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Case Report

A rare case of pleuropulmonary synovial sarcoma of the chest wall: A case report and a literature review

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ABSTRACT

Pleuropulmonary synovial sarcoma is a subtype of synovial sarcomas that commonly arises from the chest wall, pleura, lungs and the heart. They are extremely rare, with only a few cases reported in the literature. It usually affects young and middle-aged adults with no gender predilection. Chest radiographs usually show a pleural-based mass, parenchymal consolidation, or a near complete opacification of the hemithorax. On contrast-enhanced CT, synovial sarcomas of the chest wall typically appear as a well-defined, heterogeneously enhancing mass with bone destruction and infiltration of chest wall musculature. MRI usually demonstrates a heterogeneous mass with areas of both high and low T1 signal intensities representing areas of hemorrhage and necrosis. We report a case of a 39- year old African male patient who presented to our hospital complaining of chest pain of 4 months duration. The plain chest radiograph showed complete opacification of the left hemithorax. Contrast-enhanced CT and MRI were then performed revealing a large left-sided heterogeneous mass. PET-CT demonstrated avid FDG uptake in the solid components of the mass with no evidence of distant metastasis.. An ultrasound-guided biopsy was performed, and histopathology revealed a locally advanced primary synovial sarcoma of the chest wall. Treatment consisted of neoadjuvant chemotherapy followed by surgical resection.

Introduction

Synovial sarcomas are malignant spindle- cell tumors that typically arise from the para-articular soft tissues of the extremities [1,2,. They do not arise from the synovial tissues as the name implies, but from multipotent mesenchymal cells [3,7]. Primary pleuropulmonary synovial sarcomas (PPSS) are a rare subtype of synovial sarcomas and account for only 0.1%-0.5% among all primary lung malignancies. Although it is a rare tumor, it should be included in the ad a as differential diagnosis of malignant chest wall tumors especially in young

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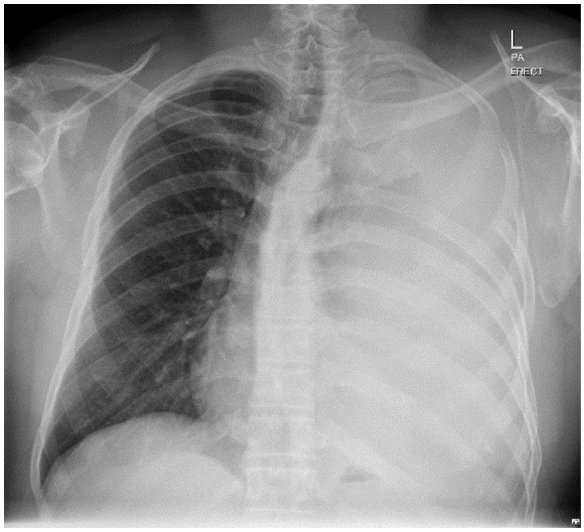

Fig. 1 – Postero-anterior chest radiograph demonstrates a near complete opacification of the left hemi-thorax with the absence of 3-6th left sided rib shadows and mediastinal shift to the right side.

and middle-age adults [2,9]. PPSS is a highly aggressive tumor with high recurrence rate and overall poor prognosis [1]. Imaging plays a crucial role in tumor characterization, distant metastasis identification, staging, presurgical planning, and treatment response assessment [2].

Case report

A 39-year old African male patient presented to our hospital with a chief complaint of left -sided chest pain localized to the posterior chest wall with associated numbness in the same region for the past 4 months. The pain had progressed gradually and worsened with breathing and exercising and did not respond to analgesics. The patient also gave a history of loss of appetite and weight loss of 7kgs over the last 4 months. He denied any history of cough, fever, night sweets, nausea, vomiting, or change in bowel habits.

He is a nonsmoker with no significant past medical or surgical history. He also denied any recent trauma to the chest. His family history was negative for cancer and for pulmonary diseases. On examination, the patient was conscious and oriented with no evidence of acute distress. Vital signs were normal. On physical exam, there was noticeable asymmetric chest wall expansion with swelling noted at the left supero-dorsal aspect. On auscultation, there was decreased breath sounds on the left side. His abdomen was soft, nontender with no organomegaly on palpation. The rest of the examination was unremarkable.

A chest radiograph was done and it showed complete opacification of the left hemithorax with mediastinal shift to the right side. In addition, the left 3rd-6th posterior ribs appeared ill-defined and obscured. The right lung was clear (Fig. 1).

Contrast enhanced CT of the chest revealed a heterogeneous mass with linear peripheral calcification causing near-total collapse of the left lung. There was also destruction of the left 3rd-6th posterior ribs, with infiltration into the adjacent para-spinal musculature. No significant hilar or mediastinal lymphadenopathy was seen (Fig. 2).

Gadolinium-enhanced MRI of the thorax showed a complex, partially solid and cystic lesion occupying most of the left hemithorax. The solid component was noted at the postero-superior aspect of the lesion and it measured approximately 12.5 × 17.5 × 13.5 cm. The mass showed areas of necrosis, hemorrhage and scattered calcifications. The mass also appeared to infiltrate and destruct the adjacent left 3rd-6th ribs posteriorly with extra-thoracic extension to left paraspinal muscles and subcutaneous tissues of the posterior chest wall causing a focal contour bulge. The solid component showed restricted diffusion on diffusion-weighted imaging (Fig. 3).

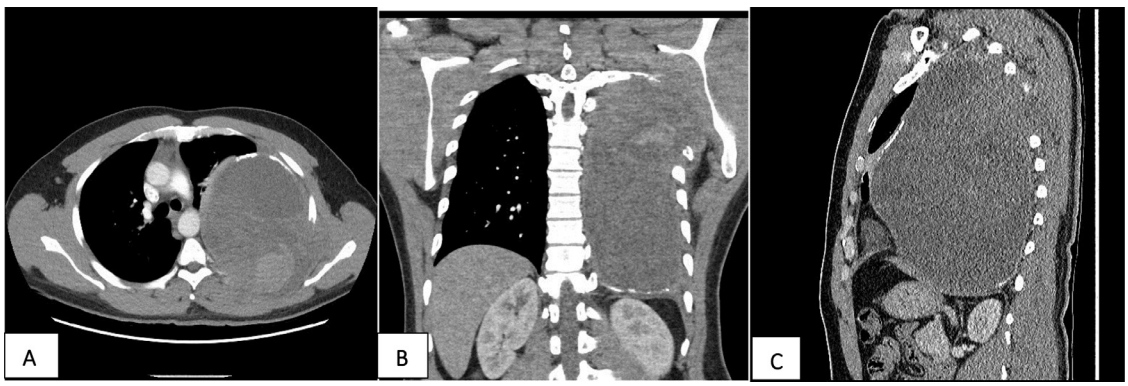

Fig. 2 – (A, B, C) Axial, coronal and sagittal contrast-enhanced CT of the chest showing a heterogeneous mass with a calcified rim. The mass is predominantly cystic with pleural- based solid enhancing component seen in the supero-dorsal aspect of the left hemi-thorax causing destruction of the adjacent 3-6th posterior ribs with infiltration of the overlying para-spinal muscles and subcutaneous tissues.

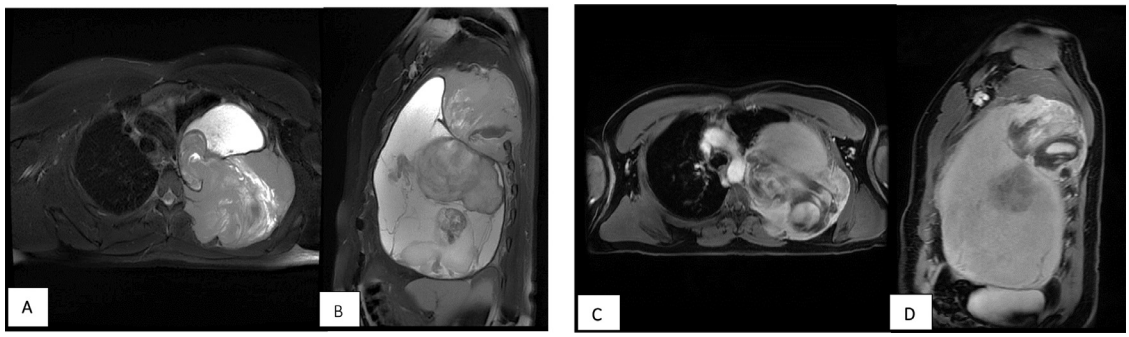

Fig. 3 – (A, B) Axial and sagittal T2 WI showing a large multi-cystic mass with solid component at the superior dorsal aspect of the left hemi-thorax(C, D) Gadolinium-enhanced axial and sagittal T1 fat saturated images showing heterogeneous enhancement of the solid component with destruction of the adjacent 3-6th left posterior ribs and infiltration of the overlying dorsal musculature.

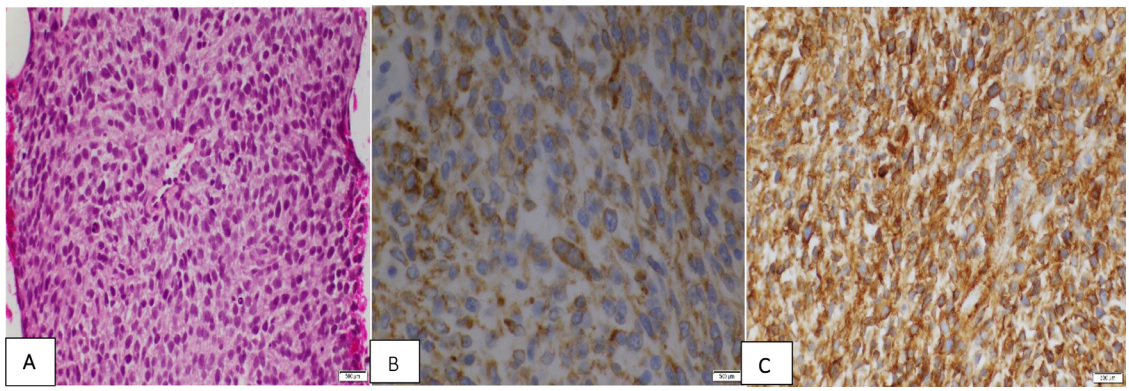

Fig. 4 – (A) Histopathology images of the heterogenous mass in x 40 magnification with hematoxylin and eosin staining showing cellular spindle cells arranged in sheets with little intervening stroma.(B,C) Immuno-peroxidase staining for BCL2 (b) and CD99(c) shows diffuse positivity in the majority of the tumor cells.

Based upon the imaging features of the heterogeneous mass a malignant neoplastic process was suspected.

Ultrasound-guided core biopsy of the solid component of the mass was obtained. Histopathology results were consistent with monophasic synovial sarcoma (Fig. 4).

The patient underwent a whole body FDG PET-CT scan, which showed intense FDG tracer uptake of the solid component of the mass with a maximum SUV of 27. Central patchy non-avid areas were seen likely representing areas of necrosis. No metastatic disease was depicted (Fig. 5).

The case was discussed in the multidisciplinary meeting and multiple cycles of neoadjuvant chemotherapy was recommended prior to surgical resection. The patient completed 6 cycles of neoadjuvant chemotherapy with Ifosfamide and doxorubicin and was referred to a specialized center for sarcoma and for reconstructive surgery.

Discussion

Synovial sarcomas are rare malignant soft tissue tumors that usually originate from the para-articular soft tissues of the upper and lower extremities [3]. They affect young and middle-aged adults with no gender predilection [1]. They are considered sarcomas as they have the sarcoma-specific chromosomal translocation. They do not arise from synovial cells, but arise from pluripotent mesenchymal cells that undergo differentiation to resemble synovial tissue on light microscopy [2,5].

Primary pleuropulmonary synovial sarcoma (PPSS is a subtype of synovial sarcomas and it usually arises from the chest wall, lungs, heart and mediastinum. PPSS represents only 0.1%-0.5 % of all primary lung malignancies [2,7,8,10]. The term pleuropulmonary was first described by Essary *et al.* [11] in reference to the difficulty to delineate the exact primary anatomical origin of the tumor from the lungs or the pleura. PPSS is a highly aggressive malignant tumor with poor prognosis and a 1-year survival rate of 55% [1,7]. Patients usually present with symptoms related to the involved structures .These symptoms include dyspnea, cough ,chest pain and back or shoulder pain [2,3].

On plain chest radiographs, PPSS may appear as a thoracic mass with sharply defined or ill-defined margins, a pleural-based mass or near-complete opacification of the involved hemi-thorax with or without associated pleural effusion [2].

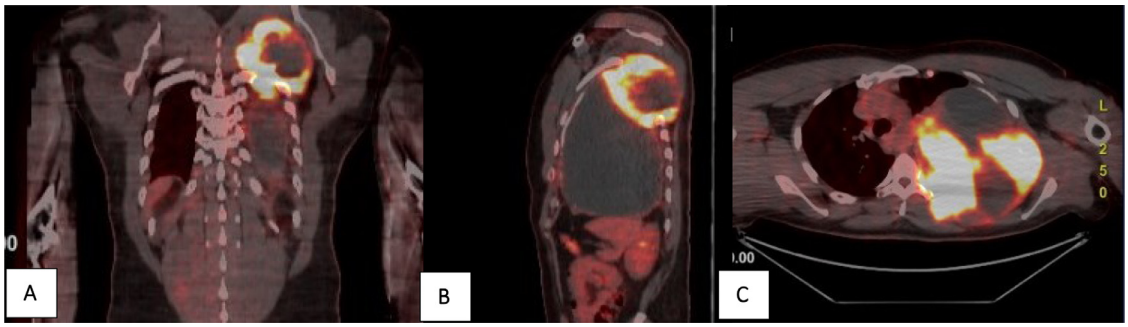

Fig. 5 – (A, B, C) Coronal, sagittal and axial FDG PET-CT images showing a thick rim of increased FDG uptake within the solid component of the mass with maximum standardized uptake value of 27.

Contrast-enhanced computed tomography most commonly demonstrates a large well-defined pleural-based mass with heterogeneous enhancement. Hypo-attenuated areas with no enhancement may represent necrosis, hemorrhage, and cystic changes [1,2,4]. Calcifications and lymphadenopathy are uncommon findings [2,4].

PET-CT imaging is performed to exclude extra-thoracic primary synovial sarcomas and for staging purposes, treatment planning and response evaluation [9]. It usually demonstrates focal increased uptake of the FDG tracer in the lesion with the absence of significant mediastinal lymphadenopathy [2,9].

Gadolinium-enhanced magnetic resonance imaging is superior to computed tomography in delineating the extent of tumor infiltration and demonstrating the nodular soft tissue and multilocular fluid -filled cystic components of the tumor. It may also depict a mass with mixed signal intensities with predominant areas of isointense signal to the chest wall on both T1 and T2 weighted sequences [2]. The presence of regions of high T1 and T2 signal intensities is consistent with hemorrhage while areas of low T1 and high T2 signal intensities correspond to necrosis [2,5]. The heterogeneous signal intensities of the lesion on T2 weighted images (high, intermediate and low signals) results in so called "triple sign'" and it is seen in 36% of soft tissue synovial sarcomas [2,12].The solid component of the tumor may show variable enhancement on post-gadolinium T1 weighted images [2].

The differential diagnosis of PPSS includes primary lung cancers, metastatic deposits to the chest, solitary fibrous tumors of the pleura, malignant mesotheliomas, other primary pulmonary sarcomas, malignant nerve sheath tumors, and hemangiopericytomas [1,8,11]. Imaging alone cannot distinguish the subtypes of sarcomas, hence histopathological correlation is essential for a more definitive diagnosis [11].

Histologically, the tumor can arise from 2 types of cells: epithelial and fibroblast-like spindle cells. Based on the cell of origin, the tumor can be classified into 4 different subtypes: biphasic, monophasic- fibrous, monophasic- epithelial, and poorly differentiated tumors [7].

There is no gold standard treatment of PPSS, a multidisciplinary management approach that includes surgical resection for local control followed by chemotherapy or radiotherapy is usually recommended [2,6]. Synovial sarcomas show 50% response rate to Ifosfamide and doxorubicin [2]. Careful follow up is mandatory since they commonly recur [2].

In conclusion, we presented a case of a 39-year-old African male patient diagnosed with a pleuropulmonary synovial sarcoma arising from the left aspect of the posterior chest wall. The radiological findings are characterized by a multicystic mass with a heterogeneously enhancing soft-tissue component infiltrating the posterior chest wall best appreciated on CT and MRI. PET-CT demonstrated avid tracer uptake in the solid component of the lesion with no distant metastasis. The management of pleuropulmonary synovial sarcomas consist of surgical resection followed by chemotherapy and/or radiotherapy. The prognosis remains unfavorable.

Consent

An informed consent was obtained from the patient for publication of this case and any accompanying images.

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