

Spindle cell sarcoma of pulmonary artery mimicking thromboembolism with lung metastasis detected in fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography

Koramadai Karuppusamy Kamaleshwaran, Pattabiraman VR¹, Sangita Mehta², Vyshakh Mohanan, Ajit Sugunan Shinto

Departments of Nuclear Medicine and Positron Emission Tomography/Computed Tomography, ¹Pulmonology, and ²Pathology, Kovai Medical Centre and Hospital Limited, Coimbatore, Tamil Nadu, India

ABSTRACT

Pulmonary artery sarcoma (PAS), although rare, must be considered in the differential diagnosis of pulmonary thromboembolism (PTE). This tumor is highly malignant and the prognosis is very poor. As much as the standardized uptake values (SUVs) at fluorine-18 fluorodeoxyglucose positron emission tomography (18F-FDG PET) have helped in differentiating between benign and malignant tumors, visualization of a low-attenuation filling defect within a pulmonary artery on contrast-enhanced chest computed tomography (CT) can be suggestive of a malignancy, such as PAS, if the lesion shows high FDG uptake at PET. We present a case of PAS that showed high FDG uptake on integrated FDG PET/CT and with lung metastasis. Patient underwent endoscopic bronchial ultrasound-transbronchial needle aspiration (EBUS-TBNA), which confirmed spindle cell sarcoma.

Keywords: 18-fluorodeoxyglucose, uptake, pulmonary artery, spindle cell sarcoma, thromboembolism

INTRODUCTION

Pulmonary artery sarcoma (PAS) is a rare malignancy arising from the mesenchymal cells of the intima of the pulmonary artery.^[1] It is frequently misdiagnosed as pulmonary thromboembolism (PTE), although chest computed tomography (CT) can help differentiate PAS from PTE by showing a low-attenuation filling defect occupying the entire lumen of the proximal or main pulmonary artery, expansion of the involved arteries, or extraluminal tumor extension.^[2-3] Therefore, a new noninvasive technique for the diagnosis of PAS is needed to avoid a delay in diagnosis and in treatment and to prevent unnecessary surgery. Fluorodeoxyglucose positron emission tomography (FDG PET) have helped to differentiate between benign and malignant

tumors.^[4] In our case, FDG PET showed increased uptake in the pulmonary artery lesion which confirmed on biopsy as PAS.

CASE REPORT

A 58-year-old female patient presented with recurrent attacks of syncope. CT chest showed focal hypodense filling defect in right pulmonary artery measuring 3.8 × 2.9 × 2 cm which extends extraluminally abutting left atrium. Eccentric filling defect in left pulmonary artery was also noted. Magnetic resonance imaging (MRI) showed enhancement of the filling defect within right pulmonary artery. With the suspicion of PTE or PAS, she was referred for whole body PET/CT. PET/CT showed an intense uptake in the hypodense filling defect [Figure 1a-d] with a standardized uptake value (SUV_{max}) of 11. Also scattered subcentimetric pulmonary nodules was noted, largest measuring 8 mm in right upper lobe [Figure 1e]. Patient underwent endoscopic bronchial ultrasound-transbronchial needle aspiration ((EBUS-TBNA) which confirmed spindle cell sarcoma of pulmonary artery [Figure 2]. She was advised surgery and lost to follow-up.

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DOI:
10.4103/0972-3919.142631

Address for correspondence:

Dr. Koramadai Karuppusamy Kamaleshwaran, Department of Nuclear Medicine, Positron Emission Tomography/Computed Tomography and Radionuclide Therapy, Comprehensive Cancer Care Centre, Kovai Medical Centre and Hospital Limited, Coimbatore - 641 014, Tamil Nadu, India. E-mail: dr.kamaleshwar@gmail.com

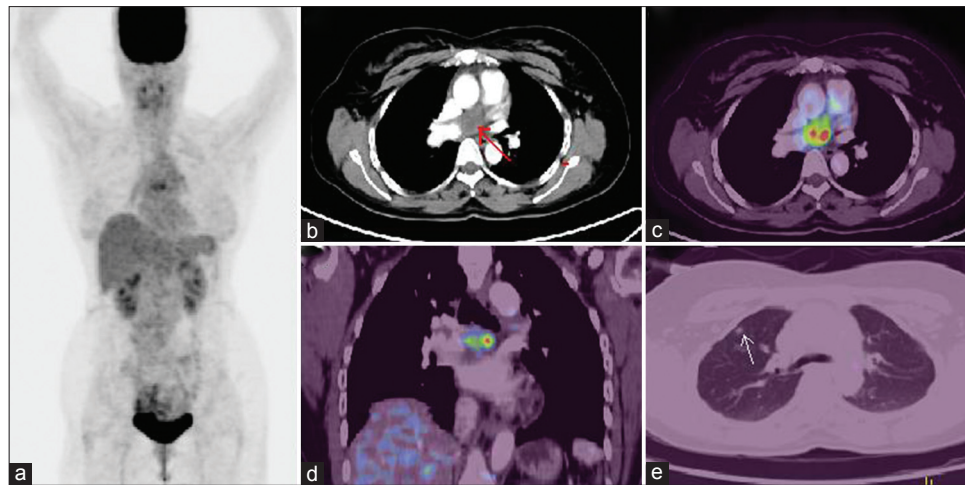


Figure 1: Whole body fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) maximum intensity projection image (a), axial CT (b) showing hypodense filling defect in pulmonary artery, axial fused PET/CT (c), and coronal fused PET/CT (d) showing uptake in pulmonary artery lesion and axial fused PET/CT (e) showing lung nodules in both the lungs suggestive of metastasis

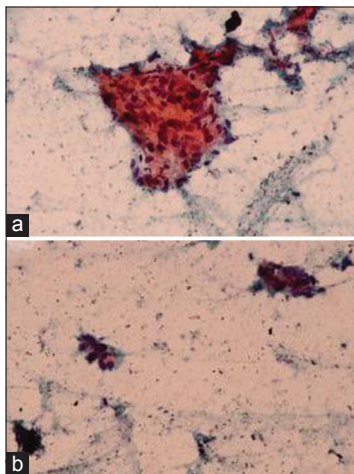


Figure 2: (a and b) Histopathological examination revealed small tiny cluster of atypical spindle cells (hematoxylin and eosin stain; original magnification ×100)

DISCUSSION

PAS, first described by Mandelstamm in 1923, is a rare and potentially lethal tumor, usually diagnosed during surgery or autopsy.^[1] It is thought to arise from multipotential mesenchymal cells of the intima and generally arises from the right, left, and main pulmonary artery. The reported age ranges from 13 to 86 years with the majority of cases occurring in the middle age. Although some studies revealed female predominance, more recent data suggest no gender preference.^[2]

Clinical symptoms are generally nonspecific and often occur at the end stage of the disease. Frequent symptoms are dyspnea, chest pain, hemoptysis, weight loss, malaise, cough, syncope, and fever.^[3] Our patient also presented with syncope as reported by Treisman *et al.*^[5] Although PAS is a rare tumor, the true prevalence is underestimated as many cases of PAS are misdiagnosed as PTE leading to inappropriate therapy. Both diseases appear as intraluminal filling defects in the pulmonary artery system on

contrast-enhanced CT scans and differentiating radiologically between PAS and PTE is relatively difficult. However, over the last decades, improvement of imaging techniques has allowed to distinguish the two entities before surgery. Yi *et al.*, reported that CT findings favoring the diagnosis of PAS included a low attenuation filling defect occupying the entire luminal diameter of the proximal or main pulmonary artery, expansion of the involved arteries and extraluminal tumor extension as shown in our case.^[3] In addition, PAS has more likely a heterogeneous appearance with areas of necrosis, hemorrhage, and ossification. It occurs frequently unilaterally, in contrast with the often bilateral involvement of pulmonary arteries in the thromboembolic disease.

PET-CT may help to make a differential diagnosis between PAS and PTE.^[6,7] According to the several case reports of FDG-PET, blood thrombi showed negative FDG uptake; whereas, a malignant tumor such as sarcoma showed positive FDG uptake.^[8,9] FDG-PET may show intense tracer activity within a PAS. While, some tracer activity within a thromboembolic disease may occur, intense tracer localization within a central pulmonary arterial filling defect strongly suggests primary pulmonary arterial malignancy. We think that the positive FDG uptake on PET-CT in the current case supports this suggestion. The cornerstone of the treatment consists of surgical resection through endarterectomy, tumorectomy, or pneumectomy. Surgical resection remains the primary treatment to provide significant palliation and offers the only chance of cure.^[10] Our case demonstrates 18F-FDG PET/CT uptake in PAS and also lung metastasis.

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How to cite this article: Kamaleshwaran KK, Pattabiraman VR, Mehta S, Mohanan V, Shinto AS. Spindle cell sarcoma of pulmonary artery mimicking thromboembolism with lung metastasis detected in fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography. *Indian J Nucl Med* 2014;29:249-51.

Source of Support: Nil. **Conflict of Interest:** None declared.

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