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Case Report

Pulmonary artery angiosarcoma mimicking chronic thromboembolic pulmonary hypertension

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Abstract

Pulmonary artery angiosarcoma (PAA) is an extremely rare malignancy that arises from endothelial cells. The pulmonary trunk is the most common anatomic site of these tumors. However, so far, there have only been a few examples of PAA reported. PAAs are aggressive and have a bad prognosis, thus early diagnosis is critical. In this study, we present the case of a 27-year-old male who was initially diagnosed with acute pulmonary thromboembolism (PTE) before being diagnosed with chronic thromboembolic pulmonary hypertension (CTEPH). Pulmonary endarterectomy was performed, and PAA was diagnosed histopathologically. The patient died three months after diagnosis.

Keywords: Pulmonary artery, angiosarcoma, CTEPH, computed tomography, pulmonary thromboembolism

INTRODUCTION

PAA is an extremely rare malignancy with a high mortality rate. Clinical and radiological manifestations can be difficult to distinguish from PTE. Radiological imaging methods may play an important role in the early diagnosis of PAA, providing appropriate treatment and long-term survival.

CASE REPORT

A 27-year-old male patient—a smoker, with no known co-morbidity—presented to the emergency department with a history of chest pain with streaky hemoptysis for two weeks, and progressive breathlessness for two days.

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In his initial examination, the respiratory rate was 28/min, pulse – 130/min, blood pressure – 110/70 mmHg, oxygen saturation–96% on room air; no other significant finding was noted. As his D-dimer was 1070 mcg/L (normal range 0-500 mcg/L) and the Wells PTE prediction score was 7, a computed tomography angiography (CTA) was performed, which showed multiple filling defects in the main pulmonary artery (MPA), in both pulmonary arteries, with extension in the bilateral lower lobar arteries and in the segmental arteries of lower lobes (Figure 1).

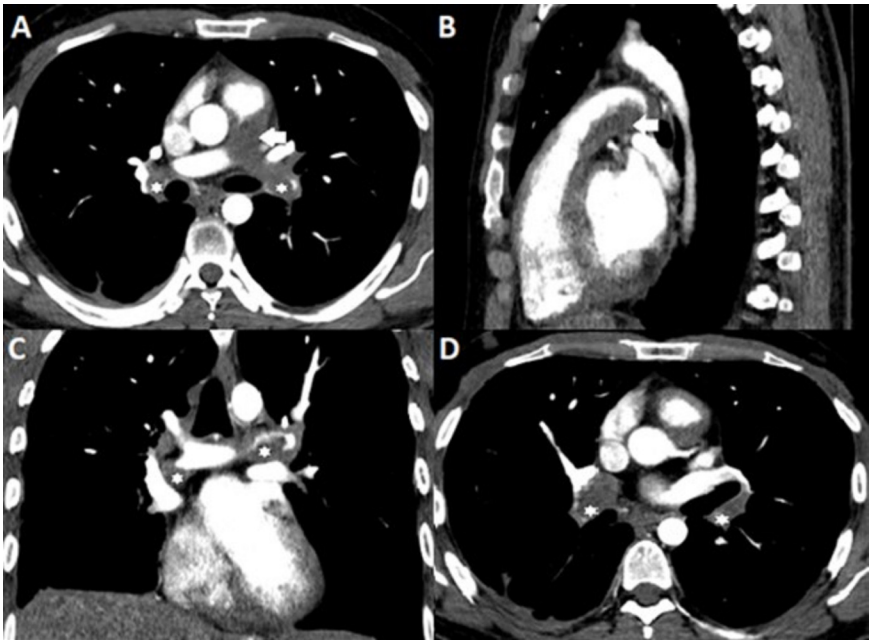

Figure 1. *Pulmonary CT angiography findings. A and D axial, B sagittal, C coronal images. Filling defects in the main pulmonary artery (arrow), in both pulmonary arteries, with extension in the bilateral lower lobar artery (Asterix).*

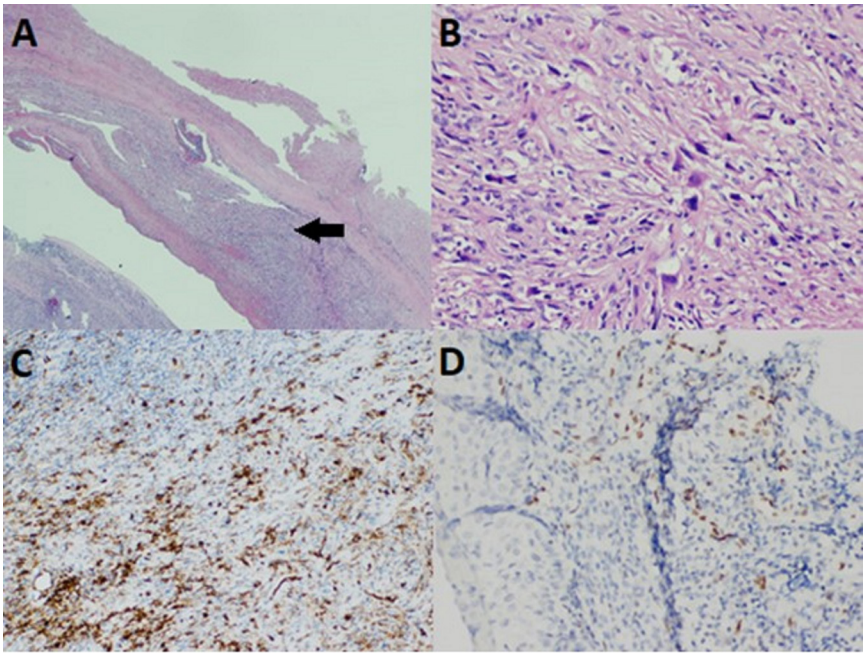

Figure 2. *A: Tumoral infiltration(arrow) in the vessel walls (H&E, X20); B: Spindle and epithelioid tumor cells showing prominent pleomorphism (H&E, x200); C: CD31 positivity in tumor cells (DAB, x100) D: FLI-1 positivity in tumor cells (DAB, x200).*

His echocardiogram demonstrated severe right ventricular (RV) dilatation with semi paradox movement in the ventricular septum, and Tricuspid annular plane systolic excursion 19 mm with moderate pulmonary artery hypertension (PAH) (mPAB 45 mmHg). Pro-BNP was 608mg/ml (normal range 0-110mg/ml). The ultrasound of the bilateral lower and upper limb was normal.

He was hospitalized as intermediate high-risk PTE, and anticoagulant therapy with 5000 IU unfractionated heparin as a bolus, followed by 18 IU/kg/hr. perfusion was initiated, with close monitoring of the coagulation parameters. As the hemodynamic parameters, oxygen saturation, and tachypnea worsened, on the second day of his admission, the patient was given tissue plasminogen activator (rtPA) iv, and later the patient was started again on an intravenous heparin infusion. His vital signs and oxygenation improved gradually. As the target aPTT was not reached on the maximum daily dose, the heparin drip was changed to a therapeutic dose of subcutaneous enoxaparin for five days. Then oral rivaroxaban was started at 20 mg/daily for long term treatment.

The patient had no predisposing factor for the disease, so he was investigated for thrombophilia, Bechet disease, and malignancy. Hypercoagulable work-up later revealed no pathologies. The upper and lower abdominal CT was normal. The patient continued to improve clinically and was discharged home on hospital day 12.

On his third month follow-up, the patient had only mild dyspnea on exertion. Physical examination demonstrated no significant abnormalities. The results of blood tests showed a normal hemoglobin level of 16.2 g/dL [normal 13.5–18] g/dL), a normal leukocyte count, and a mildly elevated C-reactive protein level of 16.7 (normal < 5.0) mg/L. Other laboratory investigations, including those for coagulation disorders, were within normal limits. Doppler ultrasonography of the legs was negative again for deep venous thrombosis.

The transthoracic echocardiogram showed a dilated right ventricle (RV) with normal RV systolic function, pulmonary arterial hypertension, and demonstrated normal left chamber size and function.

Dual-energy CTA of the chest demonstrated dilatation of pulmonary trunk and multiple pulmonary emboli, which was remarkable for a large filling defect involving the MPA and multiple progressive filling defects of the peripheral pulmonary arteries, including obstructive lesions in both lower lobar arteries. Also, he had multiple segmental perfusion defects on iodine mapping. However, the possibility of a pulmonary artery malignancy could not be ignored. He then underwent a PET scan, which revealed no 18-FDG uptake.

The patient was subsequently suspected of having CTEPH. A mean pulmonary artery pressure of 42 mmHg was found during cardiac catheterization, indicating pulmonary hypertension. He was referred for pulmonary endarterectomy.

The CTEPH committee decided on pulmonary thromboendarterectomy to remove the thrombi. A median sternotomy was performed, and a cardiopulmonary bypass was initiated. The bilateral endarterectomy was performed from the mean pulmonary artery to subsegmental branches using total circulatory arrest (TCA). For each unilateral endarterectomy, the TCA time was 20 minutes. The patient was followed up in the intensive care unit for about one month. His postoperative echocardiogram demonstrated mild right ventricular (RV) dilatation with normal pulmonary artery pressure (mPAB 25 mmHg).

The endarterectomy specimen was sent for pathological examination. The biopsy included the fragmented thrombi and vessel tissues. Some thrombi fragments were composed of tumor cells. The vessel intima and media layers were diffusely infiltrated by tumor cells. The tumor indicated epithelioid and spindled atypical cells showing severe pleomorphism and high mitotic activity. Patchy necrotic areas, poorly defined vascular channels, and extravasation of blood were seen. Immunohistochemically, there was positivity for CD31 and FLI-1 in tumor cells (Figure 2). Morphologic and immunohistochemical findings were in keeping with angiosarcoma.

The patient died three months after diagnosis.

DISCUSSION

Sarcomas are rare mesenchymal tumors. They originate from soft tissues or bones. Only 2% of soft tissue sarcomas are angiosarcomas, which originate from endothelium cells. In the literature, there are only about 250 primary pulmonary sarcomas described. These include other forms of pulmonary artery sarcomas liposarcomas, rhabdomyosarcomas and chondrosarcomas [1].

PAA is a rare malignancy that accounts for around 3.6 percent of all pulmonary artery sarcomas. PAAs usually originate from the intima and arise from the pulmonary trunk [1]. Primary pulmonary angiosarcoma has an etiology that is not well understood [2].

PTE is a rather common disease. Its annual incidence is predicted to be 112.3 cases per 100,000 adults. CTA, while highly sensitive and specific, can show a variety of diseases mimicking PTE. Tumors of the pulmonary arteries, including angiosarcomas, are unusual pathological reasons that mimic PTE on imaging.

PAA is a slow-growing malignancy. The onset of symptoms

is insidious [3]. With its nonspecific respiratory symptoms, primary pulmonary angiosarcoma is a difficult disease to diagnose [1]. The radiologic findings are similar to PTE. As a result, angiosarcomas are frequently misdiagnosed as PTE [1]. The diagnosis is usually delayed and is prone to misdiagnosis, which often leads to a poorer prognosis [4]. Angiosarcomas spread regionally to the lung and mediastinal lymph nodes, and distally to other organs such as the liver, kidneys and pancreas by hematogenous dissemination [5].

On CTA, tumors in the pulmonary vasculature can be diagnosed by low attenuation filling defects, heterogeneous enhancement of the mass in the artery lumen, and extravascular extension of the disease [6]. Contrast-enhanced magnetic resonance imaging may help distinguish between thromboembolism and tumors. Angiosarcomas, unlike thromboembolism, show heterogeneous enhancement with contrast agent [1]. FDG-PET has recently been found to be helpful for assessing the local invasion and distant metastases of malignant cardiac tumors, as well as monitoring treatment response [7]. However, it was not helpful in our case.

If total resection of the tumor is not possible, the prognosis of PAA is quite poor. With a median survival of 36.5±20.2 months, radical surgical resection seemed to have the best prognosis. A median survival of 11±3 months is provided by subtotal excision or debulking of the tumor [8]. The mean survival time without surgery is 1.5 months [9]. Unfortunately, because up to 50% of patients have metastatic disease, many are not surgical candidates [8]. The most common cause of mortality is heart failure, which is usually right-sided due to obstruction in the right ventricular outflow tract. The most extended survival period is reported as three years [10]. A recent study found that aggressive resection and multimodality treatment resulted in better survival. Chemotherapy may increase the median survival time; however, evidence is inadequate [8].

Furthermore, the diagnosis of the tumor can limit the usage of potentially harmful thrombolytic medication. As a result, accurate diagnosis is critical for improving the survival of patients with PAA.

CONCLUSION

Pulmonary angiosarcomas are difficult to diagnose. PAA is commonly mistaken for PTE, which is much more common. At the time of initial diagnosis, PAA should be considered in the differential diagnosis of PTE, particularly in patients with a low or intermediate probability of PTE, unusual symptoms, insufficient response to thrombolytic therapy. Although misdiagnosis does not increase in mortality, a considerable delay in diagnosis appears to have a detrimental effect on the outcome. Although PAA is difficult to cure, tumors that are diagnosed early and treated with surgery (total resection) and chemotherapy have the best long-term results in terms of survival.

Conflict of interests
The authors declare that there is no conflict of interest in the study.

Financial Disclosure
The authors declare that they have received no financial support for the study.

Patient Consent for Publication
A written informed consent was obtained from the both patient.

Data Sharing Statement
The data that support the findings of this study are available from the corresponding author upon reasonable request.

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