Diffuse Peripheral Pulmonary Stenosis as a Cause of Pulmonary Arterial Hypertension Case Report and Literature Review

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Case Report

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Abstract

Introduction: Peripheral pulmonary arterial stenosis is poorly recognized in the adult population that it can cause pulmonary hypertension, but it is likely that it is misdiagnosed as idiopathic pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension.

Case description: 16-year-old female, in functional class II, grade II systolic murmur pulmonary focus, saturation 96%. Chest tomography: left peribronchovascular consolidation. Catheterization: diffuse peripheral pulmonary stenosis, perfusion defect in the left lobe vasculature, probable thrombosis vs atresia, PMAP 78mmHg. Preserved biventricular systodiastolic function. Perfusion lung scan: pulmonary thromboembolism. Methodology: The Boolean method was used to search for MESH terms "segmental arteries" "pulmonary" [and] "stenosis" in the database with repositories of indexed arbitrary scientific evidence (PubMed) and not indexed (ScholarGoogle).

Discussion Patients with this disease usually have systemic pressures of the right ventricle secondary to obstruction in the lobar, segmental, and subsegmental branches. Small segmental artery stenting, particularly in patients with congenital diffuse peripheral pulmonary stenosis, has been less successful.

Conclusions: In recent years, peripheral pulmonary artery surgery has been associated with low mortality and a reduction in the RV / Ao pressure ratio of almost 50%. The current treatment of these patients remains somewhat controversial and poorly documented since there are places where they cannot be approached surgically.

Introduction

Pulmonary hypertension (PH) is defined as an increase in mean pulmonary arterial pressure (PAP) (mPAP)> 25 mmHg at rest, calculated by right cardiac catheterization (RCC). (1)

Peripheral pulmonary arterial stenosis (PPAS) is a poorly recognized condition in the adult population that can lead to pulmonary hypertension, but it is likely misdiagnosed as idiopathic pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. Peripheral pulmonary artery stenosis is a rare form of congenital heart disease frequently associated with Williams and Alagille syndromes, it can occur in one or multiple locations, in isolation or in association with other congenital heart defects. (2)

Case Description

16-year-old female with no significant family history. He began his condition at 13 years of age characterized by dyspnea, dizziness and limitation to physical activity, later paroxysmal nocturnal dyspnea was added. A diagnosis of Ebstein's anomaly was apparently made under treatment with beta blocker.

On admission to our service clinically in functional class II, rhythmic heart sounds, with grade II / VI systolic murmur in pulmonary focus, saturation of 96% without respiratory distress with bilateral audible vesicular murmur.

Chest tomography with peribronchovascular consolidation zone on the left side, Cardiac catheterization reports diffuse peripheral pulmonary stenosis, severe perfusion defect in the superior vasculature of the left upper lobe, probable thrombosis vs almost superior and segmental atresia, severe pulmonary arterial hypertension (Pmap 78 mmHg) (PCW 8). Pulmonary vein wedge with low pressure, so the main component is peripheral stenosis in segmental arteries and not a terminal precapillary change (arteriole and pulmonary microvasculature). Preserved biventricular systodiastolic function.

Rheumatology ruled out autoimmune alterations or connective tissue disease, reporting all negative antibodies, perfusion lung scintigraphy was performed with CT - MAA which reported data of pulmonary thromboembolism, the congenital heart disease service proposed angioplasty dilatation of the pulmonary arteries. An assessment was requested from the pulmonologist who determined that it is difficult to establish successful therapeutic treatment, so it was decided as a whole not to intervene with the patient.

Discussion

Patients with this disease often have systemic pressures of the right ventricle secondary to obstruction in the lobar, segmental, and subsegmental branches. Small segmental artery stenting, particularly in patients with congenital pulmonary stenosis, has been less successful.

Peripheral pulmonary artery stenosis is well described in children 3-5 but not in the adult population, where it is often underrecognized. Esta was recently included in the latest classification of PH (Nice, 2013) as part of group 5 (PH with unclear multifactorial mechanism), under the name “segmental PH.” PPAS is a rare disease that is commonly misdiagnosed as idiopathic. PPAS can present as an isolated congenital problem or, more commonly, as part surgery for congenital heart disease involving pulmonary artery reconstruction.

In addition, Takayasu arteritis and Behcet disease have been associated with pulmonary artery stenoses that required immunosuppressive therapy and pulmonary artery balloon angioplasty with or without stenting. Isolated cases of PPAS in adulthood have been seldom reported, a fact that can reflect a low prevalence of the disease in addition to underrecognition and misdiagnosis. These last two factors are of great relevance, since patients may not receive appropriate treatment (i.e., balloon or stent angioplasty) or the therapy might be inappropriate for PPAS (specific therapies or pulmonary thromboendarterectomy (3)

Treatment for PPAS includes surveillance, for those asymptomatic patients with preserved RV function and symmetric pulmonary blood flow, and balloon angioplasty, cutting balloon angioplasty, stent implantation, or surgery, for those with symptoms of right heart failure, substantial elevation of RV pressures, marked asymmetry in pulmonary blood flow (<25% of the total flow to a single lung), severe pulmonary regurgitation, or worsening hemodynamics or RV function. Recurrent stenosis is not

uncommon, as it occurs in approximately 35% of successfully dilated vessels. Stent placement in the central pulmonary arteries has been performed with success, but stenting of small segmental arteries, particularly in patients with congenital PPAS, has been less successful. In recent years, surgical intervention for PPAS has been associated with low mortality and a reduction of the RV/aortic pressure ratio in half, a hemodynamic improvement that has been maintained during follow-up.However, surgery is limited to proximal pulmonary artery lesions; distal branches are difficult for the surgeon to repair. (4)

Patients selected for balloon angioplasty are those with discrete central or multiple segmental lesions of stenosis with significant elevation of right ventricular pressures, marked inequality of blood flow and symptoms. In patients destined for univentricular circulation, pulmonary artery distortions in symptomatic patients and in those with cavopulmonary shunts are additional indications for pulmonary artery balloon angioplasty.

The vascular response to balloon dilation is complex and involves several factors including vascular endothelial growth factor, basic fibroblast growth factor, and nitric oxide. The balance between reendothelialization and smooth muscle proliferation may determine neointimal hyperplasia which leads to restenosis. Restenosis has been described after balloon dilation but little is known about its frequency, nature of occurrence, and time course.

Indications for stent therapy in the pulmonary artery tree are in those lesions not responsive to conventional balloon dilation. These are stenoses due to: kinking or tension (following the LeCompte maneuver or systemic to pulmonary artery shunts), external compression (from the neoaortic reconstruction following Norwood stage I), intimal flaps, stenoses presenting in the early postoperative period, relatively mild stenosis and restenosis following successful balloon angioplasty. Compliant (dilatable but elastic) stenotic lesions are also unlikely to respond to balloon angioplasty and would also benefit from stent therapy.

Surgical repair of peripheral pulmonary arterial lesions, in general, has been disappointing with a 50 to 60% restenosis rate at 5 years. Certain lesions however, such as supravalvular pulmonary stenosis or bifurcation stenosis of the branch pulmonary arteries are best managed by surgical repair. (5)

Conclusions

In recent years, surgical intervention in these cases has been associated with low mortality and a reduction of the right ventricular / aortic pressure ratio to fifty percent. (5)

The current treatment of patients with peripheral pulmonary artery stenosis is still somewhat controversial since the risk-benefit of the patient must always be assessed, mainly among other factors.

Declarations

Ethical approval and consent to participate

Consent was obtained from the patient.

Consent to publication

All authors approved the submission of the manuscript for publication.

Availability of data and materials

The data is available and can be used for the academic or research purposes.

Competing interests

The authors have no conflict of interest.

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Authors' contributions

All authors read and approved the final manuscript.

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Figures

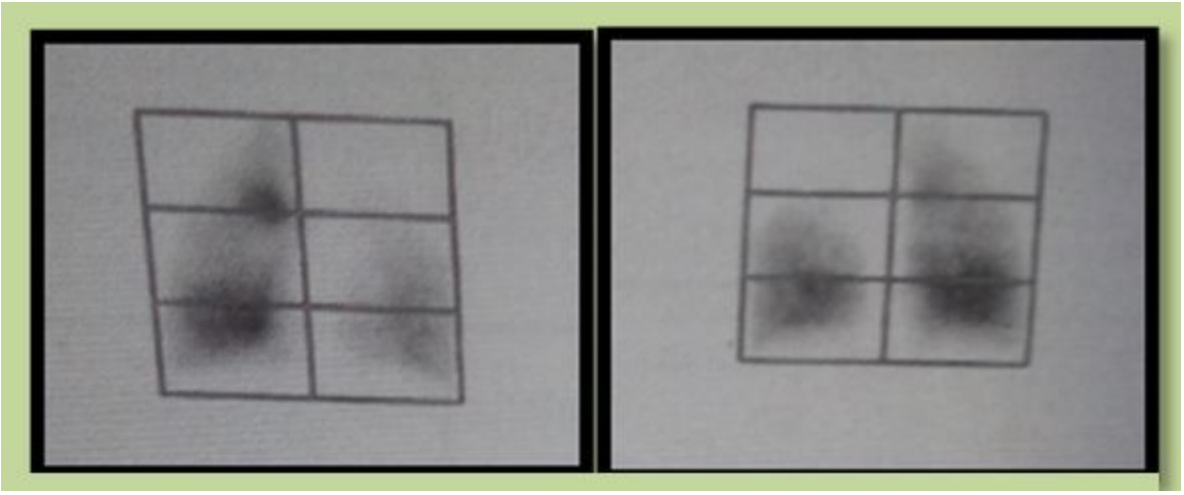

Figure 1

Perfusion lung scan with TC – MAA showing decreased perfusion in the left lung. It was also reported with data of pulmonary thromboembolism.

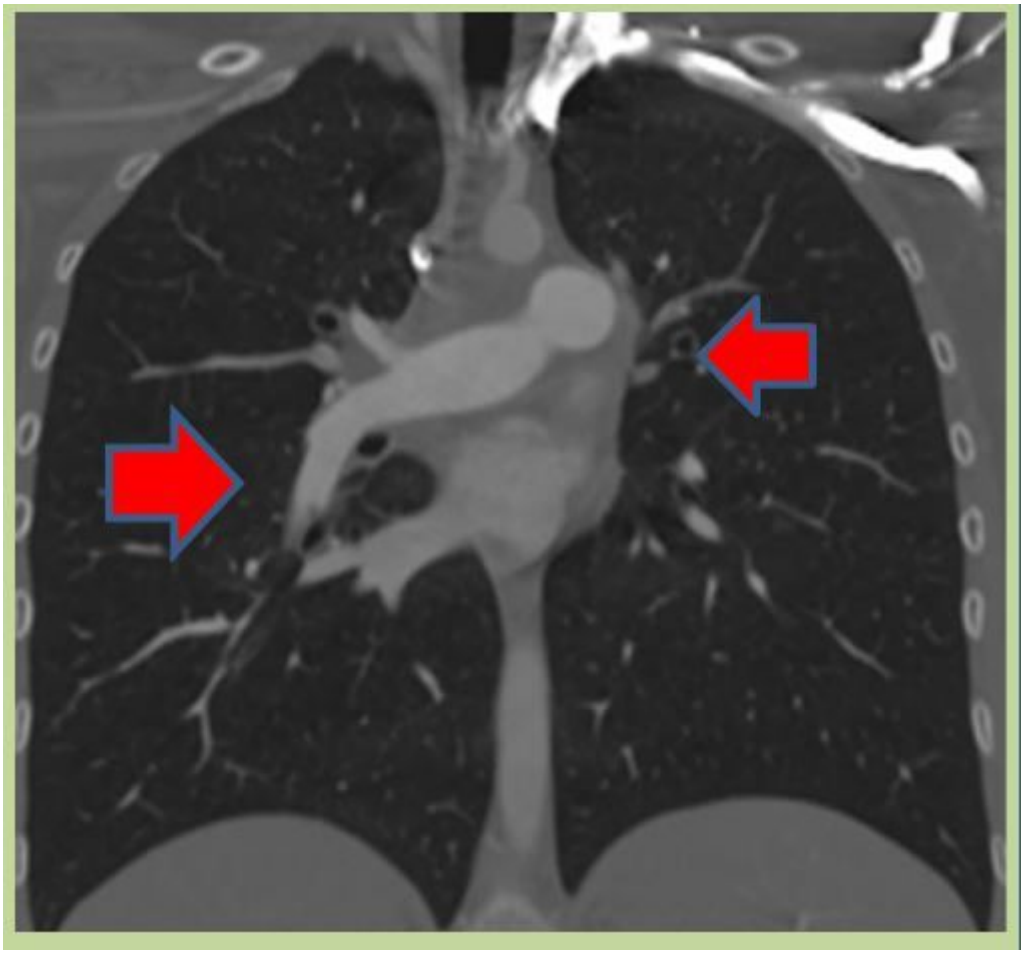

Figure 2

Chest CT scan with area of peribronchovascular consolidation on the left side and peripheral artery stenosis.

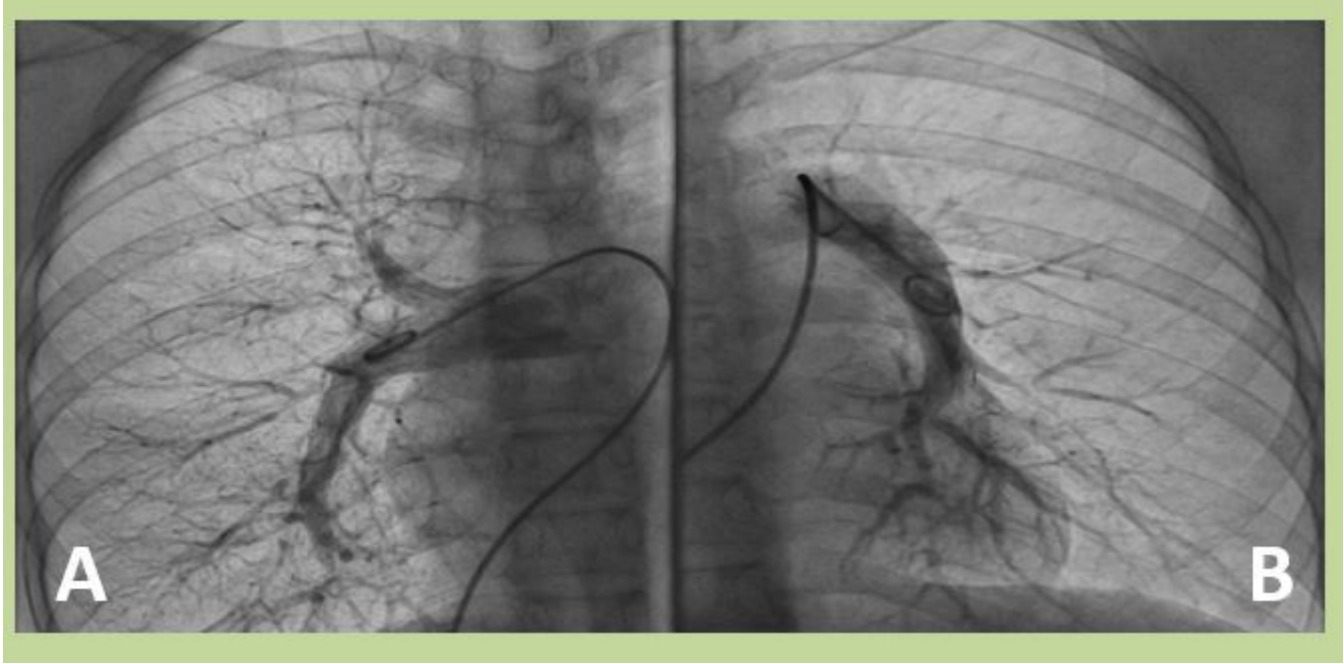

Figure 3

A) Angiography with anteroposterior projection showing stenosis of the segmental pulmonary arteries. B) Catheter in left pulmonary branch where it presents decreased distal vasculature