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Osteogenic sarcoma of the maxilla in an adolescent

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Abstract: Osteosarcoma of the maxilla is rare and occurs mainly in middle age. We present the case of 16 year old girl with osteosarcoma involving the left maxilla. She underwent maxillectomy, received chemotherapy and radiotherapy. She was in remission for 20 months when she relapsed locally.
Keywords: Osteosarcoma, maxilla, craniofacial.

INTRODUCTION

Osteosarcoma of the craniofacial region is rare and represents 6-10% of all osteosarcomas [1]. Within the craniofacial region the mandible is the most common site of the involvement, followed by the maxilla and skull [1, 2]. The average age at the onset of osteosarcoma of the maxillofacial region is about one or two decades later than that of osteosarcomas of other regions and the highest occurrence is found in the third to fourth decade of life. We present the case of a young lady with osteogenic sarcoma of the maxilla.

CASE REPORT

A 16 year old girl presented with swelling of the left maxillary region following root canal treatment. Examination showed a hard swelling on the hard palate (Figure 1).

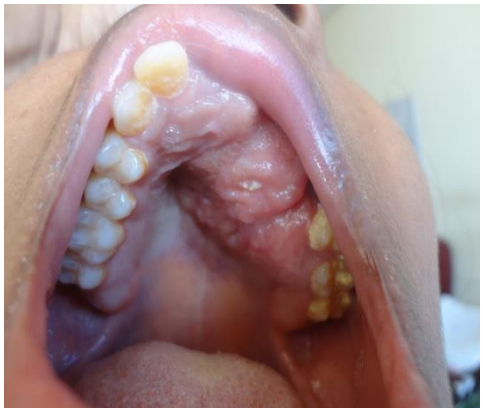

Fig 1: Photograph showing swelling on the hard palate

Computed tomogram showed a large destructive mass lesion in the left maxilla and alveolar process with a large soft tissue component and scattered foci of mineralization (Figure 2a&2b).

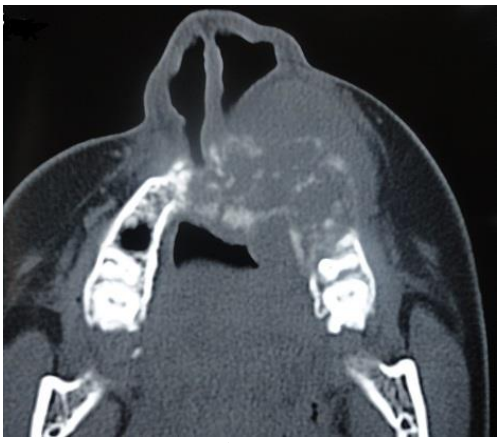

Fig-2A

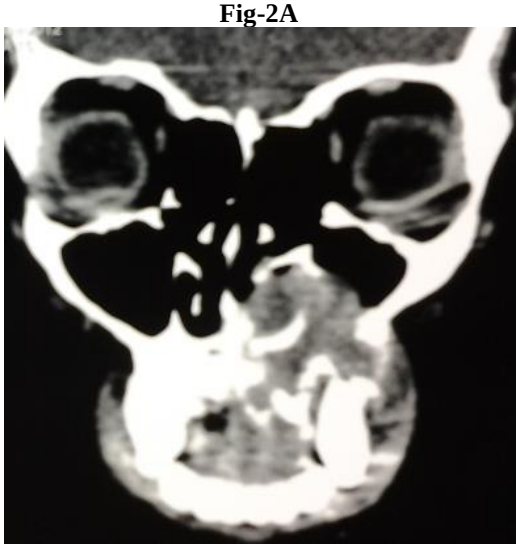

Fig: 2B

Fig 2 A & 2B: Computed tomogram showing a destructive lesion in the left maxilla and alveolar process with a large soft tissue component and scattered foci of mineralization

A Tc^{99} bone scan showed increased uptake over left maxilla. A biopsy from the mass was diagnostic of fibroblastic osteosarcoma. She received 2 cycles of neoadjuvant chemotherapy with cisplatinum, doxorubicin and ifosphamide followed by subtotal maxillectomy and adjuvant chemotherapy for 4 cycles. Since margins of resection were close, local radiation was given. She was in remission for 20 months when she relapsed locally. She refused further treatment and died 7 months later.

DISCUSSION

When compared to other locations, craniofacial osteosarcomas are less aggressive and distant metastasis is rare. The histologic types are chondroblastic (41%), osteoblastic (33%) and fibroblastic (26%) [1]. The major risk factors for the development of osteosarcoma of the jaws are similar to those for long bones namely, previous irradiation to face, fibrous dysplasia and Paget's disease.

The 'sunburst' appearance, the classic radiologic appearance of long-bone osteosarcomas, is rarely seen in jaw osteosarcomas. The radiographic appearance of osteosarcoma of the jaws has been described by Bianchi et al. as radiolucent with absence of bone formation or mottled appearance with areas of amorphous ossification and as lamellar ossification [3]. In a review on gnathic osteosarcomas, out of 33 cases, 13 cases were from maxilla [4].

Complete resection of the primary lesion is required, and hence a total maxillectomy is recommended. Patients with > 5 mm negative margins have a better survival than those with < 5mm margins [5]. Maxillary osteosarcomas do worse compared to mandibular in view of the difficulty to obtain negative margins [6]. The role of adjuvant chemotherapy is ill-defined. While adjuvant chemotherapy may improve survival, neither chemotherapy nor radiotherapy appears to impact long-term survival. Because positive margins are associated with poor prognosis, multimodal therapy significantly reduce rate of local recurrence, thereby improving survival [7, 8, 9]. Adjuvant radiotherapy should be considered for those with close or positive margins. The vast majority of recurrences are observed within 5 years. The 5-year disease-specific and overall survival rates are approximately 60% to 70% [10].

CONCLUSION

Maxilla is a rare site for osteosarcoma in children and adolescents in whom extremity osteosarcomas are common. Radiologically also, they do not have the classical appearance of extremity osteosarcomas. They require multimodality treatment.

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