Pulmonary vein occlusion: A delayed complication following radiofrequency ablation for atrial fibrillation✩✩✩

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ABSTRACT

This case reports demonstrates the rare but potentially serious complication of pulmonary vein stenosis and subsequent thrombosis diagnosed two years after radiofrequency ablation of the pulmonary veins for atrial fibrillation. Pulmonary vein stenosis can remain asymptomatic until significant occlusion occurs, after which it can present with a variety of symptoms, mimicking a myriad of cardiovascular and pulmonic pathologies. Early diagnosis and treatment rely on consistent follow up using appropriate diagnostic imaging modalities and is paramount in preventing severe complications.

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Pulmonary vein stenosis is a rare diagnosis commonly occurring secondary to radiofrequency ablation for treatment of atrial fibrillation. Varying symptoms such as dyspnea, cough, and hemoptysis develop as the result of progressive venous occlusion and subsequent pulmonary congestion. Prior studies indicate that the onset of symptoms typically develop over a period of weeks to months after the procedure. There are only a few case reports that demonstrate development of symptoms greater than one year after radiofrequency ablation. This case report reviews the unique diagnosis of pulmonary vein occlusion made almost two years after radiofrequency ablation for atrial fibrillation, emphasizes the importance of follow up surveillance, and highlights key radiographic findings of the disease.

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Case report

The patient is a 69-year-old female with a past medical history of chronic paroxysmal atrial fibrillation diagnosed in 2017. In June 2019, the patient underwent radiofrequency ablation for atrial fibrillation and has been adherent on Eliquis anticoagulation since the procedure. The patient had mild symptoms of insidious dyspnea and clinical suspicion for pulmonary vein stenosis (PVS) in the post procedural period, but was lost to follow up with surveillance screening for complications. In March 2021, the patient presented to the emergency department with a complaint of acutely worsening left sided chest pain and dyspnea over the course of three days. She did not endorse symptoms of fever, cough, hemoptysis, or wheezing. Initial chest X-ray demonstrated low lung volumes with vascular crowding and a large, ill-defined opacity in the left lung (Fig. 1). The possibility of acute pulmonary embolism based on clinical intermediate pretest probability and positive D-Dimer was raised and the patient was scheduled for a CT pulmonary angiogram of the chest with contrast.

The study was negative for pulmonary embolism but was remarkable for complete occlusion of the left superior and inferior pulmonary veins (Fig. 2). The parenchymal findings of ground-glass opacities in the left upper and lower lobes were most consistent with areas of hemorrhagic infarction and lymphovascular stasis. Following the CT imaging findings discussed above, the patient was recommended to undergo ventilation-perfusion scintigraphy (V/Q scan) for further evaluation of functional impact of the stenosis. Two weeks after initial presentation to the emergency department, the patient returned to the emergency department due to worsening shortness of breath and new onset hemoptysis. V/Q scan was performed at this time and demonstrated lack of perfusion to the entire left lung field (Fig. 3). The differential diagnosis for the V/Q scan results suggested several possible etiologies of the finding include large pleural effusion, malignancy, or central pulmonary embolism.

Same day CT scan demonstrated interval worsening of the lung parenchymal findings with a large peripheral wedge shaped, sub-solid lesions with peripheral ground glass opacities and central attenuation, characteristic of multiple large infarcts. The entire venous system including all tributaries for both the superior and inferior pulmonary veins were unopacified and directly related to the areas of lung infarction. Patient was diagnosed with pulmonary vein occlusion and resulting hemorrhagic infarction, secondary to radiofrequency ablation for atrial fibrillation. In view of clinical progression of findings and symptoms despite anticoagulation, the decision to treat the suspected underlying stenosis with balloon angioplasty and stents was made. A chest radiograph two months after the procedure demonstrated resolution of the findings (Fig. 4) and the patient’s symptoms resolved.

Discussion

PVS is characterized by the luminal narrowing of one or more pulmonary veins resulting in the progressive symptoms of dyspnea, cough, and hemoptysis [1]. It is a rare clinical condition that is associated with a bimodal age distribution [2,3]. In the pediatric population, it is usually arising as the result of a congenital abnormality [2–4]. In adults, it has been reported secondary to etiologies such as fibrosing mediastinitis, sarcoidosis, or underlying neoplastic processes [2,4,5]. However, the advent and increased prevalence of radiofrequency ablation as a treatment for atrial fibrillation has resulted in iatrogenic causes as the leading etiology for pulmonary vein stenosis in adults [2–4].

Since its introduction to clinical medicine in 1987, radiofrequency ablation has been a documented risk factor for pulmonary vein stenosis [6]. The earliest ablative techniques, involved with focal ablation within the venous ostia themselves, have demonstrated an incidence of pulmonary vein stenosis ranging from 3% to 42% [6–12]. The constrictive process is hypothesized to be the result of thermal injury to the endovascular tissue [4,13]. Injury progression demonstrates hyperplasia of the intima, fibrotic collagenous proliferation, and endovascular contraction resulting in vein stasis and congestion [10,14]. Improved understanding of risk factors for pulmonary vein stenosis and newer ablation techniques such as circumferential ablation and antral isolation has allowed for a significant decrease in pulmonary vein stenosis to about 3 percent [6,7]. However, the actual incidence rate remains uncertain due to this disease’s non-specific symptoms, delayed presentation, and lack of post-ablation surveillance screening leading to under-diagnosis and misdiagnosis of the disease [6,7].

Pulmonary vein stenosis has no specific presentation or unique characteristic, making it a challenging clinical diagnosis [6,7]. Diagnosis is further delayed due to late presentation of symptoms along with lack of routine surveillance imaging post ablation [7,12]. Iatrogenic PVS can develop over weeks
Fig. 2 – A: Axial image pulmonary CT angiogram in lung window reveals two pulmonary infarcts (arrows) in the left upper lobe, the largest with the “ground glass halo” sign. Prominent mediastinal nodes (*) are also noted. B: Left inferior pulmonary vein occlusion - CT Pulmonary angiogram, MIP double oblique reformat shows that the trajectory of the vein is completely thrombosed (arrow). A peripheral infarct in the pathway of the vein is be seen (*). On the right, the partially imaged distal right pulmonary artery (RPA) and a portion of the patent right inferior pulmonary vein (RIPV) are identified. C: Left superior pulmonary vein occlusion - CT Pulmonary angiogram double oblique MIP reformat shows tapering of the contrast column at the left atrial junction and complete lack of contrast opacification of the trajectory of the vein (long arrows). Note peripheral infarcts in the territory of the occluded veins (short thick arrows). The right inferior pulmonary vein is patent (RIPV). Mildly reactive prominent left hilar nodes (*) are also present.

Fig. 3 – Multiple spot views from the lung perfusion scan acquired after the intravenous administration of 4.0 mCi of Tc-99m MAA. Ventilation images were not acquired due to COVID-19 (Corona Virus Disease) precautions. Complete absence of perfusion is seen in the left lung which is explained by the occlusion of both pulmonary veins seen on the CT and concordant with the hyperemia seen on the chest x-ray. Normal perfusion is seen in the right lung with no perfusion defects.
to months [2]. Patients with mild to moderate stenosis, defined as less than 70 percent luminal narrowing, are typically asymptomatic [6,15]. Symptom severity is attributable to factors such as the number of veins affected, presence of collateral, and rate of progression [3,4,9,12,14,16]. Still, many of the most common symptoms, such as dyspnea, cough, hemoptysis mimic other common cardiac and pulmonary pathologies making the disease difficult to diagnose [2,5,7–9,14]. Our patient had multiple visits to the ED, cardiologist and pulmonologist for non-specific symptoms of dyspnea and chest pain. These were attributed to sleep apnea and asthma exacerbations and therefore the diagnosis was (as often seems to be the case) delayed until the acute event of complete vein occlusion and thrombosis.

In addition to the nonspecific symptom presentation, chest radiographs, non-contrast CT of the chest, and ventilation-perfusion (V/Q) scans may suggest other more common diagnoses. In pulmonary vein stenosis, pulmonary parenchymal opacities and peripheral consolidations are typically indirect signs of significant stenosis and venous occlusion secondary to alveolar infarction or hemorrhage [14]. Venous stenosis is often accompanied by regional lymphadenopathy as a result of thermal damage, as was observed in our patient [14]. On chest radiographs and chest CT, these underlying processes will typically be depicted as hazy opacities, consolidations, or pleural effusions with reactive lymphadenopathy, and are often mistaken for pneumonia or atypical infections [2,6].

Although they are non-specific, V/Q scans play a critical role in PVS. The nuclear medicine perfusion scan is done via IV administration of about 300,000 small particles of macro aggregated albumin these have average size on 20 microns (or micrometer). These particles will travel to right heart, pulmonary artery and will block distal arterioles and capillaries with average diameter of 5 microns. The occlusion affects less than 0.1% of the pulmonary arterial circulation and is not hemodynamic significant and the particles will break down after approximately 2 hours [17]. Nuclear V/Q scan are typical used to detect arterial abnormalities as pulmonary embolism. In this patient we have a venous, post capillary abnormality, causing abnormal arterial perfusion. The venous abnormality is so significant that arterial perfusion to the whole left lung is virtual absent. When significant occlusion is present, pulmonary vein stenosis will result in decreased perfusion to the part of the lung that the vein is draining [5]. Consequently, V/Q scans will depict mismatch defects due to decreased perfusion of lung fields drained by the occluded veins. This is a non-specific finding as it can be attributable to any etiology obstructing pulmonic vasculature such as pulmonary embolism or malignancy [2,6]. However, V/Q scans still play an important role in pulmonary vein stenosis by helping to determine the progression and functional significance of the occlusion in conjunction with a corresponding CT pulmonary angiogram or MRI demonstrating the stenotic lesion [2,5,7,15,16,18]. Thus, it is imperative to have a high index of suspicion for pulmonary vein stenosis in patients who have undergone radiofrequency ablation of the pulmonary veins [12].

Multislice CT with protocol optimized for visualization of the pulmonary veins is the diagnostic study of choice due to its adequate spatial and temporal resolutions [2,7,16,19,20]. Direct cross sectional imaging findings in patients with PVS are absence, abrupt cut off, or narrowing of a pulmonary vein as demonstrated by reduced flow of contrast [20]. As a result of increased venous pressure, disease progression may lead to lung edema and infarction of the region drained by the venous system. These secondary processes will present on CT imaging as indirect cross-sectional findings such as ground glass opacities, consolidations, and septal thickening [19,20]. These indirect find-
ings can be misleading, and it is important for radiologists to be aware of the patient’s clinical history of radiofrequency ablation [19,20].

MRI is another useful tool in the diagnosis of PVS. It is reliable in depicting stenotic lesions and perfusion abnormalities [2]. MRI will provide information regarding blood flow as well as right and left ventricular function [19]. Furthermore, it is free of ionizing radiation making it useful for those who need to undergo frequent surveillance screening [7]. However, its long acquisition time, respiration motion artifacts, lower spatial resolution, and metal implant contraindications make it a less favored study compared to multislice CT angiography, in the routine clinical practice.

Post procedure CT angiogram or MRI three to four months after ablation can help to identify stenosis in earlier, treatable stages [6]. Studies have indicated that symptoms of PVS take months to develop after ablation, with some studies indicating development of symptoms after one year [20]. In a large consecutive series, Di Biase et al. reported a varied duration of 3–25 months between ablation and CT diagnosis of pulmonary vein stenosis [19]. Even more, in patients with documented mild PVS a few months after the procedure, progression to severe stenosis or occlusion may occur. This was the case in our patient who complained of mild symptoms early after her procedure in 2019, but she did not undergo routine surveillance screening for pulmonary vein stenosis. Unfortunately, she was not diagnosed until two years after her procedure, at which point imaging showed extensive signs of disease progression. Currently, there is no recommendation or guideline for post surveillance screening of complications from radiofrequency ablation [7]. However, in the setting of persistent non-specific cardiorespiratory symptoms, periodic pulmonary vein imaging screening could have resulted in an earlier diagnosis and prevented the acute complications that our patient developed. Our patient underwent balloon angioplasty and stent placement in both superior and inferior left pulmonary veins. This procedure has been reported in the literature with successful results. Restenosis may however be as high as 67% in some series [21,22].

Conclusion

Although the incidence of pulmonary vein stenosis secondary to radiofrequency ablation has been greatly reduced, it is a complication that radiologists should be aware of in order to accurately diagnose the disease in its early stages. Symptoms are non-specific, and radiographic findings on plain CT, chest radiograph, and V/Q scan may overlap with etiologies such as pneumonia or pulmonary embolism leading to misdiagnosis and delay of treatment. Routine surveillance with transthoracic echocardiogram, CT pulmonary angiography or MRI may be considered periodically in patients with persistent non-specific respiratory or cardiovascular complaints. Delayed symptom presentation even years after the procedure, as it was in our patient’s scenario, should not rule out the possibility of pulmonary vein stenosis/occlusion.

Patient consent

A written informed consent was obtained from the patient for the publication of this case report.

Ethical clearance

This project did not involve any research and no ethical clearance was required.

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