Giant cell rich osteosarcoma: A rare case report

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Abstract

Giant cell-rich osteosarcoma is an extremely rare variant of conventional osteosarcoma. It accounts for only 1%-3%. It is an undifferentiated high-grade sarcoma with numerous osteoclast-like giant cells. It also contains variable amount of tumor osteoid. This article aims to present a rare case of Giant cell-rich osteosarcoma involving the right knee joint in a young man.

Keywords: Giant cell, giant cell rich osteosarcoma, giant cell tumor, osteosarcoma

Introduction

Osteosarcoma is the most common malignant bone tumor which is characterized by the formation of disorganized immature bone or osteoid tissue from mesenchymal tumor cells. It most commonly involves metaphysis of the appendicular skeleton like long bones [1]. Giant cell-rich osteosarcoma can be defined as "an osteosarcoma in which more than 50% of the tumor consists of numerous uniformly distributed osteoclastic giant cells amidst oval or spindle mononuclear cells embedded in a fibro vascular stroma" [2]. It is an extremely rare histologic variant, which accounts for only 1%-3% of conventional osteosarcoma [1]. Bathurst et al. first reported it in the year 1986 [3]. Here, we present a case of Giant cell-rich osteosarcoma in a 24-year-old man

Case Report

A 24 yrs. old man came to the department of Orthopedics with a complain of mass and pain in the right knee joint since 6 months. The mass progressively increased in size and the patient developed difficulty in walking. On local examination, a firm to hard mass of size 10 x 8 cm was palpated in the distal thigh and knee joint involving the proximal tibia. CEMRI right knee lesion was done and it revealed a large lobulated expansile heterogeneously enhancing lesion involving the proximal epiphysis and metaphysis of tibia and fibula with internal necrosis, cortical breach with soft tissue extension and abutments possibly suggesting osteosarcoma. The patient was then advised biopsy of the lesion. Biopsy was done subsequently and the tissue was sent in our department. Microscopy revealed multiple fragments of tumor tissue comprising of many osteoclast like giant cells and round to oval stromal cells with atypia. Areas of haemorrhage and necrosis and few fragments of bony trabeculae with scantly osteoid was noted.

Discussion

Giant cell-rich osteosarcoma is an extremely rare variant of conventional osteosarcoma. It accounts for only 1%-3%. Giant cell-rich osteosarcoma has been reported to originate from previous low-grade osteosarcoma with amplification of the MDM2 and CDK4 genes. Osteoclast-like giant cells originate due to differentiation of mononuclear phagocytes, and have a tendency to cause bone resorption which plays a significant role in the pathogenesis of bone tumors [1]. The highest incidence was from 10 to 25 years old. The incidence of tumors is higher in men than in women in patients over the age of 50 years. In our case too, the patient was a 24-year-old male, which was in accordance with the previous literature [4].
Patients usually present with nonspecific symptoms such as pain and palpable mass. This was seen in the present case as well, where a small swelling became significantly large within a short span of time [5].

Giant cell-rich osteosarcoma primarily arises in the medullary cavity of a growing long bone, specifically the distal femur, proximal tibia and proximal humerus, but it is exceedingly rare in the orofacial region. It presents as purely osteolytic lesion radiologically. In our case also, the lesion started in the proximal tibia [6].

The most important histopathological differential diagnosis of Giant cell-rich osteosarcoma includes fibroblastic variant of conventional osteosarcoma, telangiectatic osteosarcoma and undifferentiated pleomorphic sarcoma. Giant cell-rich osteosarcoma lacks the characteristic blood-filled spaces and has a much more uniform distribution of multinucleated giant cells, thus excluding telangiectatic osteosarcoma. The storiform arrangement of the spindloid cells is common to both Giant cell-rich osteosarcoma and the fibroblastic variant of osteosarcoma, however the clustered appearance of the giant cells in Giant cell-rich osteosarcoma helps in differentiation from the fibroblastic variant. Undifferentiated pleomorphic sarcoma is likely to have a similar picture as Giant cell-rich osteosarcoma consisting of the streaming pattern of the tumor cells, but the presence of tumor osteoid in Giant cell-rich osteosarcoma helps in its exclusion [1].

The treatment modality of Giant cell-rich osteosarcoma is same as that of traditional osteosarcoma which consists of aggressive surgical resection followed by radiotherapy and chemotherapy. The survival rate of Giant cell-rich osteosarcoma is similar to that of high-grade osteosarcoma ranging from 60% to 70% at 5 years and decreases to approximately 20%-30% in patients with metastatic disease. Long term local control can be achieved with complete resection [3].

**CEMRI Imaging**

Fig a, b, c and d: A large lobulated expansile heterogeneously enhancing lesion is seen involving the proximal epiphysis and metaphysis of tibia and fibula with internal necrosis, cortical breach with soft tissue extension and abutments
Histopathological Examination

Fig e, f (scanner view): Photomicrograph showing bony fragments and malignant spindle cell admixed with loose fibrous tissue containing multinucleated giant cells.

Fig g (low power view): Photomicrograph showing a solid proliferation of atypical mononuclear stromal cells with osteoclast-like giant cells.

Fig h (high power view): Photomicrograph showing nuclear pleomorphism of round to oval cells. Intervening lace-like osteoid deposits and multinucleated osteoclast-like giant cells.

**Conclusion**

Giant cell-rich osteosarcoma has no distinct clinical or radiological features that may aid in its recognition. It is a very rare clinical entity which poses a striking resemblance to that of a Giant cell tumor. The radiological and histopathological differentiation of Giant cell-rich osteosarcoma from other benign and malignant giant cell tumors is highly challenging. It is important to differentiate them from other aggressive giant cell tumors as the prognosis and treatment differs between them. The unusual histological appearance and the exceptional rarity of the lesion poses a great diagnostic challenge; thus, the clinical, radiological and histopathological findings should be integrated for its diagnosis and proper management.

**Conflict of Interest**

Not available

**Financial Support**

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**References**


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