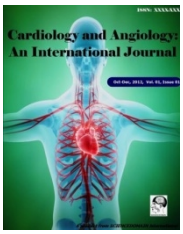

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An Unusual Cause of an Uncommon Condition: Pulmonary Vein Stenosis

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Author's contribution

This whole work was carried out by author JTL.

Case Study

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ABSTRACT

Pulmonary vein stenosis in adults is historically a rare condition, but is becoming a recognised complication, albeit an uncommon one, of radiofrequency ablation around the pulmonary veins for treatment of atrial fibrillation. It may also be due to infiltrating mediastinal processes such as neoplasm or sarcoidosis. In this case report, a 55-year-old man underwent resection of a mediastinal phaeochromocytoma involving the left atrial wall and the right inferior pulmonary vein. One year later he subsequently presented with increasing dyspnoea and atypical chest pain. Transthoracic echocardiogram showed severe pulmonary hypertension, right ventricular dilatation and dysfunction. Transesophageal echocardiogram demonstrated severe bilateral pulmonary vein stenosis with peak/mean gradients across the left pulmonary veins of about 25/20mmHg. The diagnosis was also confirmed on CT pulmonary angiography with 3D reconstruction. Open pulmonary vein stenting was planned but unfortunately the patient died suddenly before the procedure. Pulmonary vein stenosis is an uncommon but serious condition and may present with signs and symptoms indistinguishable from other conditions and may easily be missed. Clinicians should have a high index of suspicion when patients present with unexplained respiratory symptoms, especially in the context of catheter ablation or mediastinal processes such as neoplasm. Transesophageal echocardiography played an indispensible part in the correct diagnosis in our patient.

Keywords: Pulmonary vein stenosis; echocardiography; mediastinal phaeochromocytoma.

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1. CASE PRESENTATION

A 55-year-old man presented 12 months prior with hypertension and dyspnoea. Investigations revealed a catecholamine-secreting tumour. Subsequently, he was found to have a malignant mediastinal phaeochromocytoma behind the left atrium. At thoracotomy, the tumour was found to be invading the roof of the left atrium and involving the right lower pulmonary vein and the medial aspects of the left superior and inferior pulmonary veins. He underwent successful excision of the tumour with right lower lobectomy and removal of the right inferior pulmonary vein. The roof and the posterior wall of the left atrium between the orifices of the left pulmonary veins and the right superior pulmonary vein was reconstructed with a bovine pericardium patch. No distant metastasis was found and the patient did not receive chemotherapy or radiotherapy post-operatively.

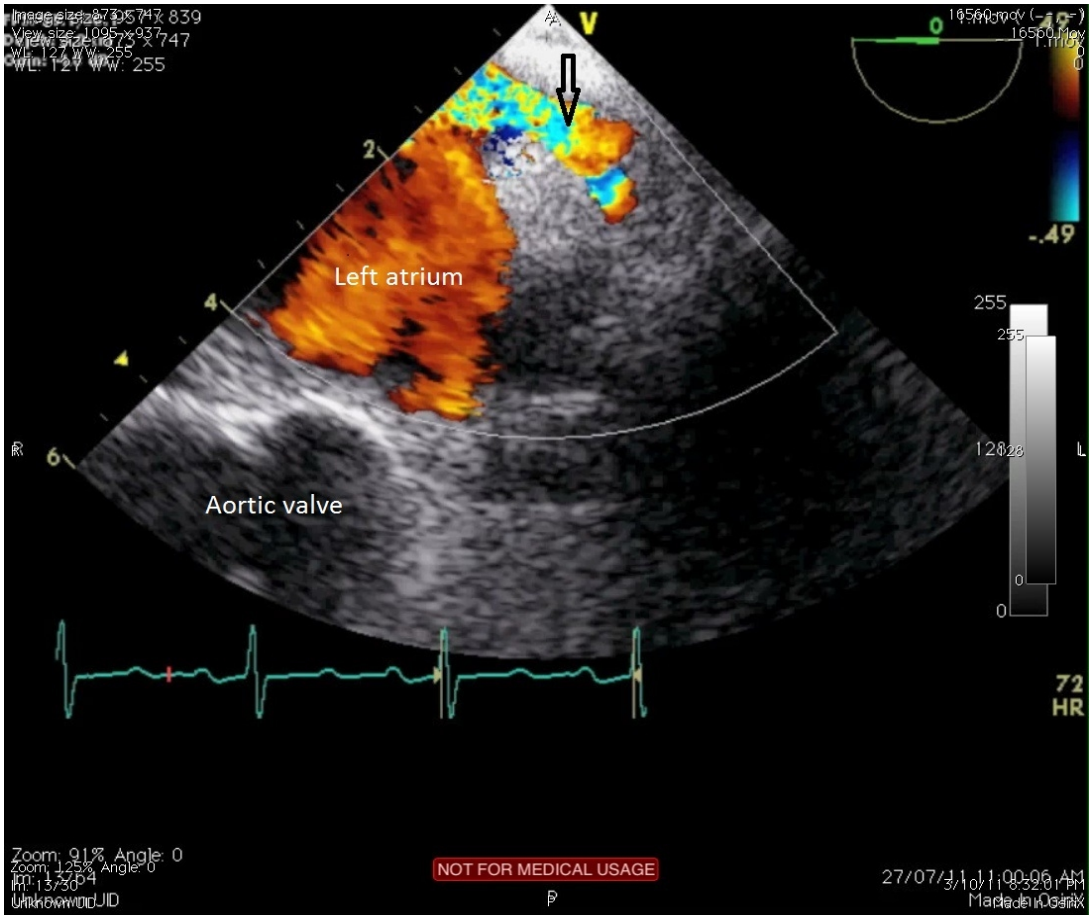

Fig. 1. Transesophageal echocardiogram with color Doppler showed the confluence of the left upper and lower pulmonary veins entering the left atrium. There is a turbulent, continuous flow (arrow) into the left atrium originating from a discrete stenosis at the left pulmonary veins

The patient presented 12 months later with increasing shortness of breath and atypical chest pain. ECG showed sinus rhythm with anterior T wave inversion and troponin T was not

elevated. Transthoracic echocardiogram showed normal left ventricular size and function with a D shaped septum. There was severe right ventricular dilatation and dysfunction with severe pulmonary hypertension. The peak pulmonary systolic pressure estimated from the tricuspid regurgitant jet was 60 mmHg. CT pulmonary angiography was negative for pulmonary embolism. High-resolution chest CT identified a moderate loculated right pleural effusion with interstitial septal thickening and linear atelectasis with patchy ground glass changes within right lower zone. Multiple mediastinal lymphadenopathies were detected. Twenty-four hour urine collection for catecholamines and metabolites showed no significant abnormalities.

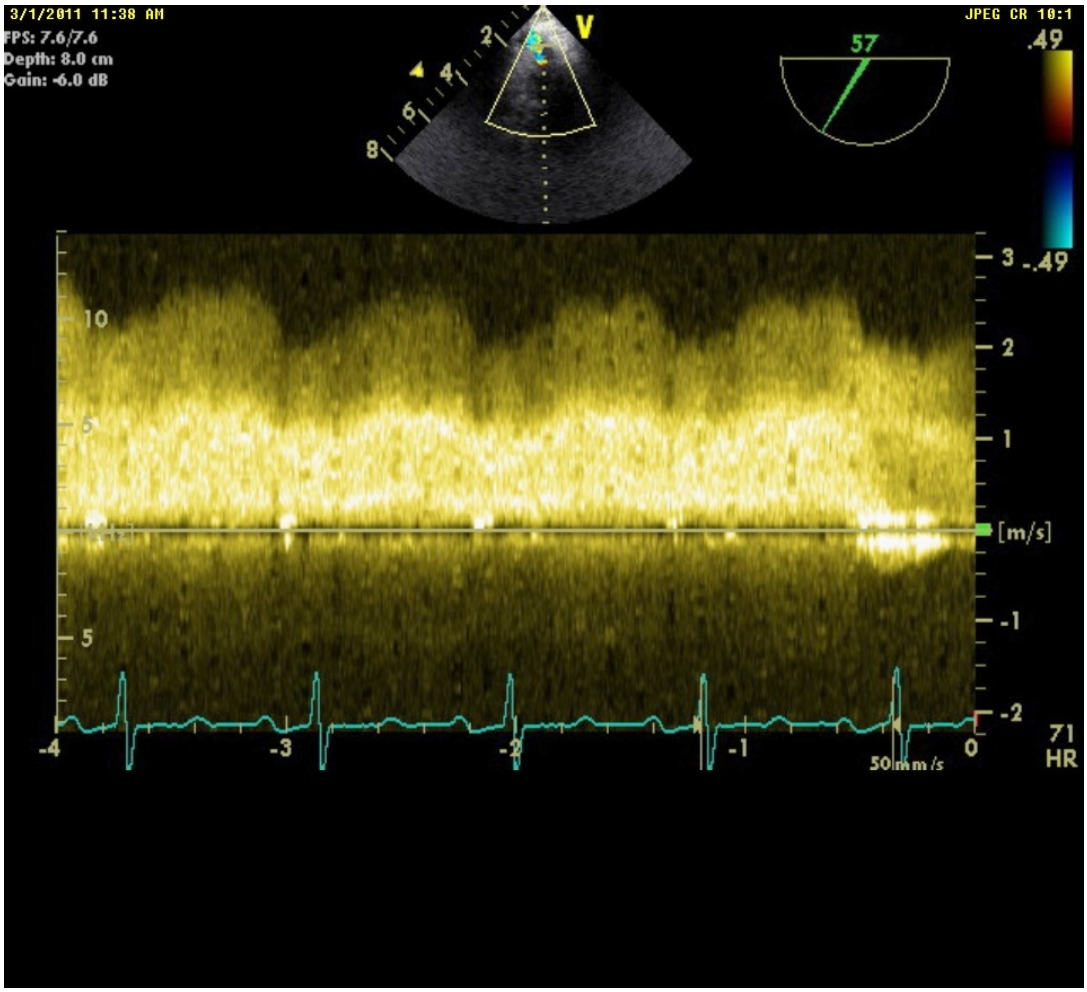

Fig. 2. Continuous wave Doppler showed a high velocity continuous flow into the left atrium with relatively little variation in peak velocities throughout the cardiac cycle. The peak velocity was about 2.5m/sec corresponding to a peak gradient of 25mmHg. The mean gradient was measured at about 20mmHg

Bilateral pulmonary venous stenosis was suspected. Transesophageal echocardiogram showed continuous turbulent flows from both the right superior pulmonary vein and the confluence of the left pulmonary veins into the left atrium Fig. 1 (above). Continuous wave

Doppler showed high velocity continuous flow with a peak velocity of 2.5m/sec on the left side Fig. 2 (above) and 1.5m/sec on the right side. The diagnosis was confirmed and the patient underwent repeat CT chest for planning intervention. A 3D reconstruction showed focal severe bilateral pulmonary vein stenoses Fig. 3 (below). No lymph nodes enlargement or masses were demonstrated on the CT to suggest tumour recurrence. The patient was planned for open stenting of the stenosed pulmonary veins. However the patient died suddenly in hospital before the procedure could be performed.

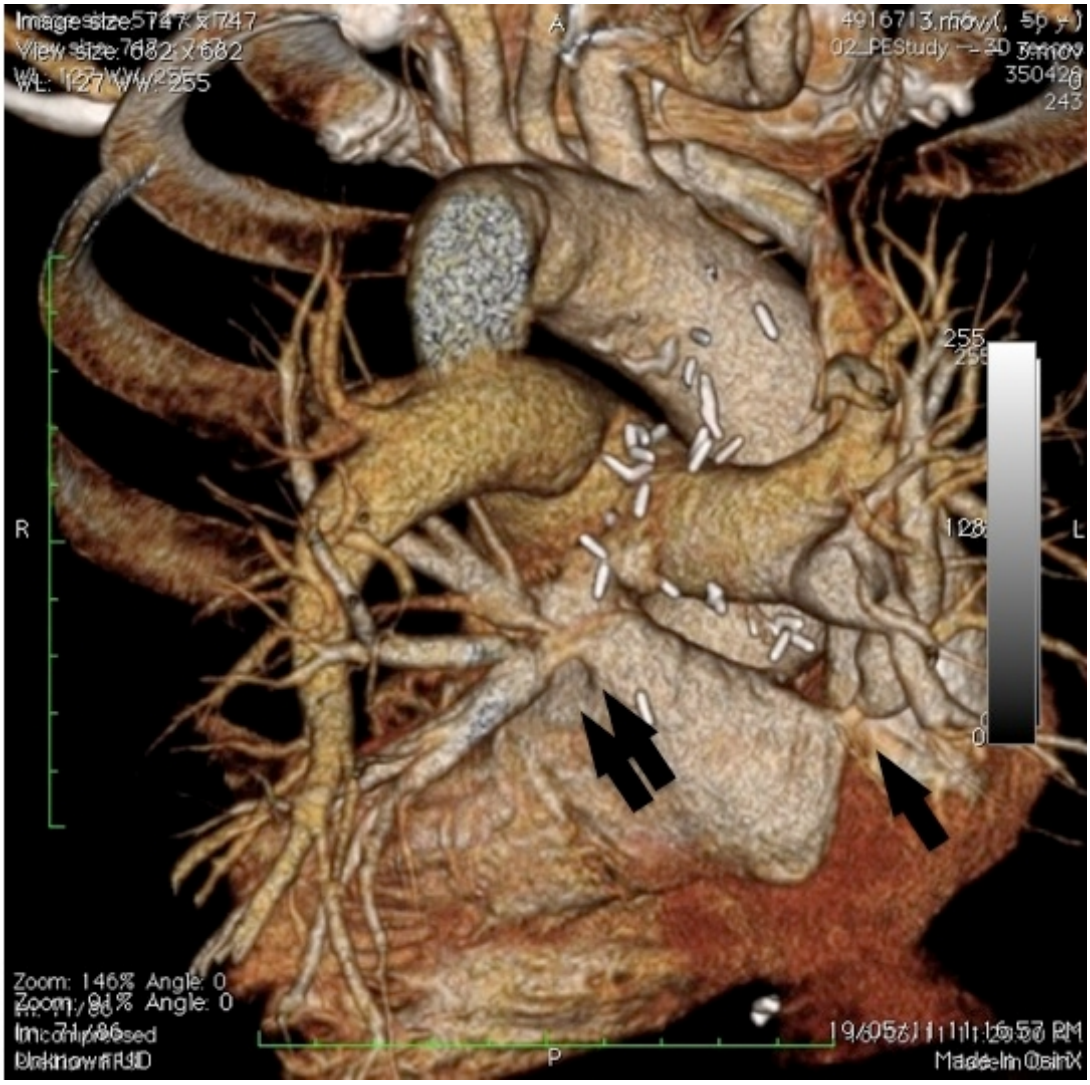

Fig. 3. Three-dimensional reconstruction from computed tomography pulmonary angiography showed the discrete stenosis at the single right superior pulmonary vein (solid arrow) and the confluence of the left superior and inferior pulmonary veins (double arrows)

2. DISCUSSION

Pulmonary vein stenosis is a rare condition [1]. Primary pulmonary vein stenosis is seen almost exclusively in children and may be associated with other forms of congenital heart disease. Acquired pulmonary vein stenosis in adults is uncommon but is increasing seen after catheter-based ablation procedures for atrial fibrillation [2]. Uncommonly, involvement of the pulmonary veins by extrinsic processes like neoplasm, fibrosing mediastinitis or after mediastinal surgery [1,3] has been described to cause pulmonary vein stenosis. The clinical spectrum of this disease is variable. Symptoms are non-specific and may be absent, depending on the number of pulmonary veins involved, the degree of stenosis and the presence or absence of collaterals. Common presenting complaints include cough, dyspnoea, hemoptysis and pleuritic chest pain. Chest x-ray findings may be indistinguishable from bronchitis or pneumonitis. This may develop into pulmonary hypertension with right ventricular dysfunction. Given the insidious nature of this condition and the fact that more common diseases such as pneumonia or interstitial lung disease have similar presentations, a high index of suspicion is required in patients with unexplained respiratory symptoms or pulmonary hypertension with a relevant past medical history. Furthermore, the correct diagnosis may easily be missed as transthoracic echocardiography may not adequately visualise the pulmonary venous flow and, as our patient has illustrated, radiologists may overlook the pulmonary venous structure on reporting CT pulmonary angiogram when pulmonary embolism was suspected.

Appropriate imaging is critical for the diagnosis and assessment of pulmonary vein stenosis. Transthoracic echocardiogram, the usual initial imaging test in these patients, may demonstrate pulmonary hypertension with or without right ventricular enlargement and dysfunction. However, pulmonary veins and their flow into the left atrium may not be adequately visualised. Transesophageal echocardiography may give the definitive diagnosis by visualising the pulmonary veins and their inflow in the left atrium. Furthermore, flow pattern and velocities can be obtained with Doppler examination allowing the assessment of pressure gradients across the stenosis. Normal pulmonary venous flow shows a distinct biphasic pattern with a systolic and diastolic forward flow and an atrial reversal wave due to atrial contraction. The flow is laminar with peak velocities <80cm/sec. With pulmonary venous stenosis of increasing severity, the forward flow velocities increase, the atrial reversal flow velocities decrease and the distinct biphasic forward flow pattern will change into a continuous turbulence with little variation throughout the cardiac cycle. As our patient has illustrated Fig. 2, the continuous wave Doppler showed a continuous high velocity flow with only minor variation in peak velocities throughout the cardiac cycle. The mean trans-stenosis gradient in case of post catheter ablation pulmonary vein stenosis was reported to be 12±5mmHg [2]. The trans-stenosis gradient in our patient across the confluence of the left pulmonary veins was up to 22mmHg. The pulmonary vein stenoses were postulated to be due to fibrosis secondary to the resection and reconstruction of the posterior left atrial wall.

CT angiogram is an excellent imaging modality in the morphological assessment of pulmonary vein stenosis. Excellent imaging quality with good spatial resolution and rapid acquisition can usually be obtained with detail visualization of the location and the extent of pulmonary vein stenosis allowing planning of intervention.

3. CONCLUSION

Severe pulmonary vein stenosis, especially when it involves all pulmonary veins, is associated with a poor prognosis [1] as our patient has illustrated. Treatment is difficult and restenosis rate high with endoluminal stenting [2]. Presenting symptoms can be non-specific and may be confused with other conditions. Diagnosis requires a high index of suspicion. Furthermore, routine surveillance for pulmonary vein stenosis may be necessary in patients after catheter ablation or after surgical treatment of mediastinal tumours

CONSENT

Consent from patient is not applicable as there is no identifiable patient information in the report.

ETHICAL APPROVAL

Ethical approval is not applicable as this is a case report and does not involve human experimentation.

COMPETING INTERESTS

Author has declared that no competing interests exist.

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