

CASE NOTE

Primary myxoid sarcoma of the pleura

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Metastatic tumours are the most common form of pleural cancer. Primary malignant pleural tumours, other than diffuse malignant mesothelioma, are extremely rare; there are few case reports and small series in the literature, and most report sarcomas of different subtypes.¹⁻³

CASE REPORT

A 62-year-old man was referred to the outpatient clinic of the Chest Surgery Department, Centre hospitalier universitaire de Sherbrooke, with progressive dyspnoea, dull pain at the left hemithorax, limitation of daily activities and 5-kg weight loss in the few months before presentation. A radiograph of his chest showed a left massive pleural effusion, a computed tomography (CT) scan showed left pleural thickening and a positron emission tomography (PET)/CT scan highlighted a large hypermetabolic mass along the mediastinum and hilar areas adjacent to the pericardium and extending into the left thoracic cavity, suggestive of cancer. On the CT scan, the heterogeneous mass was of similar density and undistinguishable from the pleural fluid in the thoracic cavity; however, the mass was easily distinguished from the pleural fluid on the PET/CT scan (Fig. 1). The primary clinical diagnosis was diffuse malignant mesothelioma. We performed a thoracoscopy to drain and biopsy the pleura; the effusion was gelatinous and could not be drained completely.

We performed talc pleurodesis, but pleural effusion recurred. The biopsy showed a myxoid sarcoma composed of epithelioid and spindle cells. The patient returned to the hospital after 3 weeks with massive left pleural effusion and progressive dyspnea. Physical examination showed edema of both lower limbs and congestion of the neck veins. A follow-up CT scan showed a progression of the neoplasia with right shift of the mediastinum. We performed a left posterolateral thoracotomy to debulk a gelatinous and friable tumour.

The histologic evaluation of the tumour revealed a diffuse spindle-cell proliferation in a myxoid stroma with some foci of epithelioid and small-cell phenotype (Fig. 2). There were no areas of chondroid or lipomatous differentiation.

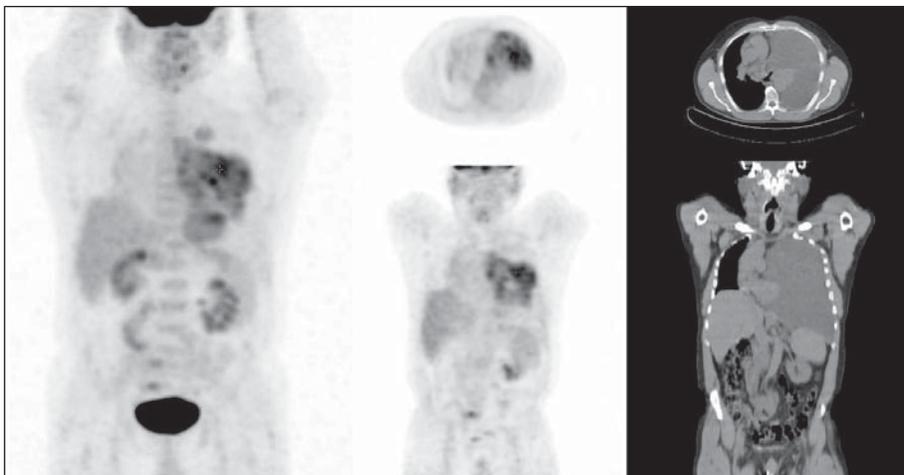


Fig. 1. Positron emission tomography (PET) maximal intensity projection image (**left**), transaxial and coronal PET image (**centre**) and corresponding transaxial and coronal computed tomography (CT) image (**right**). The PET scan was obtained 60 minutes after injection of ¹⁸F-fluorodeoxyglucose (FDG). The images show a large hypermetabolic mass along the mediastinum and hilar areas, adjacent to the pericardium and extending in the left thoracic cavity. The heterogeneous mass is of close density to that of water, appearing undistinguishable from the pleural fluid in the thoracic cavity on CT, but clearly visible on the FDG-PET images.

Mitotic figures were frequent, and there were large areas of necrosis that involved about 30% of the tumour mass. Immunohistochemistry yielded diffuse vimentin and focal CD34 and S-100 protein positivity. Results were negative for AE1/AE3 keratin, EMA, bcl-2, desmin, CD99, CD56, chromogranin and synaptophysin. The negativity of the epithelial markers excluded the possibility of a sarcomatoid mesothelioma. As the more specific immunohistochemical markers were negative, our final diagnosis was a poorly differentiated myxoid sarcoma. However, we favour an extraskeletal myxoid chondrosarcoma based on morphology. The patient's postoperative course was uneventful, and he returned home after 10 days. He received a radiotherapy regimen (3 sessions each of 18 Gy). At last follow-up, 12 months after surgery, the patient was doing well with few symptoms, including mild edema of both lower limbs, which is controlled with diuretics, and mild limitation to exercise.

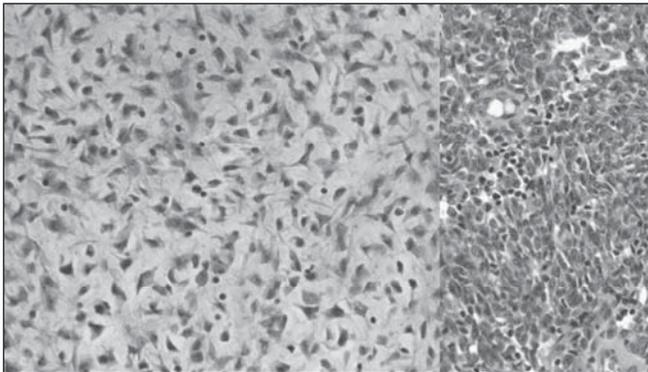


Fig. 2. Histopathology of the specimen showed (A) stellate spindle cells in a myxoid stroma (hematoxylin and eosin stain, original magnification $\times 25$) and (B) a small cell area focally observed in the neoplasia (original magnification $\times 10$).

DISCUSSION

Primary pleural sarcomas are very rare.¹⁻³ However, most of the histological types have been described in this location, including synovial sarcoma, malignant fibrous histiocytoma, primitive neuroectodermal tumour/Ewing sarcoma, angiosarcoma, chondrosarcoma, osteosarcoma and liposarcomas. Tumour diagnosis is mainly based on pathologic examination and immunohistochemistry. The prognosis of the disease depends mainly on the size and the histologic subtype of the tumour.⁴

In our patient, the pathologic evaluation showed a poorly differentiated myxoid sarcoma of no specific subtype. We referred the patient to radiotherapy after surgery based on the limited evidence found in the literature that supported aggressive surgical management followed by radiotherapy as the best treatment for this exceedingly rare condition.⁵

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