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IDIOPATHIC BILATERAL PULMONARY VEIN THROMBOSIS

Adeel Nasrullah[1], Muhammad Ibraiz Bilal[2], Khalid Malik[1], Briana DiSilvio[1], Tariq Cheema[1]

[1] Division of Pulmonology and Critical Care, Allegheny Health Network, Pittsburgh, USA
[2] Department of Internal Medicine, Allegheny Health Network, Pittsburgh, USA

Corresponding author: Muhammad Ibraiz Bilal **e-mail:** ibraiz47@hotmail.com

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ABSTRACT

A 52-year-old female with a history of chronic lymphoedema and untreated deep vein thrombosis, presented with non-specific right-sided chest pain. A CT angiogram confirmed bilateral inferior pulmonary vein thromboses (PVT). A comprehensive hypercoagulable workup and age-appropriate cancer screening were unremarkable; the lack of associated risk factors confirmed idiopathic PVT. The management strategy of systemic anticoagulation with apixaban and multidisciplinary follow-up underscores the treatment challenges of rare presentations. This case accentuates the importance of considering PVT in differential diagnoses of atypical chest pain and contributes valuable insights into the diagnosis, understanding and management of this uncommon condition.

KEYWORDS

Pulmonary vein thrombosis, idiopathic, anticoagulation

LEARNING POINTS

- Pulmonary vein thrombosis (PVT) may present as chest pain, especially in patients with a history of prior blood clots and can occur without an underlying malignancy or coagulation disorder.
- Utilising a chest CT angiogram with delayed contrast timing is effective in detecting pulmonary vein thrombus.
- Systemic anticoagulation proves effective in managing pulmonary vein thrombus; however, further data on dosage and duration are required for better guidance.

INTRODUCTION

Pulmonary vein thrombosis (PVT) presents substantial challenges in diagnosis and treatment owing to its extreme rarity and potential for fatality. PVT has previously been reported in patients with lung cancer, sickle cell disease, atrial fibrillation, and other malignancies as well as in patients who have undergone lobectomies and lung transplants[1,2]. Because there are so few cases described in the literature, PVT is under-recognised and poorly understood. To date, idiopathic PVT has been reported in fewer than 15 patients, with only one of these cases being bilateral PVT[1,3]. We report the second documented case of idiopathic bilateral PVT and discuss how contrast imaging may identify additional cases that may be otherwise overlooked. We also discuss how

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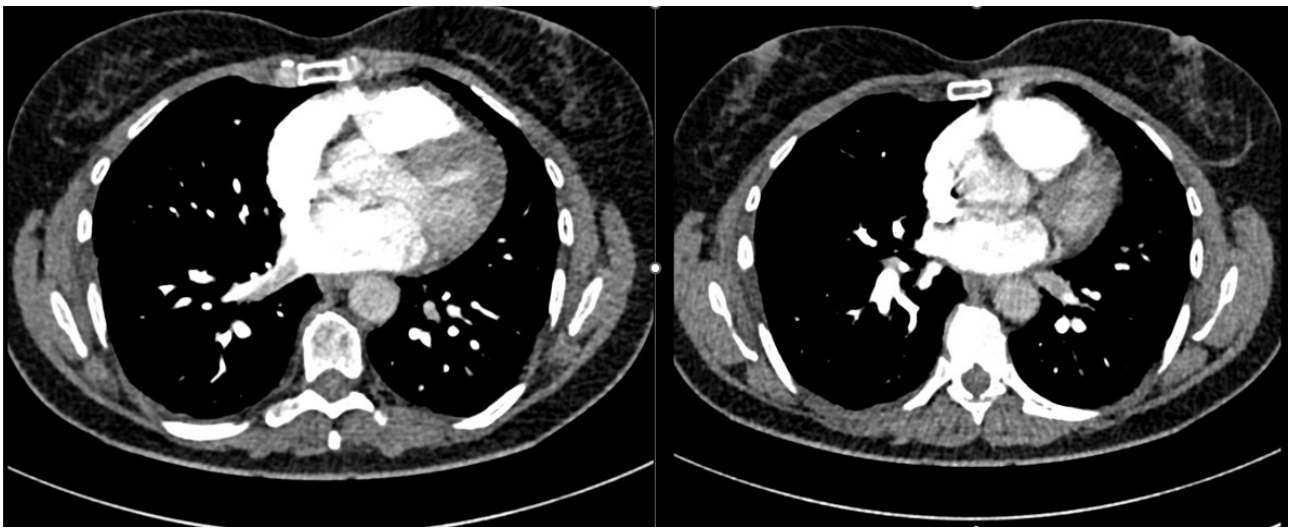

Figure 1. Parasternal long axis view: the left side of the image shows a dissection flap and enlarged aortic root, and the right side shows aortic regurgitation.

this patient was treated to facilitate the development of standardised treatment plans in the future.

CASE DESCRIPTION

A 52-year-old female was initially examined at an urgent care centre, after presenting with right-sided chest pain. Her symptoms included localised pain under the right breast that was exacerbated by deep breaths, bending and walking; it was alleviated by rest. The pain started five days prior without any associated symptoms such as shortness of breath, cough, or fever. The patient had no recent history of trauma, prolonged travel, immobility, or surgery. She had a medical history of chronic right leg lymphoedema post-appendectomy and hysterectomy, an untreated right peroneal deep vein thrombosis (DVT) in 2016, anxiety and depression. The patient's father had a history of DVT, but there was no known family history of a hypercoagulable disorder. Social history was noteworthy for a 30-pack-year history of cigarette smoking and social alcohol use.

Upon examination, the patient was haemodynamically stable, the electrocardiogram was normal and initial blood work showed normal troponins, N-terminal pro-B-type natriuretic peptide (NT-proBNP) and lactate levels. A computed tomography (CT) angiogram of the patient's chest revealed bilateral inferior PVT *(Fig. 1)*, and a heparin drip was started. The patient was transferred to our tertiary care centre for higher-level care, and venous Doppler images of both upper and lower extremities were unremarkable. A transthoracic echocardiogram (TTE) was performed which showed that the left and right ventricle function was preserved without any valvular abnormalities. However, it also showed a patent foramen ovale with right-to-left shunt. Hypercoagulable test results were unremarkable *(Table 1)*.

The patient's treatment plan included anticoagulation therapy with apixaban on discharge, and multidisciplinary follow-up with cardiology, haematology, and pulmonology specialists. This case illustrates the complexity of diagnosing idiopathic, bilateral PVT.

Test	Result
Factor V Leiden	Negative
Prothrombin gene mutation	Negative
Flow cytometry for PNH*	Negative
JAK2 V617F mutation	Negative
Beta-2 glycoprotein 1 antibody	Negative
Cardiolipin antibodies	Negative

*PNH: paroxysmal nocturnal haemoglobinuria

Table 1. Patient's hypercoagulable test results.

DISCUSSION

To date, PVT is under-recognised and poorly understood due to very few cases reported in the literature. The under-recognition of PVT is likely attributed to non-specific symptomology such as chest pain, coughing, dyspnoea and rarely, haemoptysis[1,4-6]. It has been reported in association with left upper lobectomy, malignancies, trauma, atrial myxoma, congenital narrowing of pulmonary veins, ablation for atrial fibrillation and hypercoagulable states such as COVID-19 and lung transplants[1,3,4,7-11]. In untreated cases, progressive backward congestion and upstream rise in pulmonary vascular resistance can culminate into pulmonary oedema, pulmonary infarction and right ventricular failure[4]. Similarly, a large clot burden can impair left ventricle preload, and a low cardiac output state can lead to shock or stroke with downstream embolisation[4,12].

A low index of suspicion for diagnosis and infrequent appropriate chest imaging studies to identify PVT limits its identification. Delayed contrast imaging is necessary to visualise pulmonary veins for PVT diagnosis, so a chest computed tomography angiography (CTA) often causes the condition to be overlooked[13]. Other modalities, such as a transoesophageal echocardiogram (preferable due to

proximity to pulmonary veins), cardiac magnetic resonance imaging or a pulmonary angiogram may be needed[14,15].

We established the diagnosis in our case due to the timing of CTA contrast and the patient refusing a confirmatory transoesophageal echocardiogram because of its invasive nature. Our patient lacked any of the aforementioned disease associations and had negative age-appropriate cancer screening and hypercoagulable workup, and was diagnosed with idiopathic PVT. Although the patient had normal TTE, except patent foramen ovale with right-to-left shunt and negative venous Doppler for DVT, we could not rule out paradoxical embolisation of a clot to the pulmonary veins. Stasis and sluggish flow from the pulmonary veins to the left atrium due to the right-to-left shunt may have contributed to the formation of PVT; however, a clear causality cannot be established.

Due to the rarity of PVT, there is a paucity of treatment guidelines; however, depending on the clot burden, a thrombectomy or lobe resection may be needed[4,8]. After resolving persistent or worsening symptoms, such as shortness of breath or signs of right heart failure, repeated chest imaging and/or an echocardiogram are deemed appropriate; however, it is not standard practice in asymptomatic patients who have achieved clinical resolution of symptoms. This may contribute to the underdiagnosis of PVT. In cases like the one described here, systemic anticoagulation remains the standard treatment, but the duration of anticoagulation treatment is debated[13]. This patient had a prior history of DVT, so we discussed risks and benefits before deciding on a treatment plan of lifelong anticoagulation. Practitioners must also consider extended or indefinite anticoagulation for idiopathic PVT as suggested by the latest CHEST guidelines for venous thromboembolism in cases with predisposing factors[16]. The role of antibiotics in treatment is limited to lobectomy patients with lung infarction due to a superimposed risk of infection[10,13]. Our decision to treat our patient with apixaban was guided by the only other reported case of bilateral PVT[1]. Prognosis depends on the clot burden and baseline cardiopulmonary reserve; our patient improved progressively with anticoagulation and was discharged with close outpatient follow-up.

CONCLUSION

CTA contrast imaging is a vital tool for diagnosing PVT, especially when TTE data are not available. Treatment guidelines have not been standardised due to the rarity of the condition, and continued patient monitoring is vital.

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