical radiographic diagnosis in 66.6% of cases was benign lesion (dysplastic, neoplastic), and malignant neoplasia in 33.3% of cases. None of the cases was diagnosed as OS before histologic diagnosis.[15] Similar to these findings, this case also could not secure a definitive diagnosis before the biopsy.

The differential diagnoses considered in this case were non-Hodgkin's lymphoma (NHL), metastatic tumors, central hemangioma, Ewing sarcoma, and OS.

Extranodal NHL can develop in the soft tissues, most frequently the gingiva, palate, or buccal vestibule, or they can appear centrally within the bone. NHL that appears as a growth from the extraction socket is uncommon, however, it has been seen in both HIV-positive and HIV-negative patients.[18] Signs and symptoms including tooth movement, localized edema with ulcer, inexplicable dental discomfort, or vague lytic osseous alterations are frequently present. All of these characteristics fit the circumstances of the current instance.

Most metastatic tumors to the orofacial region are seen in patients aged between 40-70 years. Lung, breast, kidney, and bone malignancies are the most typical initial causes of metastatic tumors in the oral region. The most typical initial site for tumors that spread to the jawbones is the breast. Metastatic lesions to the soft tissues of oral cavity, gingiva is the most frequently affected. It has been demonstrated that gingival metastases are polypoid or exophytic, highly vascularized, and hemorrhagic. The possibility of metastasis was ruled out due to the absence of primary malignancies.[19]

Central hemangioma was considered as it has female predilection. The body of the mandible and posterior area of the maxilla are the most frequent locations for occurrence. An erythematous nodular development in the maxillary gingiva can be a symptom. The central hemangioma's radiographic appearance is not pathognomonic and is a great mimicker. Some lesions have a honeycombed appearance and radiating spicules at the expanded periphery can prove a "sunburst" appearance as in OS.[20] Central hemangioma is ruled as the age of occurrence is younger than our case and because of the radiographic appearance.

In maxilla, the occurrence of Ewing sarcoma is rare. Localized growth is the most common presentation which may be associated with pain and paresthesia. Epistaxis is usually connected with maxillary lesions, although it wasn't present in this case. "Onion skin appearance" on radiographs is mostly found in children and young adults. In our case, a lytic lesion was found, the typical onion skin appearance was not present, and the patient's age did not favor the possibility of an Ewing sarcoma.[21]

OS is typically characterized by slow healing and edema at the tooth extraction socket site. The present case showed a rapid increase in the size of the swelling followed by extraction. This case's clinical and radiographic features, such as swelling, erythematous ulcerated nodules, and complete lack of bone development within the tumor, were in accordance with previous literature.[22]

Wide radical resection is the treatment of choice for OS of jaws with clearance margins of 1.5-2 cm. Maxillectomy is difficult to perform due to the involvement of adjacent structures like maxillary sinus, pterygopalatine fossa, and orbital fossa. A subtotal inferior maxillectomy can be done for selected malignancies located on the alveolar ridge, palate, and involving the antral floor.[23] Obturators are prescribed for the defect created.

The prognostic factors depend on the site, number of metastases, surgical resection of the metastatic disease tumor size, and females with the histologic feature of predominantly chondroblastic pattern.[13,24]

A number of potential prognostic factors have been identified which include the expression of HER2/CerbB2, tumor cell ploidy, specific chromosome gains or losses, loss of heterozygosity of the RB gene, loss of heterozygosity of the p53 locus, and increased expression of p-glycoprotein. The only feature that consistently predicts outcome is the degree of histologic necrosis following induction chemotherapy. Patients with more than 95% necrosis in the primary tumor after induction chemotherapy have a better prognosis than those with smaller amounts of necrosis.[25,26]

It has been noted that patients with skip metastases (≥2 discontinuous lesions in the same bone) have a worse prognosis. Additionally, the prognosis is poor for patients with multifocal OS (>1 bone lesion at diagnosis).[27] The overall 5-year survival rate for the primary OS of the jaws varies from 30 to 40%, and survival rates up to 80% have been reported for patients undergoing early radical resection.[28]

Conclusions

OS is an ancient disease many aspects of which are still incompletely understood. OS of the jaw is difficult to diagnose and manage due to the non-specific clinical and radiological characteristics, high rate of recurrences, and difficulties in appropriate resections because of the proximity to vital structures. However, we acknowledge the need to consider OS in the differential diagnosis when dentists encounter destructive bony lesions in the maxilla as well as the mandible.

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