Aggressive Osteosarcoma of the Mandible in a 13-Year-Old Girl

A 13-year-old girl presented with painful swelling on the lower jaw for 9 months [Figure 1]. The swelling was neglected and treated symptomatically only. Clinical examination revealed a proliferative mass from the lower jaw completely filling the oral cavity. Computed tomography scan showed destruction of the middle-third of the mandible and an associated large mass with dislodgement of the incisors [Figure 2]. Biopsy confirmed the lesion as high-grade osteosarcoma. Positron emission tomography scan showed active localised disease only. Subsequent to neoadjuvant chemotherapy based on the institutional protocol for osteosarcoma (ifosfamide, cisplatin and doxorubicin), the patient had stable disease. A middle-third mandibulectomy was performed, and the defect was reconstructed with free vascularised fibular osteocutaneous flap [Figure 3]. The post-operative histopathology confirmed an osteosarcoma with all the bony and soft tissue margins negative for the presence of tumour. The patient received adjuvant chemotherapy and radiotherapy (70 Gy). However, there was local recurrence even before the completion of chemotherapy. In view of the aggressive disease, only the best supportive care was offered. The patient and her family palliative and supportive care needs were evaluated by the institutional multidisciplinary team of palliative care specialist, psychologists and social workers. The physical issues of pain and anxiety were addressed with drugs, while the psychological and social issues involved counselling, honest communication and support groups.

The patient died two months after discharge from the hospital.

Osteosarcoma is the most common primary malignant bone tumour in children and is histologically characterised by the production of malignant osteoid. They arise from primitive mesenchymal elements and affect the long bones. Although the mandible comprises only 0.86% of total skeletal volume, osteosarcoma of the mandible constitutes 5%–13% of all osteosarcoma.[1] Due to the rarity of this tumour, the incidence in children is not known, and most studies have clubbed paediatric and adult patients together. Jaw osteosarcoma affects the mandible and the maxilla with almost equal incidence in the two bones and without any gender predominance. In the mandible, the tumour more commonly involves the body. The most common presenting symptoms are swelling with or without pain, loosening of teeth and ulceration. The patients commonly suffer for 3–4 months before seeking treatment, and the mean size of the tumour at presentation has been reported to be 4.5–8 cm. In view of the aggressive nature of the disease, most patients merit a multidisciplinary approach. Radical surgery to achieve wide negative margins with functional reconstruction is the historical mainstay. The addition of chemotherapy in the adjuvant or the neo-adjuvant setting has been shown to improve disease-free survival, especially osteosarcoma of the mandible and in the presence of positive resection margins.[2,3] Although thought to be radioresistant, there is evidence to support the use of 60–66 Gy of radiation in cases undergoing incomplete resection or resection with close margins.[4] Due to the rarity of osteosarcoma of the mandible in children, data regarding a biological difference in the nature of the disease in terms of aggressiveness and response to therapy compared to adults are deficient. Nonetheless, delayed presentation and the aggressive nature of the disease may be responsible for an early relapse while on chemotherapy and fatal outcome in our patient.



Figure 1: Clinical photograph showing a large proliferative lesion arising from the lower jaw filling the oral cavity

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

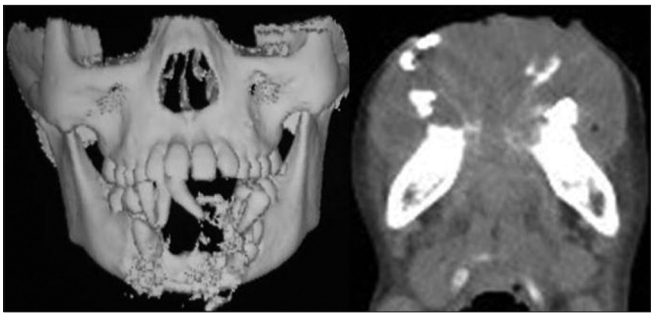


Figure 2: Computed tomogram images showing destruction of middle-third of the mandible with associated soft tissue mass and uprooted tooth within it

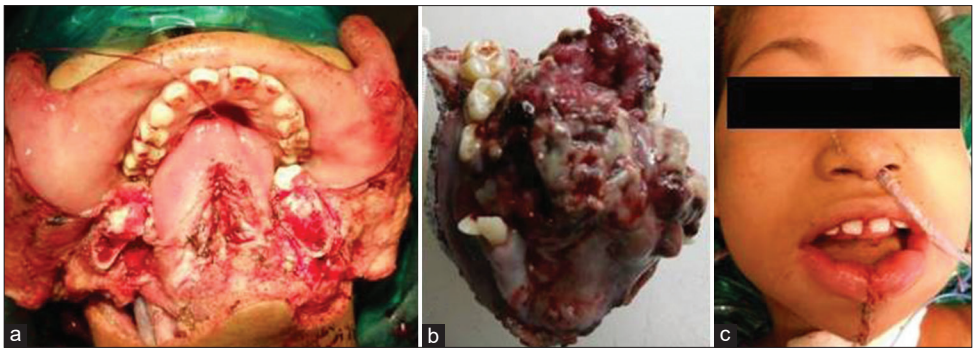


Figure 3: Intraoperative photograph showing the surgical defect (a) and the resected specimen (b) after middle-third mandibulectomy. Reconstruction with free fibular osteocutaneous flap for the surgical defect was performed (c)

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Conflicts of interest

There are no conflicts of interest.

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