Case report:

Pulmonary artery tumour masquerading as massive pulmonary embolism

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Abstract

An elderly male presented with acute shortness of breath while performing a treadmill test; he was detected to have pulmonary hypertension and subsequent contrast enhanced computeri ed tomography (CECT) chest revealed a soft tissue filling defect of the pulmonary trunk and the main branches appearing as a massive thromboembolism. However, the patient remained hemodynamically stable. Subsequent evaluations failed to substantiate embolism but diagnosed a pulmonary artery tumour that masqueraded as massive pulmonary embolism. The report summarises the clues to differentiate between pulmonary embolism due to clot and pulmonary artery tumour (angiosarcoma).

Ke words: CECT chest, pulmonary angiosarcoma, FDG-PET scan, pulmonary thromboembolism

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ABBREVIATIONS:

CECT: contrast enhanced computerized tomography

FDG-PET: fluro-deoxy-glucose positron emission tomography

PASP: pulmonary artery systolic pressure

INTRODUCTION:

Pulmonary vascular tumours are very rare entities. Here, we present a case of pulmonary artery tumour with predominant intra luminal presence. The tumour appeared as massive thromboembolism in contract enhanced computerized tomography and the patient had presented with acute shortness of breath on exercise.

THE CASE:

BC, 64 years old and an apparently asymptomatic male, could not perform the treadmill test due to acute shortness of breath in an annual health check-up programme. 2D-Echocardiography with Doppler analysis revealed a pulmonary artery systolic pressure (PASP) of 80 mm of Hg. He was suspected to have a pulmonary embolism and was started on heparin when he refused hospitalization. The gentleman had no significant past, personal, family, and treatment history except the presence of occasional cough during the change of seasons. He was admitted for relapse of acute central pain chest and shortness of breath after two weeks. The work-up for acute myocardial injury (ECG, troponin-T test and cardiac enzymes) was negative, Doppler echocardiography measured the PASP as 40mm of Hg. A contrast enhanced high resolution (HR) CT chest showed a fairly bulky looking filling defect with variegated appearance occupying the pulmonary trunk and both the right and left main pulmonary arteries (see picture-1). A tentative diagnosis of acute pulmonary embolism was made and the patient was evaluated for the source and cause of embolism but investigations were non-contributory (table-1). Interestingly, despite such extensive lesion, the patient did not have any hemodynamic compromise and the pulmonary trunk did not look wider than the adjacent aorta on the CT pulmonary angiogram, which was unexpected in a person with a high clot load. (picture-1) Additionally, the HRCT chest displayed a) left basal pleural thickening with two small pleural based nodules on the left lower pleural surface (picture-2a) and b) alveolar densities in the right middle lobe (picture-2b). The patient was anti-coagulated with heparin and thereafter continued on warfarin. He was also given antibiotics that resulted in no change in the alveolar densities on repeat selected CT Chest cuts after 10 days. The CT guided aspiration from the pleural based nodules yielded no evidence of malignancy. Now pulmonary thrombo-endarterectomy was planned and prior to that a PET CT was ordered to rule out malignancy at any part of the body, as evaluation for a risk factor for an unprovoked pulmonary embolism. To our surprise, while the pleural based nodules were not found PET avid, the so called pulmonary emboli had very strong FDG PET uptake with a central filling defect (see picture 3) at the trunk with extra-luminal extension suggesting a malignant process masquerading as pulmonary emboli.

The patient, while on preparation for further therapy, had one more episode of severe shortness of breath at his residence and passed away within minutes.

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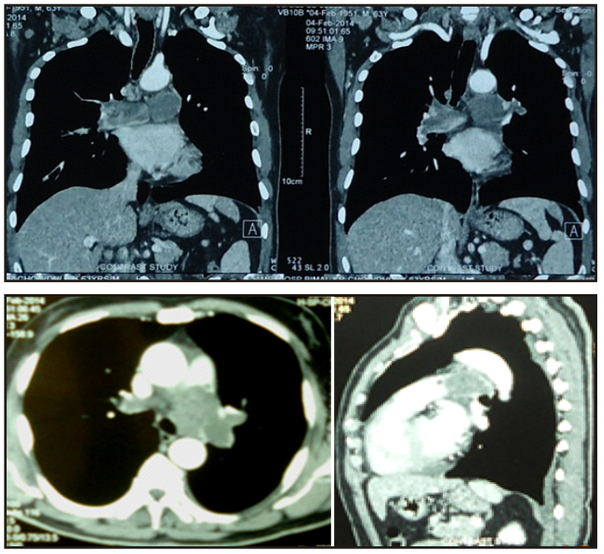
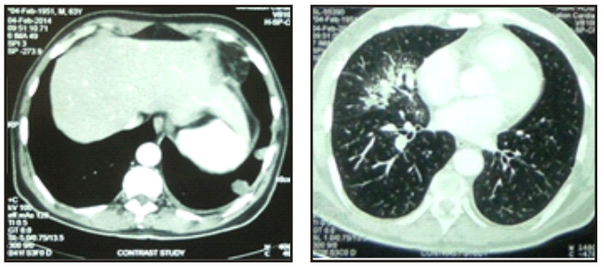
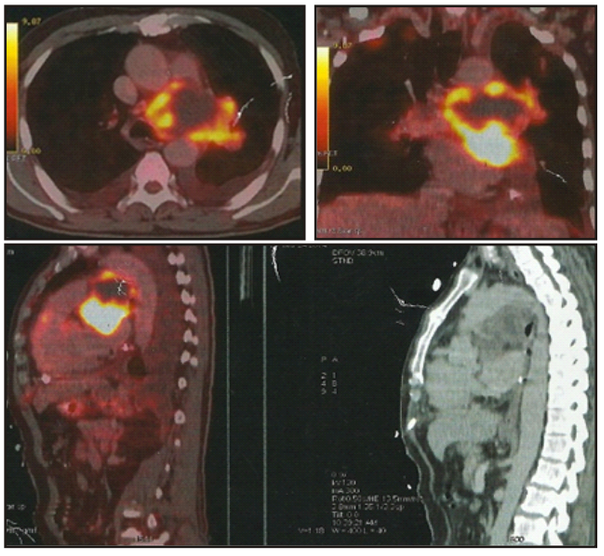


Picture 1: contrast enhanced CT thorax showing near complete filling of the distal pulmonary trunk and the pulmonary artery branches without proximal dilatation of the pulmonary trunk in different views.



Picture 2: Coronal CECT sections of chest: the picture on left shows multiple left sidedpleural based nodules and that on the right shows clusters of flame shaped alveolar densities in the middle lobe.



Picture 3: the FDG PET CT cuts that show the FDG avid lesion in the pulmonary trunk and the main pulmonary arteries with a metabolically inactive central part of the lesion and extra-luminal spread (obvious in the lateral sagittal sections being marked by black arrow).

Investigations	Result
Routine haemogram and biochemistry	Normal
Duplex doppler of the lower limb veins	Normal
Ultrasonography of abdomen	Normal
Protein C and protein S	Normal
Serum homocysteine	Normal
HIV serology (HIV I and II)	Normal
ANA, APLA, and anticardiolipin antibody	Not raised
CT guided FNAC from the pleural based lesions	No malignancy
Echocardiography twice (initially at executive check up and later when hospitalized)	Pulmonary Hypertension (PA Pressure: 80 mm at active symptoms and 40 mm later)
HRCT chest (with contrast) with CT angiography	Abnormal (described)
FDG PET scan	Abnormal (described)

DISCUSSION:

The case elaborates an example of pulmonary artery tumour masquerading as a pulmonary embolism and, therefore, the patient was anticoagulated with heparin and warfarin, and evaluated for the source and cause of embolism (see table 1). Such a presentation of a pulmonary angiosarcoma has been reported [1,2]. Although we missed the diagnosis in the first instance, multislice computed tomographic scanning has been effectively used as a diagnostic tool for pulmonary artery tumours. [3]

We noticed some odd features for massive pulmonary embolism such as (a) lack of any hemodynamic compromise (b) pulmonary artery pressure not high enough (especially the second measurement as 40 mm of Hg.) to commensurate with the appearance of massive embolism in CT angiography and (c) bulky looking clot appearing to almost expand the blocked arteries, but a possibility of a PA tumour was not entertained. Subsequently, on review, it appeared that the so-called clot like density has some variegated appearance and there is possible extra-luminal extension of the pathology. The extra-luminal extension could have been better defined by a Gadolinium enhanced magnetic resonance imaging which is also reported as a sensitive tool to differentiate between thrombi from tumours (in situ angiosarcomas or tumor emboli) [4] and the same could elaborate the heterogeneity in the appearance. [3] However, subsequent fluro-deoxy-glucose PET (positron emission tomography) done to unfold the mystery of the filling defects in the pulmonary artery and the pleural based nodules suggested that the thrombus like pathology in the pulmonary trunk and the proximal major pulmonary arteries was a tumour by nature. The FDG PET also demonstrated the extraluminal extension of the tumour (see picture-3).

Pulmonary artery tumours are rare entities. They are essentially sarcomas with several case reports described. [5, 6, 7] Most of the times, the tumours involve the right ventricular outflow tract and the pulmonary trunk although they often extend into the major pulmonary arteries. [8] Pulmonary sarcoma can be intra-luminal or extra-luminal; the former being more common and is usually constituted of undifferentiated

spindle cells; [9] leiomyosarcoma is the commonest variety of intra-luminal pulmonary artery sarcoma. In our patient, the tumour, though appears intra-luminal, has an extra-luminal component in contrast CT scan and also in the PET scan images. However, it is more likely to be intra-luminal to start with as the major bulk of the tumour and its extension remains intra-luminal only (see picture-1). It is not possible to contemplate the histological nature as we have no tissue sample of the lesion with us. Pulmonary artery sarcoma in CT image may look like an embolus or a mediastinal mass [2] and our patient had both the pictures. We could not explain the bunch of multiple flame shaped alveolar opacities in the right middle and lower lobes that remained unchanged with antibiotics (see picture-2b). This could possibly have developed from the tumour embolism. The left basal pleural based nodules could be from old pleural inflammation as they were PET negative and were associated with adjacent pleural thickening.

The pulmonary artery tumours commonly present like pulmonary embolism [1, 2] and they are often treated by anticoagulant therapy [10]; the actual diagnosis has often been made on post mortem examination [8]. The patients usually present with nonspecific symptoms such as dyspnoea, chest pain, and cough. Earlier literature reported that in 60% of cases, the diagnosis of tumour was made at autopsy.[2] The survival has been reported to be in months (2). The mean survival period has been reported to be 1.5 months without surgery and approximately 12 months with resection. [7]

In our case there was no hemodynamic compromise despite the lumen of the pulmonary artery being virtually occluded and there was no pulmonary trunk dilatation. This suggests that the process of the pulmonary artery and branch occlusion was a slow one allowing time for a collateral circulation to develop in order to compensate for the physical compromise of the pulmonary flow. The PET CT showing a filling defect in the central part of the pathology suggest that there could have been an additional clot at the site or that the tumour could have a necrosis within itself. Without a biopsy, it remains conjecture only. The cause of death is similarly unclear.

CONCLUSION:

The clues to the presence of a pulmonary artery tumour (pulmonary angiosarcoma) lie in [1] the discrepancy between the apparent large clot load and the lack of hemodynamic compromise [2] the apparently expansile appearance of the clot, which is not seen in pulmonary emboli and [3] presence of distal alveolar opacities, again different from wedge shaped pulmonary infarcts that may be seen in pulmonary embolism. The diagnosis may be confirmed by PET-CT scan which demonstrates FDG-avid intra-arterial lesions, or by MR angiography. Final confirmation may also be obtained at biopsy and the tumour may be identified only when the material obtained at pulmonary endarterectomy is sent for histopathological examination.

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