Case Report

Primary pleural liposarcoma: A rare entity

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ABSTRACT

Primary pleural liposarcoma (PPL) is a rare malignant tumor of the pleura. The diagnosis of PPL may be suspected on chest imaging based on radiologic features such as large pleural mass showing areas of fat with or without calcification. Herein, we present the case of a 32-year-old male whose contrast-enhanced computed tomography scan of the chest revealed a large, heterogeneous, hypodense, right pleural-based mass with small areas of fat and calcification within it. An ultrasound-guided biopsy was performed, which confirmed the diagnosis of a myxoid variant of pleural liposarcoma.

KEY WORDS: Liposarcoma, pleura, pleural neoplasms, sarcoma

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INTRODUCTION

Primary pleural malignant tumors, other than diffuse malignant mesothelioma, are extremely uncommon. Primary pleural liposarcoma (PPL) is one such rare malignant tumor of the pleura. Liposarcomas originate from mesenchymal cell rests and very rarely from a preexisting lipoma. Myxoid variant of PPL is the most common type of PPL.[1,2] The diagnosis may be suspected on the basis of radiologic features such as fat densities with/without calcification. However, these features are nonspecific and overlap with teratoma and hamartoma. Therefore, a histologic diagnosis is essential.

Ackerman and Wheeler reported the first case of PPL in 1942.[3] Fewer than 20 cases have been described in the literature. Herein, we present a patient with the myxoid variant of PPL.

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CASE REPORT

A 32-year-old male presented with chest pain and shortness of breath for 2 months' duration. There was no cough, expectoration, or hemoptysis. There was no swelling or lump observed in any of the limbs. There was no history of abdominal pain or weight loss. The patient had no history of tobacco smoking or any other addictions. On examination, the pulse rate was 92 beats/min, respiratory rate 20/min, blood pressure 116/78 mmHg, and oxygen saturation 95%. Chest examination revealed a dull note on percussion and absent breath sounds in the entire right hemithorax, except for the right infraclavicular area where breath sounds were heard normally. His chest radiograph showed a large, nonhomogeneous opacity involving the right hemithorax, which was denser in the middle and lower lung zones. There was

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mild displacement of the mediastinum toward the left side [Figure 1a]. Ultrasonography (USG) showed a large, heterogeneous, hypoechoic mass lesion within the right hemithorax with the presence of intratumoral vessels observed on Doppler examination [Figure 1b]. USG of the abdomen did not reveal any evidence of abdominal mass or metastasis.

Contrast-enhanced computed tomography (CECT) scan of the chest showed a large, heterogeneous mass measuring 20.8 cm × 13.6 cm × 13 cm in size (in the craniocaudal, anteroposterior, and transverse axes, respectively) in the right hemithorax. The mass was broad based toward the pleura and compressing medially onto the right lung, leading to its compressive atelectasis. However, there was no invasion of the collapsed lung, which was showing a uniform, homogeneous enhancement. The chest wall muscles also showed normal attenuation, and no erosion or lytic destruction of the ribs was seen. The mass was predominantly hypodense with an average attenuation of 10 Hounsfield units (HUs). There were small peripheral fat attenuation areas (−70–−90 HU) and few calcific foci (+650–+900 HU) within the mass [Figure 2a and b]. Based on the computed tomography (CT) findings, a diagnosis of PPL was suggested. An USG-guided biopsy was performed, which showed predominantly myxoid tissue with scattered atypical cells and lipoblasts in a myxoid background. Immunohistochemistry revealed that the tumor cells were positive for S100, whereas the stroma stained well with Alcian blue, suggesting an abundant mucinous background [Figure 3]. A diagnosis of a myxoid variant of liposarcoma was made on histopathology. Based on the radiological evidence of pleural origin and histopathological observation of liposarcoma, the final diagnosis of "PPL" was attained. The tumor was considered unresectable due to the invasion of the diaphragm. The patient was administered palliative chemotherapy and radiotherapy and is under follow-up.

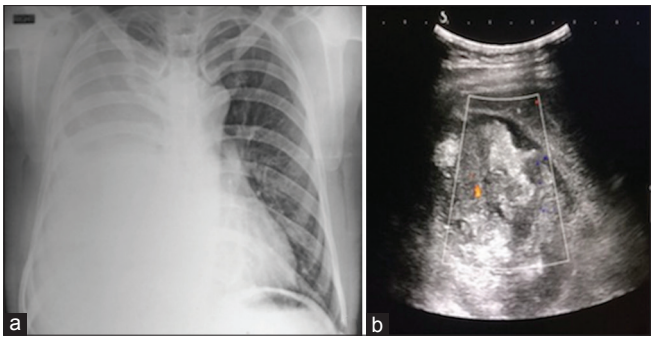

Figure 1: (a) Chest radiograph showing a large, homogeneous radiopacity involving the right hemithorax with an associated mild mediastinal shift to the left side. (b) Ultrasound with color Doppler of the right hemithorax showing a heterogeneous hyperechoic mass lesion with areas of color flow within the mass

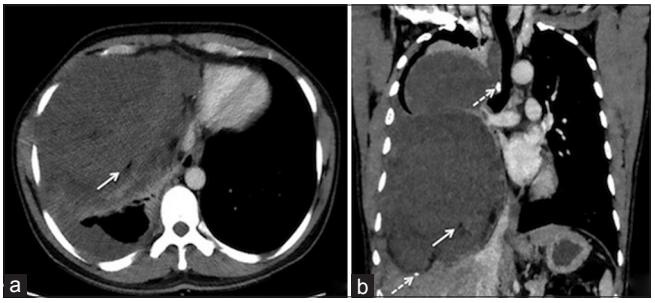

Figure 2: (a and b) Contrast-enhanced computed tomography chest, axial and coronal sections, showing a large, right pleural-based, heterogeneous, hypodense mass lesion with few fat-containing areas (solid arrows) and few calcific foci (dashed arrows). This lesion is involving the almost entire right hemithorax with the underlying collapse of the right lung and mild right pleural effusion

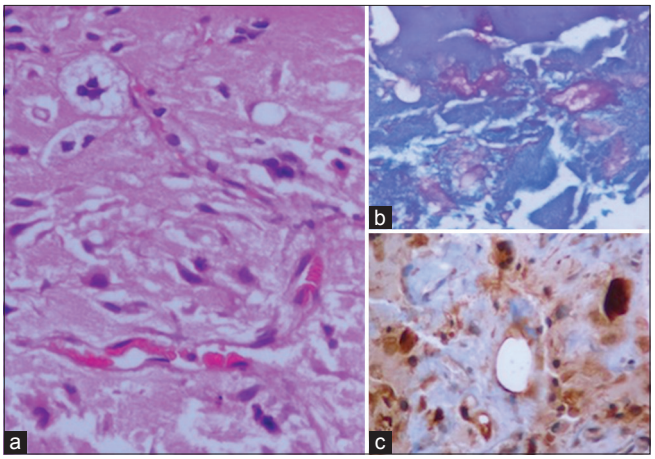

Figure 3: A panel of microphotographs showing (a) medium power view of the pleomorphic tumor comprising lipoblasts and abundant pale pink myxoid stroma (H and E, ×300). (b) Alcian blue periodic acid–Schiff staining highlighting the myxoid stroma (Alcian blue periodic acid–Schiff, ×500). (c) Microphotograph of immunohistochemistry-stained section showing the S100-positive tumor cells (peroxidase antiperoxidase, ×500)

DISCUSSION

The index case highlights the fact that PPL may grow to a large size before causing significant symptoms resulting in a late presentation. The term "intrathoracic liposarcoma" is used if it is not possible to determine the site of origin of liposarcoma on CT.[4] However, in this case, the CT scan helped in reaching the diagnosis as it revealed the pleural origin of the lesion.

Metastases are the most common malignant tumors of the pleura.[5] Primary malignant pleural neoplasms are uncommon and constitute only 10% of the pleural tumors, the most common being mesothelioma (90%). Other rare tumors that may arise from the pleura include primary pleural lymphoma and various sarcomas including liposarcoma.[6,7] PPLs are extremely rare lesions. The exact incidence is not known. Carrillo *et al*. have stated that PPL is more common in males between the age group of 19 and 80 years.[3] These produce nonspecific symptoms due to a slow growth process that causes displacement and compression of mediastinal structures.[3] Chest pain,

cough, and dyspnea are the most common presenting complaints.[8]

According to the World Health Organization classification, liposarcomas are divided into well-differentiated, myxoid, pleomorphic, and dedifferentiated subtypes, based on their complex histological components. The well-differentiated and myxoid types are the two common subtypes with the best prognosis.[6] The dedifferentiated and pleomorphic types have a poorer prognosis with local invasion and possible metastatic disease.[7,9]

The well-differentiated liposarcomas resemble lipomas on both CT and magnetic resonance imaging (MRI) and show a large amount of fat. However, fibrous septa seen in them are broader and nodular than those seen in lipomas. Myxoid liposarcoma may show atypical imaging characteristics.[10] The fat content in myxoid PPL is often <10%–25% of the tumor volume with the tumour containing predominant myxoid matrix.[11] The abundant myxoid matrix in the myxoid PPL causes diffuse hypoattenuation. These tumors also show inhomogeneous hypodensities with a lack of enhancement due to different proportions of lipoblasts, mucinous stroma, and capillary network.[11] Calcification is uncommon in the PPL, but has been reported more commonly in the myxoid type (10%). Myxoid PPL may be confused with cystic lesions or complex pleural collections on CT. However, USG with color Doppler may be useful to depict the solid nature of these tumors and the internal vascularity. On MRI, myxoid liposarcomas display a low signal on T1-weighted sequences unlike well-differentiated PPL, which shows increased signal on T1, due to the large fat component.[1] Wang *et al*. have reported CT findings similar to our case.[11] Myxoid PPL has also been reported as a large, hypodense mass without visible fat component on CT.[12] Few authors have described myxoid liposarcomas as diffusely hypodense similar to a pleural collection.[13]

The differential diagnoses of the pleural-based mass lesion with relatively chronic symptoms must include mesothelioma and other types of sarcomas. Histopathological differentials of the myxoid variant of liposarcoma include various sarcomas with myxoid changes, such as myxofibrosarcoma, myxoid variant of malignant peripheral nerve sheath tumour, myxoid solitary fibrous tumor, synovial sarcoma with myxoid changes, epithelioid hemangioendotheliomas, and desmoid tumor with myxoid changes.[14,15] The above entities are extremely uncommon in the pleura. Malignant pleural mesothelioma with myxoid changes can create diagnostic confusion. However, these tumors are cytokeratin positive and S100 negative and hence can be differentiated.[14-16]

The myxoid and well-differentiated PPLs showed 5-year survival of 71% as compared to 12.5% for the other subtypes in the series by Wong *et al*. Complete surgical excision with radiotherapy is the treatment of choice for PPL that has no radiologic signs of local invasion of the surrounding vital structures.[9] Unresectable tumors are usually treated with palliative chemotherapy and radiotherapy.[17]

The index case demonstrates the importance of considering the myxoid variant of PPL among the differential diagnoses of a low-attenuating pleural-based mass lesion with fat and calcifications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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