Case Report

Primary pulmonary artery chondrosarcoma: case report

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Background: Primary pulmonary artery sarcoma (PAS) is a rare tumor that presents like pulmonary embolism (PE), primary chondrosarcoma in the pulmonary artery is even rarer and few studies have been reported. PAS are commonly misinterpreted as in a clinical setting, many patients initially receive anticoagulant and thrombolysis therapy, but failed to respond. Management of this condition is difficult and prognosis is poor. We report a case of primary pulmonary artery chondrosarcoma that was initially misdiagnosed as PE and Inappropriate interventional therapy was performed, but with poor response. Finally, patient received surgical treatment, postoperative pathology confirmed primary pulmonary artery chondrosarcoma.

Case Description: A 67-year-old woman who had presented with cough, chest pain and shortness of breath for more than 3 months. Computed tomography pulmonary angiography (CTPA) showed filling defects were seen in the right and left pulmonary arteries, spreading to the outer lumen. The patient was initially diagnosed with PE and underwent transcatheter aspiration for pulmonary artery thrombus, transcatheter thrombolysis, and inferior vena cava filter placement at a local hospital, but with poor response. She was then referred for pulmonary artery tumor resection, endarterectomy and pulmonary arterioplasty. Histopathological examinations confirmed a diagnosis of primary PAS (chondrosarcoma). The patient developed *in situ* recurrence of pulmonary artery tumors in 10 months after surgery and received six cycles of adjuvant chemotherapy. The lesions progressed slowly after chemotherapy. The patient subsequently developed lung metastasis in 22 months and died of heart failure and respiratory failure 2 years after surgery.

Conclusions: PAS is an extremely rare and the clinical symptoms and radiological features often mimics PE, therefore When doctors make differential diagnosis of pulmonary artery mass lesions, especially when the anticoagulation and thrombolytic effects are very poor. They need to be alert to the possibility of PAS so that early diagnosis and early treatment can prolong the survival of patients.

Keywords: Case report; chondrosarcoma; pulmonary artery sarcoma (PAS); pulmonary embolism (PE)

Submitted Feb 23, 2023. Accepted for publication Apr 23, 2023. Published online Apr 28, 2023.
doi: 10.21037/tcr-23-518
View this article at: https://dx.doi.org/10.21037/tcr-23-518

Introduction

Primary pulmonary artery sarcoma (PAS) originates from the intima of the pulmonary artery or pulmonary valve. Primary chondrosarcoma in the pulmonary artery is even rarer and few studies have been reported (1). PAS is often initially misdiagnosed as pulmonary embolism (PE) because they share many clinical presentation and imaging findings. The diagnosis is challenging, a rapid and accurate diagnosis followed by appropriate therapy can improve outcomes. Herein we report a case of 67-year-old female patient with primary pulmonary artery chondrosarcoma that was initially

misdiagnosed as PE and implemented incorrect treatments, including transcatheter aspiration for pulmonary artery thrombus, transcatheter thrombolysis, and inferior vena cava filter placement at a local hospital, but with poor response. Finally, patient received surgical treatment, postoperative pathology confirmed primary pulmonary artery chondrosarcoma. We present the following article in accordance with the CARE reporting checklist (available at https://tcr.amegroups.com/article/view/10.21037/tcr-23-518/rc).

Case presentation

A 67-year-old woman was admitted to hospital in December 2018 due to "cough and chest pain for more than 3 months and chest tightness, shortness of breath, and syncope for 10 days".

In September 2018, she attended a local hospital with cough and chest pain. Chest computed tomography (CT) showed inflammation of the right upper lung, which improved after antimicrobial treatment. In December 2018, she exhibited chest tightness, shortness of breath on exertion, cyanosis of the lips, syncope, and incontinence. Repeat chest CT at the local hospital showed filling defects in the main, right and left pulmonary arteries. The possibility of PE was considered, so transcatheter aspiration for pulmonary artery thrombus, transcatheter thrombolysis, and inferior vena cava filter placement were performed. Mucous cartilage-like tissue was noted during the pathological examination of the aspirates. The therapeutic response was poor, and the patient was referred to our hospital. Physical examination showed body temperature, 37 ℃; pulse, 114 beats/min; respiration, 33/min; and blood pressure, 125/66 mmHg. The patient had the characteristic facial features of acute illness, and her lips were slightly cyanotic. Although the bilateral breath sounds were slightly coarse, no wet or dry rales were heard on lung auscultation. Her abdomen was soft and flat, and no edema was observed in either lower limb. The findings of laboratory tests included: white blood cell count, 13.05×10^9/L; and D-dimer level, 887.45 ng/mL. Color ultrasound revealed a mildly dilated pulmonary artery, and an isoechoic mass sized 23 mm × 19 mm at the bifurcation of the pulmonary artery (*Figure 1*). The deep veins of both lower limbs did not show any significant abnormalities. Contrast-enhanced CT of the abdomen suggested that the inferior vena cava was patent, and the implanted filter was visible. On CT pulmonary angiography (CTPA), filling defects were seen in the right and left pulmonary arteries, spreading to the outer lumen, which was suggestive of PE, but the possibility of malignant lesions could not be ruled out (*Figure 2*). She underwent pulmonary artery tumor resection, endarterectomy and pulmonary arterioplasty on December 13, 2018 (*Figure 3*).

The postoperative pathological examination revealed that the tumor was composed of cartilage and fibers under light microscopy, and some chondrocytes were pleomorphic. Mitotic images were visible (*Figure 4*). Immunohistochemical assay (9-item) showed seven positive indicators, vimentin, S-100, Fli-1, PGP9.5, CD99, EGR (partially positive), and Ki-67 [Ki-67-positive cells were unevenly distributed, with ≈60% (+) at hot spots] and two negative indicators, desmin and SOX11. These findings confirmed a diagnosis of well-differentiated chondrosarcoma of the pulmonary artery in both lungs (*Figure 5*).

The inferior vena cava filter was removed by percutaneous puncture and CTPA showed no filling defects in the main, right and left pulmonary arteries at 1 month after surgery (*Figure 6*). Positron emission tomography-CT (PET/CT) was performed 3 months after surgery and no obvious hypermetabolic signal was seen in the distribution areas of the pulmonary trunk and its left and right branches (*Figure 7*). A follow-up CTPA and PET/CT showed the patient developed recurrence of pulmonary artery tumors in 10 months after surgery and received six cycles of

Highlight box

Key findings

- This is the first report of primary pulmonary artery chondrosarcoma that was initially misdiagnosed as pulmonary embolism (PE) and underwent transcatheter aspiration for pulmonary artery thrombus, transcatheter thrombolysis, and inferior vena cava filter placement.

What is known and what is new?

- Primary pulmonary artery sarcoma (PAS) is often initially misdiagnosed as pulmonary embolism (PE).
- Computed tomography pulmonary angiography (CTPA) showed filling defects were seen in pulmonary arteries, even extra-luminal infiltration, resulting in a "worm erosion sign" is special feature of PAS and PET/CT is helpful to distinguish PAS from PE by standard uptake values.

What is the implication, and what should change now?

- The possibility of PAS should be considered in patients with clinically suspected PE but lacking PE triggers and responding poorly to anticoagulation or thrombolytic therapy. PET-CT is an effective noninvasive method for early diagnosis of PAS.

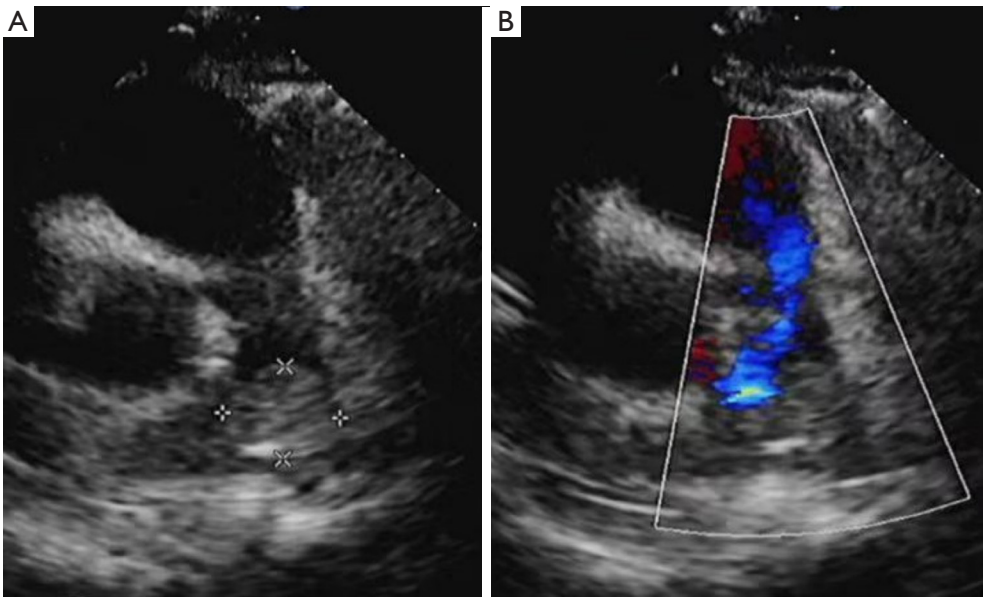

Figure 1 Color Doppler ultrasound. (A) Hypoechoic mass with irregular morphology and heterogeneous internal echogenicity (*) in the pulmonary trunk near the bifurcation of the right and left pulmonary arteries; (B) no significant blood flow signal within the mass, and blood flow in the right and left pulmonary arteries is obstructed.

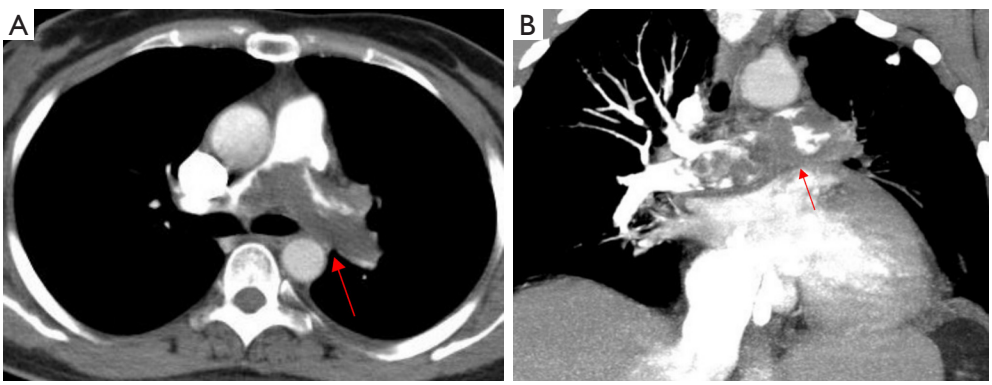

Figure 2 Computed tomography pulmonary angiography shows filling defects in the right and left pulmonary arteries, spreading to the outer lumen (red arrows) (A,B).

adjuvant chemotherapy. The lesions progressed slowly after chemotherapy and the patient subsequently developed lung metastasis in 20 months and died of heart failure and respiratory failure 2 years after surgery (*Figure 8*).

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent for publication of this case report and the accompanying images were not obtained from the patient or the relatives after all possible attempts were made.

Discussion

Primary PAS is a rare disease first described by Mandelstamm in 1923 in an autopsy, with a prevalence of 0.001% to 0.030% (2). PAS usually arises from the intima of the pulmonary artery and occurs first in the pulmonary artery trunk but can also involve the right and left pulmonary arteries. The tumor then invades smaller pulmonary

arterial branches and may affect the pulmonary valve and the right ventricular outflow tract in a small proportion of cases (3). The pathogenesis of PAS is unknown, but there are indications that the tumor originates from the reticuloendothelial system of the mesodermal structure.

PAS has many different pathological types, and further pathological subtyping is required after its diagnosis. The most common pathological types of PAS include undifferentiated sarcoma (21%), malignant fibrous histiocytic sarcoma (17%), smooth muscle sarcoma (17%), and spindle cell sarcoma (16%), which account for ≈71% of all pathological types. Other infrequent types include fibrosarcoma, fibrous mucinous sarcoma, rhabdomyosarcoma, and chondrosarcoma (4). Primary pulmonary artery chondrosarcoma is even rarer. In 1977, Hohbach and Mall reported the first case of primary pulmonary artery chondrosarcoma (5), and subsequently, sporadic cases have been reported in literature (6-8). In the current case, pathological examination of transcatheter aspirates at the referring hospital revealed the presence of a "chondrogenic component", and routine pathological and immunohistochemical examinations of the surgically resected specimen during the index admission confirmed the diagnosis of a primary well-differentiated chondrosarcoma of the pulmonary artery.

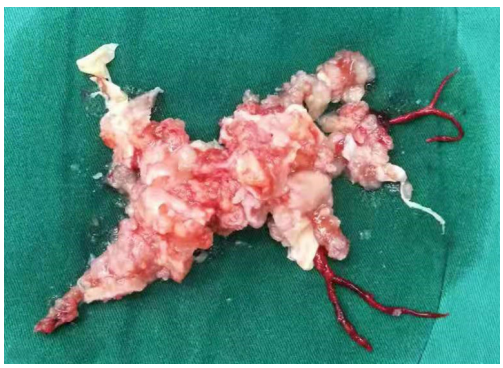

Figure 3 Several masses occluding the openings of the right and left pulmonary arteries. They are brittle in texture, with white cartilage-like tissue, and adhering closely to the pulmonary artery intima. Cord-like thrombi are visible distally.

PAS mimics PE in both clinical presentation and imaging findings, resulting in high rates of misdiagnosis and missed diagnosis. The current case was misdiagnosed as PE and treated as such, with poor response. Because our imaging findings could not rule out malignancy, the lesion was surgically treated and then pathologically diagnosed as PAS.

Although PAS is rare and difficult to diagnose, there are clues to distinguish it from PE: (I) PAS is insidious in onset and progresses relatively slowly or the course is subacute, whereas PE is a sudden illness; (II) PAS patients typically have systemic manifestations such as fever, malaise, and weight loss, often without a source of emboli (e.g., deep vein thrombosis of the lower extremity) causing PE; (III) CTPA in PAS patients often reveals large filling defects in the main, left and right pulmonary arteries, with irregular borders of the masses, lobulated or segregated nodules, or even extra-luminal infiltration, resulting in a "worm erosion sign" in the pulmonary artery wall, whereas PE is characterized by eccentric, appendicular filling defects with a more uniform thrombus density; (IV) PAS responds poorly to anticoagulation and thrombolysis; therefore, when the clinical manifestation and imaging findings cannot distinguish between these two diseases, diagnostic thrombolysis can be helpful; and (V) evidence suggests that PET/CT can distinguish PAS from PE by standard uptake

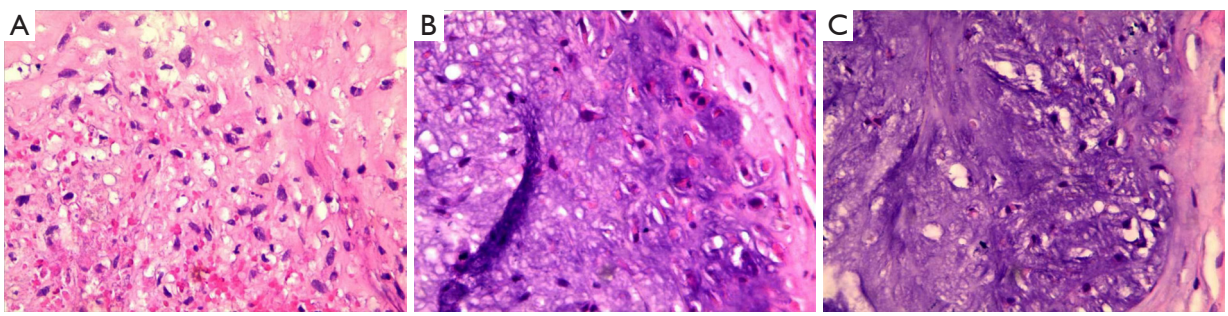

Figure 4 Light microscopy showing cartilage and fibers, with some pleomorphic chondrocytes, and mitotic figures (HE, ×200) (A-C). HE, hematoxylin and eosin.

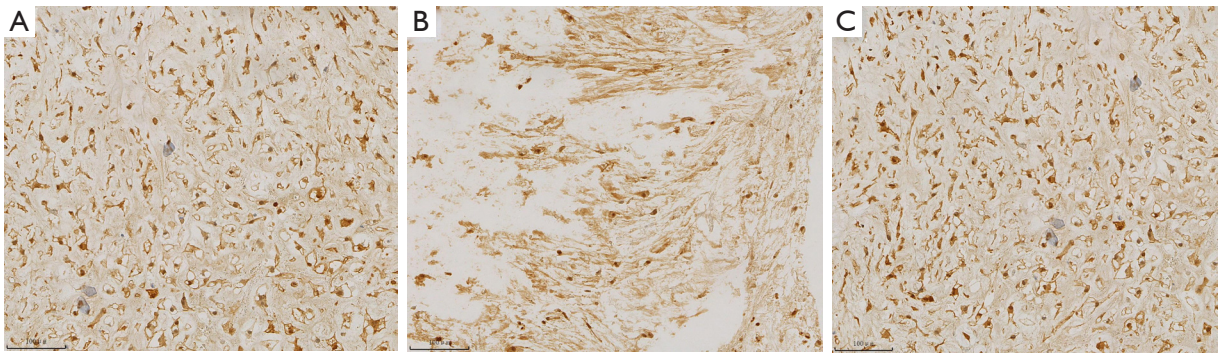

Figure 5 Immunohistochemical assays of the surgical specimens. (A) S-100 (+); (B) Fli-1 (+); (C) Ki-67 (positive). ×200.

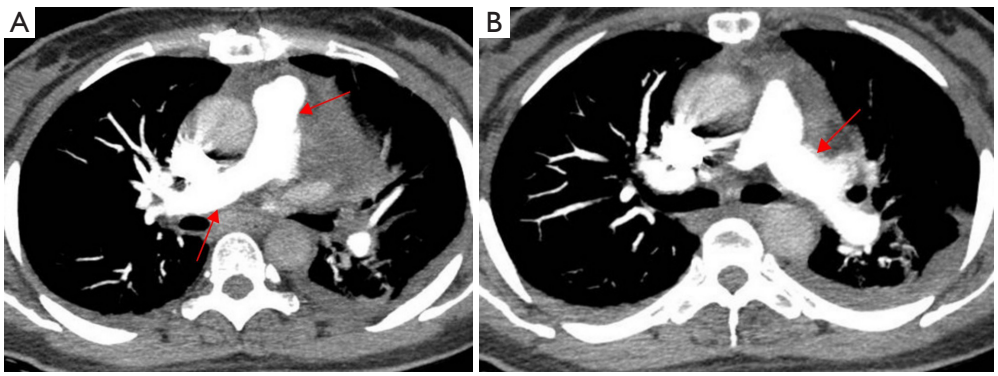

Figure 6 Computed tomography pulmonary angiography shows no filling defects in the main, right and left pulmonary arteries at 1 month after surgery (red arrows) (A,B).

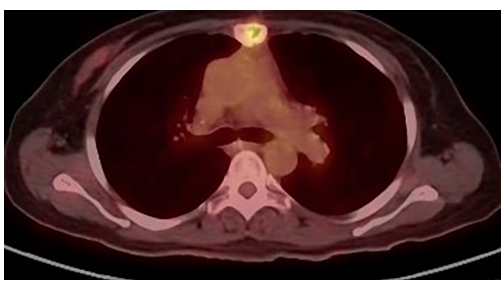

Figure 7 PET-CT shows no increased metabolic activity of the lesion in the innervation areas of the main trunk and left and right branches of the pulmonary artery. PET-CT, positron emission tomography-computed tomography.

values (SUV) (7.63±2.21 for PAS and 2.31±0.41 for PE), suggesting PET-CT may be an effective noninvasive means of early diagnosis of PAS (9).Additionally, It also needs to be differentiated from other diseases that can cause pulmonary artery embolism, such as right atrium or right ventricular myxoma and infective endocarditis of the right heart system.

PAS has a poor prognosis, and its treatment is mainly based on surgery, which can be followed by postoperative adjuvant radiotherapy, chemotherapy, molecularly targeted drugs, and immune checkpoint inhibitors (10). The survival is short (typically 1.5 months) in nonsurgical patients but can be extended up to 10 months after surgery (11). Adjuvant radiotherapy or chemotherapy may improve the prognosis. The current case had a recurrence of pulmonary artery tumor 10 months after surgery, and after adjuvant chemotherapy, she continued to survive for 14 months before dying.

Conclusions

The possibility of PAS should be considered in patients with clinically suspected PE but lacking PE triggers and responding poorly to anticoagulation or thrombolytic therapy. PET-CT is an effective noninvasive method for

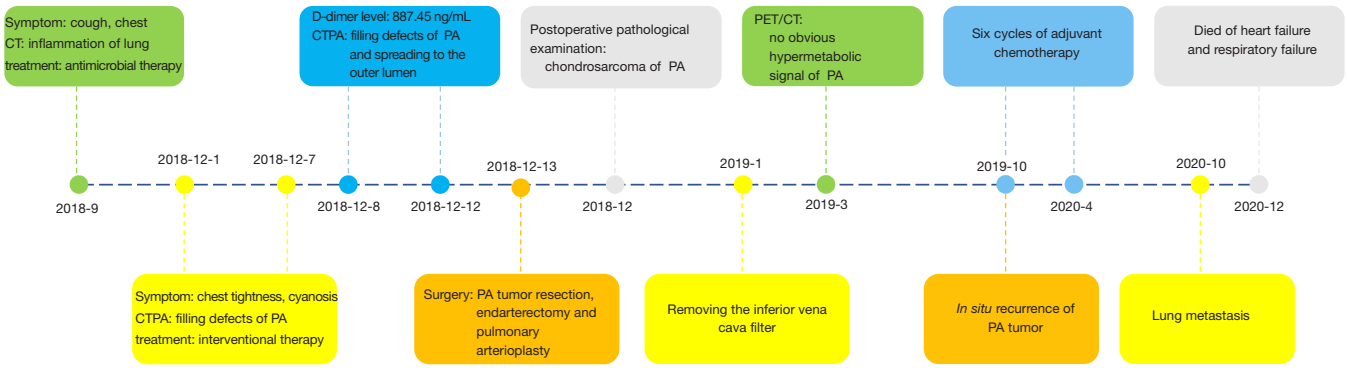

Figure 8 Timeline figure show the whole disease process of the case. Interventional therapy including transcatheter aspiration for pulmonary artery thrombus, transcatheter thrombolysis, and inferior vena cava filter placement. CT, computed tomography; PA, pulmonary artery; CTPA, computed tomography pulmonary angiography; PET, positron emission tomography.

early diagnosis of PAS. Once the diagnosis is confirmed, early surgical treatment should be scheduled.

Acknowledgments

Funding: This study was supported by Shenzhen Science and Technology Program (No. JCYJ20210324112611031).

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tcr.amegroups.com/article/view/10.21037/tcr-23-518/rc

Peer Review File: Available at https://tcr.amegroups.com/article/view/10.21037/tcr-23-518/prf

Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at https://tcr.amegroups.com/article/view/10.21037/tcr-23-518/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent for publication of this case report and accompanying images was not obtained from the patient or the relatives after all possible attempts were made.

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(English Language Editor: K. Brown)

Cite this article as: Zhang Y, Xu G. Primary pulmonary artery chondrosarcoma: case report. Transl Cancer Res 2023;12(4): 1060-1066. doi: 10.21037/tcr-23-518

