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PULMONARY EMBOLISM OR PULMONARY ARTERY INTIMAL SARCOMA? DIAGNOSTIC PITFALLS IN A RARE VASCULAR MALIGNANCY

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ABSTRACT

Pulmonary artery obstruction is commonly attributed to pulmonary embolism; however, rare malignant conditions may present with similar clinical and radiological findings. Failure to recognize these alternative diagnoses may result in inappropriate treatment and delayed management.

We report the case of a 48-year-old man presenting with acute dyspnoea and cough. Computed tomography angiography revealed a filling defect in the right main pulmonary artery, and pulmonary embolism was diagnosed. Despite appropriate anticoagulant therapy, the patient's clinical condition progressively deteriorated. The absence of deep vein thrombosis and the lack of right heart strain prompted diagnostic reconsideration. Endobronchial ultrasound-guided fine-needle aspiration revealed malignant spindle cell proliferation, and immunohistochemistry confirmed pulmonary artery intimal sarcoma.

This case highlights the importance of reconsidering the diagnosis when clinical improvement does not occur despite adequate anticoagulation. Pulmonary artery obstruction should not automatically be attributed to thromboembolism, and malignancy must be considered in the differential diagnosis.

KEYWORDS

Pulmonary artery intimal sarcoma, pulmonary embolism, differential diagnosis

LEARNING POINTS

- Pulmonary artery obstruction should not automatically be attributed to thromboembolism, as rare malignancies may present with similar findings.
- Lack of clinical improvement despite appropriate anticoagulant therapy should prompt immediate diagnostic reconsideration.

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INTRODUCTION

Pulmonary embolism (PE) is the most frequent cause of acute pulmonary artery obstruction and is routinely diagnosed when a filling defect is identified on computed tomography angiography. However, not all pulmonary artery obstructions are thromboembolic in nature. Rare entities, including pulmonary artery intimal sarcoma, may closely mimic PE both clinically and radiologically[1].

Failure to recognize these alternative diagnoses may lead to inappropriate anticoagulation and delay potentially life-saving interventions. We report a case illustrating the importance of diagnostic reconsideration when clinical evolution is inconsistent with PE[2].

CASE DESCRIPTION

A 48-year-old man presented to the emergency department with a two-week history of acute dyspnea and cough, initially treated as pneumonia without clinical improvement. He had a history of smoking and no prior episodes of thromboembolism. Based on the initial clinical assessment, pulmonary embolism was suspected, and a computed tomography (CT) scan of the chest was performed[3].

The CT scan revealed obstruction of the right main pulmonary artery, resulting in significant impairment of blood flow and leading to a diagnosis of pulmonary embolism *(Fig. 1)*.

Transthoracic echocardiography demonstrated normal left and right ventricular systolic function. Right ventricular systolic pressure could not be estimated *(Fig. 2)*.

Venous Doppler ultrasonography of the lower extremities was negative for deep vein thrombosis *(Fig. 3)*.

Despite initiation of direct oral anticoagulant therapy, the patient's dyspnoea and chest pain progressively worsened. A repeat CT scan performed two weeks later showed subtotal resorptive atelectasis of the right hemithorax associated with a moderate haemorrhagic pleural effusion.

The lack of clinical response to adequate anticoagulation represented a critical turning point in the diagnostic process and prompted reconsideration of the initial diagnosis.

Due to bleeding complicated by secondary anaemia, and in light of the massive pulmonary artery occlusion in the absence of deep vein thrombosis and echocardiographic signs of right heart strain, anticoagulant therapy was discontinued. Consequently, the suspicion of a non-thromboembolic etiology was raised.

Fine-needle aspiration of the mass was performed under endobronchial ultrasound guidance.

Cytological examination revealed malignant spindle cell proliferation *(Fig. 4)*. Immunohistochemical analysis showed positivity for MDM2 and negativity for erythroblast transformation-specific–related gene. These findings were consistent with a diagnosis of pulmonary artery intimal sarcoma[4].

Staging investigations, including chest and abdominal CT scans, bone scintigraphy, and fluorodeoxyglucose -positron emission tomography *(Fig. 5)*, revealed no evidence of distant metastases.



Figure 1. *Contrast-enhanced computed tomography scan demonstrating obstruction of blood flow in the right main pulmonary artery (arrows).*

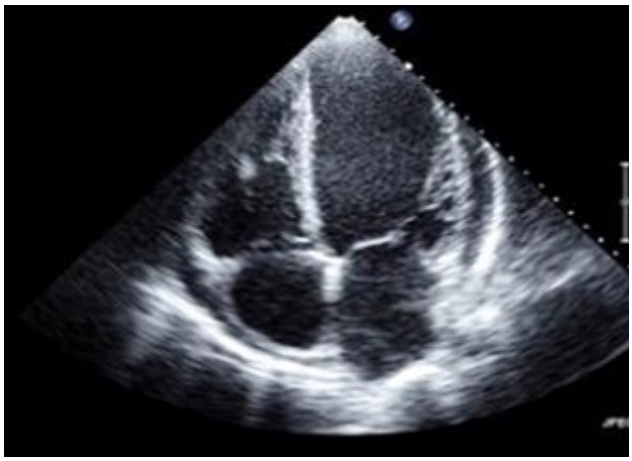


Figure 2. *Echocardiography showed no dilation of the cardiac chambers.*

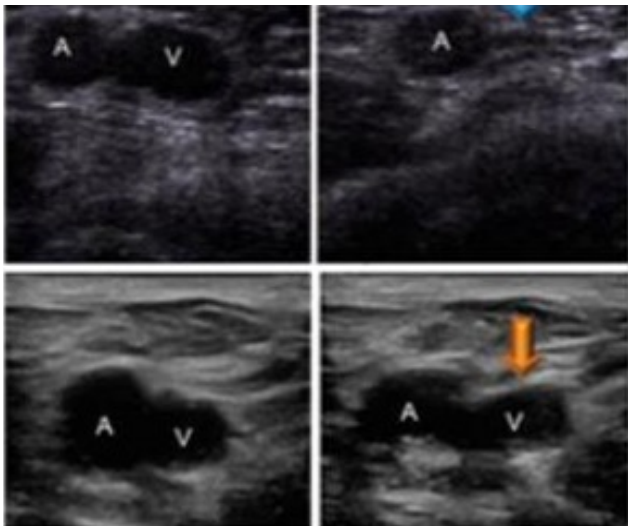


Figure 3. *Lower-extremity venous Doppler ultrasound negative for deep vein thrombosis (arrow).*
Abbreviations: A, artery; V, vein.

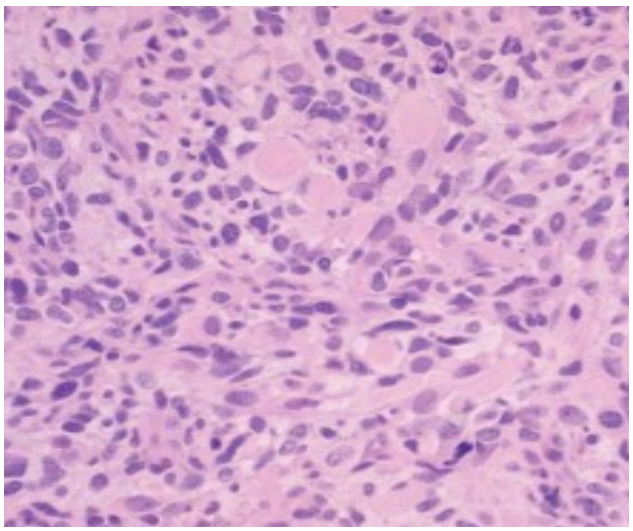


Figure 4. Atypical spindle cell proliferation.

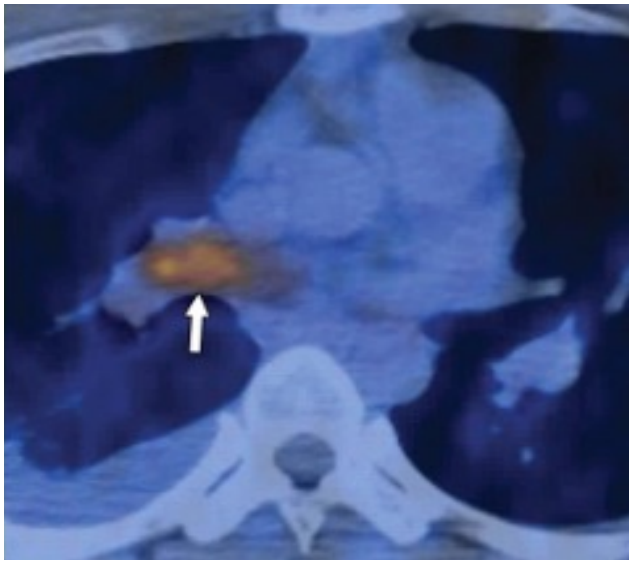


Figure 5. Fluorodeoxyglucose -positron emission tomography scan demonstrating a metabolically active lesion in the right main pulmonary artery (arrow).

The patient died two weeks after the diagnosis due to fatal local haemorrhage.

DISCUSSION

Primary pulmonary artery sarcoma represents one of the most important non-thromboembolic causes of pulmonary artery obstruction. Because pulmonary embolism is far more common, there is a natural tendency toward diagnostic anchoring when a filling defect is identified on CT imaging.

However, several features should raise suspicion of an alternative diagnosis: absence of deep vein thrombosis, lack of right ventricular strain, progressive symptoms despite appropriate anticoagulation, and imaging characteristics such as a lobulated mass or heterogeneous enhancement.

In particular, clinical deterioration or failure to improve under adequate anticoagulant therapy should prompt immediate diagnostic reassessment. Continuing anticoagulation without reconsidering the diagnosis may delay definitive treatment in malignant conditions such as intimal sarcoma.

This case underscores the need to maintain a broad differential diagnosis when evaluating pulmonary artery obstruction.

CONCLUSION

Pulmonary artery obstruction should not automatically be attributed to thromboembolism. When patients fail to improve despite appropriate anticoagulant therapy, alternative diagnoses—including malignancy—must be actively considered. Early diagnostic reconsideration is essential to avoid delayed recognition of rare but aggressive conditions such as pulmonary artery intimal sarcoma.

REFERENCES

1. Tapson VF. Acute pulmonary embolism. *Cardiol Clin* 2004;**22**:353-365. doi: 10.1016/j.ccl.2004.04.002
2. Zhao M, Nie P, Guo Y, Chen H. Pulmonary artery intimal sarcoma: A rare cause of filling defects in pulmonary arteries. *Am J Med Sci* 2022;**364**:655-660. doi: 10.1016/j.amjms.2022.05.009
3. Bhagwat K, Hallam J, Antippa P, Larobina M. Diagnostic enigma: primary pulmonary artery sarcoma. *Interact Cardiovasc Thorac Surg* 2012;**14**:342-344. doi: 10.1093/icvts/ivr087
4. Deng L, Zhu J, Xu J, Guo S, Liu S, Song Y. Clinical presentation and surgical treatment of primary pulmonary artery sarcoma. *Interact Cardiovasc Thorac Surg*;**26**:243-247. doi: 10.1093/icvts/ivx318