Management of asymptomatic pulmonary vein aneurysm

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A R T I C L E  I N F O

Aneurysm of a pulmonary vein is a rare vascular anomaly that is usually discovered incidentally as a pulmonary nodule or mediastinal mass. Most patients do not have any symptoms but some patients can present with dyspnea, hemoptysis, or cerebral thromboembolism. Proper diagnosis is crucial as to avoid unnecessary testing or surgical procedures. We highlight a case of an asymptomatic 59-year-old female with a pulmonary vein aneurysm presenting as a 1.5 cm right infralobar nodule on contrast-enhanced CT during evaluation for acute cholecystitis. Further investigation with MRA revealed that it was vascular in nature, and pulmonary angiography showed dilation of the right inferior pulmonary vein with no communication to the pulmonary artery. On serial imaging, there has been no change in the size of the aneurysm. A small non-enlarging pulmonary vein aneurysm should be managed expectantly.

1. Introduction

Aneurysm of a pulmonary vein is a rare vascular abnormality that may present as a pulmonary nodule. The earliest recorded pulmonary varix was first noted post-mortem by Hedinger in 1907 [1] and the first ante-mortem diagnosis was proposed via pulmonary angiography by Mouquin et al., in 1951 [2] which was the gold standard for assessing the anatomy of the pulmonary vasculature. Pulmonary vein aneurysm (PVA) is a focal dilation that communicates with normal vein through a single channel and it does not involve a varicose segment [3]. The true incidence of PVA is not known and its natural history has not been well documented in the literature.

2. Case report

The patient is a 59-year old female with hypertension who was incidentally found to have a lung ‘lesion’ during evaluation for cholecystectomy in October 2014. A computed tomograph (CT) of the chest with contrast showed a homogenously enhancing 1.5 cm nodule in the right infralobar region, closely related to the right inferior pulmonary vein (Fig. 1A–B). Magnetic resonance angiography (MRA) revealed that it was a vascular lesion interposed between a small branch of the right inferior pulmonary vein and posterior basal segmental branch of right lower lobe pulmonary artery, suspicious for arteriovenous malformation or venous aneurysm (Fig. 1C). She denied cough, shortness of breath, pain, hemoptysis and weight loss. It was recommended that patient undergo diagnostic angiography with Interventional Radiology with a plan for angioembolization if it were revealed to be an arteriovenous malformation. Right pulmonary arteriography showed a 1.5 cm dilation within the right lower lobe vein with no communication with the pulmonary artery (Fig. 1D), so embolization was not indicated. She underwent surveillance chest CT with contrast six months later, and the pulmonary vein aneurysm size was stable. Due to published correlations between pulmonary vein aneurysms and mitral valve regurgitation, it was recommended that she have an echocardiogram. Transthoracic echocardiogram showed trace mitral regurgitation with no indication for intervention.

3. Discussion

Pulmonary vein aneurysm can present in an asymptomatic patient as an incidentally found pulmonary nodule or mediastinal mass on routine imaging. Symptomatic patients typically present with cough, dyspnea, palpitation, chest tightness, orthopnea, hemoptysis, and cerebral embolus [4–7]. The cause of PVA may be congenital or posttraumatic [8–11]. Patients with hereditary hemorrhagic telangiectasia (also known as Osler-Weber-Rendu disease) have been reported with PVA secondary to arteriovenous
malformation of the lung [12]. In addition, there is association between PVA and mitral regurgitation. A root cause was proposed and demonstrated by Yun et al. using contrast-enhanced CT in conjunction with echocardiography showing a high pressure jet stream flowing from a dysfunctional mitral valve into the aneurysmal defect of right inferior pulmonary vein [13].

PVA can be diagnosed with a contrast-enhanced CT scan. Although a CT with contrast cannot demonstrate a dynamic relationship between the heart and pulmonary vessels, it can give a precise anatomic view of the pulmonary veins [13]. The gold standard for diagnosis of PVA is pulmonary vein angiography when the diagnosis is unclear, although it is an invasive procedure that carries a small risk of thrombus and cerebrovascular accident [13].

There are few cases about the management of PVA in the literature. One case report documents a single patient with a 1.5 cm asymptomatic pulmonary aneurysm who was managed expectantly for a year [15]. Two other papers reported patients with symptomatic pulmonary vein aneurysms of 6 cm [6] and 10 cm [2], respectively, and both underwent lobectomy. In another case report, a patient with severe mitral regurgitation and a 5 cm PVA underwent mitral valve repair and demonstrated reduction in the size of the aneurysm on follow-up imaging at three months [13].

The approach to treatment for PVA is determined by the suspected etiology and related pathophysiology. While angiography is considered the gold standard for diagnosis, we believe that contrast CT and echocardiogram are usually sufficient for diagnosis and surveillance without having to subject patients to the risks of an invasive diagnostic test. Normal pulmonary veins vary in diameter from 9 mm to 13 mm with aneurysm defined as a 50% increase from normal size [16]. Several papers have reported aneurysms greater than 6 cm, all of which have undergone resection. If there is no underlying cardiac pathology and the PVA is small, it should be monitored for possible growth with contrast CT scan every 6 months for one year. If stability is demonstrated over one year, surveillance interval may be extended or be performed as needed, depending on individual patient factors. If the lesion increases in size over time, if it is a large aneurysm, or if it is symptomatic, the patient should undergo surgical resection to manage symptoms and to decrease the risk of thromboembolism and stroke. While cases of rupture of venous aneurysms have not been reported in the literature, it must be considered as a possible complication. Patients with small asymptomatic PVA may be managed conservatively with serial imaging.

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References
[1] E. Hedinger, Demonstration einer Lungen varix, Dtsch. Gesellsch Pathol. 11 (1907) 303–308.
[2] A. Emmert, A.F. Jelwan, K. Schmidt, M. Hinterrthaler, H. Bohnenberger, M. Bahr, et al., Aneurysm of the pulmonary vein: an unusual cause of stroke, Ann. Thorac. Surg. 98 (5) (2014) 1841–1843.
[3] R. Gabrielli, M.S. Rosatt, A. Siani, L. Irace, Management of symptomatic venous aneurysm, ScientificWorldJournal (2012) 386478. PMCID: PMC3329879.
[4] W.N. Liao, C.C. Huang, J.K. Huang, S.L. Shih, Pulmonary venous aneurysm mimicking a right infralobar tumour, Eur. J. Cardiothorac. Surg. 42 (6) (2012) e172.
[5] J. Narula, K.K. Talwar, A. Bharani, S. Mukhopadhyaya, M. Rajani, M.L. Bhatia, Pulmonary varix associated with mitral valve disease, Cathet. Cardiovasc. Diagn. 13 (6) (1987) 411–413.
[6] D.A. DeBoer, M.L. Margolis, D. Livornese, K.A. Bell, V.A. Livolsi, J.E. Bavaria, Pulmonary venous aneurysm presenting as a middle mediastinal mass, Ann.
[7] F.A. Hipona, A. Jamshidi, Observations on the natural history of varicosity of pulmonary veins, Circulation 35 (3) (1967) 471–475.

[8] S. Sirivella, I. Gielchinsky, Pulmonary venous aneurysm presenting as a mediastinal mass in ischemic cardiomyopathy, Ann. Thorac. Surg. 68 (1) (1999) 241–243.

[9] B. Hubler, J.P. Earls, K. Stevens, Traumatic pulmonary arterial and venous pseudoaneurysms, AJR Am. J. Roentgenol. 169 (5) (1997) 1354.

[10] S.P. Christow, R. Dietz, Severe mitral regurgitation after valve replacement as cause of pulmonary venous aneurysm, Circulation 102 (17) (2000) 2159–2160.

[11] C.S. Restrepo, A.P. Carswell, Aneurysms and pseudoaneurysms of the pulmonary vasculature, Semin. Ultrasound CT MR 33 (6) (2012) 552–566.

[12] K.L. Swanson, U.B. Prakash, A.W. Stanson, Pulmonary arteriovenous fistulas: Mayo Clinic experience, 1982-1997, Mayo Clin. Proc. 74 (7) (1999) 671–680.

[13] Y. Mou, Y. Cheng, Q. Feng, C. Ni, Etiology of pulmonary venous aneurysm diagnosed by a combination of echocardiography and contrast-enhanced computed tomography: a case report, J. Cardiothorac. Surg. 9 (2014) 132. PMCID: PMC4172824.

[14] E.P. Vonken, B.K. Velthuis, F.H. Wittkampf, B.J. Rensing, R. Derksen, M.J. Cramer, Contrast-enhanced MRA and 3D visualization of pulmonary venous anatomy to assist radiofrequency catheter ablation, J. Cardiovasc. Magn. Reson. 5 (4) (2003) 545–551.

[15] N. Loqman, F. McGuire, T. McLemore, Case report: identification of pulmonary vein aneurysm using radial endobronchial ultrasound and electromagnetic navigation bronchoscopy, J. Bronchol. 15 (2) (2008) 116–117.

[16] Y.H. Kim, E.M. Marom, J.E. Herndon 2nd, H.P. McAdams, Pulmonary vein diameter, cross-sectional area, and shape: CT analysis, Radiology 235 (1) (2005), 43–9; discussion 9–50.