Case Report - Surgical Pathology

Juxtacortical osteosarcoma of mandible

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ABSTRACT

This article presents a case of juxtacortical (paraosteal) osteosarcoma in a 43-year-old female, which is rare malignant mesenchymal tumor. As per the literature, it accounts for less than 4% of all osteosarcomas. Due to its rare variety, this tumor can cause diagnostic dilemma and clinicians should be aware of it. Due to its clinical picture, it can be confused with peripheral fibro-osseous lesions. Success rate of treatment are good if the tumor is detected early and resected with wide margins giving a negligible chance for recurrence.

Keywords: Juxtacortical osteosarcoma, osteosarcoma, paraperiosteal osteosarcoma

INTRODUCTION

Osteosarcomas (OS) are the most common primary malignant tumors of bone or mesenchymal tissue that histopathologically osteoid.[1,2] It is reported that 4–13% of the total involves the jaw.[3,4] Primary OS represents a heterogeneous group of malignant bone tumor characterized by diverse histological features, clinical, and biological behavior.[5]

OS of the jaws are different from that of long bones in several aspects:

1. Seen most commonly in 3rd and 4th decade (10-15 years older than mean age of occurence).[5]
2. No bimodal distribution
3. More common in males [5:3].[6]
4. Swelling is the major complaint in OS of jaws when compared with long bones, where pain is the most common presenting complaint.
5. Have better prognosis than lesions of long bones.[7,8]
6. Metastasis (6–51%) much less than in the long bone counterpart (78–90%), but when they do occur it is to the lung in the first 2 years after surgery.[9]

Interestingly, OS almost always arise in the craniofacial bones with intra membranous ossification and practically never occurs in the bones of the base of the skull that are formed by endochondral ossification.[10] The alveolar ridge of maxilla and the body of the mandible are the most common sites of involvement.

Swelling is the chief complaint in most of the cases. Other symptoms include paresthesia and loosening of teeth. Nasal obstruction and symptoms mimicking sinusitis are found in cases involving maxilla. Duration of symptoms is partly related to the rate of growth.[10]

World health organization recognizes several variants which differ in location, clinical behavior, and degree of cellular atypia.[2]

1. Majority are intramedullary (conventional) OS which originates and grow within the central and intramedullary portion of bone. These lesions have a poor prognosis than surface lesions.[2]
2. Surface osteosarcoma (juxta cortical) is further subdivided into paraosteal, periosteal, and high-grade surface osteosarcoma.[8] Juxtacortical OS are rare neoplasms accounting for less than 4% of all osteosarcomas.[1]

Periosteal OS are characterized by peak occurrence at around 20 years of age. The tumor is often lobulated and well defined.[9] Radiographically, the cortex is intact but thickened. There may be minimal intramedullary involvement.[11]

Paraosteal osteosarcomas are extraskeletal mostly soft tissue lesions with female preponderance. They present as a very

dense lobulated mass attached by broad base to the underlying bone. The rarer juxtacortical lesions have a better prognosis than central lesions.

The histopathological variants of OS are many and include osteoblastic, chondroblastic, and fibroblastic types.[1] The rarer variants are telangiectatic, small cell, malignant fibrous histocytoma like, osteoblastoma like, granular cell rich, and epitheloid OS. It is a tumor characterized by direct bone or osteoid formation by the malignant neoplastic cell. It is most heterogeneous from a histological perspective. The amount of osteoid and ossified tissue can vary tremendously. Osteoid can again be well-defined structures that mimic trabecular bone or be irregular deposits.[10]

The radiographic picture is determined by the amount of calcified content which dictates the density of the lesion. It can vary from a dense sclerosis to a mixed sclerotic and radiolucent lesion to an entirely radiolucent lesion. Borders are often ill defined due to destructive nature of the lesion. The classic "sun burst" appearance is seen only in one-fourth of the cases.

Root resorption can lead to a tapered narrowing of the root of involved teeth called "spiking resorption." Tumor infiltration along the periodontal ligament space leads to symmetrical widening of the periodontal ligament space of one or several teeth.[10]

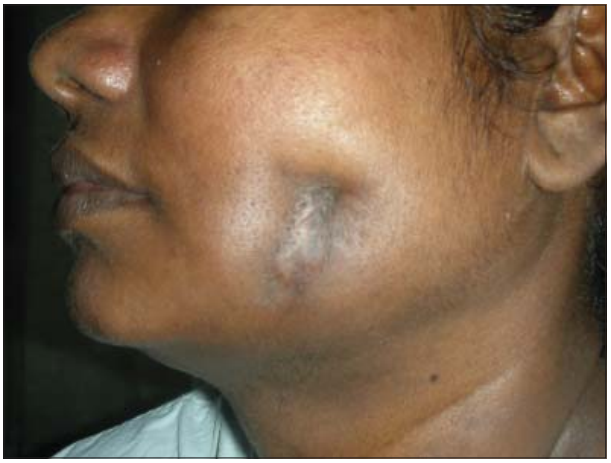

Figure 1: The puckered skin with a healed scar which was probably due to the previous surgery

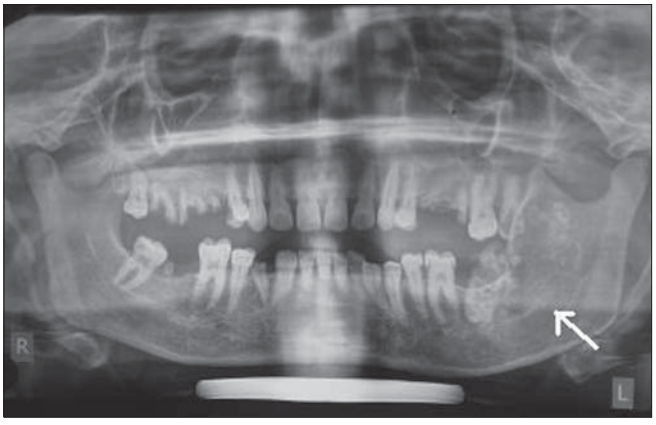

Figure 2: OPG showing scattered calcification superimposed over the left third molar and ramus region on the left side is seen

CASE REPORT

A 43-year-old female patient had come with the complaint of pain and swelling in the left angle of mandible which was progressively increasing in size since the last 4 months. Pain was continuous and was aggravated while having food and while opening the mouth. Patient had been operated 15 years back for swelling at the same site, records for which were not available.

On inspection, a swelling was seen in the left angle of the mandible measuring around 7 cm × 5 cm in diameter. The skin over the swelling was puckered with a healed scar which was probably due to the previous surgery [Figure 1].

On palpation, there was a firm to hard tender swelling in the left angle of mandible extending from left first molar to the third molar region which was fixed to underlying bone. The involved teeth showed no signs of mobility or resorption. There was no paresthesia in the area supplied by inferior alveolar nerve. The mucosa over the swelling was normal and showed no secondary changes.

OPG showed scattered calcification superimposed over the left third molar and ramus region on the left side [Figure 2]. CT scan showed a well-encapsulated juxtacortical mass in left angle of mandible with areas of calcification [Figures 3 and 4]. There was erosion of cortical bone in the third molar and the anterior ramus region with some intramedullary extension. A well-defined swelling overlying the left mandibular body with calcified/ ossified areas was seen in the soft tissue.

Incisional biopsy was done intraorally and the extrabony component of the mass was removed. The extraosseous mass was well encapsulated but fixed to the underlying bone which was eroded in the third molar region where the mass was found infiltrating into bone and was not separable. The section of

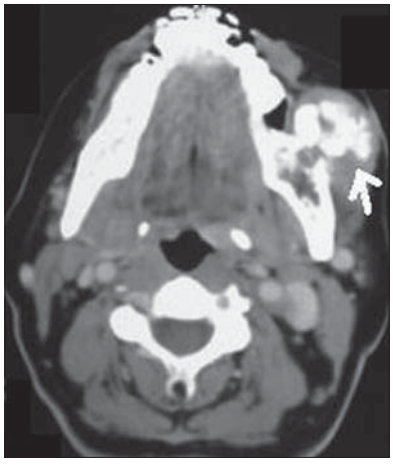

Figure 3: CT axial section showing well encapsulated juxtacortical mass in left angle of mandible with areas of calcification

specimen showed multiple irregular calcified masses some of them measuring almost 2 cm × 3 cm in diameter.

H and E-stained section showed highly cellular areas of connective tissue in which lace-like pattern of neoplastic osteoid tissue was seen. There was no demarcation between tumor osteoid and rest of connective tissue cells. Osteoid tissue was surrounded by abnormally plump osteoblast and immature bone. Few osteoblasts exhibited abnormal mitosis. Rest of the connective tissue showed plump, pleormorphic, and haphazardly arranged cells with vesicular nuclei [Figure 5]. Moderate vascularity and areas of hemorrhage were also seen. Overall picture was suggestive of a low-grade osteosarcoma involving the mandible.

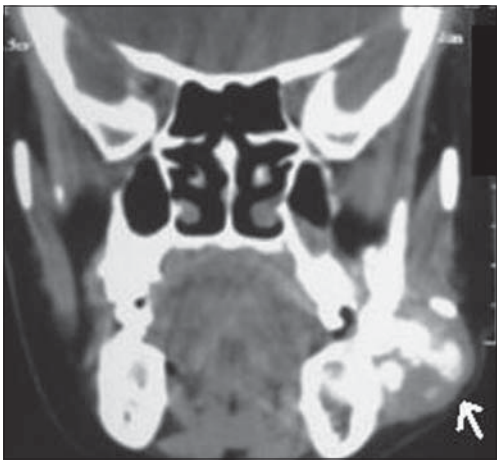

Figure 4: CT coronal section showing erosion of cortical bone in the third molar and the anterior ramus region with some intramedullary extension

The patient was subjected to a whole body examination and relevant radiographs were done to rule out primaries elsewhere. The radiographs were normal and it was confirmed as primary osteosarcoma of the mandible.

Within 2 weeks of the biopsy, the patient developed trismus and increasing pain in the same region. The skin over the lesion was fixed and nonpinchable. Repeat CT scan showed involvement of masseter muscle along with the skin.

Patient underwent a hemimandibulectomy with wide excision of the tumor mass with involved skin. Involved skin paddle was marked with a circular incision which was connected anteriorly

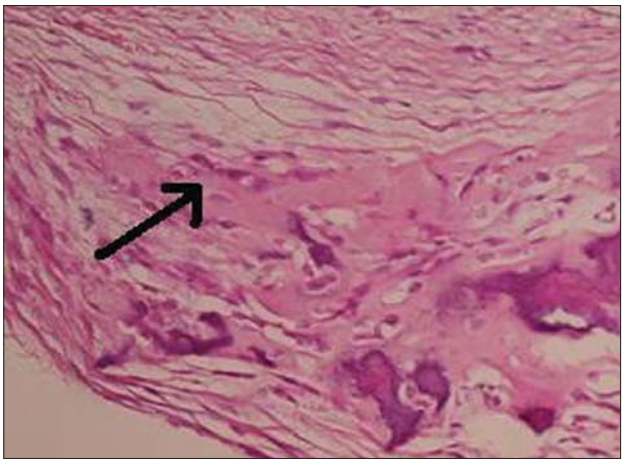

Figure 5: Histopathology slide showing highly cellular areas and plump osteoblasts (H&E, 40×)

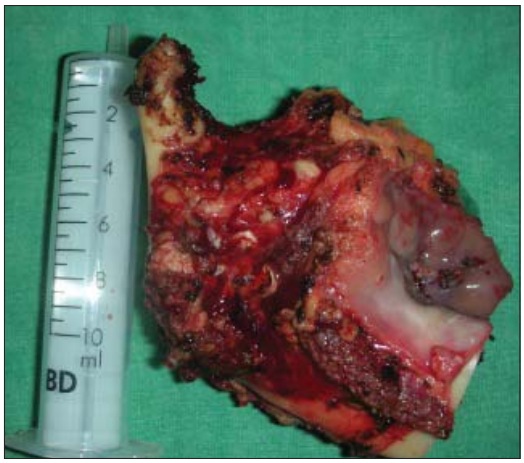

Figure 6: The excised tumor mass along with the overlying skin and involved soft tissue and hemimandible on that side from canine to condyle

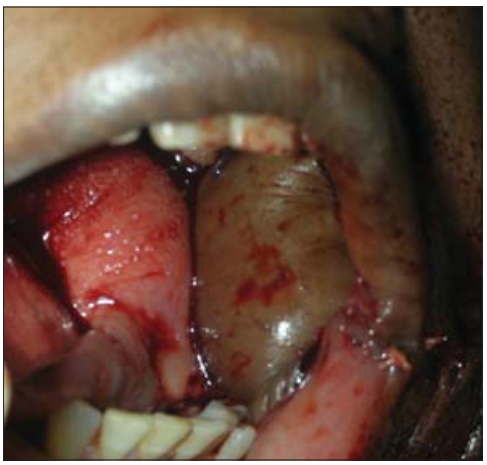

Figure 7: PMMC flap in position

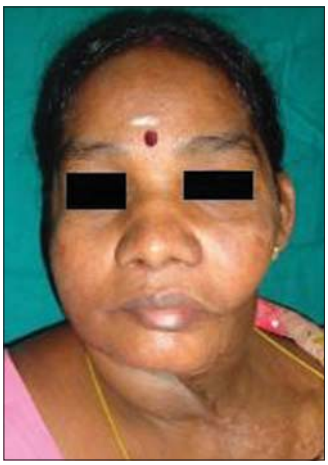

Figure 8: One year postoperative picture of the patient

to the left commisure of the lip. It was then extended along the paramedian line all along the neck to the clavicle where it was extended horizontally. Blunt dissection was done through layers. Lower border of mandible was exposed. The tumor mass along with the overlying skin and involved soft tissue and hemimandible on that side from canine to condyle was excised [Figure 6]. Adequate clearance was achieved on all aspects and was confirmed with fresh frozen section. A pectoralis major myocutaneous flap was raised and used to line the intraoral defect, while the overlying skin defect was covered with cervical rotation flap [Figure 7]. Postoperative period was uneventful and patient after regular follow-up for 1 year was symptom free with no signs of local recurrence or distant metastasis [Figure 8].

DISCUSSION

Primary OS represent a heterogeneous group of malignant bone tumors with diverse features posing a diagnostic dilemma.

Age can be an important factor in the degree of differentiation of OS. According to them older patients have a better prognosis due to increased resistance to the tumor.[7,12]

Some preexisting conditions that can lead to development of OS are previous exposure to radiation, fibrous dysplasia, Pagets disease, and local trauma. This suggests an association between the occurrence of this neoplasm and increased cellular activity.[5]

In this particular case, although there did not seem to be any obvious predisposing factors, there might have been some underlying fibrosseous lesion which was operated 15 years back. This previous surgery could have triggered off secondary changes and malignant transformation. However, histopathological reports of previous surgery were unavailable to draw any conclusion.

Considering the fact that this case presented with a lobulated sessile mass with only minimal intramedullary involvement, no mobility or resorption of roots of involved teeth, no evidence of periodontal ligament space or mandibular canal widening on plain radiographs, it could probably be a Juxtacortical osteoblastic variant of low grade osteosarcoma with a predicted good prognosis and 5 year survival rate. As evident in the coronal CT the lesion was predominantly extraskeletal with an intact cortex, showing breach only in the third molar region, with minimal intra medullary extension from where the biopsy was performed. Considering the above radiographic and the CT findings, it was concluded as a case of paraosteal variant of juxtacortical osteosarcoma. The current trend is to give more importance to the histological grading than the tissue pattern as the former seems to have greater bearing on the prognosis.

Differential diagnosis of reactive bone lesions, fibromyxoid chondroma, and peripheral fibrosseous lesions were considered before concluding it as a low-grade osteosarcoma.[10] These all were ruled out histopathologically, as it did not show any chondroid or myxoid tissue and showed malignant cells.

Literature review showed that patients treated initially by wide local excision like hemimandibulectomy fared better. The higher success rate associated with such aggressive surgical resections could be due to two reasons:

1. Wide excision of adjacent bone and periosteum prevents local recurrence by removing small satellite lesions and
2. Overcomes the risk of local spread of tumor along bone surface.

Chemotherapy does have a role to play in modern management of OS by improving primary control and eradicating systemic disease. As this particular case was a low-grade juxtacortical type, radical surgery was considered sufficient and chemotherapy was reserved for recurrence if any in future.[13]

Accurate diagnosis is considered the key to establish an effective therapeutic regimen that will improve the survival rate of patients with OS. As local recurrence is reported as the most frequent complication of this variant, meticulous long-term follow-up is mandatory for all patients.

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