Case Report

# Osteosarcoma of jaw - Case report and review of literature

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## ABSTRACT

***Osteosarcoma is a bone tumor and can occur in any bone, usually in the extremities of long bones, near the metaphyseal growth plates. Osteosarcoma of the jaw bones represents a distinct group of lesions from the conventional type commonly occurring in long bones. The emphasis should be laid on the aggressiveness of this lesion which warrants an early identification and diagnosis of the lesion followed by prompt treatment. True synchronous multicentric osteosarcomas of the jaws are extremely rare but, like other osteosarcomas of the jaws, have a favourable outcome, and palliative resection of such lesions, though challenging, can therefore lead to an enormously improved quality of life and self-image, and may even offer the opportunity for cure. We present a case of osteosarcoma of mandible in a young male treated by radical excision and reconstruction using free fibula bone flap.***

***Key Words: Bony swelling, osteosarcoma, jaws, radical surgery, reconstruction***

## Introduction

Osteosarcoma refers to a group of primary malignant neoplasms affecting bone. Osteosarcoma of jaws is a rare, aggressive malignant mesenchymal tumor which is characterised by the formation of osteoid tissue. It constitutes 5%-13% of all cases of skeletal osteosarcomas. [1, 2] Jaw osteosarcomas usually occur in 4th decade of life. The maxilla and mandible are equally involved. The mandibular tumors arise more frequently in the posterior body and ramus, whereas the maxillary tumors are discovered more commonly in the alveolar ridge, sinus floor and palate. [1-4]

Clinically, osteosarcoma of jaws presents as swelling associated with pain and loosening of adjacent teeth. The other clinical features include trismus and paresthesias in the case of mandibular tumor and nasal obstruction in maxillary tumors. [1, 2]

Radiologically, it manifests as mixed, radiolucent/radioopaque lesions mainly. The other radiological features include PL widening, radioopaque masses with moth eaten appearance, codman's triangle and sunburst appearance. [4] The etiology remains debatable and they have been regarded as infection associated lesions and pseudo or true neoplasms. [3] However, ionizing radiation, fibrous dysplasia and pagets disease of bone are considered to be predisposing factors. The current knowledge on jaw osteosarcoma has indicated that certain other factors which appeared to co relate with the

occurrence include linear bone growth and genetic and environmental factors. [4, 6]

Osteosarcomas are categorised histlogically into osteoblastic, chondroblastic, fibroblastic subtypes depending upon the predominant type of extracellular matrix present. [7] Early diagnosis and adequate surgical resection are the keys to high survival rates. [6] The treatment protocols for osteosarcoma include radical or conservative surgery which is complimented by radiotherapy and/ or chemotherapy. [8]

We are reporting a case of osteosarcoma involving the mandible in a young male which was initially being treated as an abscess, so presented late to us in course of disease. The patient was managed by radical excision and reconstruction using free fibula bone flap

## Case Report

An 18 year old male patient reported to department of dentistry, PIMS medical college and hospital, Jalandhar with chief complaint of swelling in right lower jaw since 3-4 months which had been slowly increasing in size. It was associated with pain, which was severe, pricking and continuous in nature. According to the patient the swelling was evident extraorally since 1 month. He also expressed difficulty in eating since 1 month. On examination, an extra oral swelling 6*5 cm in size ovoid in shape was present in lower jaw on right side. (Fig. 1) On palpation, the swelling was hard in consistency, smooth, tender on touch, non mobile, and non pulsatile. Intraorally, there was a buldge on alveolar ridge in relation to lower right region extending from 1st molar to ramus, approximately 6 x 3.5 cm in size. The swelling had distinct margins, a smooth surface and normal colour of overlying mucosa.

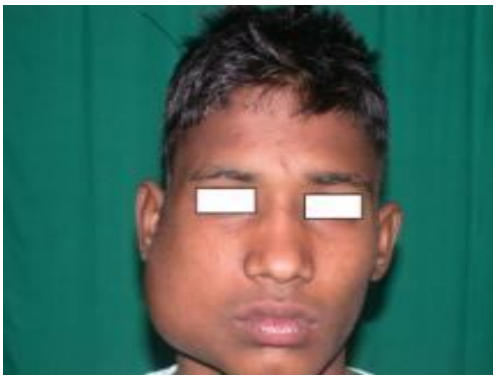

**Fig.1 Extra oral swelling on right side of mandible**

On palpation, buccal and lingual cortical plate expansion was evident and swelling was found to be hard, tender and immobile. The radiographic evaluation included a panoramic radiograph and non contrast computerised tomogram with 3D reconstruction of face. OPG showed a large radiolucent lesion extending from lower 1st molar and involving whole of ramus and condylar neck with loss of trabecular pattern. (Fig. 2)

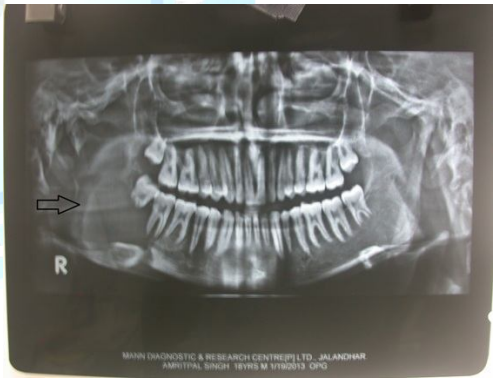


**Fig. 2 OPG showing radiolucent lesion right side with destruction of mandible**

CT scan revealed an expansile mass involving the right side of mandible with destruction of cortex in ramus of mandible. (Fig 3) FNAC was done and report was suggestive of presence of malignant cells. The patient underwent right hemimandibulectomy (Fig. 4) with free fibula reconstruction in the same sitting.

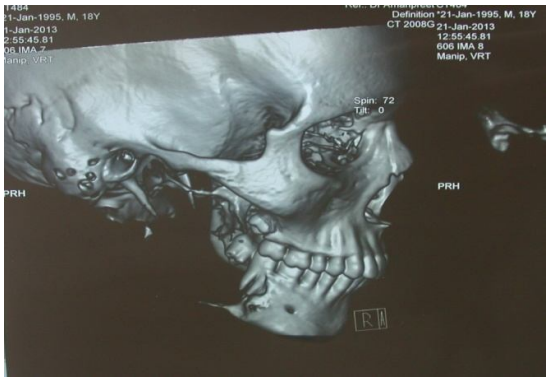


**Fig. 3 CT scan showing extent of bony destruction**

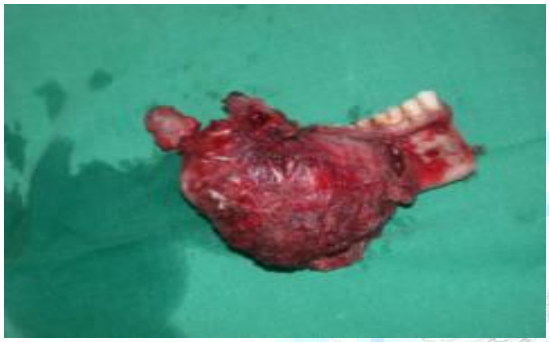

**Fig. 4 Right hemimandibulectomy specimen**

HPE revealed round to ovoid tumor cells arranged in the form of sheets or lobules in a scanty stroma. (Fig. 5) Stroma exhibited numerous blood vessels packed with red blood cells and foci of tumour osteoid.

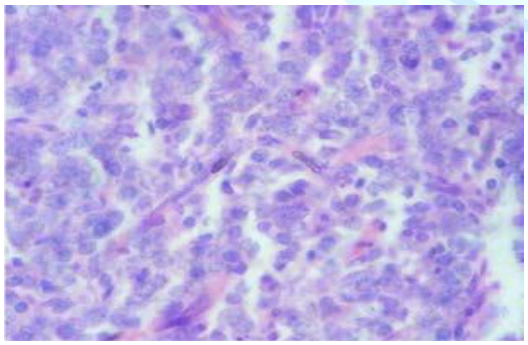

Fig. 5 - Microphotograph showing sheets of round cells with indistinct cellular margins (H & E 100x)

## Discussion

Osteosarcoma is a rare tumor in maxillofacial region, its occurrence is 1 in 1, 00,000 persons per year. The lesions of the jaw despite the histopathological similarities with osteosarcoma of the long bones are biologically different. [1]

They have been classified according to their site of origin into conventional type i.e arising within the medullary cavity, juxtacortical tumor arising from the periosteal surface and intraskeletal osteosarcom. They arise rarely in soft tissues. Juxtracortical osteosarcoma is a rare neoplasm, comprising less than 4% of all osteosarcomas and occurs less commonly than the conventional tumor. [9] The clinical characteristics of the case reported here were in agreement with those of previous studies. [1-4] There is a need to be more conscious while such patients are being diagnosed, as they often go undiagnosed for a significant period of time. Further, its presentation is similar to that of osteomyelitis with proliferative periosteitis, suppurative osteomyelitis, ossifying fibroma, osteoblastoma and even fibrous dysplasia, which has often caused an osteosarcoma to be delayed in its diagnosis. [6, 8]

The radiological features of the osteosarcoma in the present case showed a radiolucent lesion, with expansion of cortical plates. The typical feature of osteosarcoma is a sunburst pattern but it is not a sensitive or specific feature. The extent of tumor was best appreciated in CT scan. CT scan of an osteosarcoma often shows the formation of irregular endosteal and extracortical bone as well as a destroyed or obliterated cortex. However, CT cannot differentiate between osteosarcoma and fibrous dysplasia.

It was observed that the lesion in this case was a radiolucent- radiopaque in appearance which was in accordance with the findings of Clark et al classification. [10] The radiographic differential diagnosis included peripheral osteoma, myossitis ossificans.

Histologically, osteosarcoma can be classified according to their cellular differentiation as osteoblastic, chondroblastic and fibroblastic. A majority of tumors are heterogenous reflecting the pleuripotency of proliferating mesenchymal cells.

The other histological types include a malignant fibrous histiocytoma like osteosarcoma which shows spindle anaplastic cells. In large cell predominant osteosarcoma there are large cells with prominent nucleoli. This type of tumor may be confused with a giant cell tumor.
In small cell or round cell predominant type osteosarcoma, osteoid producing small malignant cells and primitive bone tissue are the characteristics whereas in telangiectatic osteosarcoma anaplastic cells are present along with osteoid. In our case, histologically the tumor was composed of hyperchromatic cells and pleomorphic cells.

The decalcified tissue section showed vital bone with marrow spaces which exhibited the proliferation of angular cells some of which were in lacunar spaces, some in association with an amorphous eosiniphilic material which was suggestive of chondroblastic osteosarcoma. [9, 10]

The treatment for osteosarcoma has been well established in the long bones but it is not well understood when the condition involves the mandible or the maxilla. [3, 7] It is clear that chemotherapy is beneficial for osteosarcoma of long bones leading to significant changes in disease free survival rate. The improvement did not include osteosarcomas of jaws, due to its rare occurrence and due to lack of standardized chemotherapy protocols, which made it difficult to evaluate the effectiveness of adjuvant therapy. [2]In most of cases, the treatment of choice is radical surgical excision since it provides improved survival rates of over 80%. Chemotherapy, it seems does not have much impact on the survival rates of patients with osteosarcoma of jaws. This can be explained on the basis of the fact that metastasis was rare and late, and the local recurrence of the lesions was the leading cause of death. [2] Unni K K has reported a 40 % 5 year survival rate for jaw osteosarcoma compared to conventional osteosarcoma (20%). [13]

In the present case, the patient had a radical surgical excision (with free fibula reconstruction) of mandible with tumor free margins. The prognosis of jaw osteosarcoma is better than that of long bone. This could be due to the histologically better differentaiation of jaw osteosarcoma than that of long bone osteosarcoma. [12] The prognosis for patients with metastatic disease appears to be determined largely by the site, no of metastasis, as well as surgical respectability of metastatic disease. It appears more favourable for patients with unilateral than bilateral pulmonary metastasis, and for patients with fewer nodules rather than many nodules. Patients with multifocal osteosarcoma (>1 bone at diagnosis) have a poor prognosis. [14, 15]

## Conclusion

The patients of osteosarcoma usually report to the dental professional first, due to clinical presentation of pain and swelling. It is a challenging role, as the maxillofacial surgeons / plastic surgeons come across osteosarcoma cases which have reached advanced stages and the prognosis in such patients is very poor.

It can be concluded that misdiagnosis is very common in osteosarcoma of jaw. Osteosarcoma is an ancient disease, many aspects of which are still incompletely understood. There have been a plethora of discussion and

also controversies about the nature, aggressiveness, behaviour and various treatment modalities of this entity. However, for purpose of management, emphasis should be laid on the aggressiveness of lesion which warrants an early identification and diagnosis of lesion followed by prompt treatment.

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