Chondroblastic Osteosarcoma in A 13 Year Old Child

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ABSTRACT:

Primary neoplasms of human skeleton are rare, accounting for 0.2% of overall human tumor burden. Osteosarcoma is a malignant mesenchymal tumor, predominantly occurring in long bones and occasionally in the maxillofacial region. Osteosarcoma accounts for 15-35% of all primary bone tumors and represent 4-8% of osteosarcomas of jaw. Here a case of chondroblastic variant of osteosarcoma in a 13 year old child on the left side of palate is reported.

Key words: Mesenchymal tumor, woven bone, osteosarcoma, long bones.

INTRODUCTION:

Osteosarcoma (OS) is an mesenchymal malignant bone tumor characterized by formation of disorganized immature woven bone or osteoid tissue with a reported incidence of 1:100,000 per year.[1,2] Osteosarcomas account for 15-35% of all primary bone tumors and constitute 4-8% in the jaw. It is the most common non-hemopoietic primary malignant bone tumor of children and adolescence.[3] Osteosarcomas of jaw are diagnosed two decades and later than sarcomas of long bone with a peak incidence between 20-40 years. Osteosarcomas of the head and neck originate from soft tissues and have lower metastatic rate than osteosarcomas of long bones.[2] OS of jaw occurs mostly in males with a ratio of 1.5:1 and peak incidence is observed in 3- 4th decade.[3] Common site of occurrence is body of mandible followed by alveolar ridge of maxilla and maxillary sinus.[4] Typically the patient presents clinically as a swelling, pain, paresthesia and loosened teeth.[5] Computed tomography helps in better diagnosis than conventional radiography.[4] Osteosarcomas should be considered in differential diagnosis of expansile lesions of jaws. Early diagnosis and adequate surgical resection are the keys to high survival rates.[4]

CASE REPORT:

A 13-year-old male patient presented with a chief complaint of pain and swelling on the left side of the face with a corresponding swelling inside the oral cavity since one month. History revealed that swelling started one month ago on the left side of the palate spontaneously and gradually increased in size and associated with pain. There was no significant medical, dental or family history.

On inspection, a single diffuse extra-oral swelling is seen on the middle third of the face on left side (figure 1). The swelling was round to oval in shape measuring about 3x4cm.The swelling extended superiorly up to 2cm below the lower eyelid and inferiorly 2cm lateral to the corner of the mouth. It extended anteriorly up to the alae of nose, posteriorly 4cm upto infront of the ear. The surface of swelling was smooth. There was no evidence of ulceration or sinus formation.

On intra-oral examination, swelling was found on the left side of the palate crossing the midline. Swelling was noted extending anteriorly from palatal rugae and posteriorly upto the maxillary tuberosity of size 5x6 cm (figure 2) and laterally obliterating the buccal vestibule in relation to 25,26,27 region. The surface over swelling was normal and swelling was tender on palpation. The conventional radiograph revealed a typical sun-burst appearance (figure 3). There was widening of the periodontal space around the involved maxillary teeth, suggestive of tumor infiltration into PDL space. Provisional diagnosis was given as central giant cell granuloma.

Incisional biopsy was done and the tissue was sent for histopathological examination (figure 4). The Hand E stained decalcified section showed areas of woven bone with irregularly arranged deeply stained osteoblasts. Sheets of tumor osteoid with malignant chondroid tissue showing cellular pleomorphism (figure 4 and 5) and mitotic figures were evident. Areas showing primitive mesenchymal tissue were also evident in the sections. Based on histopathological findings, a diagnosis of chondroblastic osteosarcoma was given. After diagnosing the lesion as chondroblastic osteosarcoma, the case was referred to higher oncology center for further evaluation and treatment.

DISCUSSION:

Osteosarcoma(OS) is a true malignant neoplasm of bone, which may occur as a central, juxtacortical or peripheral lesion.[6] OS is characterized by production of osteoid tissue by mesenchymal tumor cells.[3] It is the second most common malignant bone tumor after multiple myeloma.[3] OS accounts for 15-35% of all primary bone tumors followed by chondrosarcoma and Ewing's sarcoma.[1] OS of jaw bones are rare and account for 5-6% of all osteosarcomas with an estimated incidence of 0.07 cases per 100,000 per year.[5]

OS are generally classified into two types; primary and secondary.[6] Etiology of primary OS is unknown and considered as may be due to genetic influence or environmental factors. Secondary OS occurs in older patients and predisposing factors include paget's disease, fibrous dysplasia of bone, trauma, chronic osteomyelities, bone infracts and late sequelae to craniofacial irradiation.[1,7] It also has been associated with metallic implants, joint prosthesis and in some genetic syndromes, Li - Fraumeni syndrome, hereditary retinoblastoma and Rothmund- Thomson syndrome.[1] Genetic mutations in tumor suppressor gene P53 and mutated retinoblastoma gene also found to be an etiological factor for the development of OS. This suggests an association between the occurrence of this neoplasm and increased cellular activity.[8] OS occurs more commonly in males with peak incidence of 3-4 decade. Mandible has been reported to be a more likely location than the maxilla in many extensive reports.[9] Primary OS represents a heterogenous group of malignant bone tumor characterized by diverse histological features, clinical, and biological behavior.[10]

OS of the jaws differ from long OS of long bones in several aspects: A) seen mostly in 3-4 decade (10-15 years later than mean age of occurrence of OS of long bones). B) No bimodal distribution. C) More in Males (In bones, equal predilection). D) Swelling is major complaint in OS of jaw where as pain in OS of long bones. E) Better prognosis. F) Less metastatic spread (6-51%) in jaws and metastasis in long bones(78-90%).[11]

Nearly 70% of OS display a multitude of cytogenetic abnormalities including haploidy in chromosomes 1p11-p13,1q11-q12,1q21-q22,11p14-p15,14p11-p13,15p11-p13,17p, and 19q13; gain of chromosome 1; loss of chromosome 9,10,13 and 17; and amplification of chromosomes 6p12-p21,17p11 and 12q13-q14.[12]

Variants of OS recognized by WHO differ in location,clinical behavior and degree of cellular atypia in that majority are intramadullary (conventional) OS and remaining are surface OS which are further subdivided into paraosteal, periosteal and high grade surface OS.[13.14]

Osteosarcoma being a rare presentation in the oral cavity can easily be misdiagnosed.[4] Clinically present with pain, swelling and tooth mobility. Radiographic findings vary from sclerotic to mixed

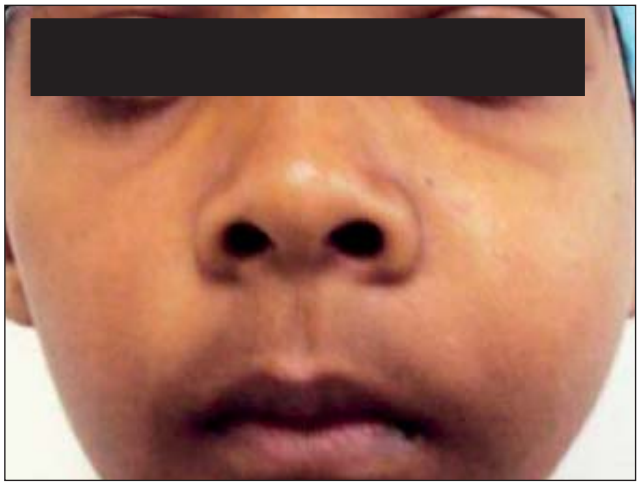

Figure 1: A single diffuse extra-oral swelling is seen on the middle third of the face on left side

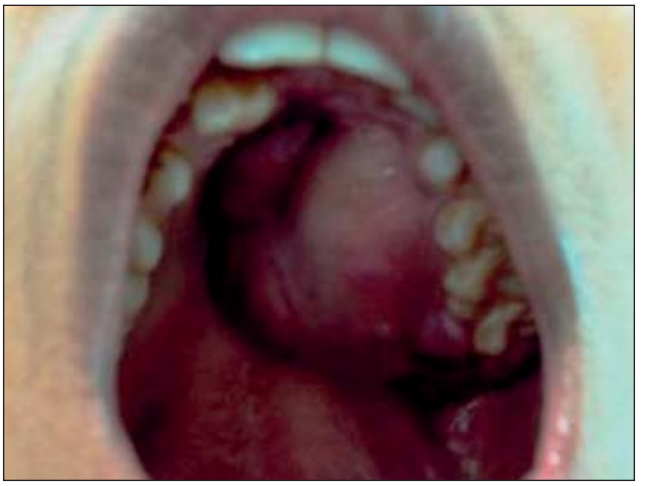

Figure 2: Swelling noted on the left side of palate extending from mid palatal raphae to the maxillary tuberosity.

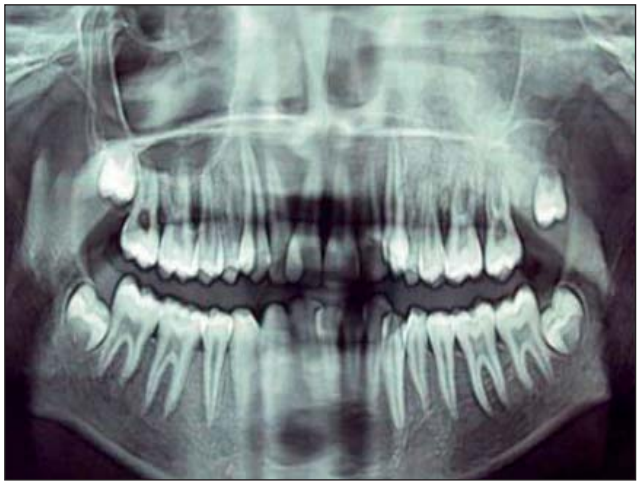

Figure 3: OPG Showing a typical sun-burst appearance

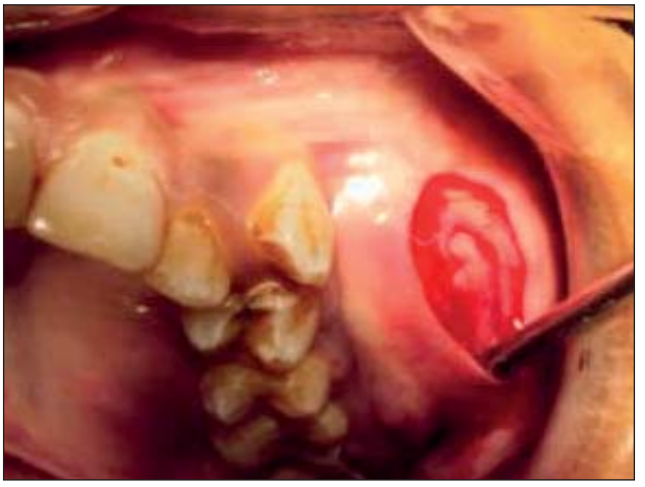

Figure 4: Surgical region where incisional biopsy was carried out.

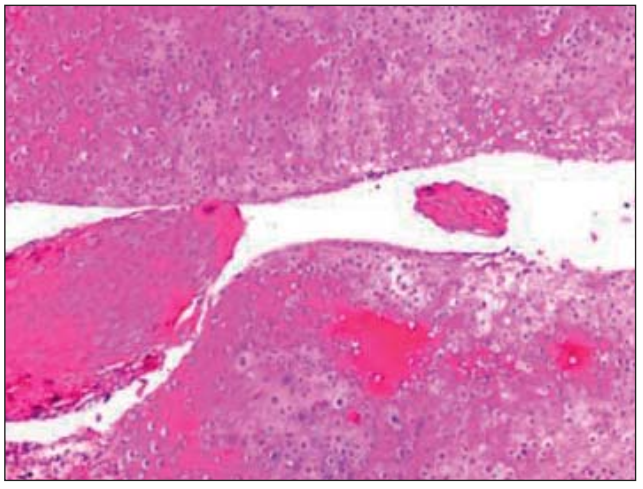

Figure 5: H&E section showing areas of woven bone with irregularly arranged deeply stained osteoblasts

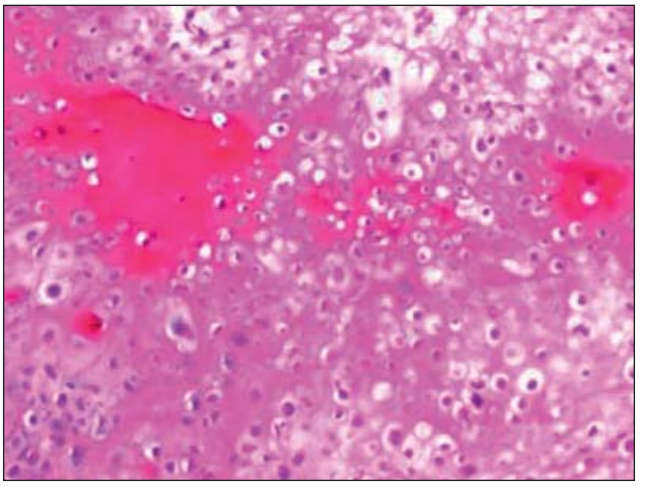

Figure 6: H & E section showing sheets of tumor osteoid with malignant chondroid tissue showing cellular pleomorphism (X10).

sclerotic to radiolucent. "classic" sunray or sunburst appearance due to osteophytic bone production best demonstrated in occlusal radiograph and CT Scan.[15] Panoramic radiograph may show Garrington's sign - widening of periodontal space around affected teeth with tapered resorption of tooth roots due to infiltration of tumor.[16]

Histologically OS of jaws are classified into osteoblastic, chondroblastic and fibroblastic depending upon the amount of the osteoid, cartilage or collagen fibres produced by the tumor.[17]

Staging of the tumor incorporates the degree of differentiation, distant metastasis and to estimate the prognosis of the patient.[18] The system used most commonly for bone sarcomas is Enneking System which is based on grade (G) of the tumor, local extent of the primary tumor (T), and metastasis (M).[6]

Immunhistochemistry (IHC) helps in differentiating chondrosarcoma from chondroblastic osteosarcoma as it is positive for vimentin, EMA, S100 and rarely cytokeratin where as chondrosarcoma is positive for vimentin and S100 only.[19]

Most common site for metastasis is lung which accounts for 20%. Risk of distant metastasis with OS of the head and neck varies, but death usually results from extension into the skull.

OS can be treated by radical or conservative surgery complemented by radiotherapy and/or chemotherapy. Overall prognosis is 25-50% with 5 year survival rate.[17]

CONCLUSION:

Osteosarcoma is an ancient disease in which many aspects still not understood. Excluding hematopoietic neoplasms, osteosarcoma is the most common type of malignancy which originates within bone and produce osteoid or immature bone. OS should always be considered in the differential diagnosis of expansile lesions of the jaws. Only early detection and proper treatment is the key to reduce morbidity and significantly improves prognosis. For proper management, emphasis should be laid on the aggressiveness of the lesion for early identification and diagnosis followed by prompt treatment.

Identifying the genes and signal transduction pathways through more molecular research may help in the development of newer diagnostic markers and help in better prognosis and patient survival in the future.

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