Pulmonary artery intimal sarcoma mimicking pulmonary thromboembolism: A case report and literature review

YAN HUANG[1*], YIMING WANG[1*], BIKO KONG[1], FENG ZHENG[2], YI DING[3], BOAMAH HAYFORD[4], JUNFANG GUO[1] and RONG QIN[5]

[1]Department of Cardiology, Jiangsu University Affiliated People's Hospital, Zhenjiang Clinical Medical College of Nanjing Medical University, Zhenjiang, Jiangsu 212002, P.R. China; [2]Department of Ultrasound, Jiangsu University Affiliated People's Hospital, Zhenjiang, Jiangsu 212002, P.R. China; [3]Department of Medical Imaging, Jiangsu University Affiliated People's Hospital, Zhenjiang, Jiangsu 212002, P.R. China; [4]Research Center of Fluid Machinery Engineering and Technology, Jiangsu University, Zhenjiang, Jiangsu 212013, P.R. China; [5]Department of Medical Oncology, Jiangsu University Affiliated People's Hospital, Zhenjiang, Jiangsu 212002, P.R. China

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Abstract. Pulmonary artery intimal sarcoma (PAIS) is a rare and highly aggressive form of hemangiosarcoma. The non-specific clinical manifestations, signs and routine imaging findings of this sarcoma often mimic those of pulmonary thromboembolism (PTE), resulting in frequent misdiagnosis as PTE prior to pathological confirmation in most patients. This delay in achieving an accurate diagnosis poses challenges for promptly initiating optimal treatment and further contributes to the unfavorable prognosis. The current study presents the case of a 68-year-old female who presented with acute chest tightness and dyspnea. Transthoracic echocardiography revealed the presence of atypical echogenic structures within the primary pulmonary artery, while computed tomography pulmonary angiography showed marked filling defects in the main trunk and branches of the pulmonary artery. The patient was initially misdiagnosed with PTE but did not respond well to anticoagulant therapy. Subsequent surgical resection confirmed the diagnosis of PAIS through pathological examination. Despite postoperative treatment with molecular-targeted antitumor drugs, the patient experienced tumor recurrence and intrapulmonary metastasis, ultimately succumbing to disease progression. This exceptional case is being presented to enhance the clinical understanding of PAIS, to encourage further extensive research for reliable diagnostic approaches and to provide further data that will help form efficacious therapeutic strategies to ameliorate the unfavorable prognosis of affected patients.

Correspondence to: Dr Rong Qin, Department of Medical Oncology, Jiangsu University Affiliated People's Hospital, 8 Dianli Road, Zhenjiang, Jiangsu 212002, P.R. China
E-mail: jiangsu.qinrong@163.com

Dr Junfang Guo, Department of Cardiology, Jiangsu University Affiliated People's Hospital, Zhenjiang Clinical Medical College of Nanjing Medical University, 8 Dianli Road, Zhenjiang, Jiangsu 212002, P.R. China
E-mail: zjyyxngif@163.com

*Contributed equally

Abbreviations: PAIS, pulmonary artery intimal sarcoma; PTE, pulmonary thromboembolism; TTE, transthoracic echocardiography; CTPA, computed tomography pulmonary angiography; MRI, magnetic resonance imaging; PET-CT, positron emission tomography-computed tomography; 18F-FDG, fluorine-18-fluorodeoxyglucose

Key words: PAIS, PTE, diagnosis, CTPA, prognosis

Introduction

Pulmonary artery intimal sarcoma (PAIS) is an uncommon and extremely aggressive mesenchymal tumor originating from the intimal layer of the pulmonary artery (1), with only a few hundred documented cases in the medical literature to date since being initially reported by Mandelstamm in 1923 (2). The early diagnosis of PAIS poses a significant challenge, with the sarcoma often being misidentified as a pulmonary thromboembolism (PTE) due to overlapping clinical symptoms (such as dyspnea, chest pain and hemoptysis) and imaging findings (such as pulmonary artery filling defect). This delayed and inaccurate diagnosis impedes timely intervention for patients, resulting in a poor prognosis and a marked shortening of overall survival time. Surgical excision currently remains the primary treatment strategy for PAIS. Early diagnosis and surgical resection provide substantial benefits to patients, potentially prolonging survival time (3). Furthermore, post-operative adjuvant therapies have the potential to improve clinical outcomes. Untreated PAIS results in a median survival time of only 6 weeks (3); however, advancements in surgical techniques and emerging treatment approaches hold promise for extending median survival time. Nevertheless, the overall long-term prognosis remains notably poor (4). The present study aims to deepen the clinical understanding of PAIS by

presenting a unique case report, thereby providing a valuable reference for the diagnosis and treatment protocols of healthcare professionals and ultimately optimizing patient outcomes.

Case report

A 68-year-old female patient was admitted to The Affiliated People's Hospital of Jiangsu University (Zhenjiang, China) in April 2020, presenting with chief complaints of chest tightness and dyspnea that had persisted for 4 days. The symptoms worsened post-activity and did not completely alleviate after rest, accompanied by palpitations, dizziness, fatigue and a poor appetite. The patient had sustained a right wrist fracture along with soft-tissue injuries to both lower extremities due to an accident 6 days ago that had necessitated splinting of the right wrist joint. Additionally, the patient had undergone a right-sided mastectomy 10 years prior, but had no history of hypertension or diabetes, nor any family history of hereditary diseases.

Upon the current admission, the patient presented with a blood pressure of 113/81 mmHg (normal range, 90-139/60-89 mmHg), a heart rate of 104 beats per min (normal range, 60-100 beats per min) and a respiratory rate of 19 breaths per min (normal range, 12-18 breaths per min). Oxygen saturation levels were maintained at 98% (normal range, 95-100%) while breathing room air. Physical examination revealed a grade 3 out of 6 sighing systolic murmur over the pulmonary valve area along the left sternal border. Laboratory tests showed elevated D-dimer levels at 4.7 mg/l (reference range, 0.1-0.55 mg/l), and the level of CA125 was 32.1 U/ml (reference range, 0-35 U/ml), while other parameters were within the normal ranges.

Venous ultrasound indicated no signs of thrombosis in the limbs. The electrocardiogram displayed sinus tachycardia, atrial premature contractions and ST-T wave abnormalities. Transthoracic echocardiography (TTE) detected right ventricular enlargement and an isoechoic mass measuring ~8.79 cm^2 within the main pulmonary artery (Fig. 1A), which exhibited well-defined margins and a fixed position without evidence of blood filling (Fig. 1B). Additionally, TTE imaging demonstrated severe pulmonary hypertension (mean pulmonary artery pressure, 94 mmHg; normal range, 18-25 mmHg) and marked tricuspid regurgitation (Fig. 1C). Computed tomography pulmonary angiography (CTPA) was performed to further investigate the suspicion of a PTE. The imaging revealed regions of decreased density in the main pulmonary artery, the proximal segment of the right pulmonary artery, the left pulmonary artery, the left upper lung and branches of the left lower pulmonary artery (Fig. 2A), with no nodules in the lungs (Fig. 2B). Furthermore, localized enlargement measuring a diameter of ~37 mm was noted in the main pulmonary artery. Minimal fluid accumulation was also identified in both the pericardium and right pleural cavity. The initial diagnosis was PTE, for which the patient received subcutaneous injections of low molecular weight heparin (4,000 IU every 12 h) and oral warfarin (2.5 mg per day) anticoagulation therapy. However, despite a week of treatment, the patient's clinical symptoms gradually worsened. Subsequent repeat TTE was performed to assess the efficacy of anticoagulant therapy, revealing an increase in the size of the isoechoic mass within the main pulmonary artery (~11.4 cm^2) (Fig. 1D). The patient was promptly transferred to the Department of Cardiothoracic Surgery for further intervention. Under general anesthesia and cardiopulmonary bypass, pulmonary endarterectomy was conducted at 9 days post-admission to alleviate the deteriorating clinical symptoms, revealing a 15x5-cm mass in the main pulmonary artery (Fig. 3), closely adherent to its intima and extending throughout both the left and right pulmonary arteries. The mass was successfully excised, with intraoperative transesophageal echocardiography confirming the absence of residual masses in the main pulmonary artery and no marked tricuspid valve regurgitation. The resected specimens were fixed in 10% formalin at room temperature for 24 h for histopathological examination. Histopathological staining and immunohistochemistry were performed using 4-μm thick paraffin-embedded sections, and the specific protocol was carried out in accordance with the method described by Neuville *et al* (5). The BOND-MAX fully automatic IHC and ISH staining system (Leica Microsystems) was used for immunohistochemical staining. Slides were inspected under a light microscope (Olympus BX51TF; Olympus Corporation). Hematoxylin and eosin staining revealed that the tumor cells exhibited an epithelioid and short fusiform morphology, with marked heterogeneity in cell density. Tumor cells were preferentially aggregated around blood vessels, accompanied by desmoplastic mucinous degeneration and localized hemorrhage. Notably, the tumor cells displayed pronounced atypia, with identification of pathological mitotic figures (Fig. 4A). Immunohistochemical examination revealed positive vimentin (cat. no. RTU-VIM-V9-QH; AQ Medical Technology) and Ki-67 (40%) (antibody dilution, 1:150; cat. no. NCL-L-Ki67-MIB1; AQ Medical Technology) staining, but negative staining for anaplastic lymphoma kinase (ALK; cat. no. GT226602; Gene Tech, Co., Ltd.), CD34 (cat. no. RTU-END-QH; AQ Medical Technology), CD117 (cat. no. RTU-CD117-QH; AQ Medical Technology), CD163 (cat. no. RTU-CD163-QH; AQ Medical Technology), cytokeratin (CK; antibody dilution, 1:200; cat. no. NCL-L-AE1/AE3-601; AQ Medical Technology), erythroblastosis transformation specific-1 related gene (ERG-1; cat. no. RTU-ERG-QH; AQ Medical Technology), p53 (antibody dilution, 1:100; cat. no. NCL-L-P53-DO7; AQ Medical Technology), S100 (cat. no. RTU-S100-QH; AQ Medical Technology) and smooth muscle actin (SMA; cat. no. RTU-SMA-QH; AQ Medical Technology) (Fig. 4B-L). The final diagnosis of PAIS was confirmed based on the results of the pathological examination and the specific tumor localization (6,7).

Following surgery, the patient experienced a satisfactory recovery with notable improvement in clinical symptoms. A postoperative TTE revealed no abnormal mass in the pulmonary artery and a marked decrease in pulmonary artery pressure (34 mmHg) compared with preoperative levels (Fig. 1E and F). Despite the recommendation for a positron emission tomography-CT (PET-CT) scan, the patient declined and was discharged in May 2020. Subsequently, the patient opted to undergo treatment with oral anlotinib (12 mg on days 1-14) according to the 2019 Chinese Society of Clinical Oncology treatment guidelines for soft-tissue sarcoma (8) and due to the demonstrated significant efficacy of anlotinib in

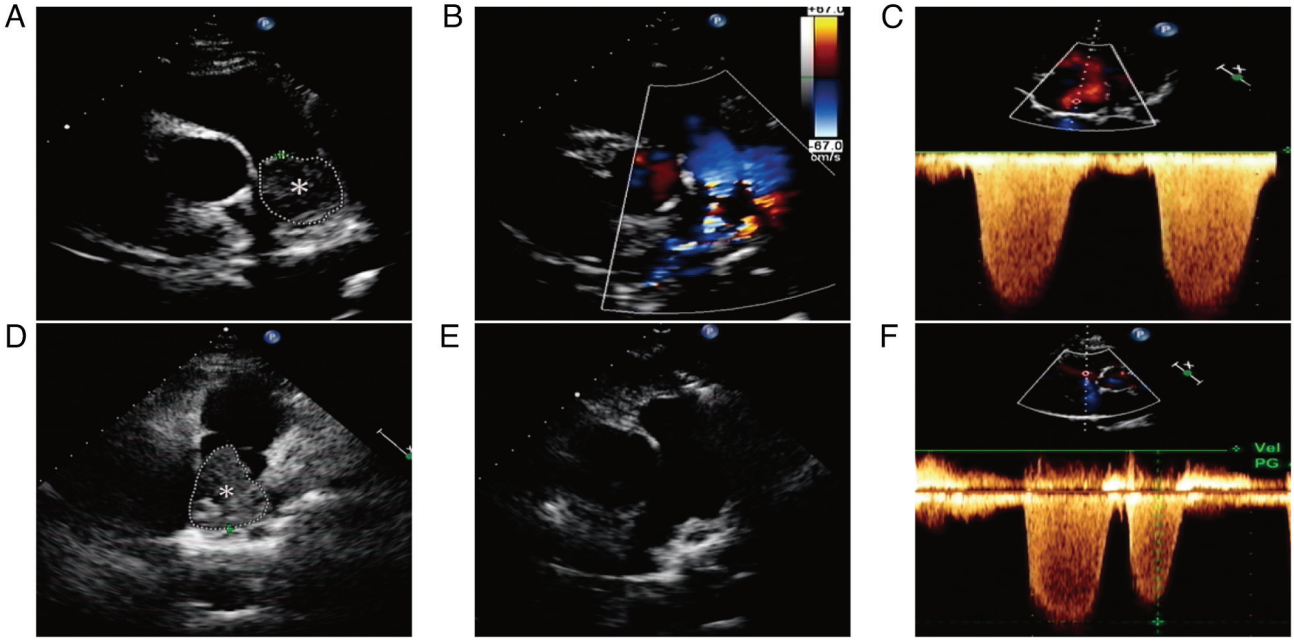

Figure 1. Transthoracic echocardiography results. (A) A large isoechoic mass (8.79 cm^2 in area) was found within the main pulmonary artery before admission, (B) with no blood flow detected in the mass. (C) Severe pulmonary hypertension estimated from tricuspid regurgitation. (D) The size of the isoechoic mass (11.4 cm^2) in the main pulmonary artery after anticoagulant therapy had increased compared with its previous state. (E) No abnormal mass echo was detected in the pulmonary artery postoperatively and (F) mild pulmonary hypertension estimated from tricuspid regurgitation. Asterisks indicate the location of the lesion.

treating soft-tissue sarcoma in multiple clinical trials (9). In December 2020, the patient reported chest tightness, dyspnea and chest/back pain. An enhanced CT scan of the chest performed on 3 days later revealed a low-density filling defect in the main pulmonary artery along with a 10-mm nodule in the left upper lobe initially suspected to be metastatic lesions (Fig. 2C and D). The patient opted to continue oral anlotinib treatment while remaining hesitant to modify the treatment regimen. A subsequent enhanced CT scan of chest and abdomen performed in March 2021 showed substantial enlargement of the low-density lesions within the main pulmonary artery (~68x67 mm), as well as multiple fresh soft-tissue masses in the left lung; however, no abnormalities were observed within the abdominal region (Fig. 2E and F). Concurrently, the levels of tumor marker CA125 were increased to 66.9 U/ml (reference value, 0-35 U/ml) and exhibited a progressive upward trend as the disease progressed. By April 2021, the level of this marker had peaked at 267 U/ml.

Despite receiving combined treatment with apatinib (250 mg orally per day) and camrelizumab (200 mg intravenously, twice), no marked improvement in the patient's condition was observed. Subsequently, the patient's condition continued to deteriorate, necessitating palliative care. The patient passed away in June 2021. The procedures for specific diagnosis and treatment are elaborated in Fig. 5.

Discussion

PAIS is a rare neoplasm characterized by intraluminal growth resulting in vascular occlusion or embolization to distant sites (6,10). PAIS originates from multipotent mesenchymal stem cells within the intimal layer of the pulmonary artery (7), which can exhibit complete undifferentiated or contain heterologous components such as rhabdomyosarcoma and chondrosarcoma (11). The reported occurrence rates range from 0.001 to 0.03% (1,12). The age of onset for the initial symptoms varies widely among individuals, spanning from infancy to late adulthood, with a mean age of onset around 48 years old (6). The prevalence of illness in women is slightly higher than that in men, with a ratio of 1.3:1 (6).

The clinical presentation of PAIS lacks specificity and is closely linked to the location and extent of tumor invasion. In the early stages, patients may be asymptomatic; however, as the tumor obstructs blood vessels, it can lead to a spectrum of respiratory issues, including dyspnea, chest or back pain, and hemoptysis. Additionally, non-specific symptoms, including palpitations, fatigue, fever and weight loss, may also contribute to the diagnosis of PAIS (13).

The clinical signs and imaging features of PAIS closely resemble those of PTE, usually leading to the misdiagnosis as PTE, which poses a significant challenge for early diagnosis and prompt, effective intervention (14,15). Radiological examination technologies, such as TTE, CTPA, magnetic resonance imaging (MRI) and PET-CT, can considerably aid in distinguishing between the two conditions (6).

TTE has been validated as a reliable modality for identifying pulmonary artery lesions, enabling visualization of their location, size, shape and activity. Additionally, it can detect blood flow signals within the mass and aid in assessing the impact of these lesions on pulmonary artery hemodynamics, as well as cardiac structure and function (16). In echocardiographic images, PAIS typically presents as areas of moderate to low echogenicity, with irregularity and potential activity; it primarily affects the main pulmonary artery, with rare involvement of the right ventricular outflow tract and pulmonary valve (10). Color Doppler imaging

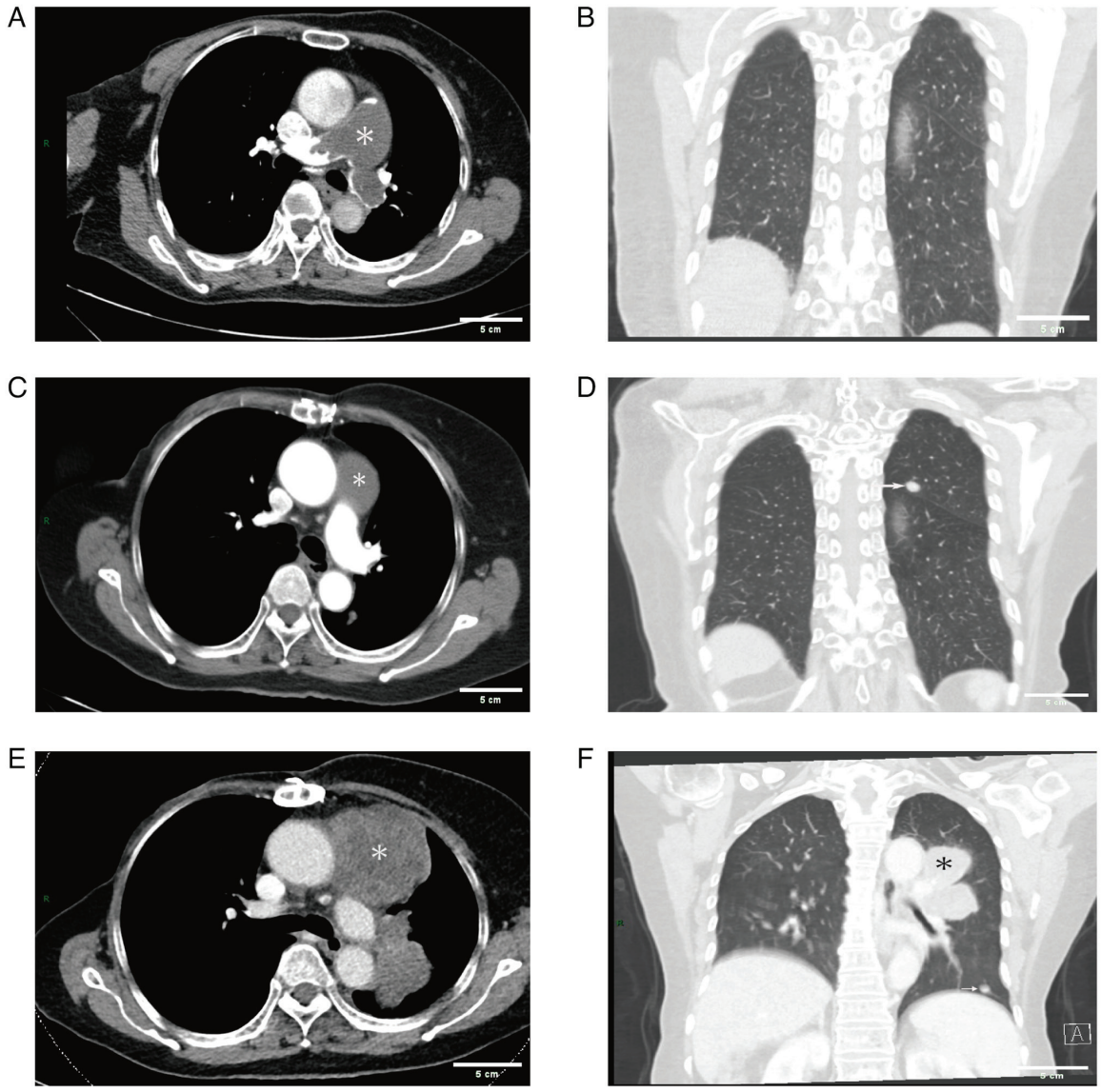

Figure 2. Chest imaging results. (A) Preoperative CTPA showed dilation of the pulmonary trunk (~37 mm), filling defects in the main pulmonary artery, as well as the left and right pulmonary arteries, (B) with no nodules in the lung. (C) Chest-enhanced CT in December 2020 showed a low-density filling defect in the main pulmonary artery and (D) a 10-mm nodule in the upper left lobe of the lung. (E) Repeat chest-enhanced CT in March 2021 showed a significant enlargement of the low-density lesions (~68x67 mm) in the main pulmonary artery, (F) as well as multiple fresh soft-tissue masses in the left lung. Asterisks indicate the location of the lesion. Arrows point to the lung nodule. CTPA, computed tomography pulmonary angiography.

can reveal discernible blood flow signals within these abnormal areas, while contrast-enhanced echocardiography can provide a more detailed display of neovascularization within the mass (7). By contrast, thrombi generally appear as fixed avascular homogeneous hypoechoic lesions, without involving the right ventricular outflow tract or pulmonary valve (16). In the present case, an isoechoic mass was present in the main pulmonary artery, rather than a hypoechoic mass, which indicated a sarcoma, which aligns with the finding of Bai and Ruan (7).

CTPA is currently the predominant imaging modality for diagnosing pulmonary artery diseases. Previous research has indicated that CTPA can differentiate between PAIS and PTE based on their location and morphological characteristics. PAIS usually affects two or more pulmonary arteries, with the main pulmonary artery trunk, and the left or right pulmonary artery being the most common sites of involvement. In rare

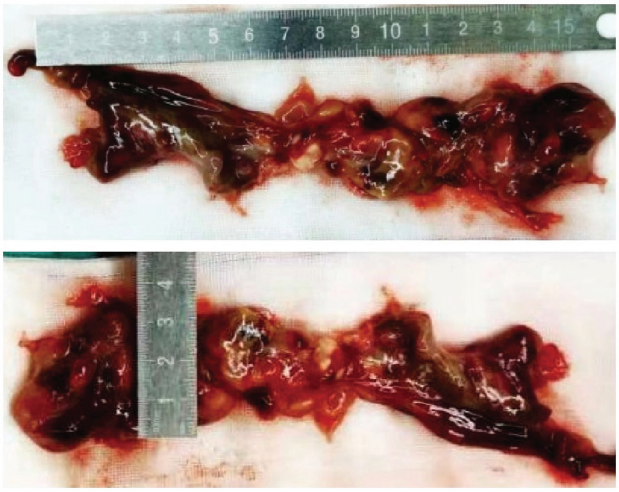

Figure 3. Surgical specimen of the pulmonary artery intimal sarcoma.

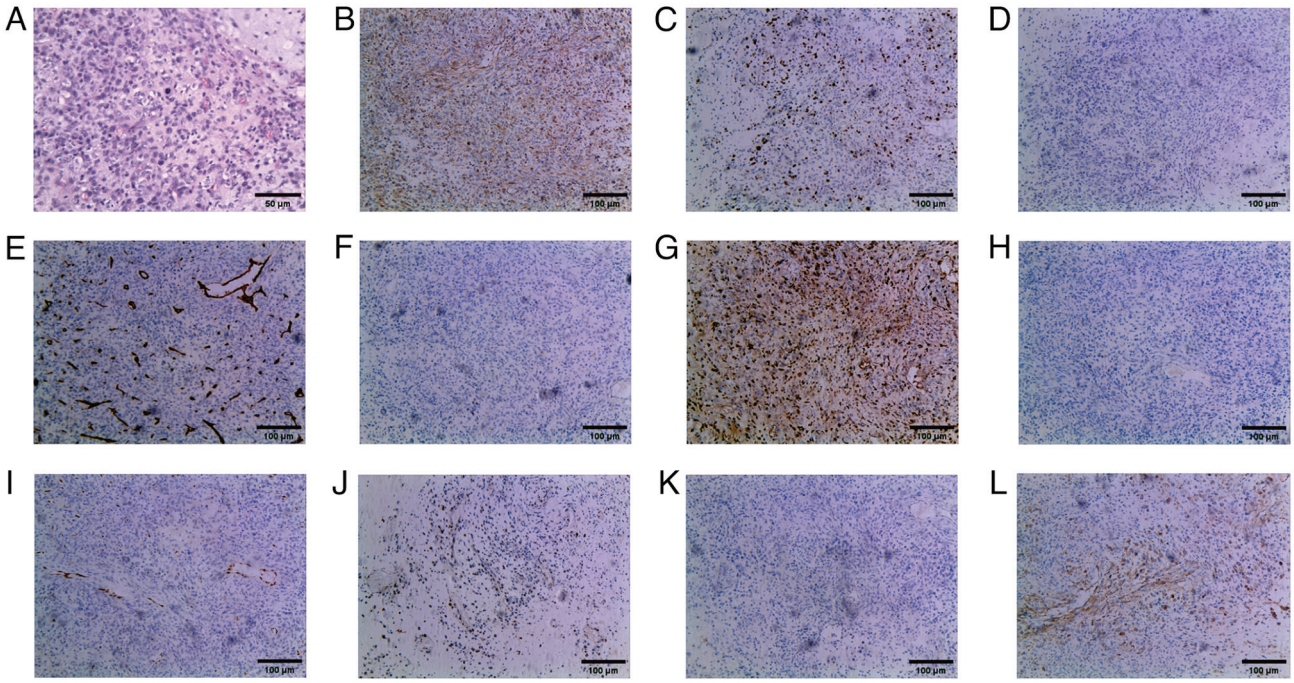

Figure 4. Microscopy examination of the pulmonary artery intimal sarcoma specimen. (A) Postoperative hematoxylin and eosin staining revealed that the tumor cells exhibited an epithelioid and short fusiform morphology, with mucinous degeneration and local hemorrhage. These cells displayed pronounced atypia, with identification of pathological mitotic figures (original magnification, x200). The immunohistochemical staining of (B) vimentin (positive), (C) Ki-67 (40% positive), (D) anaplastic lymphoma kinase (negative), (E) CD34 (negative), (F) CD117 (negative), (G) CD163 (negative), (H) cytokeratin (negative), (I) erythroblastosis transformation specific-1 related gene (negative), (J) p53 (negative), (K) S100 (negative) and (L) smooth muscle actin (negative) (magnification, x100).

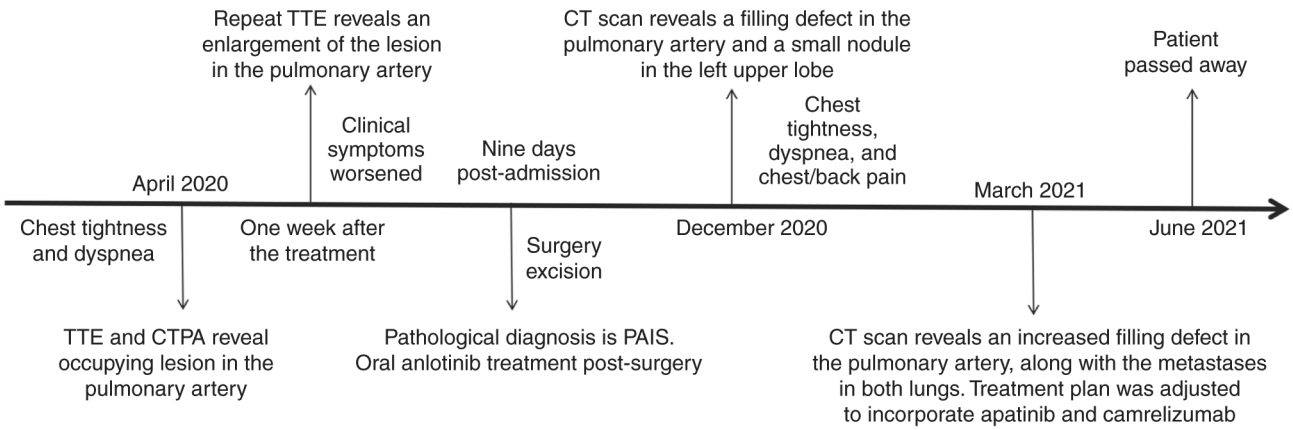

Figure 5. Timeline of clinical history for pulmonary artery intimal sarcoma. Symptoms, imaging modalities and treatment of the patient is represented. TTE, transthoracic echocardiography; CTPA, computed tomography pulmonary angiography; PAIS, pulmonary artery intimal sarcoma.

cases, it may also extend in a retrograde manner to the right ventricle and pulmonary valve (1). These lesions appear as cauliflower-like polyps with bulging or lobulating contours, showing heterogeneous attenuation on imaging, accompanied by the 'wall eclipse sign' and intratumoral vessels, along with localized aneurysm dilatation of the affected vessels (17-19). However, PTE primarily affects the right or both lower lungs and rarely involves the main pulmonary artery. PTE presents as a tubular-polypoid shape with straight proximal edges, homogeneous attenuation on imaging, but without the 'wall eclipse sign', and intratumoral vessels. In the present case, multiple filling defects were observed in the main pulmonary artery and in both left and right pulmonary arteries, along with dilatation of the affected arteries, indicative of sarcoma, which is consistent with the previous literature (18). However, no heterogeneous attenuation or 'wall eclipse sign' characteristics of the lesion were observed on the CTPA, which could potentially result in a misdiagnosis of PTE.

MRI provides excellent soft-tissue contrast and can effectively distinguish between tumors and thrombi due to its unique signal characteristics. In MRI, PAIS demonstrates restricted diffusion and heterogeneous enhancement, a condition not previously observed in PTE (20). Additionally, MRI can provide superior diagnostic information in specific sequences (fat-suppressed T2-weighted imaging) without the need for iodinated contrast agents, reducing potential

risks such as allergies and contrast-induced nephropathy for patients. However, thoracic MRI is more time-consuming compared with CT, and may be susceptible to respiratory and motion artifacts (21).

PET-CT is crucial for accurately distinguishing PAIS from PTE and serves as an invaluable tool for assessing potential systemic metastasis. PET-CT imaging reveals that a sarcoma exhibits high positive uptake of fluorine-18-fluorodeoxyglucose (18F-FDG), whereas a thrombus demonstrates no uptake (19). The study by Ren *et al* (22) demonstrated that the maximum standardized uptake value of PAIS (median, 8.0; range, 3.0-17.2) was significantly elevated compared with that of PTE (median, 1.8; range, 0.8-3.7; P<0.001). When a cut-off value of 2.9 was applied (identified using the Youden index), the sensitivity and specificity were recorded at 100.0 and 93.9%, respectively (22). This finding aligns with those from Ito *et al* (23) and Xi *et al* (24), indicating that the maximum standardized uptake value of PET-CT can serve as a critical guiding parameter for differentiating PAIS from PTE in clinical settings. However, it is essential to acknowledge that a previous study has reported lower uptake of 18F-FDG in PAIS cases (25). This may be related to the presence of sarcoma-intermingled thrombi (25). Consequently, it must be recognized in clinical practice that relying solely on low 18F-FDG uptake should not be the only determining factor for excluding PAIS.

The final diagnosis of PAIS relies on the pathological findings (6,7). Current primary biopsy methods include percutaneous intravascular aspiration, intravascular ultrasound-guided needle aspiration biopsy, percutaneous intravascular biopsy and surgical biopsy. Percutaneous intravascular aspiration has a certain failure rate due to its inconsistent ability to obtain sufficient tumor tissue (26). Intrabronchial ultrasound-guided needle aspiration biopsy enables real-time guidance and visualization of pulmonary artery flow and artery stenosis; however, the obtained tissue is also subject to limitations, such as insufficient core tissue (27). Percutaneous intravascular biopsy can be repeated to obtain multiple tissue samples, making it a relatively ideal method for biopsies. However, potential complications such as pulmonary artery perforation, bleeding and tumor dissemination still exist (28).

The prognosis for patients diagnosed with PAIS is typically fairly unfavorable. In one study, those who did not undergo surgical intervention had a median survival duration of only 6 weeks, attributable to the rapid progression of the condition. By contrast, patients who underwent surgical resection exhibited a survival spectrum ranging from 8 to 36 months, with a median postoperative survival period of ~10 months (29). Surgically excising the tumor not only relieves the clinical symptoms caused by vascular blockage, but also markedly prolongs the overall survival of the patients. Radical surgery remains the preferred therapeutic approach for PAIS due to its exceptional efficacy. However, early detection is challenging due to its subtle presentation and lack of obvious symptoms or characteristic indicators (3,6). Some patients diagnosed with metastasis may miss the opportunity for surgical intervention. Therefore, accurate and timely identification of patients with PAIS who are suitable for surgery is crucial for improving their prognosis. Studies have indicated that utilizing a combination of multiple treatment modalities, such as surgery, radiotherapy and chemotherapy, may yield superior survival outcomes compared with the use of a single treatment modality, particularly in cases where the disease is inoperable or relapse occurs after initial surgical intervention (3,30). In recent years, the use of molecular targeted therapies has emerged as a promising strategy for managing PAIS and has demonstrated favorable clinical outcomes. Sanada *et al* (31) conducted experiments using cell lines and xenograft models to investigate PAIS, and observed an upregulation of specific tyrosine kinase receptor expression in PAIS cells. The specific tyrosine kinase receptor inhibitor pazopanib displayed marked effectiveness in inhibiting the growth of PAIS cells and xenograft tumors, consistent with the findings by Funatsu *et al* (32). While Wu *et al* (33) suggested potential therapeutic effects of anlotinib for PAIS with lung metastasis, the present case study revealed that postoperative application of anlotinib did not yield the anticipated results. Therefore, further clinical trials are warranted to evaluate the precise impact of molecular targeted drugs in treating PAIS.

In the current case, the patient presented with symptoms of chest tightness and dyspnea, along with a history of prior upper limb fracture fixation preceding the onset of the condition. A slight elevation in D-dimer levels was observed, and evidence of pulmonary artery obstruction was detected by both TTE and CTPA, indicating a preliminary diagnosis of PTE. Despite undergoing anticoagulation therapy, the therapeutic response was unsatisfactory. Subsequently, surgical biopsy disclosed the presence of PAIS. Upon re-evaluation of the case, it was noted that the patient presented with non-specific symptoms, including low tolerance and fatigue. The increment in D-dimer was not substantial, and limb ultrasound examination did not reveal any signs of thrombosis. TTE demonstrated moderate echogenicity within the pulmonary artery rather than the expected low echogenicity. The presence of multiple filling defects on CTPA, coupled with a lack of imaging improvement despite anticoagulant therapy, raised the suspicion of PAIS. However, due to the rarity of PAIS in clinical practice and the non-specific nature of its hidden symptoms, diagnosis frequently hinges on histopathological tissue biopsy, resulting in a delayed diagnosis. The MRI examination could not be completed due to the patient's dyspnea. An immediate surgical resection was performed after the intraoperative diagnosis, followed by adjunctive anlotinib targeted therapy. Despite these interventions, the therapeutic response was unsatisfactory and led to tumor recurrence and local metastasis, ultimately resulting in the patient's death. It is essential to note that a gradual increase in the patient's CA125 level was observed as the disease progressed. Currently, there is a paucity of literature investigating the efficacy of tumor markers in diagnosing PAIS. Further comprehensive research is imperative to ascertain whether CA125 can be employed as a potential biomarker for PAIS.

The present case report has some limitations. Firstly, previous literature has reported that PAIS may exhibit amplification of murine double minute 2 (MDM2) and platelet-derived growth factor receptor α (PDGFRA) genes, implying potential efficacy of targeted therapy against MDM2 or PDGFRA (34,35). However, cytogenetic testing was not performed in the present case. Secondly, the therapeutic effect and mechanism of action of anlotinib in PAIS remain unclear and require further investigative clinical trials, despite the substantial clinical efficacy of anlotinib in the treatment of

advanced soft-tissue sarcoma, as supported by a number of clinical trials (9,36,37).

In conclusion, PAIS is a rare malignant neoplasm that affects the pulmonary vascular intima, leading to physiological changes and clinical symptoms similar to those observed in PTE. Due to the relatively low prevalence of PAIS, healthcare professionals may possess limited knowledge and experience in managing this specific medical condition, increasing the risk of misdiagnosis. In scenarios where clinical symptoms resemble PTE and imaging shows a blockage in the pulmonary artery, with normal or only slightly elevated D-dimer levels, but where patients do not respond well to anticoagulant therapy, alternative non-thrombotic diseases should be considered. A comprehensive analysis of the patient's medical history and imaging data is essential at this point, with particular attention given to evaluating the chest CT pulmonary window. MRI and PET-CT can serve as second-line imaging methods for differential diagnosis. If new abnormalities are detected, a timely pathological biopsy should be performed for a definitive diagnosis. This helps effectively reduce the likelihood of misdiagnosis and facilitate prompt diagnostic accuracy.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

JG and RQ conceptualized the study. YH and YW acquired and analyzed the data, and wrote reviewed and edited the manuscript. BK, FZ, YD and BH obtained medical images (such as TTE and CT scans) and analyzed patient data. BK, FZ, YD and BH confirm the authenticity of all the raw data. All authors have read and approved the final version of the manuscript.

Ethics approval and consent to participate

This study was approved by the Ethics Committee of the Affiliated People's Hospital of Jiangsu University (Zhenjiang, China; approval no. K-2022003-W). Written informed consent for participation was obtained from the patient.

Patient consent for publication

Written informed consent for publication of this case report was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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