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Congenital Pulmonary Artery Stenoses Masquerading as Chronic Thromboembolic Disease

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Abstract

Stenotic pulmonary vascular lesions, often amenable to balloon angioplasty, can erroneously appear to suggest chronic thromboembolic disease on ventilation/perfusion (V/Q) scan. We report a case of multiple peripheral pulmonary artery stenoses and describe radiologic findings and treatment options.

Introduction

Multiple peripheral pulmonary artery stenoses (MPPAS) are reported in association with congenital heart defects and rarely described with acquired diseases [1]. Pulmonary hypertension (PH) is a recognized sequella of MPPAS and progressive right heart failure can result in death [2]. Stenotic pulmonary vascular lesions, often amenable to balloon angioplasty, can show large perfusion defects suggestive of chronic thromboembolic PH on ventilation/perfusion (V/Q) scan. We report a case of MPPAS and describe radiologic findings and treatment options.

Case Presentation

A 48 year old woman presented for evaluation of PH. She was diagnosed with a small ventricular septal defect (VSD) shortly after birth not necessitating repair. On routine examination as an adult a murmur was noted, prompting echocardiography and referral to our center. She was otherwise well and employed in manual labor. Physical examination revealed a loud P2 and a 3/6 harsh systolic ejection murmur radiating posteriorly without heart failure. Transthoracic echocardiography showed a 5 mm membranous VSD with bidirectional shunt, normal chamber sizes and no tricuspid regurgitation. V/Q scan showed segmental and sub-segmental mismatched defects. On right heart catheterization right atrial pressure was 10 mmHg, pulmonary artery pressure (PAP) was 88/20 mmHg, mean 46 mmHg, and pulmonary capillary wedge pressure was 15 mmHg. Cardiac index was normal. Pulmonary angiogram showed MPPAS (Figure 1a).

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Balloon angioplasty was performed in branches supplying the medial and lateral segments of the right lower lobe and the anterior and posterior segment of the left lower lobe (Figure 1b). Follow up angiography showed good anatomic response and PAP fell to 42/18 mmHg. Four months later, the patient was symptomatically improved with a 30 m increase in six minute walk. Angioplasty of remaining lesions is planned.

Discussion

As MPPAS can present in adulthood with right heart failure suggestive of PH [2], evaluation usually requires V/Q scan to exclude chronic thromboembolic PH. The case presented here highlights the importance of further investigation of potential vascular obstruction seen on perfusion scan with confirmatory imaging when evaluating patients for PH. In patients with congenital heart disease and abnormal V/Q scans, evaluation for MPPAS with CT angiography (Figure 2) prior to right heart catheterization or pulmonary angiography may allow for better procedural planning potentially including intervention with experienced operators.

When PAP measured distal to the stenosis is low and there is an adequate vascular bed, the lesion may be treated with balloon angioplasty or stent placement [3]. In contrast to the patient presented here, a second patient severely symptomatic patient seen at our clinic with MPPAS associated with a corrected atrial septal defect (ASD) (Figure 2) had elevated pressure distal to the stenoses and was therefore not a candidate for interventional procedure. This patient died several months later awaiting heart lung transplantation. In patients with advanced disease, lung transplantation has been reported [2], however anatomically and hemodynamically suitable lesions in symptomatic patients should be considered for stent placement prior to referral to lung transplantation.

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References

1. Kreutzer J, Landzberg MJ, Preminger TJ, Mandell VS, Treves ST, Reid LM, Lock JE. Isolated peripheral pulmonary artery stenoses in the adult. *Circulation* 1996;93(7):1417–1423. [PubMed: 8641032]
2. Ulrich S, Fischler M, Pfammatter T, Vogt P, Weder W, Boehler A, Speich R. Multiple peripheral pulmonary artery stenoses in adults: a rare cause of severe pulmonary hypertension necessitating lung transplantation. *J Heart Lung Transplant* 2005;24(11):1984–1987. [PubMed: 16297810]
3. Shikata H, Sakamoto S, Ueda Y, Tsuchishima S, Matsubara T, Nishizawa H, Shono S, Kanno M, Shimizu T, Matsubara J. Reconstruction of bilateral branch pulmonary artery stenosis caused by Takayasu's aortitis. *Circ J* 2004;68(8):791–794. [PubMed: 15277740]

Figure 1a.

Figure 1b

Figure 1.

Multiple pulmonary artery stenoses are seen in the left pulmonary artery (1a) in the case presented with a small, uncorrected VSD. Arrow shows a typical stenotic lesion in the anterior segment. In 1b, the same artery after balloon angioplasty dilation is shown with good improvement of stenotic lesion highlighted by arrow. Improved flow can be seen distal to the previously stenotic lesion.

Figure 2. Three-dimensional CT angiography demonstrating multiple pulmonary artery stenoses in a patient with a corrected ASD who was not a candidate for interventional procedure due to elevated pressure distal to stenoses. Innumerable lesions are seen in the proximal pulmonary arteries with post-stenotic dilation.