Primary Pleuropulmonary Synovial Sarcoma on Fluorodeoxyglucose Positron Emission Tomography-Computed Tomography Scan

Abstract

Primary pleuropulmonary synovial sarcoma, a mesenchymal tumor of lung and pleura, is very rare and highly aggressive condition among the primary lung malignancies. As role of fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) has been established in lung malignancies in terms of staging, restaging, biopsy guidance, and treatment response evaluation, there is also role of FDG PET-contrast-enhanced CT (CECT) to raise suspicion or increase confidence in reporting of sarcomatous lung malignancy by studying characteristics of CECT scan features. We present a case of a 57-year-old female patient having large lung mass, who underwent FDG PET-CT scan and findings raised strong suspicious of noncarcinomatous pattern of lung mass and may have sarcomatous primary lung malignancy which was later proven on histopathological and immunohistochemistry report.

Keywords: *Contrast-enhanced computed tomography scan, fluorodeoxyglucose, positron emission tomography-computed tomography, primary pleuropulmonary synovial sarcoma*

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Introduction

Soft tissue sarcoma is rare in incidence, comprises <1% of all malignancies, among which synovial sarcoma is 5%–10%. Peak incidence is in third to fifth decades and predominant in males.[1] Synovial sarcoma usually arises from para-articular tissues of extremities but in rare incidences develops from abdominal wall, intestines, retroperitoneum, chest wall, lung, pleura, mediastinum, heart, neck, and esophagus.

Different subtypes of primary lung sarcoma have been identified such as leiomyosarcoma, fibrosarcoma, and synovial sarcoma.[2] Primary pleuropulmonary synovial sarcoma (PPSS) is very rare in incidence; only 0.1%–0.5% among all primary lung malignancies, very aggressive with frequent recurrences, and metastases. Surgical removal is the best treatment, and response to chemotherapy and radiotherapy is poor.

Case Report

A 57-year-old female patient with complaints of difficulty in breathing, right side chest pain and back pain. There was no history of lesion or surgery involving extremities. Her chest X-ray showed left lung opacification and lesions in the right lung and was suspected to have malignancy and referred for fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) scan. The patient was taken for scan after 60 min of 296 MBq 18-F-FDG intravenous injection. Due to her complaint of severe orthopnea, a noncontrast PET-CT scan was done on Siemens Biograph True Point 16 slice CT scanner. Contrast CT study was done after stabilizing the patient.

Combined noncontrast PET-CT scan and contrast-enhanced CT (CECT) assessment showed a very large lesion almost occupying left hemithorax with complete residual lung collapse, causing mediastinal and left hemidiaphragm displacement. The lesion had well-defined margins with internal necrotic areas, inhomogeneous low attenuation areas, and ill-defined areas of mild enhancement within. The lesion had a broader pleural base without chest wall invasion. Lesion showed heterogeneous high-grade FDG uptake in contrast-enhancing areas. Similar characteristic moderate-sized peripheral lesion with high-grade homogeneous FDG uptake was seen in the right middle lobe, and two intrapulmonary non-FDG-avid lesions were seen in the right lung upper

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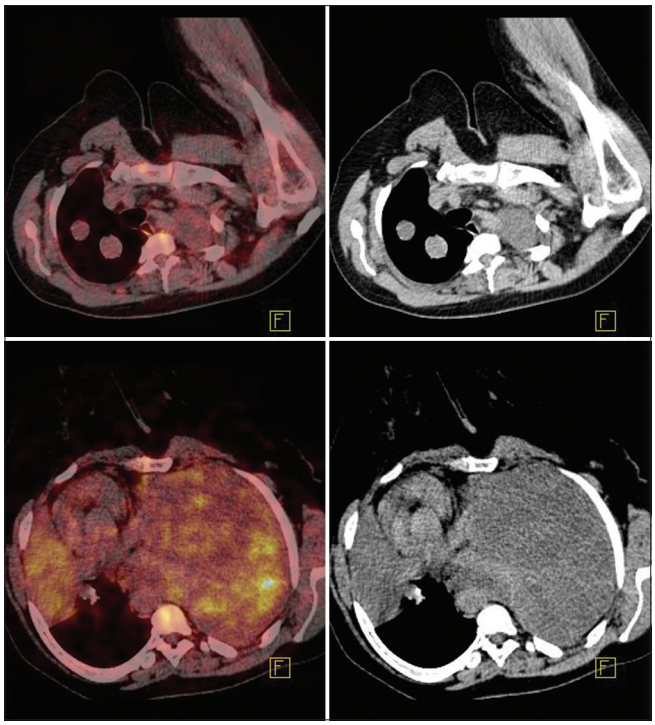

Figure 1: Positron emission tomography-computed tomography fusion and plain computed tomography scan axial images show large lesion with inhomogeneous density and internal necrotic areas and heterogeneous fluorodeoxyglucose uptake completely occupying left hemithorax with displacement of mediastinum. Similar fluorodeoxyglucose-avid right middle lobe and nonfluorodeoxyglucose-avid two right upper lobe lesions

lobe [Figures 1 and 2]. Minimal pleural effusion was seen in the left apical lung. Low-grade FDG-avid right paratracheal node was seen.

In view of CT scan characteristics [Figure 3], the lesion was suspected of sarcomatous etiology rather than lung carcinoma. Her ultrasonography-guided biopsy was done from FDG-avid part of lesion, and report was conclusive of primary PPSS FNCLCC Grade 2.

Discussion

Primary PPSS is a rare aggressive malignant mesenchymal spindle cell tumor commonly presenting with chest pain, cough, shortness of breath, hemoptysis, or ipsilateral pleural effusion.[3]

Metastatic lung sarcoma is more common than primary. Hematogenous spread from nonpulmonary primary sarcoma to the lung is the most common route of spread and lung is the most common site for metastasis. Hence, while dealing with primary PPSS, clinical examination and whole body scanning (e.g., PET-CT scan) are essential to rule out the possibility of lung metastasis from other primary site.[4]

Careful assessment of CT characteristics is essential to raise suspicious or confirmation of sarcomatous over carcinomatous tumor. CECT scan usually shows homogeneously enhancing or heterogeneously enhancing mass with predominant low attenuating and necrotic areas

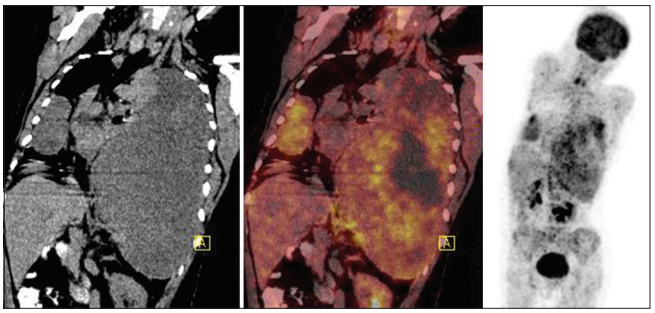

Figure 2: Positron emission tomography-computed tomography fusion and plain computed tomography scan coronal images with maximum intensity projection image of positron emission tomography scan show large lesion completely occupying left hemithorax with displacement of diaphragm and mediastinum. Similar lesion in the right middle lobe

and sometimes rim-enhancing areas within.[5,6] Areas of fluid-attenuating necrotic or hemorrhagic components may also be present. As in our case, large lesion involving hemithorax with inhomogeneous low attenuation areas and areas of mild enhancement, fluid-attenuating necrotic component may be seen.

Series of Frazier *et al*. reported wide pleural-based localization of lesion, well-delineated borders without chest wall invasion.[4] Ipsilateral pleural effusion is often present which may represent hemothorax.[5-8]

Kim *et al*. reported that most of the tumors presented as solitary (78.6%), large circumscribed masses (85.7%) with mass effect, often with calcifications and heterogeneous attenuation. Hypoattenuated regions with no enhancement corresponded to hemorrhagic, necrotic, cystic, or myxoid components of pathologic findings. Well-enhancing solid tissue components with high attenuation and intratumoral enhancing vessels were very common internal features (92.9%). Frequent extrapleural fat or chest wall extensions (64.3%) without bony involvement was seen. The tumors showed a propensity to rupture in pleural space, which resulted in hemothorax or pleural effusion (50.0%). However, hilar or mediastinal lymphadenopathy was very rare. Most common anatomic location was lung (42.9%).[9] Because of the large size of the lesion, its origin may be uncertain and appear to be located within lung or pleura.[4]

In our case, intralesional and interlesional heterogeneity in FDG uptake in left lung primary lesion and contralateral lung metastatic lesions without extrapulmonary lesion confirmed it to be primary PPSS.

Polverosi *et al*. reported primary PPSS cases presenting with high-grade FDG-avid inhomogeneous density big masses in lungs, pleural effusion, and absence of significant mediastinal lymphadenopathy. They also emphasized the contribution of PET/CT for differentiation of primary from metastatic synovial sarcoma.[10]

Harisankar *et al*. reported one case of primary PPSS with high-grade FDG uptake.[11] Won *et al*. reported one case with non-FDG-avid large necrotic primary PPSS with

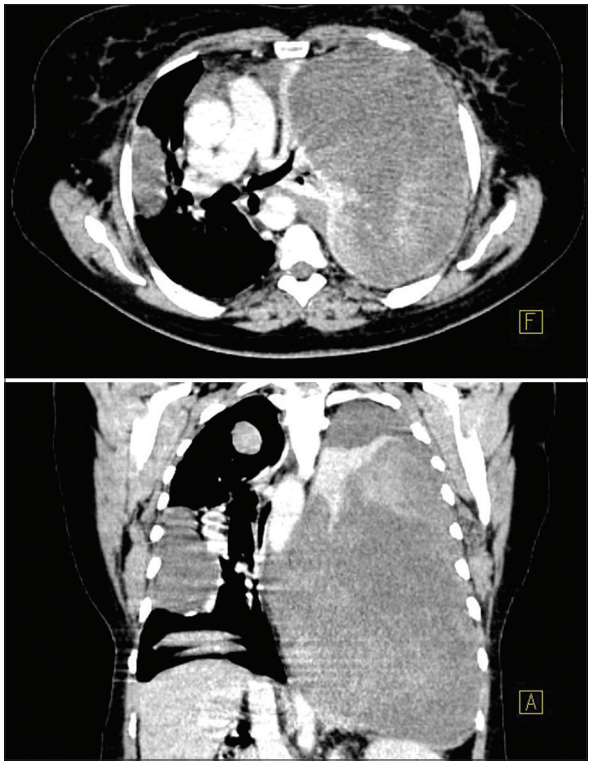

Figure 3: Postcontrast computed tomography scan axial and coronal images show large well-defined lesion in the left hemithorax with collapsed lung medially and smaller similar lesions in the right middle lobe with inhomogeneous mild contrast enhancement

cardiac metastasis. Hence, variable degree of FDG uptake in lesion is found among reported cases primary PPSS.[12]

Conclusion

As rarity of primary PPSS, FDG PET-CECT may act as first line imaging modality of choice and helps to guide biopsy site selection for definite pathological characterization of lung mass, differentiate primary versus metastatic pulmonary malignancy, staging, treatment planning, response evaluation, and follow-up to early localization of metastatic spread. Even though primary lung cancer is indistinguishable from sarcoma on plain CT scan, CECT component of PET-CT scan may play an important role to differentiate sarcoma and carcinoma.

Size (>5 cm) and extensive tumor necrosis are few of the poor prognosticators of PPSS, and so, FDG PET-CECT may have role in prognostication of primary PPSS.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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