Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2014; 2(8):526-529 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2014.v02i08.008

Mesenchymal Chondrosarcoma of Fibula: Report of a Rare Case with Review of Literature

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Abstract: Mesenchymal chondrosarcoma is a rare aggressive variant of chondrosarcoma that frequently occurs in extra skeletal location and has high predilection for the head and neck region. In our case report a 30 years old lady presented with pain and rapidly growing swelling over lower lateral part of the left leg. The biopsy revealed mesenchymal chondrosarcoma of fibula composed of highly undifferentiated small round cells and islands of well differentiated cartilage involving both the bony tissue as well as soft tissue. Report of immunohistochemistry also supported our diagnosis. Here we report a rare entity that should be considered in the differential diagnosis of any calcified bony lesions, especially in young adults. Since this tumour has a high tendency of metastasis mainly to the lungs, the prognosis is worse in relation to conventional chondrosarcoma. The patient was given a course of adjuvant chemotherapy following complete removal of the tumour but the patient died within one year of follow up.
Keywords: Chondrosarcoma, fibula, mesenchymal chondrosarcoma

INTRODUCTION

Mesenchymal chondrosarcoma (MC) is one of the histomorphological variant of chondrosarcoma (CS). It is very aggressive in nature in relation to other variant of chondrosarcoma and leads to early death. It is characterized microscopically by dimorphic pattern composed of highly undifferentiated small round cells and islands of well differentiated cartilage. Although occurring at any age, the peak incidence is in the second to third decade of life. Males and females are affected equally. Skeletal tumours typically involve the facial bones and ribs, whereas extra skeletal tumours most often occur in the head and neck region. We present here a case of MC in the fibula, because MC itself is an uncommon tumour and is rarely found in the fibula. An adequate biopsy is enough for diagnosis.

CASE REPORT

A 30 year old unmarried female presented to orthopedics outpatient department with complaints of pain and rapidly growing swelling in lower part of the left leg without a history of trauma for last 5 months. X-ray of that portion revealed a mixed lucent & mineralizing lesion occupying the upper 2/3 rd of the fibula with a large soft tissue component. Her family history & medical history were uneventful. As the lesion was growing rapidly and there was clinical suspicion of a malignant neoplasm, an incision biopsy was done. Biopsy showed very few bits of well differentiated cartilaginous tissues only. Diagnosis of a well differentiated chondrosarcoma was made considering the clinical features and radiological findings. Subsequently the patient underwent a radical surgery. We received a specimen of fibula with a fusiform type of growth occupying upper 2/3 rd of the cut bone. Total length of bone was 17 cm. Growth measured 12 cm× 6 cm×4 cm. Cut surface of the growth was soft, fleshy, friable and areas of haemorrhage with attached surrounding muscles & fat, mimicking other sarcomas. During cutting a gritty sensation was also felt. The growth reached up to one end of the bone and was 5 cm from another end of the cut bone.

Microscopic examination revealed a dimorphic pattern of growth comprising of highly cellular undifferentiated mesenchymal stromal cells and abrupt presence of island of well differentiated cartilaginous tissue. These stromal cells were small in size, round to oval in shape with hyper chromatic nuclei, arranged

around slit like stag horn-shaped vascular spaces mimicking the appearance of haemangiopericytoma. Focal areas of frank spindling also found. Pleomorphism was very modest with a few mitotic figures. The chondroid islands showed foci of calcification without any ossification. Immunohistochemically the small undifferentiated cells showed strong positivity for vimentin whereas the chondroid areas were weakly positive for S-100. A final diagnosis of mesenchymal chondrosarcoma was established. In view of the disease the patient was given a course of radiotherapy and a combination of chemotherapies. But patient died within one year of diagnosis in spite of all modalities of treatment.

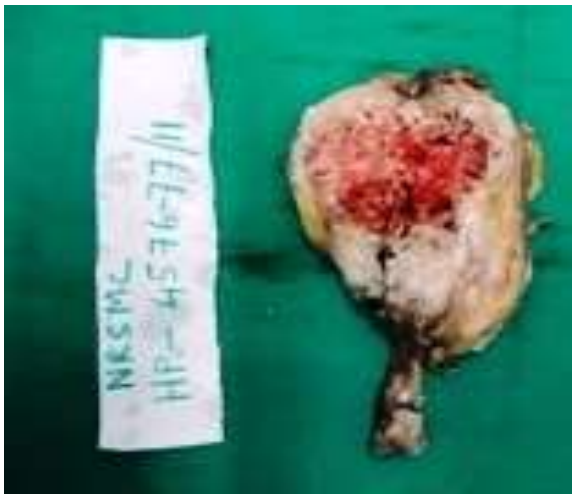

Fig. 1: Gross picture of cut section of one end of the fibula with a growth showing friable, necrotic and fleshy in appearance

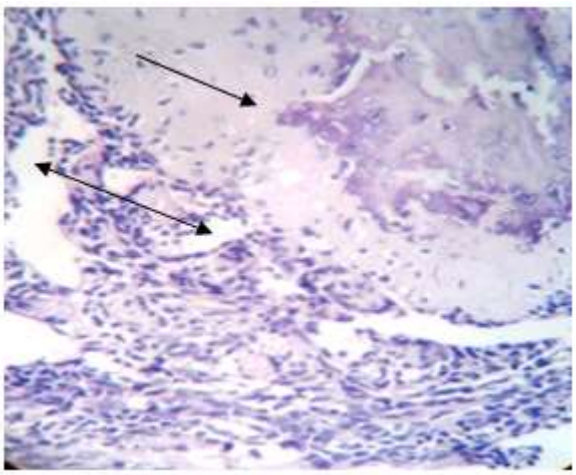

Fig. 2: Low-power-field photomicrograph revealing areas of haemangiopericytoma-like spaces (double arrow) surrounded by hypercellular undifferentiated mesenchymal cells which are densely stained. Relatively pale staining chondroid components are also shown (arrow) (haematoxylin & eosin, ×100)

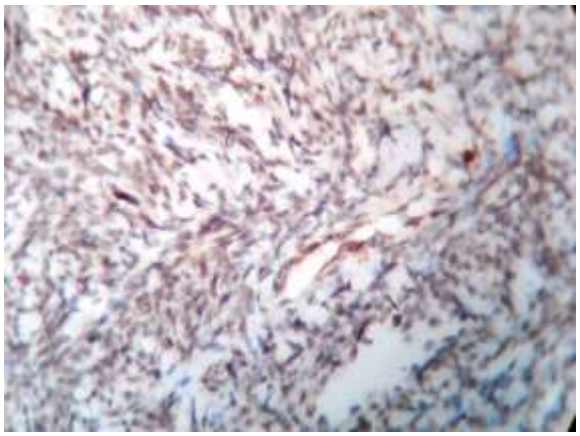

Fig. 3: Immunohistochemically the small undifferentiated cells were vimentin positive (haematoxylin & eosin, × 400)

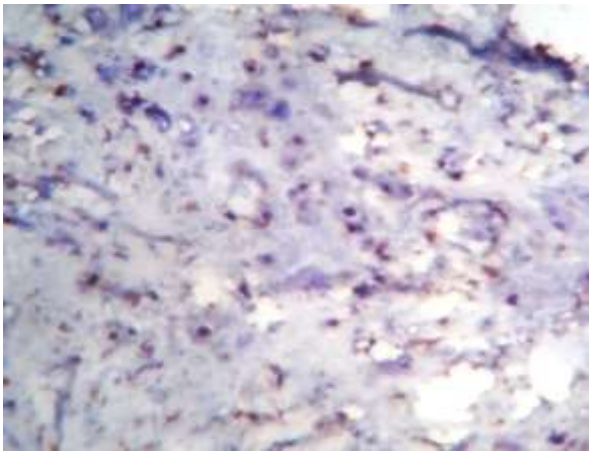

Fig. 4: Immunohistochemically the chondroid areas were weakly positive for S-100 (haematoxylin & eosin, ×400)

DISCUSSION

Mesenchymal chondrosarcoma (MC) is a rare tumour of bones and extra osseous tissues, most commonly occurring in the third decade of life compared to conventional CS [1]. Our patient too was a 30 year old female. This rare entity was first described by Lichtenstein and Bernstein in 1959 as a distinct entity [2]. There are several histological variants of chondrosarcoma including clear cell, dedifferentiated, myxoid and mesenchymal [1]. MC represents less than 2% of all cartilaginous malignancies [3]. Compared to conventional chondrosarcomas, which are rarely extra skeletal and occur later in life, MC frequently has an extra skeletal location and an earlier age at presentation [3]. Skeletal tumours typically involve the jaw and ribs [3, 4]. Whereas extra skeletal tumours most often occur in the head and neck region including the orbit [5], meninges [6] and in the lower extremities, thigh being the most common site [7]. Huvous *et al.* [8] in their study have reported 14% cases of MC occurring in extra osseous tissues like upper arm, para-spinal soft tissues, inguinal region, thigh and lower leg. Whereas

Niven *et al*. [9] found 36 cases of MC in maxilla and the most common reported symptom was swelling/mass in 68% of cases, followed by nasal obstruction (32%), epistaxis (32%) and tooth mobility (24%). However, MC in any location is extremely rare reported by the diagnosis of just 15 cases among 6,221 benign and malignant tumours gathered by the Mayo Clinic [10]. The largest review was also done by Nakashima *et al*. [3] in 1986 with 111 patients of MC to date.

Radiologically, the skeletal lesions are primarily lytic and destructive with poor peripheral margins [16]. The radiolucent area often contains scattered foci that are caused by calcification or ossification of cartilage matrix [17]. Cortical destruction with breakthrough and extra osseous extension into soft tissue are common.

However, according to the study by Jaetli *et al*. [11], these radiological features of MC are not characteristic. Histopathologically mesenchymal chondrosarcoma characteristically displays a dimorphic pattern, comprising islands of mature cartilage and cellular undifferentiated mesenchymal cells. The main differential diagnoses are Ewing's sarcoma, Primitive neuro ectodermal tumour (PNET), lymphoma/leukemia, haemangiopericytoma, rhabdomyosarcoma, small cell osteosarcoma and malignant melanoma, particularly in biopsy specimens [19]. However, presence of relatively well differentiated cartilage helps to make a specific diagnosis of MC [12]. Immunohistochemically, the small cell components are positive for vimentin, CD99 and Leu7 but not for S-100 protein [13]; the latter is found instead in the chondroid areas [14]. Not much is known about the pathogenesis and the molecular events resulting in the development of this tumour. Unlike other types of chondrosarcoma, which have a tendency to grow more slowly and rarely develop metastasis, mesenchymal chondrosarcoma is a fast growing tumour and often metastasizes [20]. Metastasis of MC occurs by haematogenous route and the most common site of involvement is the lung [15].

MC requires surgical excision with wide margins [21, 22]. Pre and postoperative chemotherapy or radiation may be a choice of treatment, although their effectiveness is unclear [23]. Therefore continuous long term follow-up should be carried out in all patients with MC.

CONCLUSION

Although MC is quite rare, it should be kept in mind while dealing with malignant cartilaginous tumour specially in young individuals. Since MC is a rapidly growing malignant tumour that may involve the long bones of the extremities with popensity to metatasize, early diagnosis and initiation of treatment is mandatory.

ACKNOWLEDGEMENT

The author is grateful to Mr. Animesh Hazra, Assistant Professor, Department of Computer Science & Engineering, Jalpaiguri Government Engineering College, Jalpaiguri, W.B, India, for the provision of valuable technical support.

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