A Rare Case of Multicentric Primary Pulmonary Artery Sarcoma: Eliminating the Masquerade with Multimodality Imaging

Kavya S. Kaushik[1]   Chandresh Karnavat[1]   Ritu Kakkar[1]   Shrinivas Desai[1]

[1] Department of Radiology, Jaslok Hospital and Research Centre, Mumbai, Maharashtra, India

Address for correspondence Kavya S. Kaushik, DNB, Department of Radiology, Jaslok Hospital and Research Centre, Mumbai, Maharashtra, India (e-mail: kavya.k91@gmail.com).

Abstract

A 68-year-old male presented with a short history of exertional dyspnea and a provisional diagnosis of pulmonary thromboembolism was made. However, chest radiograph and further investigations in the form of computed tomography pulmonary angiogram, magnetic resonance imaging of thorax, and whole body fluorodeoxyglucose (FDG) positron emission tomography-computed tomography revealed a large mass arising from the distal left pulmonary artery extending into adjacent lung and another lesion near the root of the main pulmonary artery, both of which showed post-contrast enhancement and intense FDG uptake. Tissue sampling by transthoracic computed tomography-guided biopsy and immunohistochemistry confirmed the diagnosis of pulmonary artery angiosarcoma. Here, we present such a case of very rare occurrence which, in view of multicentricity and substantial extension into adjacent lung, is the first of its kind to be reported, to the best of our knowledge.

Keywords
► pulmonary artery sarcoma
► sarcoma
► angiosarcoma
► wall eclipsing sign
► PAS

Key Message

Multimodality imaging approach should be made use of in the setting of an atypical clinical presentation to arrive at the diagnosis of pulmonary artery sarcoma and thereby initiate prompt treatment.

Introduction

Primary pulmonary artery sarcoma (PAS) is a rare disease with poor prognosis, having an incidence of ~0.003%, with only close to 400 cases having been reported in literature till date. Angiosarcomas form ~3.6% of all the histological subtypes. They have an atypical clinical presentation and, more commonly, mimic pulmonary thromboembolism, thereby implicating the essential role of multiple noninvasive imaging modalities in distinguishing them and aiding in their

early diagnosis. The treatment consists of surgical resection and chemotherapy. Here we report a rare case of multicentric primary pulmonary artery angiosarcoma with substantial extension into lung.

Case Presentation

A 68-year-old male presented with 2 weeks history of breathlessness on exertion and weight loss of approximately 10 kg over the last 2 years. There were no complaints of chest pain, palpitation, or fever. He was a known diabetic and hypothyroid, and was on regular medications. On examination, vitals were stable. Blood investigations were normal except for an elevated C-reactive protein (75 mg/L).

The clinical diagnosis was of pulmonary thromboembolism (PTE). However, the chest radiograph revealed a well-defined opacity in the left parahilar region (►**Fig. 1**).
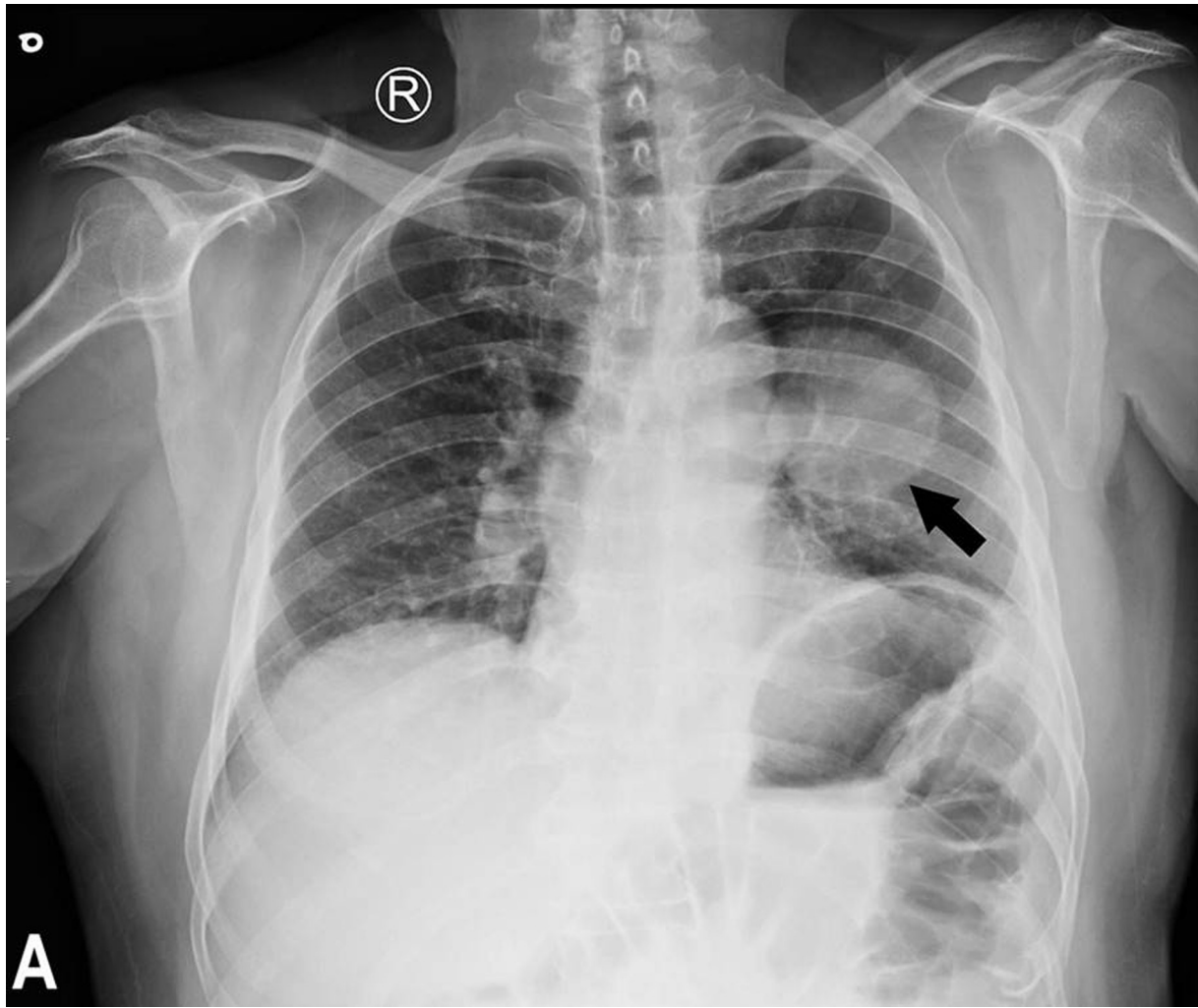
Fig. 1 Frontal chest radiograph shows a well-defined rounded opacity in the left parahilar region (black arrow).

Computed tomography (CT) pulmonary angiogram revealed a well-defined, 5.6×5.3 cm sized rounded lesion in the superior segment of the left lower lobe, just extending into the apicoposterior segment of the left upper lobe of lung causing complete obliteration of the left pulmonary arterial tree from distal left pulmonary artery onward. There was patchy, eccentric, minimal enhancement of this lesion (Hounsfield Unit [HU] of 21 on plain and 41 on contrast) (►**Fig. 2**). Small ground glass nodules were seen in the inferior aspect of this lesion. A focal filling defect sized 2.7×1.3 cm was seen at the root of the main pulmonary artery, which also revealed enhancement (HU of 25 on plain and 61 on contrast) (►**Fig. 2**). The main pulmonary artery was dilated, suggestive of pulmonary hypertension. There was mild volume loss of the left lung with oligemia (►**Fig. 2**). With these findings, the possibilities considered were primary pulmonary artery sarcoma (PAS), primary lung malignancy, and thrombosed pulmonary artery aneurysm.

In view of the conflicting nature of the lesion, magnetic resonance imaging (MRI) was done. The lesion was hypo-isointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images (T2WI) with central hypointensity on gradient recalled echo images and mild

restricted diffusion (►**Fig. 3**). There was enhancement in the central and medial part of the lesion on arterial phase with progressive diffuse enhancement of the lesion on delayed phases. The lesion at the root of the pulmonary artery, just distal to the pulmonary valve, appeared mildly hyperintense on T2WI and showed subtle post-gadolinium enhancement (►**Fig. 4**).

On further evaluation with fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT), the lesion revealed heterogeneous intensely increased FDG uptake with SUV_{max} of 10.7. Central necrotic areas with no FDG uptake were seen. Moderately increased FDG uptake was seen in the filling defect at the root of the pulmonary artery with SUV_{max} of 6.3 on the baseline images that increased to 9 on the delayed scan (►**Fig. 5**). No other FDG avid area was seen. These findings were highly suggestive of neoplastic etiology, most likely a multicentric angiosarcoma.

To confirm the same, a CT-guided transthoracic biopsy of the larger lesion was done under local anesthesia by posterior approach and sent for histopathological analysis (►**Fig. 6**). The microscopy of the biopsy cores revealed high-grade spindle cell sarcoma, showing cytologic atypia, mitotic figures, and foci of necrosis. Immunohistochemistry showed
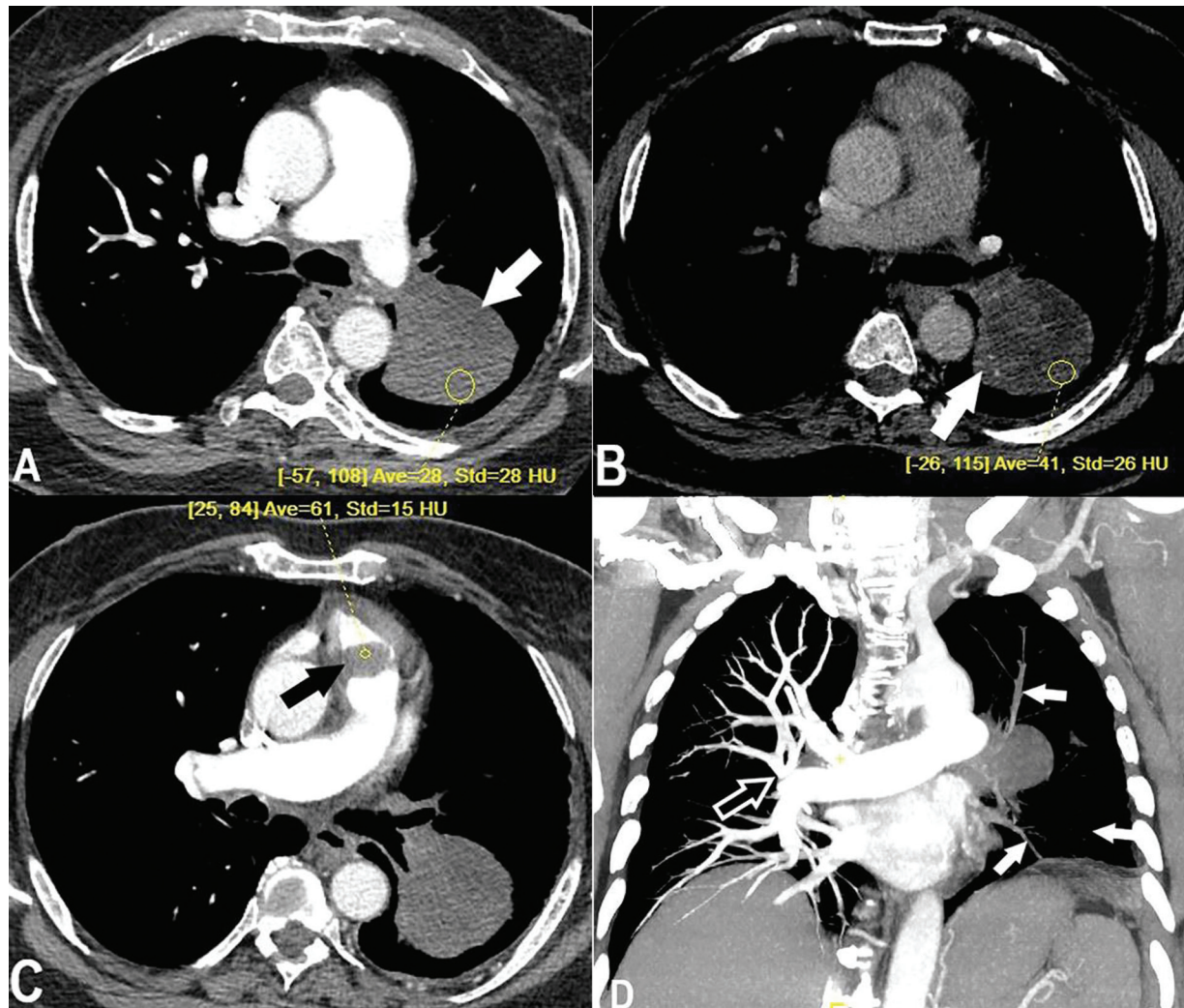
Fig. 2 (**A, B**) Axial computed tomography pulmonary angiogram (CTPA) images show patchy minimal peripheral enhancement (white arrows) of the large mass lesion with Hounsfield Unit (HU) of 28 and 41 in the early and delayed phases, respectively, occluding the distal left pulmonary artery. (**C**) Axial CTPA image shows minimally enhancing filling defect in the root of the pulmonary artery with HU of 61 (black arrow). (**D**) Coronal maximum intensity projection image showing normal right pulmonary arterial tree (black arrow) and complete nonopacification distal left PA onward with corresponding left lung oligemia (white arrows).

focal expression of epithelial membrane antigen, ETS-related gene, and CD31 and was negative for cytokeratin (AE1/AE3), thyroid transcription factor-1, P 63, desmin, smooth muscle antigen, h-caldesmon, S-100 protein, and CD34. These histopathological features were suggestive of a high-grade spindle cell sarcoma, consistent with an angiosarcoma.

Thus, with two separate enhancing, FDG avid lesions and with histology as described above, the diagnosis of multicentric primary pulmonary artery angiosarcoma was established.

Discussion

PAS are neoplasms with origin from the mesenchymal cells of the intimal layer of the pulmonary artery. The occurrence of primary PAS is so rare, only ~0.001 to 0.03%,[1] that though the first case was described in 1923, thereafter its description has been limited to case reports and case series. A total of ~391 cases have been described thus far in literature.[2] Among the sarcomas of the great vessels, PAS are the most common, affecting the trunk (80%), the right pulmonary artery (57%), and the left pulmonary artery (58%), both the arteries in 37% and the pulmonary valve in 29%.[3] Angiosarcomas are estimated to have an incidence of ~3.6% among the various types of PAS.[4]

PAS can present with varied symptoms that may mimic PE. However, they can even be completely asymptomatic initially and cause short duration of symptoms only at an advanced stage, like in unilateral extension into the lung from peripheral involvement of the pulmonary artery,[5] as observed in our case. Shortness of breath is the most common presenting symptom, others being chest pain, cough, hemoptysis, weight loss, fatigue, dizziness, and fever.[2] Lack of predisposing factors for thromboembolism, persistent symptoms, recurrence despite adequate anticoagulation, and unilateral massive perfusion defect should raise the possibility of tumoral obstruction.[6] The differential diagnoses include pulmonary thromboembolism and lung neoplasm.
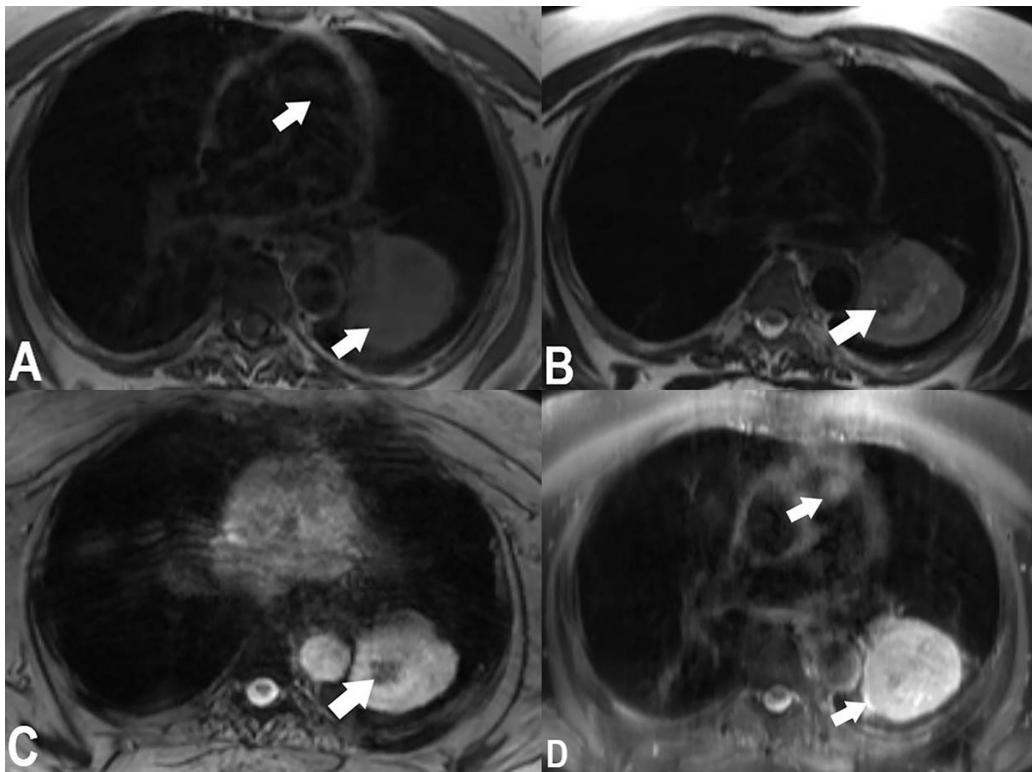
Fig. 3 Magnetic resonance imaging thorax. Arrows show the two lesions appearing isohypointense on T1-weighted image (**A**), heterogeneously hyperintense on T2-weighted image (**B**), with central gradient recalled echo hypointensity (**C**) and hyperintense on PDFS images (**D**).
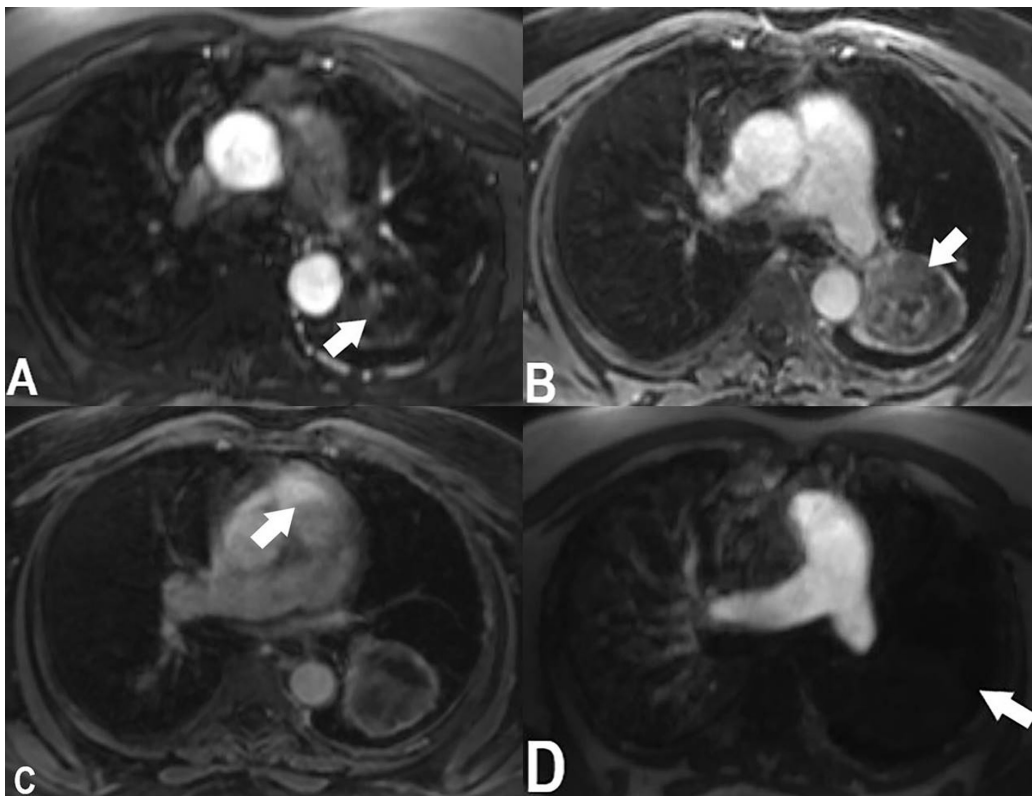

Fig. 4 Post-contrast magnetic resonance images: (**A, B**) Medial and central lesional enhancement (white arrows) in the arterial phase with progressive diffuse enhancement on the delayed phases with occlusion of the left main pulmonary artery distally. (**C**) Small enhancing lesion within the root of the main pulmonary artery (arrow). (**D**) Classical demonstration of left lung oligemia secondary to occlusion of the left main pulmonary artery (arrow).
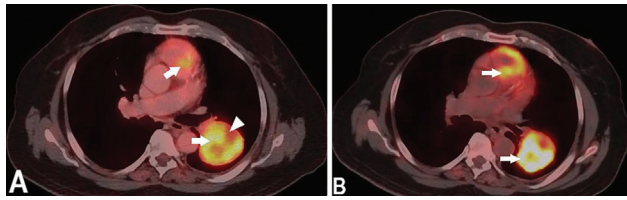
Fig. 5 Fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT): (**A**) Initial and (**B**) delayed. Arrows show a large lesion with heterogeneous increased uptake and another lesion at the root of the pulmonary artery, with increased uptake on delayed images. Arrowhead marks central non-FDG avid area of necrosis within the lesion.

Potential benefits of metabolic imaging using F-18 FDG PET-CT have been sporadically described in the literature. Unequivocal positive findings with mean SUV_{max} of 13.1 have been described for PAS.[10] The mean SUV_{max} of 7.6 was significantly higher in PAS than PE.[11] Our case also revealed significantly higher FDG uptake in both the lesions that favored the diagnosis of PAS. PET-CT being a whole-body examination helps to decide on the possible malignant nature of a filling defect and also serves as a staging modality.[10] Thus, FDG PET-CT should also be considered as part of the imaging workup in a case of suspected PAS.
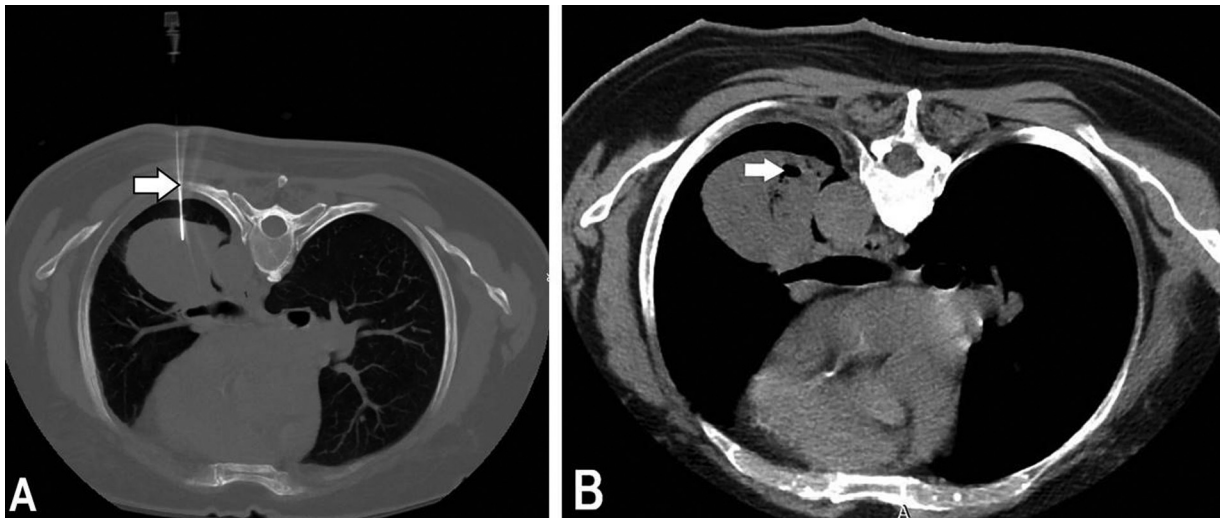

Fig. 6 Computed tomography (CT)-guided biopsy of the mass lesion by posterior approach. (**A**) Needle tract. (**B**) Post-biopsy plain axial CT with air foci within (arrows).

CT helps differentiate PAS from PE by indicating a low-attenuation filling defect occupying the entire luminal diameter of the proximal or main pulmonary artery, expansion of the involved arteries, or extraluminal tumor extension.[7] In our case, filling defect was seen in distal left pulmonary artery with extraluminal tumor extension. PAS on CTPA has been demonstrated to show eclipsing of the pulmonary artery wall before it infiltrates beyond the artery, absent in PE, termed as the wall eclipsing sign and defined as almost full occupation of the lumen of the pulmonary trunk, left pulmonary artery or right pulmonary artery by a low-density mass; protrusion of the proximal end of this mass toward the right ventricular outflow tract; and eclipsing of one or both walls of the artery by the lesion.[8] Though this could be a useful sign assisting in the diagnosis of PAS, it was not visualized in our case, most likely due to substantial extension of the tumor into the adjacent lung, beyond the left pulmonary artery.

Multiparametric MRI findings of PAS include hyperintense filling defect in the main pulmonary artery on fat suppressed T2-weighted and diffusion-weighted imaging and contrast enhancement.[9] These findings were also observed in our case.

Conclusion

Thus, in a clinical setting of high index of suspicion, multimodal imaging approach should be made use of, so that prompt diagnosis and timely institution of appropriate treatment can be made, thereby improving the prognosis. The definitive diagnosis, however, is achieved only by histopathology.

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