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CLINICAL STUDY

Maxillofacial osteosarcoma successfully treated with surgery and adjuvant chemotherapy in a child

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Abstract: Maxillofacial osteosarcoma constitutes a minor percentage of all the head and neck tumors. We describe a 10 year-old girl presenting with swelling and pain in left maxillary region and diagnosed as low grade osteosarcoma. The patient was operated and given a chemotherapy protocol consisted of Cisplatin and Doxorubicin. After six courses of chemotherapy the patient was in complete remission and she is well with no evidence of disease for five years. Since high local recurrence rates have been reported in craniofacial osteoarcoma and we know the deleterious side effects of radiation therapy in children, we believe that best management strategy for osteosarcomas in maxillofacial region in children is radical surgical excision and postoperative chemotherapy *(Fig. 3, Ref. 11)*. Full Text in PDF *www.elis.sk.*
Key words: maxillofacial osteosarcoma, treatment, children.

Osteosarcoma is a primary malignant tumor of bone, deriving from primitive bone-forming mesenchyme and constitutes the most common malignant bone tumor of the childhood. Although primary bone tumors are rare in childhood they are sixth most common group of malignant neoplasms in children. The tumor occurs most frequently in the long bones, especially around the knee (1). Maxillofacial osteosarcoma constitutes a minor percentage of all the head and neck tumors (2). We describe a child with osteosarcoma located in maxillofacial area with a review of treatment modalities.

Case

A 10 year-old girl was admitted to another hospital for pain and swelling in left part of the upper jaw. Despite aspiration of the swelling and antibiotic therapy the complaints of the patient continued. After biopsy of the left palatinal region pathologic examination had been reported as osteoblastoma. The parents brought the child to our hospital for treatment.

On physical examination there was a swelling in her left maxillary region. The computed tomography (CT) of the paranasal sinuses (Fig. 1) revealed an ill-defined and expansive mass at the left maxillary alveolar process, extending to the palate and to the maxillary sinus. The mass had a diameter of 3.5 cm, had chondrogenic matrix and was causing bone destruction The CT of the chest was normal. Bone scintigraphy with 99mTc-MDP demonstrated no metastasis.

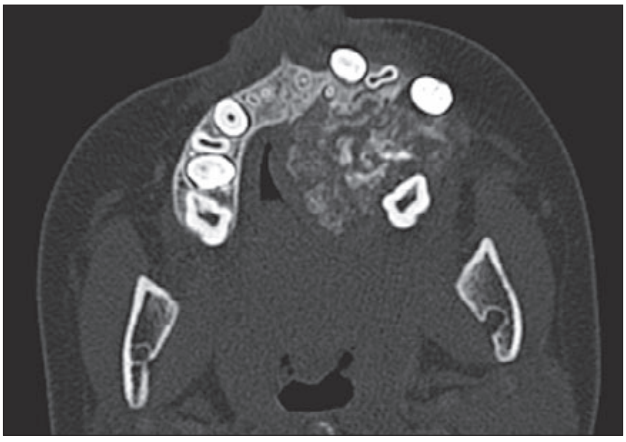

Fig. 1. CT of the paranasal sinuses demonstrating an expansive, ill-defined mass centered to the left maxillary alveolar process, destructing the bone.

Maxillar biopsy taken by otolaryngology department revealed mesenchymal neoplasia. Subsequent left inferior maxillectomy revealed low grade osteosarcoma and tumoral cells in surgical margin (Fig. 2). During the surgery a mass of approximately 3x4 cm was seen in hard palate, at the left of the midline. Hard palate was extracted with inferior nasal concha and floor of the nose. An obturator prosthesis was inserted to the palatal defect.

In the patient a chemotherapy protocol was started consisting of Cisplatin (120 mg/ m^2) on day 1, and Doxorubicin (30 mg/ m^2) on days 1–3 with three weeks interval. After six courses of

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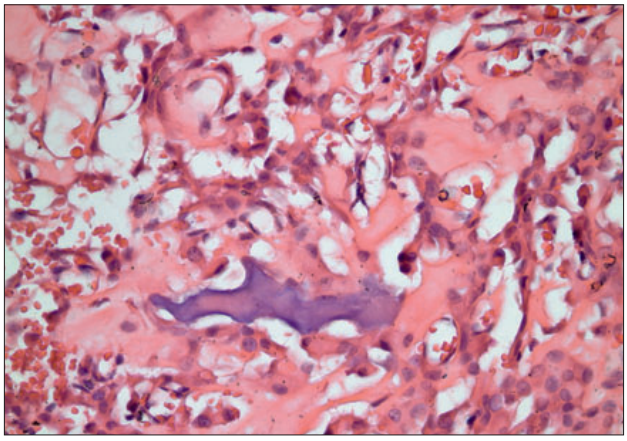

Fig. 2. Malignant mesenchymal neoplasm with osteoid production. Osteoid is seen as eosinophilic lace-like material mixed with malignant mesenchymal cells (hematoxylin&eozin, x400).

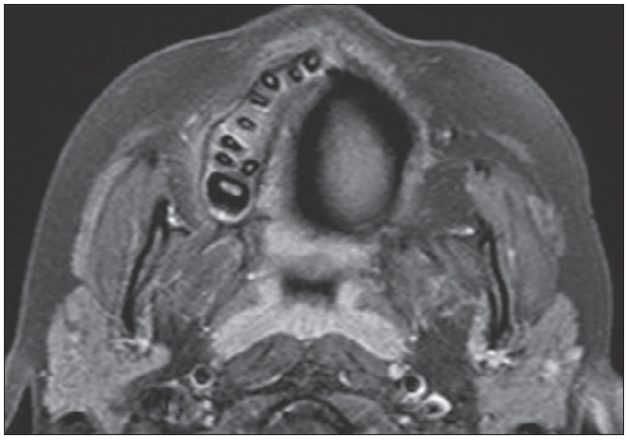

Fig. 3. Axial post-contrast and fat-supressed T1 weighted image of the follow-up study, performed upon completion of the chemotherapy reveals post-operative changes of left inferior maxillectomy. There is no evidence of residual or recurrent tumor.

chemotherapy the physical examination and MRI investigation of the patient showed no signs of residual or recurrent tumor (Fig. 3). Bilateral ototoxicity developed secondary to cisplatin administration. Except ototoxicity we did not encounter any side effects related to chemotherapy. The therapy was ceased in September 2006. She is in complete remission and well with no evidence of disease for five years.

Discussion

Osteosarcoma of the face is one of the most rare tumors encountered in pediatric oncology practice. Additionally, primary osteosarcomas of the head and neck in children not associated with retinoblastoma or previous irradiation are extremely rare. Previously in a review of the literature Gadwall et al (2) reported that maxillofacial osteosarcomas accounted for 4–9 % of all osteosarcomas. Most of the patients are associated with retinoblastoma or previous irradiation of this region. The rarity of these tumors makes it impossible to conduct prospective studies on behaviour of the tumor and to compare the effectivity of treatment modalities. Instead, the information about the issue in literature exclusively stands on retrospective series and sporadic case reports.

Paget disease of bone, fibrous dysplasia and genetic predilection are also among the other etiologic factors proposed (3). There was no significant finding in our patient's medical and family history in this manner.

The majority of patients with osteosarcoma present with pain over the involved area with an associated soft tissue mass. Cheek swelling, facial pain, mass in the hard palate and previous dental extraction are among the most frequent presenting features in patients with maxillofacial osteosarcoma. Patients may also present with the complaints of epistaxis, trismus, nasal obstruction, proptosis or reduction in vision (4). Our patient was admitted to us with the complaints of swelling and pain in left maxillary region.

Osteosarcoma of the facial region is also a malignant tumor that is difficult to manage. The mainstay of the therapy is wide surgical resection with negative margins. It was shown that radical surgery in young patients with a craniofacial osteosarcoma gives long-term disease-free survival (5). Since head and neck region is not always accessible for wide resection, patients with tumor in these areas had significantly decreased survival. Therefore adjuvant therapy was strongly suggested for osteosarcomas of the head and neck (6). Radiotherapy is suggested only to prevent local recurrence if surgery is not complete. Adjuvant chemotherapy can be employed after radical surgical removal. The most frequently used chemotherapy protocol is a combination of doxorubicin with either cisplatin or cyclophosphamide and vincristine (6) or high dose methotrexate (7). In a systematic review of 201 patients with craniofacial osteosarcoma Smeele et al (8) have reported improved survival with complete resection and adjuvant chemotherapy. Early radical resection was also recommended as the primary treatment with the aim of local control in two recent series of patients with head and neck sarcomas especially located in the maxillofacial region (9, 10).

High local recurrence rates have been reported in craniofacial osteosarcoma. Risk factors for local recurrence in a series including children and adults have been reported as younger age, tumor size >4 cm, previous diagnosis of retinoblastoma and positive surgical margins (11). Kassir et al (3) have shown that patients treated with surgery tended to have better prognosis compared to patients treated with surgery and adjuvant chemotherapy and/or radiation therapy. However, this was probably due to reserving adjuvant therapy for more advanced tumors.

The adjuvant chemotherapy selected in this patient because the patient had positive surgical margin. It is the most appropriate option to manage such patients with adjuvant therapy after surgical resection, to prevent local recurrence. Our patient had moderate risk of recurrence since she had 2 of the 4 mentioned risk factors namely positive surgical margin and tumor size of 3.5 cm. The patient is in complete remission and there is no recurrent disease with MRI evaluation 5 months after diagnosis. We believe that adjuvant chemotherapy following radical surgical excision is the

best practice for osteosarcomas in maxillofacial region, taking into account high local recurrence rates and long term side effects of radiotherapy in children.

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