

CASE REPORT**Childhood Facial Osteo Sarcoma: a Case Report**

Hamid Farhangi, MD; Mahdi Farzadnia, MD; Ali Alamdaran, MD

*Research performed at Dr. Sheikh Pediatric Hospital, Mashhad University of Medical Sciences, Mashhad, Iran**Received: 25 November 2014**Accepted: 28 February 2015***Abstract**

Osteosarcoma (OS) is the eighth common cancer of childhood and its incidence is 4 cases in one million in children younger than 14. Facial OS incidence is estimated between 8 and 10% of OS cases. The main etiology of OS is unknown, but various predisposing factors are proposed such as radiation, radiotherapy, some benign bone diseases like Paget's disease or fibrous dysplasia. There is a 5-year survival rate of 68% and it decreases with the increase of age. Positive history of radiotherapy is the main predisposing factor for childhood OS. There is some evidence about the X-ray induced mutation in genomic DNA that leads to osteosarcoma. In the present paper we present a 19-month old girl with a mass located in the inferior margin of the left cheek and orbit. Our case is unique with regard to her young age and sex. Moreover, the tumor was located in an uncommon site and her disease was progressive and resistant.

Key words: Bone tumor, Childhood, Facial bone, Osteosarcoma

Introduction

Osteosarcoma (OS) is one the most frequent malignant tumors that is derived from mesenchymal cells with bone formation capacity (1). This tumor is more frequent in the black race (2). Male to female ratio is estimated at 5.4:4 in some reports (3). But other studies suggest that OS is more common in females (4). Osteogenic sarcoma prevalence is 1-3 in one million, annually (1). The disease has two peaks of incidence: first in children between 10 and 14 years and second in older ages. OS is the tumor of long bones, and occurs in the metaphysis of the long bone near the epiphyseal plate. Distal femur and proximal tibia are the main sites (5). The 5-year relative survival rate after diagnosis of OS is estimated about 68% and it decreases in older ages (6, 7). OS is the eighth common form of cancer in childhood (2). This malignant tumor is more frequent in boys, and tall children are more at risk for developing this malignancy. Positive history of radiotherapy is the main predisposing factor for childhood OS. There is some evidence about the X-ray induced mutation in genomic DNA that leads to osteosarcoma (8). We present a baby with facial OS.

Case Report

A 19-month old girl was referred to our tertiary academic hospital with a facial unilateral mass and swelling. The mass was located in the inferior margin

of the left cheek and orbit. Clinical features indicated rhabdomyosarcoma.

She had a history of upper respiratory infection 20 days ago that lasted for two days. She was initially treated for orbital cellulitis with the antibiotic co-amoxiclav. She was referred to our pediatric center after no observed changes in clinical presentation. She was the second child of non-relative parents, with an uneventful pregnancy and vaginal delivery. Her growth status was normal and there was no history of malignancy or exposure to ionizing radiation in the family. Moreover, the parents' first child was healthy.

Head and neck computed tomography scan (CT scan) was performed; plain X-ray study (water radiography) was suggestive for soft tissue sarcoma with invasion of the bone. The chest X-ray, chest CT scan and bone scan showed no metastasis. Abdominopelvic ultrasound study was also normal.

There was a painless mass with a 3*4 cm dimension lesion in the inferior orbital rim and was fixed to the zygoma. The globe had inward to upward deviation. Ophthalmic examinations were normal, including those of the optic nerve, retina and anterior chamber. Chemosis was observed in the lower eyelid conjunctiva.

Laboratory tests were normal except for lactate dehydrogenase (LDH) level that was 850 (normal range: 140-280 UI/L) and alkaline phosphatase (ALP) was 590 (normal range in children < 350 U/L).

Corresponding Author: Ali Alamdaran, Department of Radiology, Dr Sheikh Pediatric Hospital, Mashhad University of Medical Sciences, Mashhad, Iran.
Email: AlamdaranA@mums.ac.ir



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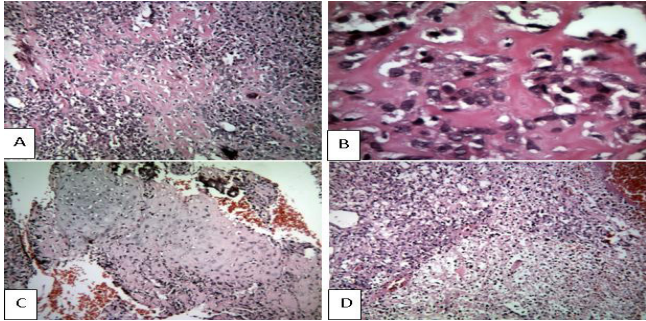


Figure 1. Highly pleomorphic cells and haphazard deposits of osteoid (A & B), a few foci of neoplastic cartilage (C) and necrosis (D).

A biopsy was performed of the lower eyelid conjunctiva and histopathological assessment was suggestive for osteosarcoma. In the histopathological evaluation the hematoxylin and eosin (H&E) stained section revealed proliferation of highly pleomorphic spindle cells exhibiting considerable variation in the size and shape. They showed large hyperchromatic nuclei with a high nuclear to the cytoplasmic ratio and high mitotic activity arranged in a disorderly fashion with neoplastic bone formation including thin irregular trabeculae and osteoid. Also, a few foci of neoplastic cartilage and necrosis were seen [Figure 1].

Computed tomography scan of the orbital and paranasal sinuses indicated unilateral bulging due to a soft tissue mass with zygomatic bone origin. The compression effect was noted on the globe because of the intraorbital component. In the bone window, a mixed lytic and sclerotic lesion was seen in the zygomatic bone with sunburst periosteal reaction [Figure 2]. Metastasis work-up revealed no other site of invasion.

Standard chemotherapy (EURAMOS Clinical Trial) was started for this child, and despite initial regression, after two chemotherapy sessions, she did not show a good response. The child was readmitted due to tumor progression after 14 days. Due to tumor expansion and high risk progressive osteosarcoma, removal surgery was impossible. Treatment protocol was changed to high risk OS (MAPIE), but this shift did not stop the disease from progressing (9). Tumor remained inoperable after neoadjuvant chemotherapy. The chemotherapy course was continued for 10 sessions to control local growth. Palliative radiotherapy was administered, but response to treatment was tragic and she died after one year.

Discussion

Our case was unique with regard to sex and its presentation in a very young age. Moreover, the disease was progressive and resistant with a rare location.

Although osteosarcoma is one of the most frequent bone malignancies, head and neck OS is very rare. Facial OS incidence is estimated between 8 and 10% of OS cases (10). Maxilla and mandible is the main site of facial OS. In various articles mean age of patients with jaw OS was reported at about 40 (10).

The main manifestations of OS are pain, swelling and

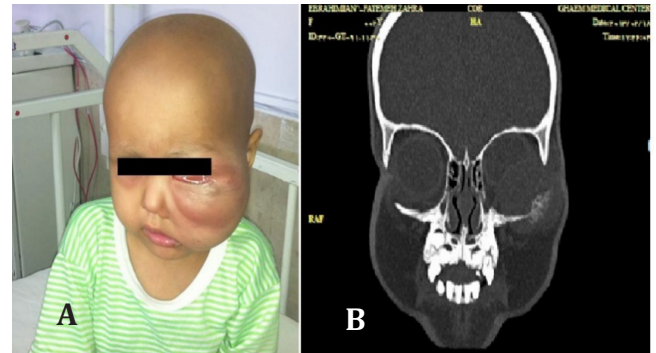


Figure 2. (a) Clinical and (b) radiologic features.

ulceration (11). In our patient OS was presented with swelling without pain, ulceration was not observed during therapy, the mass was located in the zygoma, and the tumor did not invade the mandible or palate. This painless condition might have occurred because the tumor did not invade the optic or facial nerve.

Osteosarcoma radiologic findings include erosion and sunray spicules with a calcific center (5). Our patient's CT scan showed mixed lytic and a sclerotic lesion in the zygomatic bone with soft tissue mass and a sunburst and all of this were suggestive of OS.

Jaw OS has a better survival rate in comparison with other OS diseases, and has less metastasis and lymph node involvement (12). In the introduced case there was no evidence of metastasis or invasion. But she died one year after disease diagnosis due to resistance and having a progressive form of OS.

The main etiology of OS is unknown, but various predisposing factors are proposed such as mutation in the P53 or retinoblastoma gene, expression of PCNA and Ki-67 gene, radiation or radiotherapy, some benign bone diseases like Paget's disease or fibrous dysplasia (13). Our patient had no predisposing factor during the gestational period and after birth and her family history was negative for malignant diseases. Although, noting patients past medical history is very important in malignancy, there are cases without risk factors.

Differential diagnoses of OS include chondrosarcoma, rhabdomyosarcoma, Ewing sarcoma, bone metastasis, and osteomyelitis (13). Our patient was initially diagnosed as cellulitis due to her past history of an upper respiratory tract infection. After further investigations, her primary radiologic study was suggestive for rhabdomyosarcoma. Finally, osteosarcoma was confirmed with a CT scan and histopathological findings.

Microscopic examination showed cellular stroma contained osteoid and chondroid, all of which confirmed the diagnosis of OS. Several studies revealed that there is no relation between histopathological classification and patients' survival. Also, no relation was found between immunohistochemical markers and prognosis (10). Facial OS is a difficult tumor to manage, and surgical resection is the main treatment option in OS. However, surgery leads to impairments of socio-occupational outcomes

and other long term complications in OS patients. With the invention of adjuvant and neoadjuvant therapies in 1980, OS treatment progressed, and some studies have shown that chemotherapy might improve patients' outcome (14). The main cause of treatment failure in OS patients is local recurrence that happens in 1.1 to 40% of cases (15). In our case, the tumor progressed after two session of chemotherapy. Surgery was impossible in this patient due to tumor expansion; so, she received chemotherapy and palliative radiotherapy.

In conclusion, although facial OS is very rare in childhood, it should be considered in patients who have a fixed swelling mass and radiologic studies are the first step for diagnosis.

Hamid Farhangi MD
Department of Pediatric Hematology-Oncology, Dr Sheikh Pediatric Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

Mahdi Farzadnia
Department of Pathology, School of Medicine, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

Ali Alamdaran
Department of Radiology, Dr Sheikh Pediatric Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

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