Case report

Spontaneous idiopathic pulmonary vein thrombosis successfully treated with Warfarin: A case report and review of the literature

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ABSTRACT

Pulmonary veins (PVs) are the most proximal source of arterial thromboembolism. Pulmonary vein thrombosis (PVT) is an uncommon clinical condition that can be fatal. Its incidence or prevalence is unclear as existing cases are case reports. It is often seen as a complication of malignancy, lobectomy, atrial fibrillation and less commonly idiopathic. It can be diagnosed using different types of non-invasive imaging studies. We present a 68-year-old woman who was undergoing treatment for recurrent urinary tract infection (UTI) but was incidentally noted to be dyspneic and intermittently hypoxic. She was found to have idiopathic pulmonary vein thrombosis that was successfully managed with systemic anticoagulation.

1. Case presentation

A 68-year-old obese woman with history of poorly controlled Type 2 diabetes mellitus, recurrent urinary tract infections (UTI), heart failure with preserved ejection fraction who presented to the emergency room with weakness and abdominal pain after a failed outpatient treatment for UTI with 5 days of Nitrofurantoin. She was contacted by her primary care physician who recommended hospital admission as her urine culture grew Pseudomonas resistant to Nitrofurantoin. During her hospitalization, she was noted to be dyspneic and hypoxicemic with oxygen saturation of 80%. Patient denied chest pain or discomfort, paroxysmal nocturnal dyspnea, orthopnea, dizziness, syncope or pre-syncope, slurred speech or focal weakness. Physical examination demonstrated diminished basal breath sounds, and trace pedal edema. A 12-lead electrocardiogram demonstrated sinus rhythm. Chest CT with contrast revealed left lower pulmonary vein thrombosis extending almost into the ostia draining the left atrial chamber (Fig. 1a and b) but no pulmonary artery embolism. Transesophageal echocardiogram (TEE) showed normal global left & right ventricular systolic function with probable moderate functional PFO. A follow-up transesophageal echocardiogram (TEE) revealed known PFO but did not identify thrombus in the left atrium (LA) and proximal left pulmonary vein. Chest/Abdomen/pelvis CT and breast ultrasound was negative for deep vein thrombosis. A gated cardiac CT done 6 weeks after revealed interval resolution of left lower lobe pulmonary vein thrombosis without residual thrombus (Fig. 2).

2. Discussion

Clinically detectable PVT is an uncommon underdiagnosed clinical condition with potential to be life-threatening. Its incidence is unknown as existing literature is from case reports. Its rare occurrence is partly explained by the presence of a rich network of venous collateral vessels that drain the lung. It has been occasionally reported as a postoperative complication of pulmonary lobectomy, lung cancer, radiofrequency catheter ablation (RFCA) for atrial fibrillation and lung transplant [1]. Less common conditions associated with PVT include atrial myxoma, congenital pulmonary venous narrowing [1,2]. There have been case reports of PVT associated with sickle cell disease/hemoglobinopathy [3–5], large hiatal hernia [6]. To our knowledge, only 3 cases of spontaneous idiopathic PVT have been reported in the literature [3,7].

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Possible mechanisms of thrombosis include mechanical nature, vascular torsion, or direct injury which is thought to be the most probable precipitating factor [1]. PVT is often seen after lung transplant involving the pulmonary venous anastomotic site due to thrombus formation [1]. Existing postulations of PVT pathogenesis from a tumor include direct extension of the tumor into the vein, compression of the vein by the tumor, hypercoagulable state or epithelial damage from tumor invasion [8]. 

Patients often have non-specific symptoms such as cough, dyspnea, hemoptysis, hypoxemia and interstitial infiltrate in transplanted lung, hypercapnia which poses difficulty to clinical diagnosis [1]. Diagnosis is often made using a combination of imaging modalities such as CT with contrast, TTE, pulmonary angiography, TEE, chest magnetic resonance imaging (MRI). Chest X-rays do not aid diagnosis [1]. 64-slice multidetector CT scan is thought to be superior to TTE in depicting thrombi in pulmonary vein and LA [9]. Chest MRI can differentiate a bland thrombus from a tumor thrombus [4]. TEE visualizes the extension of a thrombus into larger distal veins and LA [4], it also measures the blood flow velocities in pulmonary veins which indirectly suggest PVT [10]. Due to the high accuracy & sensitivity of TEE in evaluating pulmonary veins and its ability to provide real 2-dimensional imaging of the LA intraoperatively during double-lung transplant, some authors have recommended its use routinely [11]. Treatment of PVT is based on pathologic finding as there are no published treatment guidelines. Existing options include antibiotic therapy, anticoagulation, thrombectomy and/or pulmonary resection. Systemic anticoagulation is used in all cases; however, the choice or duration of anticoagulation is unclear [1]. However, short and long term anticoagulation have been successfully used [12]. Use of Warfarin has been reported [13] which was used in our patient, achieving resolution of PVT. Making a diagnosis of PVT is important due to its grave complications if undiagnosed. Common complications include pulmonary infarction, pulmonary edema, right ventricular failure, allograft failure [6,14]; less common cases include limb ischemia & stroke [15] from peripheral embolism, renal infarction [16].

### 3. Conclusion

The non-specificity of pulmonary vein thrombosis presentation poses a challenge to making an early diagnosis clinically particularly in spontaneous, idiopathic cases. Fortunately, it can be detected with available imaging studies especially with TEE which maybe the initial diagnostic study soon [13]. PVT should be borne in the mind of clinicians in patient who have risk factors as its mortality depends on underlying etiology. Our case adds to existing literature of successful treatment with Warfarin.

### Table 1

| Demographics | Presentation | Diagnostic Modality | Treatment                           |
|--------------|--------------|---------------------|-------------------------------------|
| 80-year-old male | Acute shortness of breath | Chest CT scan | Warfarin                             |
| 34-year-old female | Fever, Rales in both lung fields, then became pulseless and unresponsive | TEE | Median sternotomy, Extracorporeal membrane oxygenation (ECMO) though patient later passed away |
| 51-year-old male | Hypoxemia | TEE | Median sternotomy, ECMO though patient later passed away |
Conflicts of interest

The authors declare that there is no conflict of interest regarding this article.

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