Respiratory Medicine Case Reports 26 (2019) 296–298

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

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

Benedicta Nneoma Nnodum*, Madhura Manjunath, Ameet Kumar, Tony Makdisi

Berkshire Medical Center, Pittsfield, MA 01201, USA

ARTICLE INFO

Keywords:
Pulmonary vein thrombosis
Warfarin
Idiopathic
Systemic anticoagulation
Spontaneous

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 hypoxemic 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. Bilateral lower extremity ultrasound was negative for deep vein thrombosis. Transthoracic echocardiogram (TTE) 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 did not reveal occult malignancy. Fortunately, patient remained hemodynamically stable throughout hospitalization. Patient was started on oral anticoagulation with Warfarin while bridging with Heparin until therapeutic INR was achieved.

Pertinent diagnostic tests done upon discharge to evaluate underlying hypercoagulable and rheumatological etiology such as anti-nuclear antibody (ANA) titer, rheumatoid factor, cyclic citrullinated peptide IgG, c-ANCA, p-ANCA, proteinase 3 antibody (Ab), myeloperoxidase Ab, SS-A/Ro Ab, SS-B/La Ab, Scl-70 scleroderma Ab, anti-smooth muscle Ab; D-dimer, lupus anticoagulant, LA sensitive aPTT, Dil Russell viper venom, protein C functional activity, free protein S antigen, functional protein S, total protein S, and factor V Leiden were unremarkable. 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]

* Corresponding author.
E-mail address: bnnodum@bhs1.org (B.N. Nnodum).

https://doi.org/10.1016/j.rmcr.2019.02.020
Received 9 December 2018; Received in revised form 21 February 2019; Accepted 21 February 2019

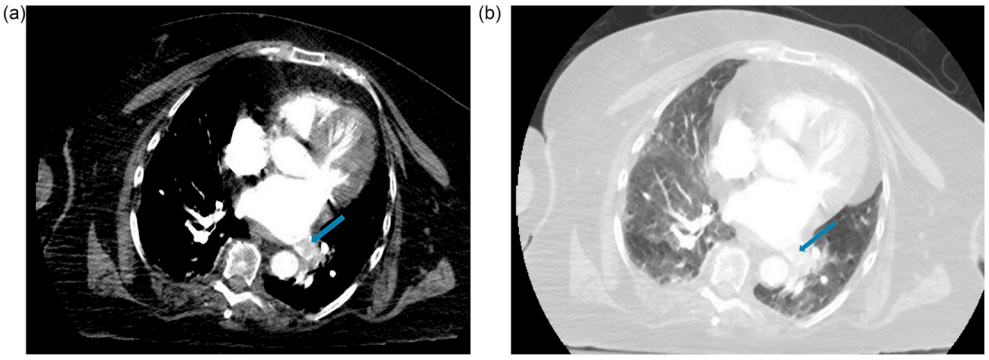

Fig. 1. a & 1b: Chest CT demonstrating a filling defect within the confluence of left lower pulmonary vein at the junction with the left atrium seen on both chest/abdomen view (Fig. 1a) and lung view (Fig. 1b).

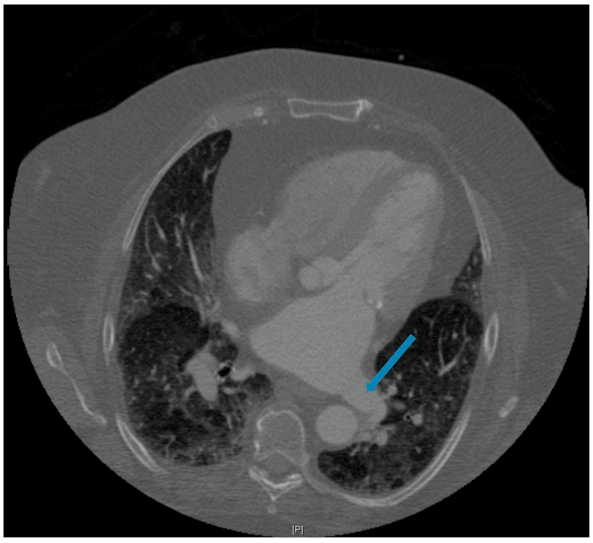

Fig. 2. Chest CT 6 weeks later showing resolution of left lower lobe pulmonary vein thrombosis.

(Table 1). 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
Summary Table of Reported spontaneous pulmonary vein thrombosis.

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.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial or not-for-profit sectors.

References

[1] G. Chaaya, P. Vishnubhotla, Pulmonary vein thrombosis: a recent systematic review, Cureus J. 9 (1) (2017).
[2] R.A. Cavaco, S. Kaul, T. Chapman, et al., Idiopathic pulmonary fibrosis associated with pulmonary vein thrombosis: a case report, Cases J. 2 (9156) (2009).
[3] N. Mumoli, M. Cei, Idiopathic pulmonary vein thrombosis, J. Emerg. Med. (2012) 182–183.
[4] S.D. Selvidge, M.L. Gavant, Idiopathic pulmonary vein thrombosis: detection by CT and MR imaging, Am. J. Roentgenol. (1999) 1639–1641.
[5] D.V. Porres, O.P. Morenza, E. Pallisa, et al., Learning from the pulmonary veins, Radiographics (2013) 999–1022.
[6] J. Saoraya, P.C. Inboriboon, Pulmonary vein thrombosis associated with a large hiatal hernia, J. Emerg. Med. (2013) e299–e301.
[7] A.S. Shah, R.E. Michler, R.J. Downey, et al., Management strategies for pulmonary vein thrombosis following single lung transplantation, J. Card. Surg. (1995) 169–178.
[8] O. Akiode, G. Prakash, Pulmonary vein thrombosis associated with metastatic carcinoma, Fed. Pract. (2014) 26–28.
[9] H. Takeuchi, Diagnosis of a left lower pulmonary veing thrombus by 64-MDCT, BMJ Case Rep. 2013 (2013) bcr-2013-010256.
[10] J.B. Cywinski, L. Wallace, B.M. Parker, Pulmonary vein thrombosis after sequential double-lung transplantation, J. Cardiothorac. Vasc. Anesth. (2005) 225–227.
[11] D.W. Leibowitz, C.R. Smith, R.E. Michler, et al., Incidence of pulmonary vein complications after lung transplantation: a prospective transesophageal echocardiographic study, J. Am. Coll. Cardiol. (1994) 671–675.
[12] S. Belok, L. Parikh, J. Robertson, A case of stroke due to pulmonary vein thrombosis, R. I. Med. J. (2016) 40–41.
[13] N.H. Kim, C.A. Roldan, B.K. Shively, Pulmonary Vein Thrombosis, Chest, 1993, pp. 624–626.
[14] L.L. Schulman, T. Anandarangam, D.W. Leibowitz, et al., Four-year prospective study of pulmonary venous thrombosis after lung transplantation, J. Am. Soc. Echocardiogr. (2001) 806–812.
[15] L. Bonnet, N. Raposo, N. Blot-Souletie, et al., Stroke caused by a pulmonary vein thrombosis revealing a metastatic choriocarcinoma, Circulation (2015) 2093–2094.
[16] S. Manabe, Y. Oshima, M. Nakano, et al., Renal infarction in a patient with pulmonary vein thrombosis after left upper lobectomy, Case Rep. Nephrol. Urol. (2014) 103–108.