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ABSTRACT

Introduction

Osteosarcoma is the most common primary malignant bone tumor excluding hematopoietic neoplasms. It is thought to arise from a primitive mesenchymal bone-forming cell and is characterized by production of osteoid. Despite osteosarcomas of the jaws being less aggressive than those occurring in the long bones, local recurrence after surgical therapy is a major complication.

Case Report

We report a case of recurrent osteosarcoma of maxilla, within three years after surgical resection presenting as a rapidly growing fungating mass in a 24 year old female patient.

Conclusion

Osteosarcoma affecting the maxilla is a relatively rare condition when compared to the long bones. Early diagnosis and treatment can improve the survival rate. Management of super added fungal infections associated with chemo-radio therapy can prevent further complications and improve the general health of the patient in recurrent lesions.

Keywords: Osteosarcoma of maxilla, local recurrence, secondary fungal infection.
TITLE: Recurrent osteosarcoma of maxilla presenting as fungating mass- a case report

INTRODUCTION

Osteosarcomas of the jaws are uncommon and represent 6% to 8% of all osteosarcomas [1]. It occurs most often in the third and fourth decades of life, with the mean age being 33 years. As in the long bones a slight male predominance is noted in gnathic osteosarcomas. The maxilla and mandible are involved with about equal frequency. Maxillary lesions are seen commonly involving the inferior portion (alveolar ridge, sinus floor, palate) than the superior aspects (zygoma, orbit) [2]. Although it is generally believed that osteosarcoma of the jaws is less aggressive than those of the long bones, current clinicopathologic studies suggest that osteosarcomas of the jaws are aggressive neoplasms. The ability to achieve initial complete removal of the lesion with clear surgical margins of greater than 5 mm is said to demonstrate a better survival, fewer local recurrences, and less metastasis [3]. Osteosarcomas arising from the maxilla are often more difficult to resect completely than mandibular lesions reflecting in a relatively high recurrence rate [4, 5]. Here we present a diagnostically challenging case of osteosarcoma with a unique clinical presentation.

CASE REPORT

A 24 year old female reported to the clinic with the chief complaint of a large swelling on the right side of face. She also complained of foul smell, inability to open mouth and multiple ulcers over the swelling. History revealed that the patient developed a right maxillary swelling 2 years back for which an intra-oral incisional biopsy was performed. The lesion was then given a diagnosis of osteosarcoma. The patient was treated with hemimaxillectomy and chemotherapy with Adriamycin 90 mg and Ifosfamide 3000 mg, with addition of Cisplatin 60 mg to the regimen on the second cycle. She received post-operative adjuvant radiotherapy of 60 Gy/30# by IMRT using 6 MV over a span of one month and nine days. Presently patient reported to the clinic with a swelling in the same region which rapidly increased in size. On examination, a large extra oral swelling involving the entire right side of face with
multiple superficial polypoid masses was seen. The overlying skin appeared dark and erythematous, with multiple large polypoid masses exhibiting superficial ulcerations. Intra-oral examination revealed huge mass involving the entire oral cavity with necrotic areas causing dysphonia and dysphagia [Figure 1].

Radiographically, AP view shows right hemimaxillectomy and a mixed radiolucent-radiopaque lesion with ill-defined borders [Figure 2]. CT scan revealed large infiltrating heterogeneous isodense mass with areas of cystic density and haemorrhagic foci. The lesion was causing erosion of the right side of the facial structures involving lateral wall of orbit, zygomatic arch, ethmoid sinus, hard palate. The lesion was extending to the right aspect of nose displacing nasal septum to left and was protruding into oral cavity. Right submental enlarged lymph node of size 1x10.9 cm was observed. Tc$^{99m}$-MDP bone scan revealed an abnormal increased radiotracer uptake in the right maxillary bone and rest of the skeletal system appeared normal.

An incisional biopsy was performed from the intra oral lesion and was sent for histopathological examination. Microscopic examination revealed numerous fungal hyphae and spores along with large areas of necrotic debri [Figure 3]. A repeat biopsy after antifungal therapy was performed. The histopathology revealed numerous ovoid to polygonal neoplastic cells showing nuclear pleomorphism with increased mitosis. Osteoid formation [Figure 4] and few tumor giant cells were noticed within the stroma and the lesion was diagnosed as high grade recurrent osteosarcoma [Figure 5].

The patient was put on chemotherapy with Doxyrubicin (60 mg), Ifosfamide (3000 mg), Methotrexate (12 mg) and Cisplatin (60 mg). The patient expired during course of the treatment.

**DISCUSSION**

Osteosarcomas are malignant connective tissue tumours originating from undifferentiated mesenchymal cells that are able to form bone or osteoid tissue. It can occur in any bone, the most common site being the long bones of the extremities near metaphyseal growth plate. Osteosarcomas of the craniofacial region are relatively rare. Although mandible is reported as the most common site of
involvement in the craniofacial region, equal frequencies of maxillary osteosarcomas have also been reported [6]. Mean age of onset of osteosarcomas of craniofacial region is in the third to fourth decade of life. There is usually a predilection for occurrence in males. However some report it to be more frequent in females or with an equal gender predilection [6].

Although it is generally believed that osteosarcoma of the jaws is less aggressive than those of the long bones, current clinicopathologic studies suggest that osteosarcomas of the jaws are aggressive neoplasms. Our case is an example of such an aggressive neoplasm in the maxilla. The multiple large polypoid masses seen on the surface of the lesion were unique to our case. These superficial masses were due to the secondary infection by opportunistic fungi. The patient’s compromised immune status due to the previous chemotherapy and radiotherapy could have contributed to the fungal infection, which in turn can cause serious morbidity and mortality. The radiographic appearance of osteosarcoma is variable ranging from osteolytic to mixed osteolytic-osteoblastic to predominantly osteoblastic lesions [7]. It depends on the amount of tumor bone synthesized by the malignant osteoblasts. Our case showed mixed radiolucent- radiopacity with ill-defined borders. The common histological types of osteosarcomas are chondroblastic, osteoblastic and fibroblastic. The typical histological criteria for the diagnosis of a high grade osteosarcoma include bizzarely arranged tumor cells, immature bone formation (osteoid), atypical fibroblasts and infiltration of adjacent soft tissues. In our case the histopathological examination revealed a high grade osteoblastic type of osteosarcoma.

Clear surgical margins play a role in the eradication of the disease and limitation of intramedullary extension. However, due to the complexity of the anatomical location and proximity to vital structures, osteosarcomas arising from the maxilla are often more difficult to resect with a sufficiently safe margin. This is reflected by a relatively high local recurrence rate which is a major complication after surgical therapy, as presented in this case. Study by Delgado et al suggested that when surgical margins are not free of disease, the use of radiation does not improve the outcome [8]. The recurrence in our case was noted within three years. In the study by Tabone et al, the median interval between the diagnosis of the primary osteosarcoma and the
first recurrence was found to be 21 months. They concluded that the most important
prognostic indicator at first recurrence seems to be the possible complete resection
of disease. Patients not amenable to surgery and patients with a second or a third
recurrence have a poor prognosis [9]. If surgical resection is not attempted or cannot
be performed, progression of the disease can be fatal [10]. The overall 5 year survival
rate of osteosarcoma patients is found to be 62%. The ability to achieve initial
complete removal of the lesion with clear surgical margins of greater than 5mm is
said to demonstrate a better survival, decreased recurrences and less metastasis
than those with margins less than 5mm [3].

CONCLUSION
Osteosarcoma affecting the maxilla is a relatively rare condition when compared to
those in the long bones. Anatomic limitations in the orofacial region cause difficulties
in achieving uninvolved margins and for this reason local recurrence of the lesion is
high. Superficial fungal infections can cause alterations in both clinical and
histopathological presentation. A regular follow up of the patients after maxillary
osteosarcoma resection is advised to rule out further recurrence.

CONFLICT OF INTEREST
The authors declare no potential conflict of interests.

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Proof reading and special stains

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Collection of data and assisting in manuscript preparation

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REFERENCES


FIGURE LEGENDS

Figure 1: Clinical picture showing a large extra-oral swelling with multiple superficial polyloid masses. Intra-orally the mass is seen to be filling the entire oral cavity.

Figure 2: Antero-posterior view showing right hemimaxillectomy and a mixed radiolucent-radiopaque lesion with ill-defined borders in the same side.

Figure 3: Histopathology (H &E stain, 40 x) Dense masses of necrotic debris, fungal hyphae and spores masking the underlying cellular area.

Figure 4: Histopathology (H &E stain, 40 x) proliferation of neoplastic cells with associated tumor osteoid formation.

Figure 5: Histopathology (H &E stain, 40 x) Numerous multinucleated tumour giant cells within the cellular connective tissue.
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