



Case Report

Osteosarcoma of orbital bone-chondroblastic morphology mimics to chondrosarcoma-A rare case report & review of literature.

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Abstract

Conventional osteosarcoma, a subgroup of intramedullary osteosarcoma (OS) is the most common osteosarcoma that occurs in adolescents and early adulthood. We present a case of 22-year-old male patient with the chief complaint of proptosis, pain and swelling in the left orbital region & diagnosed as chondroblastic osteosarcoma (COS) of left orbital bone. COS is very rare at orbital location, this report aimed to discuss clinical, radiographic, histopathologic, IHC findings and diagnostic pitfalls & management of COS in view of the literature.

Keywords: osteosarcoma, orbit, chondroblastic osteosarcoma

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INTRODUCTION

Osteosarcoma (OS) is an intramedullary high-grade sarcoma in which the tumour cells produce bone.[1]. The age distribution of osteosarcoma is bimodal, with the first peak occurring in adolescence and the second peak occurring in later adulthood. It usually appears in the metaphyses of the long bones of the proximal tibia, humerus, and distal femur. It has a preference for long bones. [2,3] When the underlying bone is normal, the tumor is primary; when the bone is damaged due to radiation therapy in the past, Paget's disease, an infarction, or any other condition, it is secondary OS.[1] The annual incidence rate of OS is about 4.4 cases per 1 million population for people aged 0-24 years. Males are affected more frequently (M:F ratio: 1.3:1).[1]

It is a relatively rarer entity in the craniofacial region consisting of about 6.5-7% of all OS.[2] We present a rare case in a 22-year-old male patient diagnosed as chondroblastic osteosarcoma (COS) of the orbital bone. An extensive search of literature has revealed, no case of COS of orbital bone. This article reviews the pertinent literature and describes our experience with a very unusual instance of chondroblastic osteosarcoma of the left orbital bone in a male patient who is 22 years old.

CASE PRESENTATION

A 22-year-old male patient presented with the chief complaint of pain and swelling in the left orbital region since 6 months. He had history of trauma in left eye 6 years back, which caused a swelling in the left orbit. No significant past medical & family history was found. Routine laboratory investigation was within normal limit. On the computed tomography (CT scan), well defined enhancing lytic lesion 3.5×3.2cm arise from lateral wall orbit, mild exophytic extension seen on lateral aspect abutting temporarily muscle. Upon full body radiological workup, there was no evidence of distant metastasis or any signs of other abnormality or disease. Clinical suspicion were bone tumour, chondroma or chondrosarcoma or chondromyxoid fibroma and was advised incisional biopsy and histopathological evaluation.

After obtaining the patient's written consent, an incisional biopsy was done from the left orbital region. Grossly two grey white firm to hard tissue bits, largest measuring 1.1×0.8cm was obtained. On Haematoxylin and eosin (H and E) stain, section showed a proliferation of atypical chondrocyte in lobular pattern surrounded by neoplastic osteoid matrix admixed with spindle cells in fibrous stroma. At places mineralization of osteoid, myxoid changes, bony trabeculae surrounded by chondrocytes were observed [Fig-1 & 2]. On the basis of above findings, a diagnosis of chondroblastic osteosarcoma was arrived at and was advised to be confirmed by Immunohistochemistry (IHC). It showed as SATB-2 positivity, leads to confirmation as osteosarcoma having chondroblastic morphology (chondroblastic osteosarcoma).

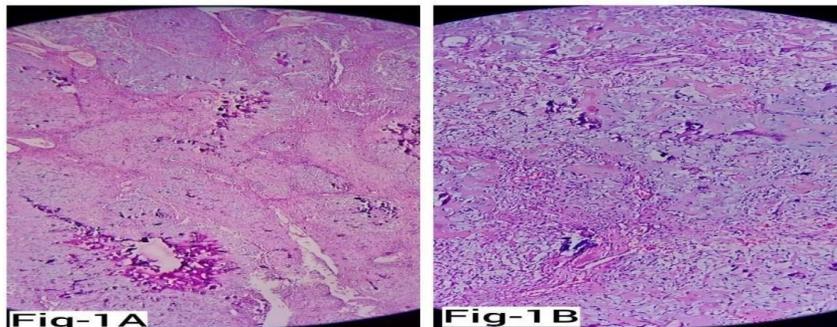


Figure-1A & 1B: Hematoxylin & eosin stain, H & E stain (200x) atypical chondrocyte in lobular pattern surrounded by neoplastic osteoid matrix & myxoid changes

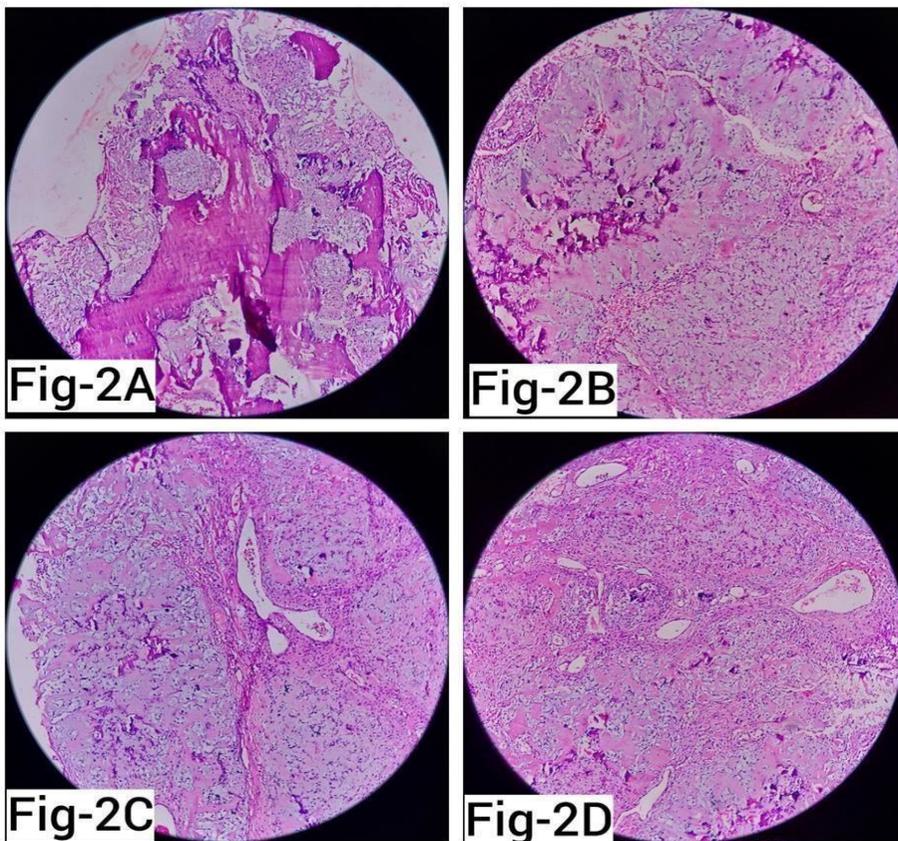


Figure-2A, B, C, D: Hematoxylin & eosin stain, H&E stain (200x) shows atypical chondrocyte in lobular pattern surrounded by neoplastic osteoid matrix, mineralizing osteoid & myxoid changes, spindle cells in fibrous stroma & few dilated thick wall vessels.

DISCUSSION

Osteosarcoma is the most common primary bone malignancy of childhood and adolescent. It primarily affects the mandible and maxillary bones in the craniofacial region. Rather than the superior aspect (zygoma, orbital rim) of the maxillary bone, it typically affects the inferior aspect (alveolar ridge, sinus floor, palate). [3]

The main clinical manifestations of OS of orbit is pain of variable intensity, swelling of bone & compression of adjacent soft tissues leading to proptosis. The results of radiography can vary, although they frequently show cortical damage and extend into the nearby soft tissue. In the literature, very rare cases of extra skeletal

osteosarcoma (EOS) & metastasis of OS to orbit have also been described. Patients with skeletal OS typically present in their earlier years, while those with EOS typically do so in their fourth or fifth decade of life. [4]

In the 2020 WHO classification of soft tissue & bone tumour, osteosarcoma has been sub-classified into 6 subtypes: (1) low-grade central osteosarcoma, (2) osteosarcoma NOS (not otherwise specified), includes Conventional osteosarcoma (COS), telangiectatic osteosarcoma, and small cell osteosarcoma (3) parosteal, (4) periosteal, (5) high-grade surface, and (6) secondary osteosarcoma. [1]

Conventional osteosarcoma (COS) may have different histological patterns. Currently, there is no correlation between prognosis, treatment, and histology patterns. Histological pattern depends on the basis of predominant matrix into osteoblastic (76-80%), chondroblastic (10-13%), and fibroblastic (10%) subtypes.[1]

On histomorphology of COS, essential to the diagnosis is the identification of the permeative pattern encasing bony trabeculae & lace like arrangement of neoplastic bone formation. The cells can be spindled, clear, epithelioid, plasmacytoid or anaplastic giant cells. In our case of chondroblastic osteosarcoma, the predominant component is hyaline cartilage with severe cytological atypia, myxoid chondroid matrix, spindling of tumour cells at the periphery of the nodules which closely mimics to chondrosarcoma.

When a biopsy results in mainly high-grade malignant cartilage, it is important to strongly suspect chondroblastic osteosarcoma, especially in younger patients with orbital bone disease (in whom chondrosarcoma is much less common). On small biopsy specimen, distinguishing chondrosarcoma from chondroblastic osteosarcoma can be challenging & difficult. This distinction is critical in selecting appropriate treatment modality, as surgery+adjuvant chemotherapy is the standard treatment for OS, whereas chondrosarcoma generally treated by surgery alone.[5]

Interesting & unique characteristic of the case was, small biopsy sample in a young male, orbital bone is rare among craniofacial OS, it contained malignant chondroid tissue closely mimicking chondrosarcoma, is prone to misdiagnosis & subsequent mismanagement. Since chondrosarcoma and chondroblastic osteosarcoma have radically different forms of therapy, IHC was crucial in confirming the diagnosis. Smith et al. evaluated 496 cases of the head and neck OS and found that the, higher-grade tumors were more commonly encountered in the skull and other craniofacial bones.[8] Takahama junior et al., in a study with 25 cases of OS, observed that, according to the histological type of the tumor, patients with the chondroblastic type had a higher survival rate when compared to patients with the osteoblastic type.[6] Ezrin is a useful IHC marker for differential diagnosis between COS and conventional CS with a specificity of 100%.[7]

The prognosis of osteosarcoma is also influenced by histological response to neoadjuvant chemotherapy, tumour stage, anatomical location, and adequacy of surgical resection margins.[1]

CONCLUSION

Since the orbital border and craniofacial bone are complex anatomical structures, orbital OS remains a diagnostic and therapeutic challenge. For an early diagnosis and to select the best course of therapy, surgeons and pathologists must be aware of this common bone tumor in a rare location in young patients

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Conflicts of interest

There are no conflicts of interest

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