

CASE REPORT

Extraskkeletal mesenchymal chondrosarcoma in an elderly male - A rare entity

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Abstract

Mesenchymal chondrosarcoma (MCS) is a rare high grade malignant tumor having both skeletal and extraskkeletal manifestations. This rare variant of chondrosarcoma occurs in the deep soft tissues, usually of the limb girdles or extremities, but may occur in the trunk wall, abdomen and breast.

Unlike conventional chondrosarcomas, mesenchymal chondrosarcomas occur with greater frequency in young adults. This form of cancer often metastasizes to other areas of the body and can cause life-threatening complications.

This report describes the case of a 60 years old male, who presented with purely extraskkeletal MCS involving the right side of his chest wall.

Key words

Mesenchymal chondrosarcoma, Chest wall, Elderly male

1 Introduction

Mesenchymal chondrosarcoma (MCS) is a rare aggressive malignancy accounting for 3-10% of all primary chondrosarcomas. This tumor is characterized by propensity for local recurrence and distant metastasis.

It usually appears in the second and third decades of life, more frequently in females than males (F/M = 1.4/1) and can involve bones as well as soft tissues ^[1-3]. Due to these unique features, MCS deserves to be differentiated from conventional chondrosarcoma and a host of other close mimics.

We describe the case of an elderly male who presented with this tumor on the right side of his chest wall and also briefly review the relevant literature.

2 Case presentation

A 60 year male, presented with painless firm, lump on the right anterior chest wall since 6 months duration without any history of shortness of breath. He was treated with anti-inflammatory medication; however, there was no alleviation of symptoms.

Chest radiograph depicted a soft tissue lobulated mass in the right anterior chest wall (Figure 1). Computed Tomography scan showed an 18 cm × 15 cm × 10 cm heterogeneous soft tissue mass in the right anterior chest wall with no involvement of underlying bony structure, which was further confirmed by MRI (Figure 2). The patient was operated upon with wide excision of the irregular mass lesion with grossly lobulated outer surface, which was situated anterior to the ribs, without its involvement or the pleura. The cut section revealed solid grey white firm surface with specks of haemorrhage (Figure 3). Histological sections showed a lobulated mass displaying scattered atypical chondroid cells with hyperchromatic small ovoid cells with smudgy chromatin and inconspicuous nucleoli. Occasional cells revealed binucleation and prominent pleomorphism in a background of chondroid matrix (Figure 4). The tumor cells showed negative staining for CD99 and myogenin and focal desmin positivity. The chondroid cells showed positive nuclear staining with S100.

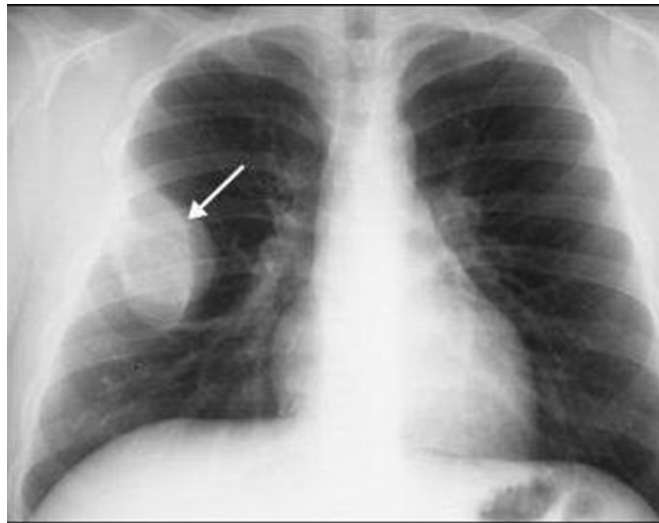


Figure 1. MCS: Chest radiograph shows a soft tissue lobulated mass in the right anterior chest wall



Figure 2. MCS: MRI image shows an 18 cm × 15 cm × 10 cm, soft tissue lesion in the right anterior chest wall with no change in the underlying bony structure



Figure 3. MCS: Gross photograph shows a well circumscribed solid, firm, glistening white growth

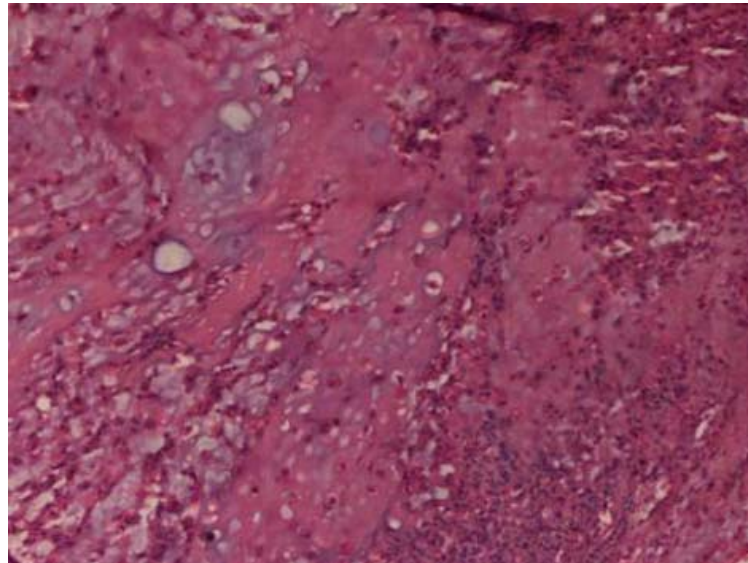


Figure 4. MCS: Tissue section shows well differentiated chondroid areas, and undifferentiated neoplastic cells with small, round nucleus and clear cytoplasm. Hematoxylin & Eosin 100X

Postoperatively, a metastatic workup was performed with staging CT imaging of the head, chest and abdomen which did not show any evidence of metastatic disease. The patient received 6 cycles of chemotherapy: Vincristine 1 mg/m², day 1-2, Ifosfamide 1000 mg/m², day 1-5, Doxorubicin 65 mg/m², day 1-2, Etoposide 125 mg/m², day 1-5. As the resection margins were positive for malignancy under the microscope, Cobalt-60 radiation therapy consisting of 54 Gy in 30 fractions was also administered at the site of the resected lesion. The patient is doing well after 18 months of follow up period.

3 Discussion

Mesenchymal chondrosarcoma (MCS) is a rare high grade malignant tumor. It was first classified as a distinct variant of chondrosarcoma of the bone by Lichtenstein and Bernstein ^[2]. Subsequently Dowling described the extraskelatal occurrence of this tumor ^[3]. The incidence of MCS is high in the second and third decades of life, usually within 15-35 years, although any age group can be affected ^[1]. Nakashima et al have observed equal gender distribution of MCS, in their study on 111 cases of this tumor ^[4].

Around 66% cases of primary MCS are osseous in origin while the rest arise in soft tissues. Bones of the axial skeleton are principally affected, namely cranium, mandible, ribs, ilium and vertebrae. Among the extraskelatal locations, meninges are the most favoured site followed by head & neck region, orbit, nasopharynx, extremities, trunk and retroperitoneum ^[4, 5]. Computed Tomography (CT) imaging shows a well-defined mass lesion with multiple areas of fine and coarse calcifications in MCS. CT scan in our case showed a massive soft tissue lesion in the right anterior chest wall, without any change in the underlying bone structure.

Patients suffering from MCS present with gradually enlarging mass usually accompanied by pain. Lesions in orbit can cause proptosis and visual disturbance, while those arising from meninges result in symptoms of raised intracranial tension and compressive myelopathy. The duration of symptomatology is variable, ranging from less than 6 months to 2-3 years, as reported in different studies ^[4, 5]. Plain X-rays of skeletal MCS show destructive lytic lesions with spotty calcification while the extraskelatal counterparts tend to exhibit soft tissue masses with variable amount of mineralization ^[1, 6].

The most striking feature of MCS is its biphasic histological appearance. One component is made up of islands of well differentiated cartilage while the other consists of sheets of anaplastic small round to oval cells, often arranged around thin vascular channels resembling a haemangiopericytoma like growth pattern. The proportion of these two components is highly variable amongst different tumors, but the presence of both these components is essential for the diagnosis of MCS. Areas of calcification and rarely ossification may be present ^[1, 4, 5].

Immunohistochemically, the undifferentiated small cells of MCS stain positive for CD99, vimentin, Leu7 while the cartilage can exhibit S-100 positivity. These features are also expressed by other small round cell tumors and hence seldom help to diagnose MCS. However, Muller et al have demonstrated that presence of type II collagen in extracellular matrix is unique to MCS and is not expressed by other small cell sarcomas ^[7]. Also the ability of MCS to consistently express Sox-9 gene can aid in its distinction from other small round cell tumors ^[8].

It is imperative to remain aware of the differential diagnosis of MCS, especially when small biopsy specimens with selective sampling are interpreted or the tumor has a peculiar small round cell histology; as in Ewing's sarcoma, small cell osteosarcoma, synovial sarcoma, lymphoma, leukemia deposits (granulocytic sarcoma), neuroblastoma and rhabdomyosarcoma. In tumors with prominent vascular component, haemangiopericytoma should also be considered in the differential diagnosis. Presence of chondroid lobules separate Ewing's sarcoma and haemangiopericytoma. Lymphoma and leukemia usually show leukocyte common antigen, CD43 positivity and variable CD20 expression. Antibodies to cell surface antigen, HbA 71 is more specific to identify lesions with neural origin (Ewing's sarcoma and neuroblastoma). Translocation (X;18) is seen in synovial sarcoma and t(1;13) and/ or t(2;13) in rhabdomyosarcoma. In contrast, MCS is associated with Robertsonian translocation [der (13; 21) (q10; q10)]. Thus, a combined approach involving morphology, immunohistochemistry and molecular genetics can comprehensively distinguish between MCS and its mimics ^[7-10].

MCS is an aggressive malignancy with marked tendency to local recurrence and distal metastasis, principally to the lungs and less commonly to lymph nodes. Current treatment options include wide surgical excision with chemotherapy or radiotherapy and long term follow up. However, prognosis remains poor despite early diagnosis and adequate treatment ^[1, 5]. Nakashima et al, have stated that extensive surgical resection has a better survival rate due to lesser rate of

recurrence, with a 5-year and 10-year survival rates as 54.6% and 27.3% respectively^[4]. Chemo-irradiation alone, leads to complete resolution of this tumor with eradication of micrometastases and tends to offer a good prognosis^[11-13]. Our case is doing well after 18 months of follow up after surgical excision and adjuvant chemo-irradiation therapy.

4 Conclusion

Because of its rarity, the natural history of MCS is still poorly understood and requires further research. For a pathologist, the potential diagnostic pitfalls include small biopsies and selective sampling. Hence presence of both cartilaginous and anaplastic small cell components should be confirmed for correct diagnosis.

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