EJCRIM
European Journal
of Case Reports
in Internal Medicine

PULMONARY ARTERY INTIMAL SARCOMA MIMICS RECURRENT PULMONARY ARTERY EMBOLISM

Styliani Ioakeimidou[1,2], Frank-Peter Stephan[3], Emanuel Pluess[1], Marcus Seeman[4], David Srivastava[1]

[1] Department of Internal Medicine, Buergerspital Solothurn, Solothum, Switzerland
[2] Department of Internal Medicine, University Hospital of Zurich, Zurich, Switzerland
[3] Department of Cardiology, Buergerspital Solothurn, Solothum, Switzerland
[4] Department of Radiology and Nuclear Medicine, Buergerspital Solothurn, Solothum, Switzerland

Corresponding author's e-mail: Styliani.ioakeimidou@usz.ch

Received: 15/02/2025
Accepted: 28/04/2025
Published: 03/06/2025

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent was obtained from the patient.
This article is licensed under a Commons Attribution Non-Commercial 4.0 License

How to cite this article: Ioakeimidou S, Stephan FP, Pluess E, Seeman M, Srivastava D. Pulmonary artery intimal sarcoma mimics recurrent pulmonary artery embolism. *EJCRIM* 2025;**12**:doi:10.12890/2025_005150

ABSTRACT

Deep vein thrombosis and pulmonary embolism (PE) are the third most common cardiovascular condition. Pulmonary artery intimal sarcoma (PAIS) is a rare tumour and is often misdiagnosed as a PE.

This case report presents a 50-year-old man with fever, dyspnoea, cough and thoracic pain since a month. The patient had a history of two PE episodes. After diagnostic screening for tumour, including laboratory parameters, transthoracic echocardiography, computed tomography scan and positron emission tomography-computed tomography scan, PAIS was diagnosed. Unexplained recurrent PE should raise suspicion of a tumour or PAIS.

KEYWORDS

Pulmonary embolism, cardiovascular events, angiosarcoma

LEARNING POINTS

- Recurrent pulmonary embolism should raise suspicion for tumours.
- Imaging and histopathology play a crucial role in diagnosis of pulmonary artery intimal sarcoma.
- Surgical resection as the primary treatment for pulmonary artery intimal sarcoma.

INTRODUCTION/BACKGROUND

Deep vein thrombosis (DVT) and pulmonary embolism (PE) are the third most common cardiovascular condition. The annual incidence of PE ranges from 39 to 115 per 100,000, and DVT from 53 to 162 per 100,000[1-3]. Pulmonary artery intimal sarcoma, a rare tumour with a prevalence of 0.001-0.003%[4] (1-3/100.000) is often misdiagnosed as PE, resulting in inappropriate anticoagulation therapy and delayed diagnosis. This case report presents a 50-year-old man, with fever, dyspnoea, cough and thoracic pain since a month. The patient had a history of two PE episodes. After diagnostic screening for tumour, including laboratory parameters, transthoracic echocardiography, computed tomography (CT) scan and positron emission tomography (PET)-CT scan, PAIS was diagnosed. This case underscores the diagnostic challenges presented by PAIS. Unexplained

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

recurrent PE should raise suspicion for a tumour or PAIS, leading to a better prognosis of the patient.

CASE DESCRIPTION

A 50-year-old man was admitted to the hospital with fever, dyspnoea, cough, and thoracic pain. These symptoms had been recurring for a month. The patient reported night sweats, fever and chest pain on the left side while breathing since 7 months. Clinical findings included rales over the left lung. Laboratory analysis showed an elevation of inflammatory parameters.

The patient had experienced two episodes of PE, the first 6 months previously, and the second one 5 months previously. After his initial visit to the general practitioner 6 months ago, treatment for pneumonia with amoxicillin-clavulanic acid was initiated. However, the symptoms worsened. A CT scan of the chest *(Fig. 1)* showed infiltrates indicating pneumonia and a PE in the left pulmonary artery 6 months earlier. Thus, the patient was given moxifloxacin and apixaban was started. Five months before presentation, the patient had been diagnosed again with PE during a trip to his country of origin. After this diagnosis, the patient had pain on the left side of the chest, so that he could not lie on the left side. Since the onset of symptoms, he also noticed exertional dyspnoea, which sometimes occurs when walking on level ground.

Three months before presentation, lung function tests showed normal dynamic and static lung volumes with normal lung volumes with normal CO_2 diffusion capacity. Blood gas analysis results were also normal, although the PaO_2 had decreased significantly compared to the previous examination. On CT-scan *(Fig. 2)*, there was a persistent occlusion of the left lower lobe pulmonary artery, where infiltrates were detected in the previous examinations, there were currently pleural consolidations, which were probably related to the chronic reduced perfusion in this area and caused persistent pleuritic symptoms. On ultrasound examination, the pleural effusion in the left basal region did not appear completely anechoic. Therefore, contrast-enhanced ultrasound was performed. In the absence of contrast uptake, there was no indication of malignancy in that area.

Tumour screening, including gastroscopy, colonoscopy, dermatologic examination, and prostate-specific antigen, was unremarkable. The patient had been on enoxaparin until the bridging in phenprocoumon, because of a potential diagnosis of thrombophilia, due to unexplained thrombosis under anticoagulation. Chronic thromboembolic pulmonary hypertension (CTPEH) or chronic thromboembolic pulmonary disease (CTPED) was a potential diagnosis. During hospitalization, transthoracic echocardiography *(Fig. 3)* showed normal left ventricular function and no right heart strain, with pulmonary pressure at the upper normal limit (s/m/dPAP 38/19/6 mmHg). The pulmonary thrombus was deemed old, and catheter-based thrombolysis was not performed due to high interventional risk.

A detailed CT scan review identified a localized solid

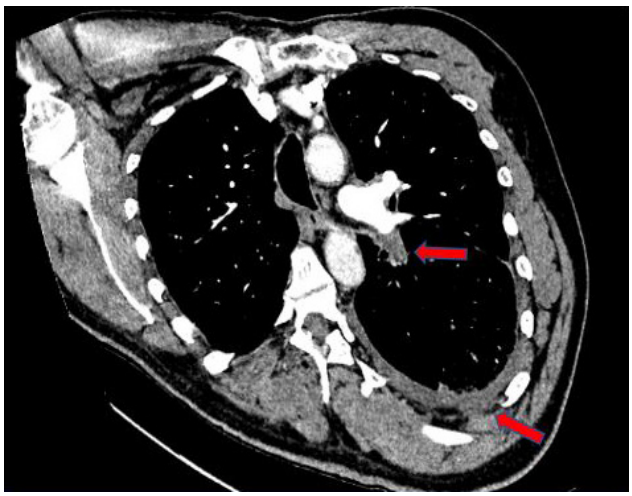


Figure 1. Computed tomography pulmonary angiography 09/2021. Thrombotic occlusion of the left inferior pulmonary artery with a small pleural effusion (arrows).

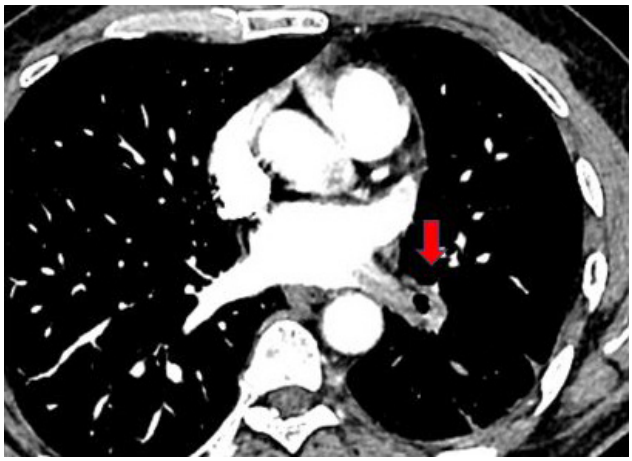


Figure 2. Computed tomography pulmonary angiography 12/2021. Persistent thrombotic occlusion of the left inferior pulmonary artery, new findings: partial thrombotic occlusion of the left superior pulmonary artery (arrow).

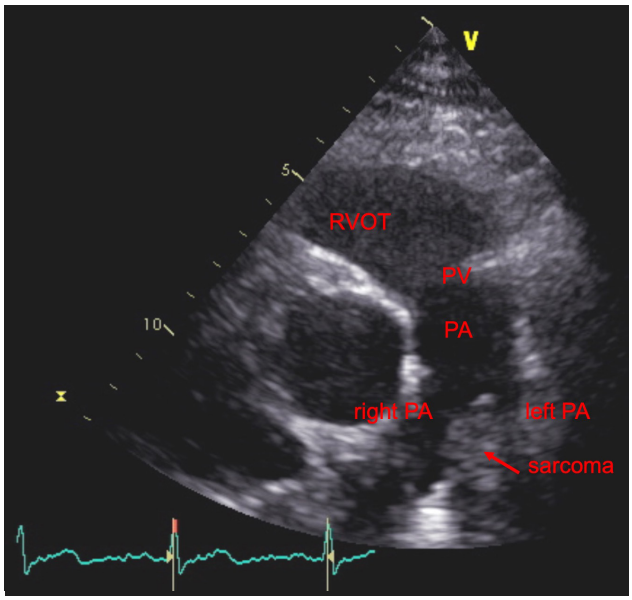


Figure 3. Transthoracic echocardiography 03/2022 (parasternal short axis). RVOT right ventricular outflow tract. Abbreviation: PV, pulmonary valve; PA, pulmonary artery.



Figure 4. Computed tomography pulmonary angiography 03/2022. Complete occlusion of the left pulmonary artery.

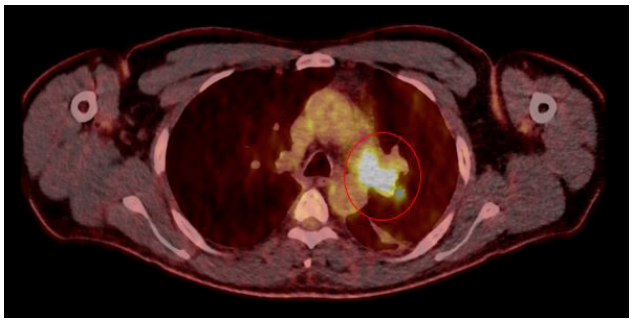


Figure 5. F-18 FDG positron emission tomography-computed tomograohy03/2022. Tumour-related malignant metabolic increase in the left pulmonary artery (red circle).

lesion near the left bronchus causing truncus pulmonalis constriction. Follow-up CT *(Fig. 4)* showed rapid lesion progression. PET-CT scan *(Fig. 5)* indicated metabolic activity in the solid lesion and ipsilateral lymph nodes, suggesting malignancy. Due to its location and poor bronchoscopic access, a pulmonary embolectomy was performed, revealing a pulmonary intimal sarcoma. Subsequent pulmonary artery resection and left pneumonectomy followed.

The histology revealed a pulmonary intimal sarcoma. Therefore, resection of the pulmonary artery and pneumonectomy on the left side followed. The patient had an Eastern Cooperative Oncology Group (ECOG) score of 1 after surgery. He was unable to return to work due to the deterioration of lung function. In an interdisciplinary discussion, radiotherapy and chemotherapy were recommended.

DISCUSSION

PAIS is a rare, aggressive tumour originating from the pulmonary artery's intimal layer. Classified by the World Health Organization as a tumour within blood vessel lumens, PAIS often mimics PE, leading to diagnostic challenges due to non-specific symptoms. Median age at onset is around 48 years[4].

Our patient had persistently elevated D-dimer levels (initially 1720 µg/l, later 432–470 µg/l), complicating differentiation between PE and PAIS. This detail was not discussed by Tuft et al.[5] and highlights how D-dimer may mislead diagnosis in recurrent PE cases. Troponin T was <4 ng/l and B-type natriuretic peptide <10 ng/l, indicating no significant cardiac strain, despite significant pulmonary artery obstruction. In contrast, Tuft et al.[5] report elevated troponin values, suggesting variable cardiac involvement. In addition, this case underscores the limitations of relying on D-dimer in recurrent PE and offers new clinical and radiologic insights that may aid in earlier suspicion of PAIS. In contrast to Tuft et al.[5], we emphasize biomarker interpretation in the diagnostic algorithm and provide additional imaging characteristics that support the diagnosis.

Despite advances in imaging, differentiating PAIS from thromboembolic diseases remains difficult. CT pulmonary angiography (CTPA) is crucial for identifying pulmonary artery lesions, but distinguishing PAIS from thromboembolic diseases is challenging[6]. The literature indicates that 25-40% of PEs are unprovoked[7,8], highlighting the need to consider malignancy in unexplained recurrent embolic events.

Key diagnostic features were observed including the "wall eclipsing sign" and non-dependent intraluminal defects, consistent with the findings of Tuft et al.[5]. This case further revealed additional findings such as complete luminal obstruction by a low-attenuation lesion, arterial expansion, and extraluminal tumour extension—features not fully detailed in previous reports.

The link between malignancy and thromboembolism is well-documented. This case emphasizes the need for thorough malignancy evaluation in recurrent thromboembolic events. Surgical resection is the primary treatment for PAIS, potentially extending survival despite variable outcomes. Post-surgical survival ranges from weeks to over 10 years. Our patient's successful tumour and pulmonary artery segment resection resulted in significant clinical improvement, with the patient surviving 1 year post-surgery with an ECOG score of 1.

The complexity and rarity of PAIS necessitate increased clinical awareness and consideration of this diagnosis in atypical PE presentations. Early diagnosis and prompt surgical intervention are crucial for improving patient outcomes. Further research and case studies are needed to enhance understanding and management of this challenging condition.

REFERENCES

1. Raskob GE, Angchaisuksiri P, Blanco AN, Buller H, Gallus A, Hunt BJ, et al.; ISTH Steering Committee for World Thrombosis Day. Thrombosis: a major contributor to global disease burden. *Arterioscler Thromb Vasc Biol* 2014;**34**:2363-2371. doi: 10.1161/ATVBAHA.114.304488
2. Wendelboe AM, Raskob GE. Global Burden of Thrombosis: Epidemiologic Aspects. *Circ Res* 2016;**118**:1340-1347. doi: 10.1161/CIRCRESAHA.115.306841
3. Keller K, Hobohm L, Ebner M, Kresoja KP, Münzel T, Konstantinides SV, et al. Trends in thrombolytic treatment and outcomes of acute pulmonary embolism in Germany. *Eur Heart J* 2020;**41**:522-529. doi: 10.1093/eurheartj/ehz236
4. Rabbani M, Hafiz A, Algadheeb M, Tugaleva E, Bergin ML, Ray Guo LR. Pulmonary Artery Intimal Sarcoma: A Deadly Diagnosis in Disguise. *CJC Open* 2020;**2**:711-715. doi: 10.1016/j.cjco.2020.07.008
5. Timmers L, Bové T, De Pauw M. Intimal sarcoma of the pulmonary artery: a report of two cases. *Acta Cardiol* 2009;**64**:677-679. doi: 10.2143/AC.64.5.2042701
6. Wong HH, Gounaris I, McCormack A, Berman M, Davidson D, Horan G, et al. Presentation and management of pulmonary artery sarcoma. *Clin Sarcoma Res* 2015;**5**:3. doi: 10.1186/s13569-014-0019-2
7. Penel N, Taieb S, Ceugnart L, Dansin E, Hoguet D, Vanseymortier L, et al. Report of eight recent cases of locally advanced primary pulmonary artery sarcomas: failure of Doxorubicin-based chemotherapy. *J Thorac Oncol* 2008;**3**:907-911. doi: 10.1097/JTO.0b013e318180720d
8. Aharoni M, Horesh N, Rogowski O, Kremer A, Mayan H, Justo D. Unprovoked pulmonary embolism in older adults: incidence and prognosis. *Arch Med Sci* 2021;**17**:337-342. doi: 10.5114/aoms/90673