

## Case Report

# Osteosarcoma of jaw, common entity at an uncommon site: a rare case report

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## ABSTRACT

Osteosarcoma is the most common primary malignant bone tumor. Jaw is an uncommon site. The etiopathogenesis is still unknown. We present a case of 47-year-old female who underwent left partial maxillectomy as the lesion increased in size over one month after removal of a tooth. Prior to the surgery, computed tomography (CT) scan was suggestive of left maxillary sinus carcinoma and biopsy report was suggestive of spindle cell lesion. Partial maxillectomy specimen received in which size of the tumor was 5.5×5.5×4 cm with a pearly-white, solid, homogenous cut surface with gritty sensation on cutting. On histopathology it turned out to be osteosarcoma of the jaw. Mouse double minute 2 homolog (MDM2) and special AT-rich sequence-binding protein 2 (SATB2) was done to confirm the diagnosis of osteosarcoma. The patient was given chemotherapy after the confirmation and at present the patient is doing well.

**Keywords:** Osteosarcoma, Jaw, Bone tumor, MDM2, SATB2

## INTRODUCTION

Osteosarcoma is the most common primary malignant bone tumor. It is characterized by osteoid synthesis of malignant osteoblasts. Long bones are the most common site. Jaw is an uncommon site. Incidence of osteosarcoma of jaw is 6 to 7% of all the bone tumors.<sup>1</sup> Clinical features such as swelling of jaw associated with pain and tooth mobility is the most common presenting feature.<sup>2</sup> It is considered to be less aggressive with lower incidence of metastasis and has better prognosis as compared to the one occurring in the long bones.<sup>2</sup>

## CASE REPORT

A 47-year-old female came to the out-patient department with complaints of lesion in the oral cavity which was increasing in size after removal of a tooth one month back (Figures 1 and 2). Patient was afebrile and routine blood investigations were done. Magnetic resonance imaging (MRI) of face and neck (plain) revealed a large lobulated

altered signal intensity mass measuring 4.9×5.6×6.5 cm involving the maxillary sinus causing destruction of the floor of the maxillary sinus and mandibular bone with extensions suggestive of neoplastic etiology. Her computed tomography paranasal sinus (CT PNS) with contrast report showed a 5×4 cm hyper density bony lesion in the anterior and lateral walls of left maxillary sinus with soft tissue component seen in the left maxillary antrum causing erosion of the lateral and interior walls of the maxillary sinus extending along the maxillary alveolus suggesting a left maxillary sinus carcinoma (Figure 3). A biopsy of the mass was reported as spindle cell lesion probably benign in nature.

The patient underwent left partial maxillectomy surgery. The surgery was uneventful. The mass was sent to the histopathology department. We received a left partial maxillectomy specimen measuring 6×5.5×5.5 cm (Figure 4). The specimen measures 5.2 cm along upper alveolus and had two teeth in-situ. A 5.5×5.5×4 cm exophytic mass was noted over the roof of the oral cavity and was seen

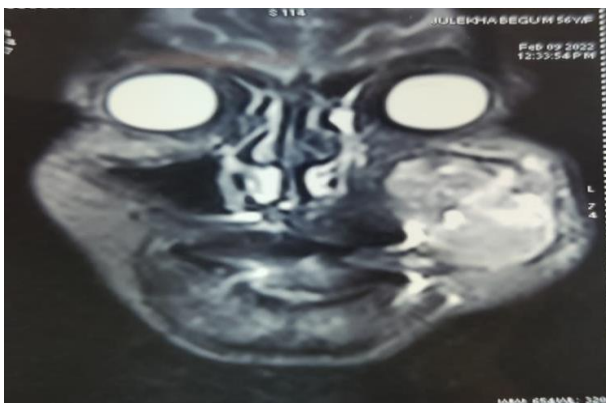
infiltrating the maxilla and extending into maxillary antrum. The cut surface of the mass was pearly-white, solid, firm, homogenous with gritty sensation on cutting (Figure 5). No cystic areas or hemorrhage was noted.



**Figure 1: Swelling over the left jaw.**



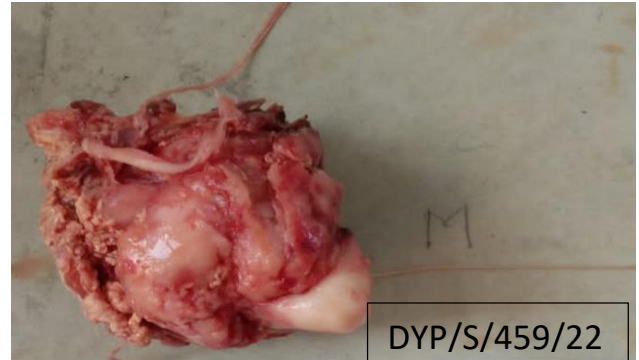
**Figure 2: Lesion in the oral cavity.**



**Figure 3: Bony lesion in the anterior and lateral walls of left maxillary sinus.**

Microscopically sections studied from tumor shows focally ulcerated stratified squamous epithelial mucosa (Figure 6). The submucosa shows a malignant tumor arranged in fascicles and in a haphazard manner. Individual tumor cells were spindled with mild to moderately pleomorphic round to oval vesicular nuclei, prominent nucleoli and moderate eosinophilic cytoplasm.

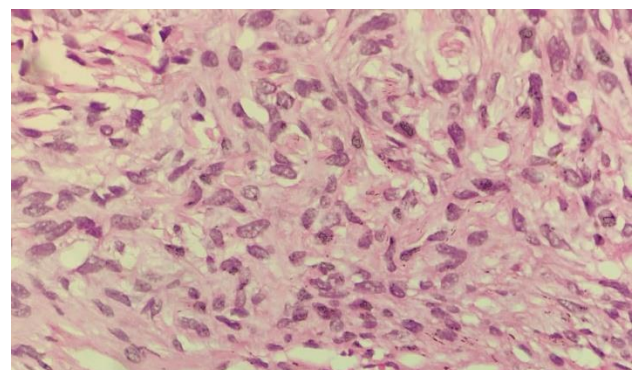
Bizzare cells were also seen. The tumor cells were seen infiltrating and destroying the maxillary bone. Lace like osteoid matrix was seen which at places was mineralized. Tumor cells are also seen surrounding the lace like osteoid matrix giving a whorled appearance (Figure 7a and b). Areas of necrosis are also seen. The immunohistochemistry studies of diffuse and strong nuclear positivity (SATB2) and focal and strong nuclear positivity (MDM2) confirmed the diagnosis of osteosarcoma (Figures 8 and 9). A histopathology diagnosis of high-grade osteosarcoma of left maxilla was given.



**Figure 4: External surface: left partial maxillectomy specimen.**

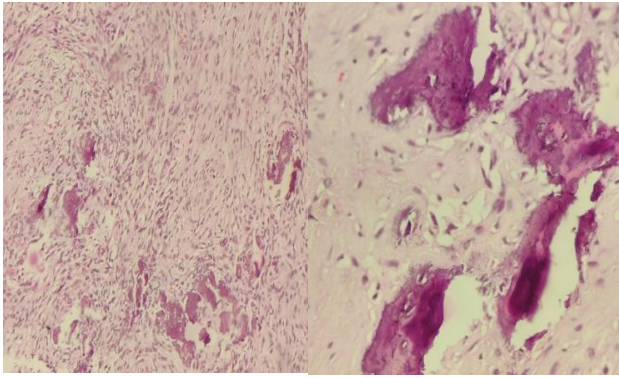


**Figure 5: Cut surface: pearly-white, solid, firm, homogenous.**

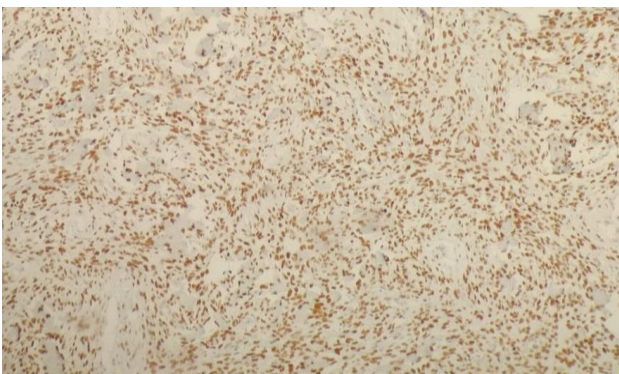


**Figure 6: Spindle cells with mild to moderate pleomorphism, prominent nucleoli and eosinophilic cytoplasm (40x H&E).**

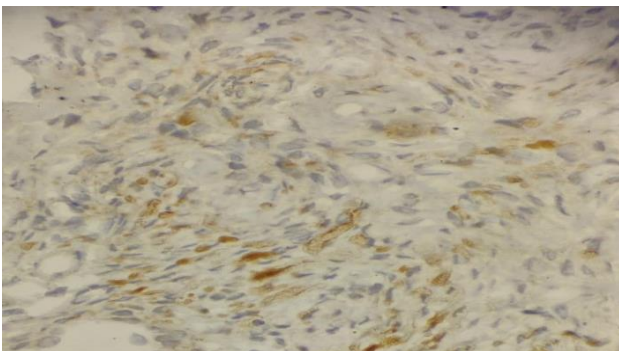




**Figure 7: (a) Tumor cells surrounding the lace like osteoid matrix (10x H&E), and (b) osteoid matrix (40x H&E).**



**Figure 8: SATB2 (diffuse and strong nuclear positivity) (10x).**



**Figure 9: MDM2 (focal and strong nuclear positivity) (40x).**

## DISCUSSION

Osteosarcoma of long bones is most common. In this case tumor was in the jaw. Osteosarcoma more commonly occurs in males than in females. The patient was a female in this case. Osteosarcoma usually present with swelling, pain, ulcerations. The most common presenting symptom is pain, swelling and loosening of teeth also noted.<sup>2</sup> Symptoms such as nasal obstruction, ulceration, epistaxis,

gingival inflammation and numbness of lower lip is also seen.<sup>1</sup> The early lesions of this usually occurs as a lobular mass which involves the surface of the bone. Radiological features of osteosarcoma are of bone destruction and bone formation which are dependent on tumor behavior.<sup>2</sup> Lytic, sclerotic or mixed lesion with soft tissue extension seen in most of the cases.<sup>1</sup> It gives a sun ray appearance on X-ray. Sun ray appearance is due to many thin irregular spicules of new bone which may develop outward and seen perpendicular to the surface of the lesion.<sup>1</sup> But this sun ray appearance is usually absent in osteosarcoma involving the jaw. Mandible is more commonly affected as compared to maxilla.<sup>1</sup> In mandible they occur usually in the body and ramus whereas in maxilla they occur in premolar and canine region.<sup>1</sup> Special investigations such as CT scan and MRI play a major role in assessing the size of lesion for staging, for tumor calcification and for knowing about the invasion of tumor into the adjacent tissues.<sup>1</sup> Histopathologically, osteosarcoma of jaw is osteoblastic, along with osteoid matrix and minimal atypia.<sup>4</sup> It is characterized by well differentiated lesions.<sup>4</sup> In the present case, tumor cells were oval and spindle along with the surrounding lace like osteoid matrix giving a whorled appearance, with minimal atypia, and few mitoses. It was a typical osteoblastic type.<sup>4</sup> Therapeutic modalities in a primary treatment of osteosarcoma is chemotherapy and radiotherapy.<sup>4</sup>

## CONCLUSION

Though osteosarcoma is commonest tumor of long bone, jaw is a rare site. Clinical and radiological presentation can be misleading. Hence, histopathology and immunohistochemistry remain gold standard diagnostic approach.

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