CASE REPORT

Mesenchymal chondrosarcoma – A rare cause of pleural effusion

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SUMMARY

Mesenchymal chondrosarcoma is a malignant neoplasm arising from cartilaginous bone or soft tissue. It is uncommon yet devastating. Our patient was a 21-year-old man who presented with pleuritic chest pain and weight loss. His chest radiograph showed left pleural effusion. His pleural effusion analysis was consistent with exudative pleural effusion. Tuberculosis workup was negative. Pleural fluid cytology did not yield malignant cells. Subsequently, his computed tomography of thorax showed left rib sclerotic lesion with soft tissue component. Biopsy of the soft tissue eventually confirmed the diagnosis of mesenchymal chondrosarcoma. He succumbed to his illness before the diagnosis was confirmed. We hope that through this case report, we are able to provide some insight into this rare condition.

KEY WORDS:
Mesenchymal chondrosarcoma, malignant neoplasm, pleural effusion

INTRODUCTION

Mesenchymal chondrosarcoma is a rare malignant cartilaginous neoplasm arising within bone or soft tissue. It represents only 2-10% of all chondrosarcoma and it has a prevalence of 0.2 to 0.7 cases per 100,000 population.[1] It can arise from bone or extraskeletal tissue. Common sites of tumour occurrence include axial skeleton, femur, craniofacial bone and rib.[2] Cases of extraskeletal involvement such as meninges, parapharyngeal and orbits have also been reported. Clinically, patients present with pain, bony swelling and rarely pleural effusion. Here, we report a case of mesenchymal chondrosarcoma presented with recurrent pleural effusion.

CASE PRESENTATION

A 22-years-old man with no past illness presented with chest pain, dyspnoea and loss of weight for 2 weeks duration. He denied fever, cough or haemoptysis. There was no family history of tuberculosis or malignancy. On examination, there was reduced breath sound over left hemithorax, associated with stony dullness on percussion. There was no palpable lymph node. Systemic examination showed marked cachexia with no other positive findings.

His full blood count, renal profile and liver function test were normal. His chest radiograph showed left pleural effusion. His sputum was negative for acid fast bacilli. A diagnostic and therapeutic thoracocentesis was done. His pleural fluid findings was consistent with exudative pleural effusion. Both pleural fluid adenosine deaminase and pleural biopsy were negative. There were no malignant cells seen from his pleural fluid cytology.

Subsequently, we proceeded with computed tomography (CT) of thorax which showed left 7th rib sclerotic lesion with enhancing soft tissue component. Biopsy of the soft tissue showed island of cartilage and short spindle cells with myxoid background. The immunochemistry showed positivity to CD99 with negative S100, CD45, CKAE1/3. The biopsy confirmed the diagnosis of rib mesenchymal chondrosarcoma. Unfortunately, he succumbed to his illness before the diagnosis was obtained.

DISCUSSION

Mesenchymal chondrosarcoma is a rare neoplasm of cartilaginous bone and has an aggressive disease course. Despite growing numbers of reported cases, it still remains a challenge to clinicians both diagnostically and therapeutically. The tumour was first reported by Lichtenstein and Bernstein back in 1959 with its distinct histopathological features. Overall, mesenchymal chondrosacroma represents less than 10% of all chondrosarcoma. The common age group involved is second and third decade of life and it has similar gender preponderance.[1]

Clinically, the presentation is usually dependent on the sites involved. Patients may complain of bony swelling, mass, bone pain and occasionally pathological fractures.[2] Occasionally, extraosseous tissue can be involved such as orbit, dura, trunk and retroperitoneum.[3] Patient may present with cranial nerve palsy, headache, etc. Our patient's presentation was rather atypical. He presented with pleuritic chest pain and exudative pleural effusion.

It is quite common to have patients who present with pleural effusion in our daily practice especially in the primary care setting. It is therefore important to evaluate and identify the aetiology of pleural effusion. In chest radiograph, clinicians need to examine the effusion, lung tissue and of course not

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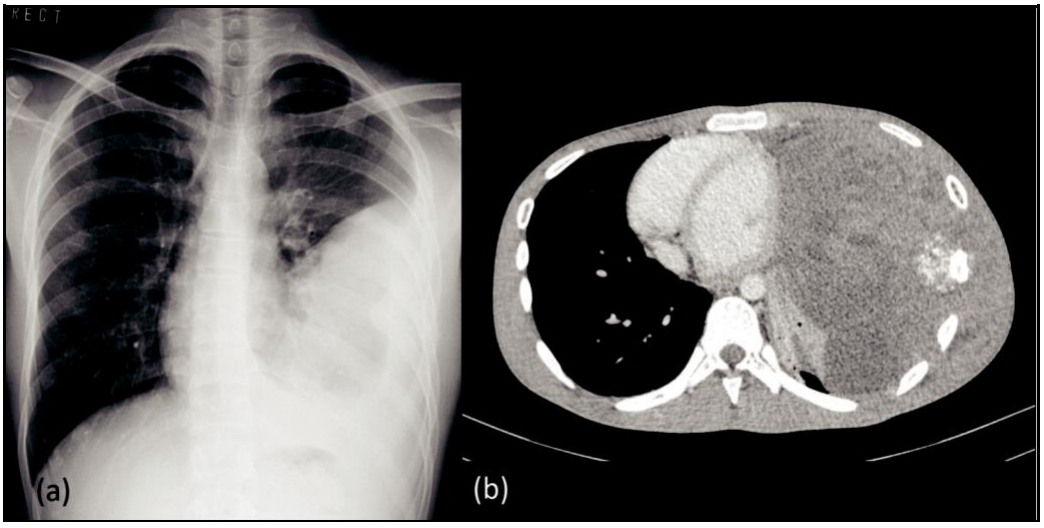

Fig. 1: a). Initial Chest Radiograph of the patient. b). Computed tomography of thorax showing lesion at left 7th rib with soft tissue component and infiltration to surrounding structure

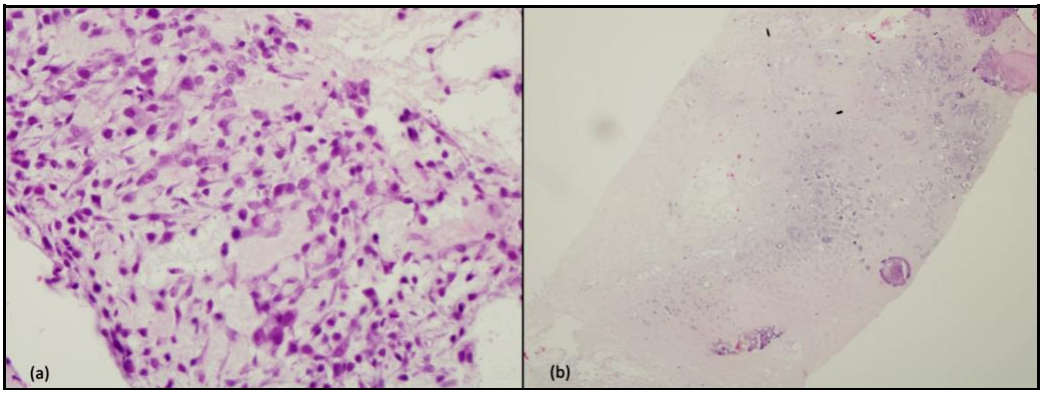

Fig. 2: a). Histopathological examination of biopsy showed aggregate of small spindle cells with hyperchromatic nuclei and scanty cytoplasm. b). Histopathological slide showing hyaline cartilaginous elements

forgetting the ribs. Plain radiography in mesenchymal chondrosarcoma usually show osteolytic lesions with calcification within.[4] In plain computed tomography, the tumour may appear well defined or lobulated or there could be stipple, arc, ring or coarse dense calcification. It usually shows intense enhancement after contrast.[5] In magnetic resonance imaging, the tumour may have heterogenous appearance or T1 and variable T2 weighted signal.[1] However, there are no unique features on imaging to differentiate mesenchymal chondrosarcoma from other chondrosarcoma.

The diagnosis of mesenchymal chondrosarcoma can usually be confirmed by biopsy and histopathological examination. Macroscopically, it may appear greyish-white or pink, soft to firm in consistency. Microscopically, the tumour has a characteristic dimorphic pattern, the presence of highly cellular and undifferentiated mesenchymal cells and islands of well-differentiated cartilage.[3] The main differential diagnosis include Ewing sarcoma, hemangiopericytoma and dedifferentiated chondrosarcoma. Immunohistochemistry may play a role. Mesenchymal chondrosarcoma usually shows immunopositivity for vimentin, S-100 and Mic-2 (CD99) and negativity for cytokeratin, EMA and CD34.

The optimal treatment for mesenchymal chondrosarcoma remains unknown. The available treatment options are surgical resection of the tumour, chemotherapy and radiotherapy.[5] Wide local excision is the recommended treatment of choice when surgical resection is possible. In large case series, it is shown that the resection improves the outcome of the patient especially when combined with radiotherapy. Radiotherapy can improve survival of patients when complete resection is not possible. The role of chemotherapy is rather controversial. Some studies demonstrate benefits of chemotherapy in treating mesenchymal chondrosarcoma. The ambiguous response to chemotherapy may be due to specific histology pattern. Ewing-like microscopic feature may respond better to chemotherapy.[4]

The prognosis of mesenchymal chondrosarcoma is rather poor as the disease sometimes has a very aggressive course. The overall 10-year survival is reported to be less than 30%. Our patient died before histological diagnosis was obtained. This case highlighted that mesenchymal chondrosarcoma is a rare cause of pleural effusion. When approaching a case of pleural effusion, we should always be vigilant and bear in

mind that pathology of the rib can be a cause of pleural effusion. Early diagnosis is important so that treatment can be initiated.

CONCLUSION

In conclusion, mesenchymal chondrosarcoma is a very rare and aggressive malignant neoplasm. It can occur in skeletal and extra-skeletal tissue. Our case highlights the difficulty in obtaining the diagnosis of mesenchymal chondrosarcoma. Given its rarity, clinicians must have high index of suspicion in order to achieve early diagnosis as this is often associated with better outcome.

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