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CASE REPORT

Pulmonary Artery Sarcoma - Multimodality Imaging

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ABSTRACT

Pulmonary artery sarcoma (PAS) is a rare and fatal disease. PAS can often be misdiagnosed as pulmonary thromboembolism. Moreover, the correct diagnosis is frequently delayed due to nonspecific signs and symptoms. The prognosis of patients with PAS is poor. We report a case of a woman with a primary PAS who was initially diagnosed with pulmonary thromboembolism.

Key words: Computed tomography, positron emission tomography, pulmonary artery, pulmonary artery sarcoma, pulmonary thromboembolism, sarcoma

INTRODUCTION

Pulmonary artery sarcoma (PAS) is uncommon, and the prognosis of patients with PAS is poor. Early diagnosis of PAS is very difficult since there are no specific diagnostic tests including imaging modalities for PAS. High degree of clinical suspicion is required to make the diagnosis of PAS.

CASE REPORT

A 76-year-old woman with hypertension was referred to our hospital for the evaluation of dyspnea, fever, and weight loss. The patient underwent anticoagulant therapy for pulmonary thromboembolism for 2 weeks but had no clinical improvement. A physical examination revealed tachypnea (respiratory rate 22/min). Blood pressure was 100/80 mmHg, Cardiovascular evaluation was unremarkable, and there was no cyanosis. Electrocardiogram was normal with sinus rhythm. Chest X-ray revealed patchy multifocal consolidations, calcific granulomas, bronchiectasis, and volume loss with pleural thickening in the right lung [Figure 1]. D-dimer was slightly higher than the upper limit of normal. Arterial blood gas analysis showed a pH of 7.49, $PaCO_2$ 21 mmHg, PaO_2 93 mmHg, HCO_{3-} 16.0 mmol/L, and SaO_2 98% on room air. Transthoracic echocardiography showed echogenic mass and no color flow in the right pulmonary artery [Figure 2 and Videos 1, 2]. Right ventricular systolic pressure gradient was 38 mmHg. Chest computed

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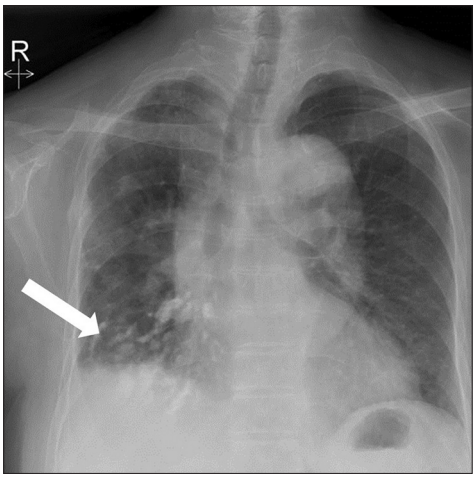

Figure 1: A 76-year-old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. Chest X-ray shows previous inflammatory fibrosis, cicatricial bronchiectasis, and volume loss with pleural thickening in the right lung (arrow).

Video 1: A 76 year old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. Echocardiography shows oval shaped echogenic mass in the right pulmonary artery in the subcostal view.

tomography (CT) revealed a low-attenuation filing defect that occupied the entire luminal diameter of the right pulmonary artery [Figure 3]. However, there was no definite invasion into the lung. Bronchoscopy did not demonstrate evidence of tumor invasion into the lungs. Ultrasound evaluation did not demonstrate deep vein thrombosis in the legs. Positron emission tomography (PET) showed the tumor occupied the majority of the right pulmonary artery. It showed an intense hypermetabolism in the right proximal pulmonary artery, suggesting the presence of malignancy [Figure 4]. The patient underwent percutaneous trans-lung biopsy. A biopsy was confirmed malignant spindle cell neoplasm, consistent with sarcoma [Figure 5]. The patient refused the operation and received four courses of chemotherapy with paclitaxel. However, the tumor size increased, and the disease progressed. She died 6 months after diagnosis due to shock.

DISCUSSION

PAS is rare and arises from the intimal layer of the pulmonary artery. The estimated incidence of PAS is 0.001%, with a

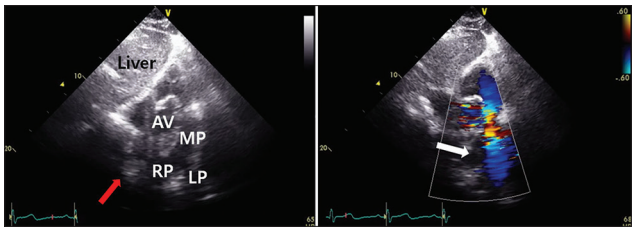

Figure 2: A 76-year-old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. Echocardiography shows oval shaped echogenic mass in the right pulmonary artery in the subcostal view (red arrow). There is no color flow Doppler in the right pulmonary artery (white arrow). AV: Aortic valve, MP: Main pulmonary trunk. RP: Right pulmonary artery, LP: Left pulmonary artery.

Video 2: A 76 year old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. Echocardiography shows no color flow Doppler in the right pulmonary artery.

slight preponderance in women (1:1.3).[1] The common symptoms are dyspnea, cough, hemoptysis, weight loss, fever, and constitutional symptoms.[2] As such, PAS can frequently be misdiagnosed as pulmonary thromboembolism.[3] Early diagnosis of PAS is very difficult because there is no specific diagnostic test including imaging for this disease.[4] Echocardiography is used to investigate pulmonary hypertension noninvasively and in cases of PAS echocardiography may show a dilated right ventricle with obstruction in the ventricular outflow tract or the pulmonary arterial trunk.[5] CT and PET are valuable tools to assist diagnosis of PAS. Chest CT can reveal some features of PAS such as contrast enhancement.[6] Contrast-enhanced CT can detect a low-attenuation filling defect that occupies the entire luminal diameter of the proximal or main pulmonary artery, the expansion of the involved arteries, and extraluminal tumor extension. These CT findings are nonspecific as they can also be seen in cases of extensive pulmonary thromboembolism.[7] There is one study that has shown that fluorodeoxyglucose positron emission tomography (FDG-PET) is useful for diagnosing intimal sarcoma.[8] FDG-PET may be useful in differentiating pulmonary thromboembolism and PAS, as blood thrombi do not take up ^{18}F-FDG, whereas a malignant tumor, such as PAS and does.[9] However, some reports indicate that PASs may not always show high FDG uptake.[10] Lung

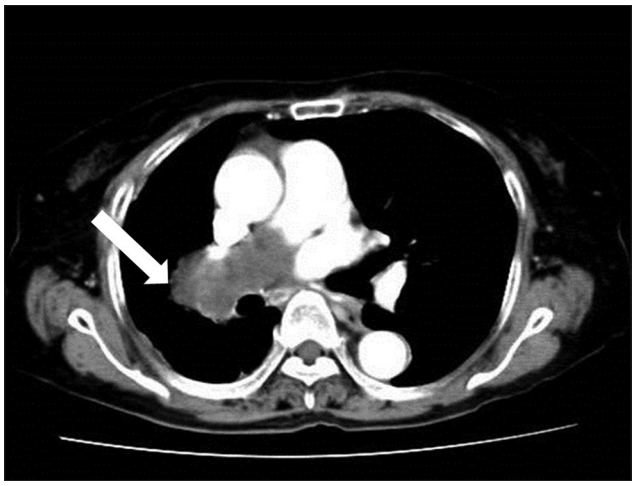

Figure 3: A 76-year-old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. CT of the chest reveals the tumor is occupying the right pulmonary artery (arrow).

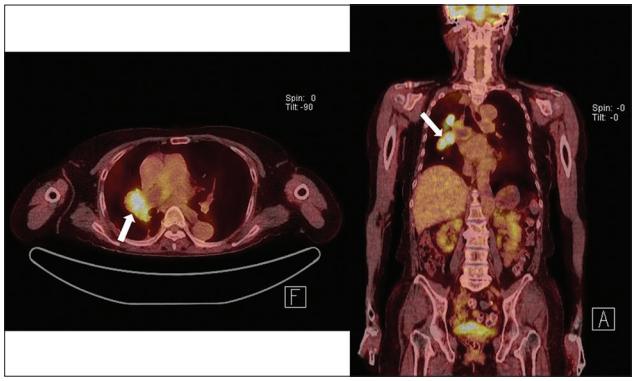

Figure 4: A 76-year-old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. Positron emission tomography demonstrates a hypermetabolic lesion in the right proximal pulmonary artery (arrows).

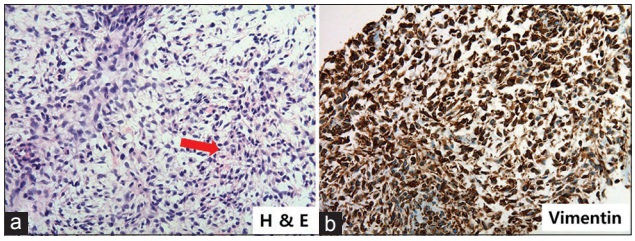

Figure 5: A 76-year-old woman with hypertension and pulmonary embolism presented with dyspnea, fever, and weight loss. (a) Histopathology shows atypical spindle cells (red arrow) with frequent mitotic figures (H and E, ×400). (b) Immunohistochemical stains reveal that the tumor cells were negative for epithelial cell marker (cytokeratin) and strongly positive for mesenchymal cell marker (Vimentin).

metastases occur in 50% and distance organs in 16%, including kidney, lymph nodes, brain, and skin, often detected at the time of diagnosis.[7] The prognosis of PAS is poor, and survival is usually 12–18 months from the time of diagnosis. Mean survival of PAS is <2 months in patients who do not undergo surgery. Surgical resection decreases the clinical symptoms and improves the survival time of patients.[1] Efficacy of chemotherapy or radiotherapy for PAS remains controversial.

CONCLUSIONS

Persistence of the filling defect in the pulmonary artery despite proper anticoagulation therapy in patients with pulmonary thromboembolism, PAS should be considered in the differential diagnosis. Echocardiography, CT, and PET are useful in differentiating pulmonary thromboembolism from PAS.

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Conflicts of interest

There are no conflicts of interest.

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