Idiopathic pulmonary vein thrombosis?

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Abstract
Idiopathic pulmonary vein thrombosis (PVT) is a rare disease which is likely under-diagnosed because of nebulous presentations. Accurate diagnosis is essential to prevent complications.

Introduction
We describe a patient with pulmonary vein thrombosis (PVT) who presented with non-specific chest pain, evaluated with computed tomography (CT) angiography, and discuss the limited cases in the literature, providing guidance for clinicians.

Case Report
A 26-year-old female gravida 5 para 5 presented to the emergency room with a single complaint of pleuritic chest pain. She denied shortness of breath or exertional dyspnoea. Past medical history was unremarkable. Chest pain began one week previously and progressed in severity over three days before presentation. Afebrile, she denied complaints of infection, rash, cough, haemoptysis, leg swelling, or contact with the sick. No illicit drug use, smoking, or regular alcohol consumption was reported. No family history of vasculitis or clotting disorders was noted. Physical examination revealed normal vitals with normal heart and lung sounds. Extremities exhibited no rashes, Homan’s sign, or petechiae. The remaining examination was unremarkable. Haemogram revealed hypochromic microcytic anaemia and thyroid stimulating...
Cardiac chambers are labelled with partial occlusion (arrows) within the right inferior pulmonary vein.

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Figure 2. The CT angiography revealing a well-deﬁned ﬁlling defect with partial occlusion (arrows) within the right inferior pulmonary vein. Cardiac chambers are labelled.

hormone (TSH) of 10.24 (range 0.35–5.00 uIU/mL). Pulmonary function tests were normal.

Chest X-ray showed bilateral inﬁltrations in lower lobes in an interstitial pattern, inconsistent with pneumonic processes. Chest angiogram (Figs. 1–2) revealed right hilar mass or lymph node causing encasement of the right main pulmonary artery and inﬁltrates in the right middle and lower lobes, consistent with PVT. Positron emission tomography showed metabolically active focal consolidation in right middle lobe at lung periphery; standardized uptake (SU) value was 4.7. Serologic workup for vasculitis and connective tissue disorders and thrombophilia panel were all negative. ANA, lupus anticoagulant, and C-ANCA were all negative. Erythrocyte sedimentation rate (ESR) was slightly elevated, 49 (range 0–20 mm/h by manual technique), and P-ANCA was 25 AU/mL (range 0–19 AU/mL). Electro-navigational bronchoscopy with EBUS biopsy was negative for malignancy, lymphoma, endobronchial tumours, and granulomatous process. Small blood vessel biopsy was negative for vasculitis.

The patient was started on oral anticoagulation. After discharge, the patient had no difﬁculties. However, repeat CT scan after two months post discharge demonstrated residual narrowing of the right pulmonary vein. Oral anticoagulation with weekly monitoring of INR was continued. The patient was subsequently lost to follow-up.

Discussion

Idiopathic PVT is a very challenging diagnostic entity because of non-speciﬁc presentations and limited image modalities. Known aetiologies of PVT include lung cancer; postoperative complications after lobectomy and lung transplantation; ﬁbrosing mediastinitis; hypercoagulable states, such as protein C and S deﬁciencies; and Factor V Leiden mutations. [1] Only seven cases of idiopathic PVT have been reported, including the current case (see Table 1). [2–6] Males and females appear equally affected; mean age in the series is 48, the current case representing the youngest in the series. Symptoms are non-speciﬁc and include dyspnoea, cough, haemoptysis, chest pain, and fever. Although touted as idiopathic, several patients had an underlying myocardial infarction and another had elevated homocysteine levels. Our patient exhibited positive P-ANCA. No predilection was noted for thrombus location. Delayed presentation or diagnosis was associated with lobectomy. Oral anti-coagulation appears to be the mainstay of treatment. A high level of suspicion is needed to conﬁrm the diagnosis.

Imaging modalities of choice are pulmonary angiography (CTA), ventilation-perfusion scan, transesophageal echocardiography (TEE), and cardiac gated magnetic resonance imaging (MRI) which can also distinguish bland from tumour thrombus [2]. TEE is more accurate for distal pulmonary veins. Hurwitz et al. [7] have suggested treatment with antibiotics to prevent complications associated with thrombosis and have also showed that collaterals play a major role in the presentation of symptoms.

Thrombi that are small and non-occlusive can be treated with systemic anticoagulants. Antibiotics are also necessary due to the high risk of secondary infection of inﬁltrated tissue [8,9]. Symptomatic patients or those with large thrombus should be treated with thrombolitics or surgical thrombectomy [8]. The current patient was empirically treated for pneumonia with antibiotics and warfarin was started after diagnosis. Duration of anticoagulation is always questioned. Some patients may initially present with complications, such as pulmonary gangrene, peripheral embolization, and myocardial ischaemia [4].

In conclusion, idiopathic PVT is a rare disease with ambiguous presentations. PVT is challenging to diagnose and treat. A detailed evaluation including risk factors, complications, and beneﬁts of intervention appears to best serve treatment. Idiopathic PVT complications in literature reported cases include delayed diagnosis and gangrenous lung, transient ischemic attacks, stroke, peripheral embolization and chronic lung conditions. While deﬁnitive treatment of idiopathic PVT is unknown given its rarity, lung consolidation and pulmonary gangrene may require antibiotics. Anticoagulation may prevent clot propagation, embolization, TIA, and stroke. Accurate diagnosis is essential to prevent debilitating and fatal complications.

Disclosure Statements

Appropriate written informed consent was obtained for publication of this case report and accompanying images.
| Author, year | Age/sex | Symptoms/presentation | Past medical history | Imaging | Location | Hypercoagulability | Treatment(s) |
|-------------|---------|------------------------|----------------------|---------|----------|-------------------|--------------|
| Selvidge SD et al. [2] | 33 F | Acute left sided abdominal pain, nausea and vomiting | Sickle cell anaemia | Contrast enhanced helical abdominal CT and confirmed with ECG-gated MRI | Right inferior pulmonary vein (RIPV) thrombosis extending to left atrium | Not reported | Oral anticoagulation but non-compliant. Two months later PVT still present but smaller |
| Alexander et al. [3] | 47 F | Massive haemoptysis with three days of chest pain and dyspnoea. Complete consolidation of the LLL | Unremarkable CT (high-resolution) scan. Intraoperative finding | Intraoperative finding. Left inferior pulmonary vein (LIPV) | Unknown | LLL lobectomy. No long-term treatment |
| Komatsu et al. [4] | 57 M | Chest pain with myocardial infarction (MI) | Dyslipidaemia | CT chest, coronary angiogram by MDCT | Bilateral lower pulmonary veins | Negative work up | Antiplatelet for CAD and warfarin |
| Mumoli et al. [5] | 80 M | Chest pain with MI and heart failure (EF-40%) | Dyslipidaemia | CTA | Left superior pulmonary vein | Homocysteinemia, otherwise negative | LMWH then to warfarin |
| Wu et al. [6] | 30 M | Intermittent left chest pain for six months | Unremarkable CT angiography (CTA) chest | Left inferior pulmonary vein thrombosis extending to atrium | Negative along with normal connective tissue tests | Left atrial mass resection and left lower lobectomy plus long-term anticoagulation |
| Rana et al., 2016 | 63 M | Sudden onset of central chest pain | Unremarkable | CTA chest, TEE | Pulmonary vein | Negative | Oral anticoagulation |
| Kollipara 2016 (current case) | 26 F | Pleuritic chest pain | Gravida 5 para 5 | CTA chest | Right inferior pulmonary vein | Positive for P-ANCA | Oral anticoagulation |
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