

PRIMARY OSTEOSARCOMA OF MANDIBLE – A CASE REPORT

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ABSTRACT

The garden variety of Osteosarcoma occurring in long bones, also affects the mandible and maxilla. It's a proportionately uncommon lesion, contributing to just 2% to 10% of all osteosarcoma cases incidence. It occurs in the core of the bone and is classified into three subtypes based on the prevailing cell type: osteoblastic, chondroblastic, and fibroblastic. These tumors bring a variety of bone modifications on radiographs, from well-defined boundaries to lytic bone degradation with undefined margins and irregular cortical bone loss, as well as sclerotic bone in some cases. Surgery, chemotherapy, and radiotherapy are the common methods used to treat osteosarcoma, depending upon the age, histological type, and tumor location. The following case report describes a 25-year-old male patient, who developed Primary Osteoblastic Osteosarcoma of the right angle of mandible. A brief discussion of its clinical, radiologic, histopathologic features and treatment are included in this report.

**Keywords:** Osteosarcoma, Mandible, Neoplasms, Bone Tissue, Bone Formation.

INTRODUCTION

Osteosarcomas are rare bone malignancy which has been reported with high risk of mortality. They belong to the group of connective tissue neoplasms that grows in expense of mesenchymal cells and capable of forming bone or bone-like material. <sup>1</sup> It is most common in rapidly growing bones and has an average occurrence between the second and fourth decades. Males have always been thought to have a greater prevalence of osteosarcoma than females, with males showed 5.4/M affected per year compared to 4.0/M incidence in females. Primary bone sarcomas are very rare cancers that make up less than 0.2% of all cancers. The three most prevalent sarcomas of bone are osteosarcoma (35%), chondrosarcoma (30%), and Ewing's sarcoma (16%).

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The metaphysis of long bones are the most frequent site, with the area immediately near the knee, responsible for nearly half of all instances and an average of 7% in jaws.<sup>2</sup> Jaw tumors are most prevalent between the ages of 30 and 39, with a significant male predilection in the mandible and female predilection in the maxilla.<sup>3</sup> Roentgenographic evaluation of osteosarcoma shows a spectrum of bone mass, and the characteristic "sunray" pattern can be seen at the tumor's periphery.<sup>4</sup>

### CASE REPORT:

A Male patient of age 26 years presented with a swelling in the lower right posterior region of jaw for 6 months. he gave a history of swelling that developed after food impaction in the left lower jaw that was larger and painful 6months ago. It reduced spontaneously on taking medications to present size. And associated with pain which was dull and intermittent, aggravated during mastication. His medical history was not contributory but revealed that the patient's sibling had succumbed to the treatment of brain cancer at the age of 16 years. His vitals and general appraisal were normal without any abnormalities with the other systems. On extraoral examination, a diffuse swelling was evident on the right mandibular angle, extending anteriorly

three cm behind the angle of mouth and posteriorly few mm behind the angle of mandible; superiorly, two cm below the line joining angle of mouth and tragus of the ear and inferiorly one cm below the lower border of mandible approximately 8×7 cm in size. Overlying skin appeared normal color, firm and tender on palpation. No regional lymph nodes were palpable (Figure-1). On intraoral examination, a diffuse swelling was evident on right mandibular body extending from mesial side of 34 to retromolar trigone, irregular in shape, approximately measuring - 7×6 cm with obliteration of mucobuccal fold, overlying mucosa was smooth and appeared normal in color and surrounding mucosa was normal, no discharge was evident. The swelling extended to the ramus of mandible and bicortical expansion was evident. Palpatory findings revealed tenderness, hard consistency, no discharge was evident and the borders were ill-defined (Figure-2). The associated teeth were normal. The differential diagnosis according to clinical presentation, considered were Periapical cyst, Dentigerous Cyst, Odontogenic Keratocyst and Ameloblastoma.

The panoramic view revealed periapical region of 48 showed resorption of apical third of mesial and distal roots with loss of lamina dura. Altered trabecular pattern

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seen on the angle of mandible and ramus on right side. Loss of cortical border of angle and ramus of mandible. Inferior alveolar canal border is indistinct (Figure-3). Ultrasonography of the angle of mandible revealed, heteroechoic lesion noted along the posterior aspect of the body, ramus and mandibular angle on right side, measuring = 6x4cms, extending anteriorly beneath the masseter muscle on right side, Interiorly the lesion extends into the submandibular region (Figure-4). Cone Beam Computed Tomography (CBCT) examination of jaws revealed, an ill-defined destructive radiolucent lesion is noted extending from 47 region (body of the mandible) till mid-ramus region. Loss of lamina dura is noted in 47 and 48. 48 shows spike-like root resorption on both mesial and distal root. Corticated border of inferior alveolar canal appears to be destructed and poorly visualized. Destruction of the lingual cortical plate is noted at the level of mandibular angle which appears to extend till the lingula. Osteophytic periosteal reaction is noted as sun-ray/ sun-burst appearance in the angle and ramus region. Multiple foci of destruction/ radiolucency are noted in the corticated border of right condylar head (Figure-5). Magnetic Resonance Imaging (MRI) revealed, Large lytic expansile T2, STIR hyperintense lesion seen involving the condylar process, ramus and body of

mandible on right side with marked surrounding solid sunburst periosteal reaction. The lesion shows cortical breach along the inner cortex in the ramus portion. No significant soft tissue component. The lesion shows mass effect in the form of mild compression on the right submandibular gland, masseter muscle and the remaining masticator space muscles ipsilaterally. No infiltration into the muscles. The lesion measures 8 x 7:7 x 4-5 cm. Craniocaudally the lesion extends from the level of the condylar fossa to the level of 1st molar. Medially the lesion is seen abutting the alveolar process to maxilla and the lateral pterygoid plate. The inferior dental nerve is spared. No significant increase in size when compared to previous sea Few enlarged lymph node seen in the right Ia, Ib and level II lymph node station. Histopathology shows tumor composed of variably sized tumor cells having spindled to round oval nuclei showing marked pleomorphism and hyperchromatism with lace like disorganized woven bone intimately associated with tumor cells surrounded by highly atypical cartilaginous matrix with mitotic figures and focal areas of empty lacunae with ghost cells seen with tumor cells infiltrating through the cortex into adjacent skeletal muscle fibres. No lymphovascular or perineural invasion was

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seen (Figure-6). Finally, definitive diagnosis of **Low-Grade Osteogenic Osteosarcoma of Right Angle of Mandible- (MSTS-Ia)** was given. Primarily, Radical Hemimandibulectomy along with functional neck desiccation was done. Later, patient underwent 60 Neo-adjuvant Chemotherapy (NACT) Cast Cycle.

### DISCUSSION:

The risk of developing conventional osteosarcoma depends on higher fluoride levels in water, radiation exposure, treatment for leukaemia. In young patients, hereditary syndromes characterized by somatic or germline defects showed a predisposition. Many significant correlations with puberty and height, as well as diseases of bone development and remodelling, were already established through epidemiological data. Whereas, osteosarcomas of jaws include chronic osteomyelitis, fibrous dysplasia, bone infarcts, trauma, high-dose radiation, paget's disease, metallic replacements, infectious infection, joint prostheses in genetic syndromes such as Li-Fraumeni syndrome, retinoblastoma, and retuximab therapy.

This patient had an unusual clinical appearance, with mild signs, swelling, and involvement of the posterior mandible.

However, the lesion's accelerated progression from the day it was first seen to the time it was recorded was not preferentially associated with the common growth trend of osteosarcomas. Osteosarcomas of jaws are clinically different from long bone osteosarcoma.<sup>5</sup>

Numbness caused by involvement of the inferior alveolar nerve(IAN) in the lesional area has been reported in osteosarcoma and can indicate a low prognosis.<sup>6</sup> Even though, numbness was not the patient's presenting symptom, he seemed to have a tingling sensation on the right side of his lower lip. Despite the patient's tingling sensation in the affected region, there was no evidence of mandibular canal invasion on radiographs. The bone swelling and loss of lamina dura in roots of 3<sup>rd</sup> molar may be due to periodontal diseases, odontogenic cysts and metastatic neoplasm. The presence of periosteal reaction most likely aided in diagnosis. In jaw lesions, the typical 'sunray' or sunburst appearance and the Codman's triangle formations are less common unlike osteosarcoma of the long bones.<sup>7</sup>

Based on the tumor cells matrix, osteosarcoma can be subclassified into osteoblastic, fibroblastic and chondroblastic. The osteoblastic variant is

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characterized by the presence of immature bone surrounded by haphazardly arranged fibroblast-like cells. Abnormal hyaline chondroid material can be seen in the chondroblastic variant. The cartilage may be the most prominent feature or it may be dispersed around the tumor. The fibroblastic variant has spindle-shaped neoplastic cells that are arranged in a herringbone pattern. The histologic type and grading of the tumor, in addition to the extent of the lesion, are essential considerations in deciding the prognosis.<sup>8</sup> Bone sarcomas are staged based on grade, size, and the presence and location of metastases.

**TABLE-1 Musculoskeletal Tumor Society Staging System (MSTS)**

Stage	Grade	Local extent	Metastases
I A	Low	Intracompartmental	None
I B	Low	Extracompartmental	None
II A	High	Intracompartmental	None
II B	High	Extracompartmental	None
III	Any	Any	Present

Unlike long bone osteosarcomas, the treatment of choice for jaw osteosarcomas is radical surgical excision, which has a 5-year survival rate of over 80% theoretically. Since jaw osteosarcomas form at a younger age, the patients are less likely to develop metastases. Local recurrence and metastasis of jaw osteosarcomas are just 18%. Jaw osteosarcomas have a higher prognosis than long-bone osteosarcomas in general. This may be because of the jaw osteosarcomas have a greater histological differentiation than long bone osteosarcomas.<sup>9</sup>

The two significant considerations in improving the prognosis of jaw osteosarcomas are early intervention and thorough tumor resection. Radical surgery is being used to treat osteosarcoma. The prognosis of osteosarcoma is improved by tumor-free margins in surgery, chemotherapy, and radiotherapy following surgery.<sup>10</sup> For long bone Osteosarcomas, External Beam Radiotherapies are usually provided five days a week for several weeks.

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**FIGURE-1** Extraoral appearance



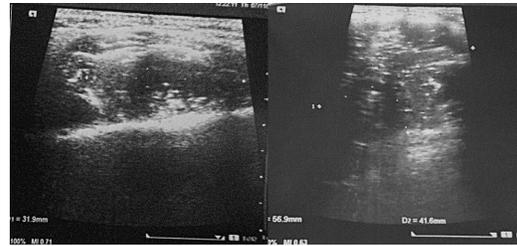
**FIGURE-2** Intraoral picture



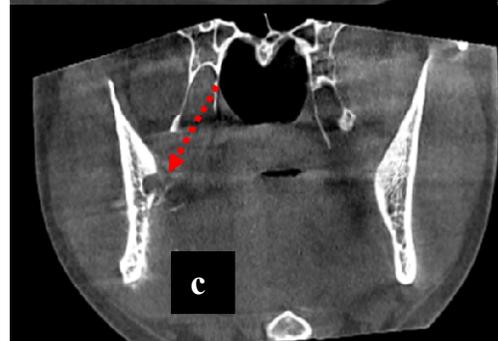
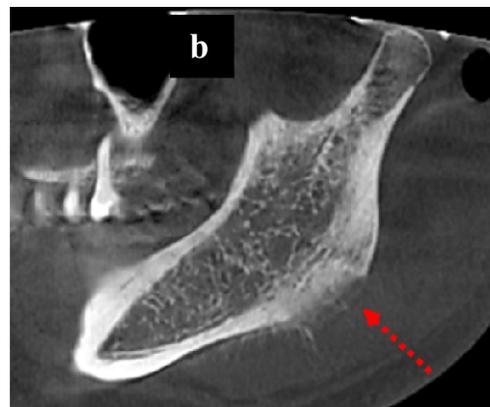
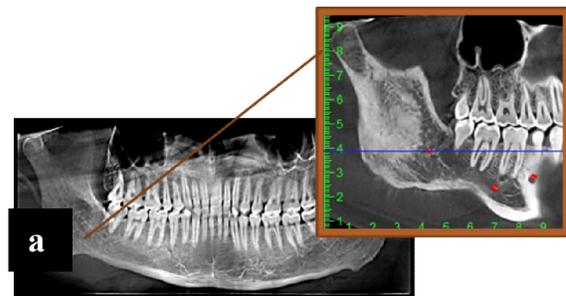
**FIGURE-3** Orthopantomograph



**FIGURE-4** Ultrasonography



**FIGURE-5** a-CBCT panoramic reconstruction, b-sagittal view, c- coronal view.



**FIGURE-6** Histopathology

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