Clinical Case Report

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Primary pulmonary artery sarcoma with deep vein thrombosis

A case report

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Abstract

Rationale: Pulmonary artery sarcomas (PAS) are easily misdiagnosed as thromboembolic disease of pulmonary arteries, because of rarity and presenting with nonspecific signs, symptoms, or imaging findings.

Patient concerns: A 26-year-old man was admitted to the department of invasive technology with fever and dyspnea. Blood tests showed inflammatory activity, a slight increase of D-dimer and Fibrin Degradation Product. A chest enhanced computed tomography (CT) scanning revealed multiple filling defects occurred in the main trunk of both pulmonary arteries and branches of the left pulmonary artery

Diagnoses: It was initially diagnosed with pulmonary embolism (PE) and deep vein thrombosis (DVT), but was eventually diagnosed with pulmonary artery sarcoma that was confirmed by biopsy.

Interventions: The transcatheter thrombolysis therapy, inferior vena cava filter implantation, and operation were performed.

Outcomes: The Organized mass was removed by the operation and was pathologically diagnosed as pulmonary artery sarcoma, the patient received postoperative chemotherapy according to the recommendation of oncology department.

Lessons: Coagulation markers have been reported to differentiate PAS from PE, but this case suggested that PAS can be associated with DVT and abnormal coagulation-fibrinolysis system.

Abbreviations: CT = computed tomography, CTA = computed tomography angiography, DVT = deep vein thrombosis, FDP = fibrin degradation product, PAS = pulmonary artery sarcomas, PE = pulmonary embolism.

Keywords: coagulation-fibrinolysis system, deep vein thrombosis, pulmonary artery sarcomas, pulmonary embolism

1. Introduction

Pulmonary artery sarcomas (PAS) are easily misdiagnosed as thromboembolic disease of pulmonary arteries due to rarity and nonspecific signs, symptoms, or imaging findings so that the opportunity for early diagnosis and treatment of this aggressive neoplasm is missed.[1] Some case reports suggested that PAS had the normality of coagulation-fibrinolysis system.[2–4] We present a case of PAS that was confirmed by biopsy. It was initially misdiagnosed as an embolism, because the imaging showed that the patient had deep venous thrombosis (DVT) and his coagulation-fibrinolysis system was abnormal. The patient has provided the informed consent for publication of the case

2. Case report

A 26-year-old man with fever and dyspnea was admitted to the department of invasive technology. Blood tests showed inflammatory activity (white blood cell [WBC] 13400/mL, C-reactive protein [CRP] 95.32 mg/L), a slight increase of D-dimer (2.78 μg/mL) and Fibrin Degradation Product (FDP)-20.81 μg/mL (normal range <5.0 μg/mL). The chest enhanced computed tomography (CT) scanning revealed multiple filling defects occurred in the main trunk of both pulmonary arteries and branches of the left pulmonary artery (Fig. 1: Aa, Ab, Ac, Ad). Echocardiography revealed the dilated right atrium and ventricle, impaired right ventricular systolic function, and abnormal echo in the pulmonary artery and its branches (Fig. 2: A, B, C). Color-Doppler ultrasound showed venous thrombosis of the lower extremity and its blood flow stagnation (Fig. 2: D, E). Because of his symptoms, high blood coagulation, the CT scanning and deep venous thrombosis revealed by the angiographic, the patient was treated with the transcatheter thrombolysis therapy and inferior vena cava filter implantation, under the diagnosis of pulmonary embolism (PE) and DVT. However, there was no relief of symptoms after transcatheter thrombolysis therapy, and the

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XL and QQ contributed equally to this work.

The written consent for publication of this case report and accompanying images were obtained from the patient.

The authors have no conflicts of interest to disclose.

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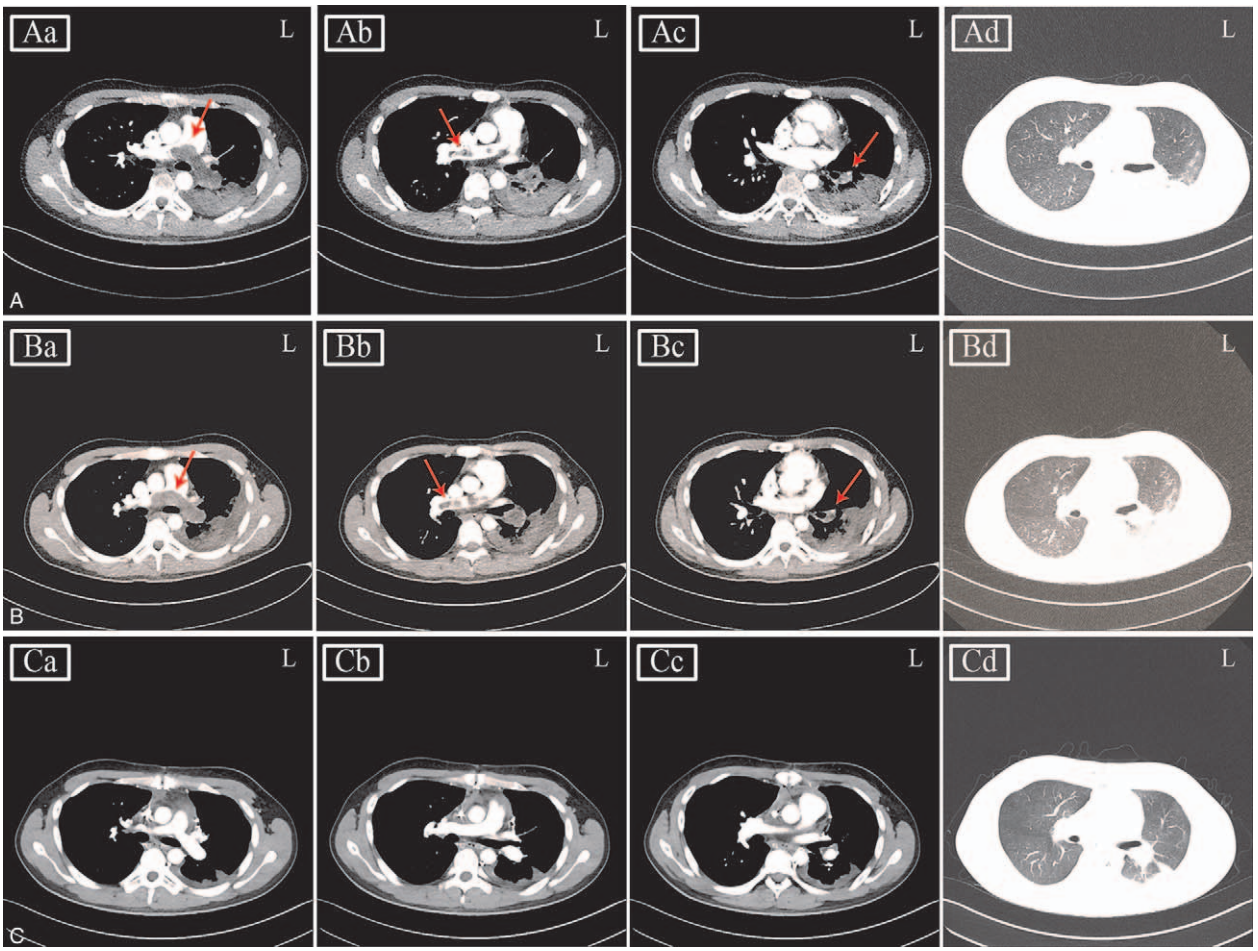

Figure 1. Aa-Ac (the first chest enhanced CT) showed that filling defects were found in pulmonary aorta, left and right pulmonary artery, and left inferior pulmonary artery (red arrow), Ad (the first chest enhanced CT) showed that the left lung had a partial infarction. Ba-Bd (pulmonary artery CTA after transcatheter thrombolysis therapy) revealed multiple filling defects as same as the last CT scanning (red arrow), and there was no significant difference in a partial infarction of the left lung. Ca-Cd (the chest enhanced CT after the operation) showed that no other tumor tissue was found and a partial infarction of left lung was recovering. CT = computed tomography, CTA = computed tomography angiography.

pulmonary artery computed tomography angiography (CTA) revealed multiple filling defects as same as the last CT scanning (Fig. 1: Ba, Bb, Bc, Bd).

Based on the diagnosis of pulmonary thromboembolism, the patient was transferred to the department of cardiovascular surgery for pulmonary thromboendarterectomy. Under general anesthesia (GA) and cardiopulmonary bypass (CPB), midline sternotomy was performed, and main pulmonary artery was opened. Grey white tissue mass in thrombosed areas was removed by separating it from the main pulmonary artery, bilateral pulmonary artery branches, and pulmonary valves. The diameter of the mass was 1.5 to 5 centimeter (cm), and the total length was about 20 cm. In histopathology, the tumor is composed of spindle to epithelioid cells exhibiting marked pleomorphism with vascular growth pattern. Strange nuclei, giant nuclei, and multiple nuclei can be seen in tumor cells. Interspersed mitotic figures and areas of focal myxoid change and fibrinoid necrosis showed features of high-grade sarcoma. The immunohistochemistry results of tumor cells were positive for EMA, vimentin, and CD68, focally weakly positive for CR, SMA, ckp and bcl-2, and negative for WT1 and GFAP. The final diagnosis was primary pulmonary artery high-grade sarcoma (Fig. 3). The enhanced CT scanning of head, chest and belly revealed that no other tumor tissue was found (Fig. 1: Ca, Cb, Cc, Cd). Up to now (2 months), the post-operative period was uneventful. The patient underwent 4 cycles of postoperative chemotherapy according to the recommendation of another cancer hospital (we are not informed of the specific chemotherapy regimen and its curative effect). We carried out follow-up visit of the patient for 6 months and he was still alive.

3. Discussion

PAS is of mesenchymal cell origin, and its etiology is still unknown. It has been found in the bronchus (20%), lung parenchyma (70%), or pulmonary arteries (10%) and is considered rare, accounting for only 0.5% of all lung malignancies.[5] The patients with PAS often have a poor prognosis. The median survival time of PAS without therapy is around 1.5 months, and the reported longest survival period was 3 years.[6] Surgical resection is the most common treatment option for it and the effect of adjuvant chemotherapy or radiotherapy remains unclear.[7]

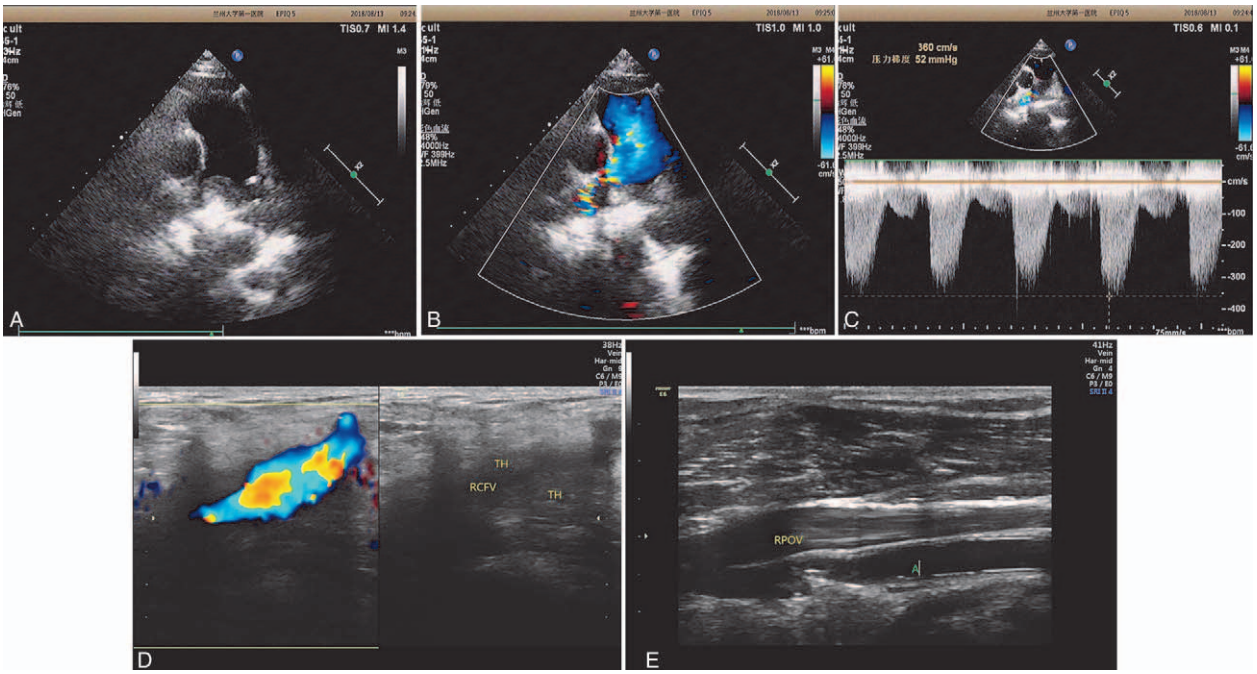

Figure 2. A, B, C (Echocardiography) revealed the partial obstruction and increased arterial pressure of the left pulmonary artery. D, E (Color-Doppler ultrasound) showed the right femoral vein thrombosis and early right popliteal vein thrombosis.

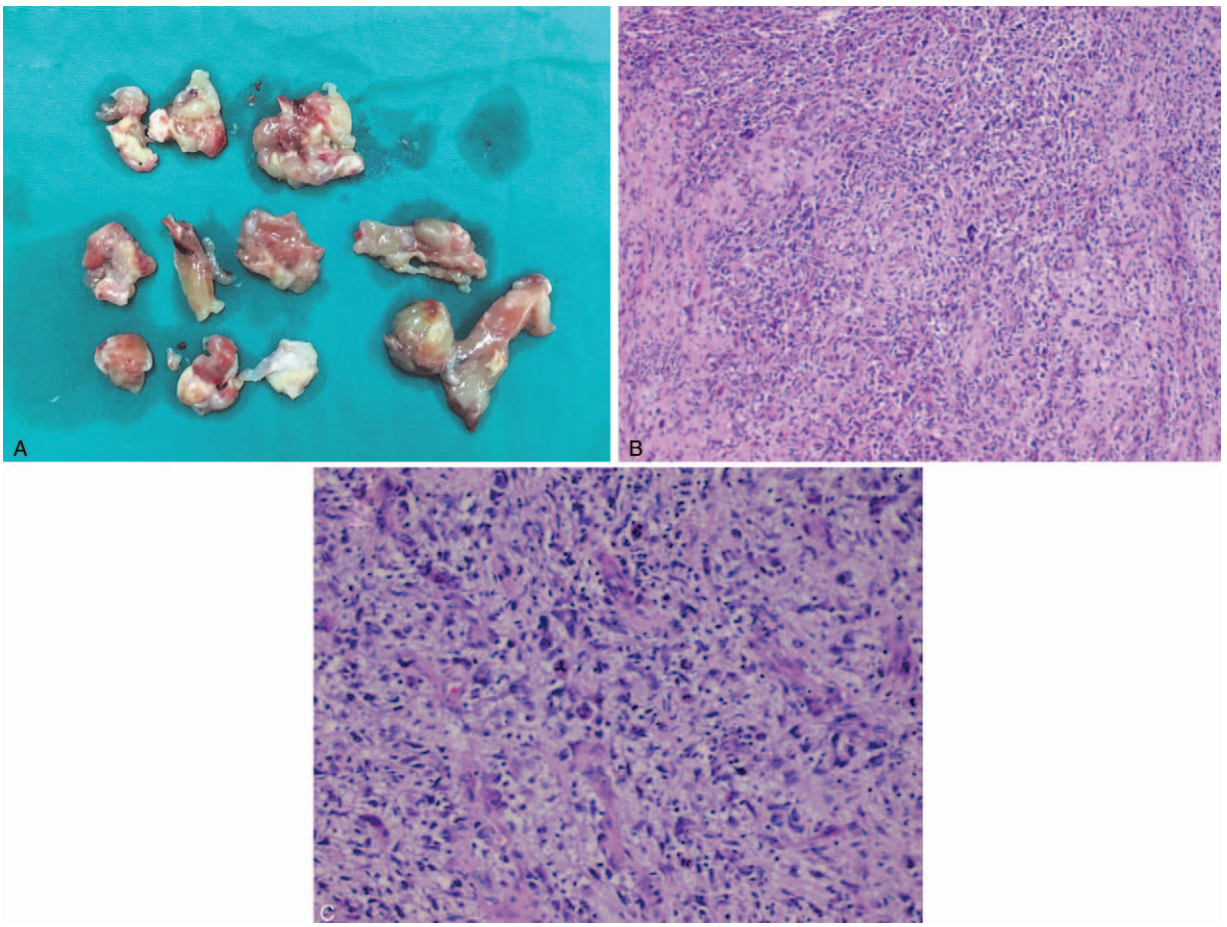

Figure 3. A was the organized mass in thrombosed areas by separated from the main pulmonary artery, bilateral pulmonary artery branches and pulmonary valves. B, C was pathological section image of it.

PAS, which should be suspected when a patient presents hemoptysis, is difficult to diagnose clinically because of nonspecific respiratory symptoms.[8–10] Other studies reported such symptoms as weight loss, fever, and digital clubbing for the diagnosis of PAS.[8] Many reports suggested that PAS had the normality of coagulation-fibrinolysis system, which is an important point to suspect PAS.[2,11,12] Guo et al reported that D-dimer values were within the normal range in all the 9 PAS cases.[2] In addition, we failed to find a case which reports PAS with DVT and abnormal coagulation-fibrinolysis system.

In this case, the coagulation-fibrinolysis system (D-dimer and FDP) of the patient was hyperactive, and the patient suffered from PSA and DVT at the same time. We cannot define the causal relationship between DVT and abnormal coagulation-fibrinolysis system in this case - DVT leads to abnormal coagulation-fibrinolysis system or vice versa, though it is generally believed that DVT can cause secondary hyperfibrinolysis. Koshiro et al reported PAS could combine with abnormal coagulation-fibrinolysis system without DVT, and they speculated that the reason for it might be the long-term inflammatory response induced by PAS or a thrombus surrounding PSA.[13] Despite the rapid development of imaging technology, including CT and MRI, it is still unable to diagnose PSA clearly.[14] The chest enhanced CT or CTA of our patient only revealed massive filling defects, but it could distinguish from PSA or PE. Further examinations are needed for the patients with these manifestations and imaging results to diagnose PAS. Some studies reported that the endovascular catheter biopsy, which can be accomplished through angiography, percutaneous core needle aspiration (PCNA), or endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA), might be the final and decisive diagnostic tool.[15,16] In view of the above-mentioned facts and the importance of an early diagnosis of PAS for the patient's prognosis, we suggested that the diagnosis of PAS should be based on overall findings, especially the endovascular catheter biopsy, rather than coagulation markers.

Author contributions

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Methodology: Xin Li.
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Resources: Shuai Dong.
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Writing – original draft: Xin Li.
Writing – review & editing: Xin Li, Quan Qi.

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