CASE PRESENTATION

Succesfull management of a giant pulmonary artery aneurysm associated with severe congenital pulmonary valve stenosis in an adult patient

Raluca Sosdean[1], Adina Ionac[1], Alina Stan[2], Sorin Pescariu[1], Adrian Ursulescu[1,2]

Abstract: Pulmonary artery aneurysms are rare entities usually associated with congenital anomalies that increase blood flow/pressure in the pulmonary circulation, like left to right shunts and/or severe pulmonary valve pathologies. Clear recommendations about their management are lacking. We report on a case of severe congenital pulmonary valve stenosis associated with a giant pulmonary artery aneurysm, secondary right ventricular outflow tract obstruction and right to left atrial shunt. We discuss the diagnostic pathway, therapeutic approach and patient evolution. The case report proves that a complete imaging of the heart and blood vessels is mandatory with severe pulmonary valve stenosis, in order to better describe its repercussions on the right ventricle and pulmonary circulation. They might be under detected by simple transthoracic echocardiography but their presence has important therapeutic implications.
Keywords: pulmonary artery aneurysm, pulmonary valve stenosis, right ventricle hypertrophy, resection, pulmonary homograft, reconstruction.

Rezumat: Anevrismele de arteră pulmonară sunt afecțiuni rare, de obicei asociate cu anomalii congenitale care cresc fluxul/presiunea în circulația pulmonară, precum șunturile stânga-dreapta și/sau patologii valvulare pulmonare severe. Recomandările cu privire la managementul lor sunt precare. Noi raportăm un caz de stenoză pulmonară valvulară severă asociată cu un anevrism gigant de arteră pulmonară, obstrucție secundară a tractului de ejecție al ventriculului drept și un șunt atrial drepta- stânga. Sunt discutate metodele de diagnostic, atitudinea terapeutică și evoluția pacientei. Prezentarea de caz demonstrează că o investigare imagistică completă a cordului și a vaselor mari este obligatorie la pacienții cu stenoză pulmonară severă, pentru a descrie cât mai exact repercusiunile acesteia asupra ventriculului drept, respectiv asupra circulației pulmonare. Acestea pot fi nedetectate prin ecocardiografie transtoracică însă prezența lor are implicații terapeutice importante.
Cuvinte cheie: anevrism de arteră pulmonară, stenoză valvulară pulmonară, hipertrofie ventriculară dreaptă, rezecție, homograft pulmonar, reconstrucție.

INTRODUCTION

Pulmonary artery aneurysms (PAA) represent a dilation of the main pulmonary artery of more than 40mm[1]. They are very rare entities; studies show a prevalence of 0.007% with 89% in the pulmonary artery trunk[2]. Cases have been described mostly in association with congenital defects like left to right shunts (ventricular/atrial septal defects, persistent ductus arteriosus) but also with pulmonary valve stenosis, regurgitation or absence[1,3]. In these cases, PAAs are the result of a significantly increased blood volume in the pulmonary artery and/or of the elevated wall shear stress due to the high pressure pulmonary flow[1,4,5]. They may also be secondary to acquired diseases like infections, autoimune diseases, vasculitis, and pulmonary hypertension- because of decreased resistance of the pul-

[1] Department of Cardiology, Institute of Cardiovascular Diseases, „Victor Babes" University of Medicine and Pharmacy, Timisoara, Romania
[2] Department of Cardiovascular Surgery, Robert-Bosch-Hospital, Stuttgart, Germany

Contact address:
Adina Ionac, Institute of Cardiovascular Diseases, 13A Gheorghe Adam Street, 300310, Timisoara, Romania.
E-mail: adina.ionac@gmail.com

monary artery wall- as well as iatrogenic or idiopathic. Cystic medial necrosis was detected in many samples after surgical resection of the PAA, but also normal samples were described[1,3]. Pulmonary stenosis has been asociated with elastic fibres deficiency in the pulmonary artery wall, predisposing to dissection during balloon valvuloplasty[6].

Because of their low frequency, data in the literature is still scarce regarding the PAAs pathophysiology and evolution, especially regarding dissection risk, and specific evidence based indications regarding their management are lacking[1]. Their diagnosis also has important implications in the associated lesions' treatment. These aspects are very important as PAAs may sometimes be hard to detect with conventional imaging techniques.

Our aim is to report the case of an adult patient with an unusual association between PAA and congenital pulmonary valve stenosis (PVS) with an excessive right ventricular outflow tract (RVOT) hypertrophy with significant systolic narrowing and a right to left (R-L) atrial shunt, describing the diagnostic pathway, therapeutic approach and patient evolution.

CASE PRESENTATION

A 47-year-old female patient, with known congenital pulmonary valve stenosis for which she initially deferred surgery, was admitted for advanced right heart failure symptomatology and signs, exacerbated in the last couple of months. Her medical history revealed, besides the congenital pulmonary stenosis, a grade II arterial hypertension and chronic C virus hepatitis. The heredo- collaterals were insignificant. Physical exam revealed grade II obesity, digital clubbing, perioral cyanosis and a palpable systolic impulse within the second left anterior intercostal space, with a grade 3/6 systolic murmur consistent with the pulmonary stenosis.

The ECG showed sinus rhythm with complete right bundle branch block and RV hypertrophy (Figure 1) and the postero-anterior thoracic radiography revealed cardiomegaly. The laboratory tests revealed erythrocytosis, hypoxemia and hepatocytolysis.

Transthoracic echocardiography (TTE), in spite of the low quality image, revealed a significantly hypertrophied RV without dilation (free wall of 8mm, RV end-diastolic diameter of 25mm) with a preserved systolic function (TAPSE of 21mm), severe pulmonary valve stenosis with a peak/mean pressure gradient of 62/31mmHg and an acceleration time of 300ms, and a large interatrial septum aneurysm. A subvalvular stenosis could not be excluded at this point. The right ventricle outflow tract was significantly hypertrophied and consecutively narrowed. Color Doppler revealed flow turbulence starting from the subvalvular area. Pulsed-wave Doppler at this level revealed a late-peaking dagger-shaped appearance of the signal, with a flow pressure of 16mmHg (Figure 2). No other significant problems could be found. The LV was normal sized with a preserved ejection fraction and a type I diastolic dysfunction, mild mitral and tricuspid valve regurgitation, and slight dilation of the right atrium. Transesophageal echocardiography (TEE) detected a small right to left shunt through a patent foramen ova-

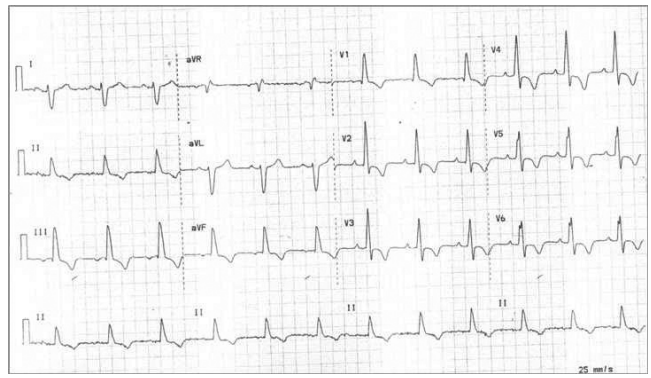

Figure 1. Twelve lead ECG revealing sinus rhythm with RBBB and RV hypertrophy.

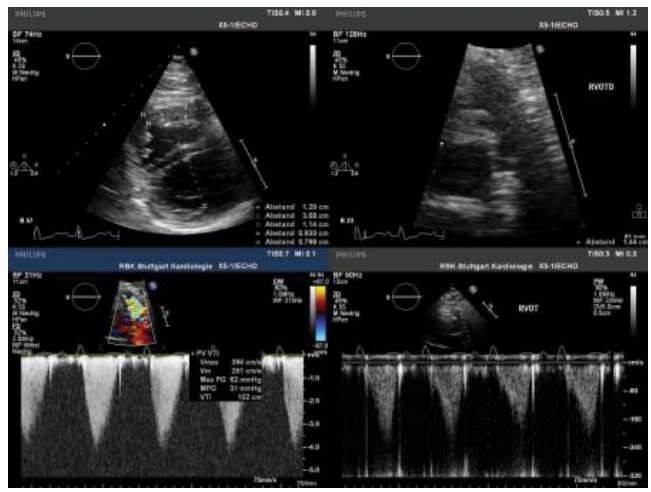

Figure 2. Transthoracic echocardiography. Upper-left: parasternal short axis view- hypertrophied RV with a D-shaped LV secondary to RV pressure overload; Upper-right: parasternal short axis view-narrowed RVOT; Lower-left: short axis view, Continuous Wave Doppler- transpulmonary valve gradient of 62mmHg; Lower-right: short axis view, Pulsed Wave Doppler- RVOT (subvalvular) gradient of 16mmHg.

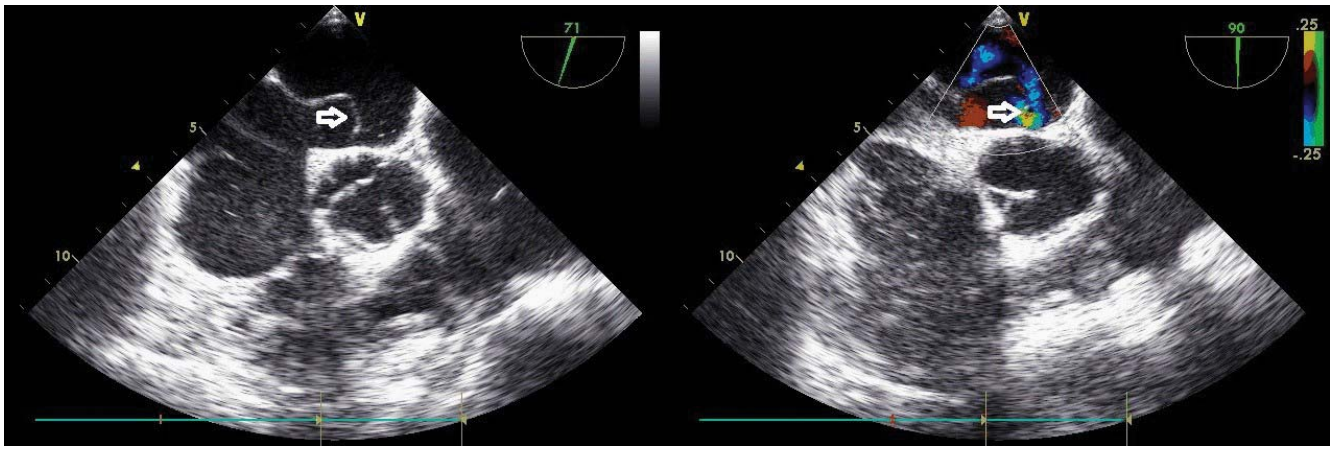

Figure 3. Transesophageal echocardiography. Left: interatrial septum aneurysm (arrow); Right: permeable foramen ovale with a small right to left shunt (arrow).

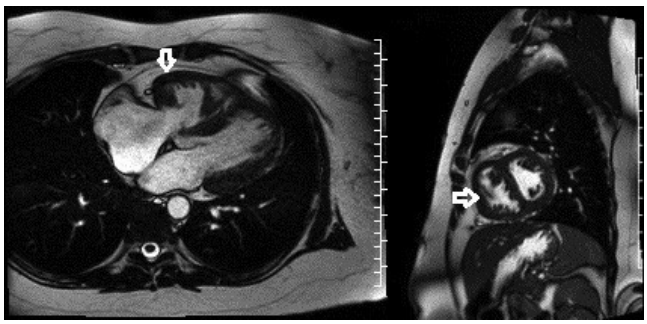

Figure 4. Cardiac magnetic resonance, True FISP Cine- Sequences. Left: transverse image-right ventricle hypertrophy (arrow), right atrial dilation; Right: sagittal image- right ventricle hypertrophy (arrow).

le, without any other supplemental findings (Figure 3). The pulmonary artery could not be properly visualized by echocardiography.

For proper and complete imaging of the actual disease, cardiac magnetic resonance (CMR) was performed. It showed a PV with significant doming and an effective opening area of 0.8cm^2 with flow acceleration starting in the RVOT in the presence of an important RVOT narrowing, as a result of excessive RV hypertrophy. It also revealed an important unknown PAA with a diameter of 58mm. The flow direction pointed to the area of maximal dilation (Figure 4 and 5). The other findings were concordant with the echo results.

Coronary catheterism revealed normal epicardial coronary arteries.

Surgical treatment was preferred over the percutaneous PV dilation despite the concern for unpredictable complications, as it offered the possibility of a complete therapy.

The procedure was optimally performed by an interdisciplinary team consisting of cardiac surgeons, an interventional cardiologist and an anesthesiologist. A classical median sternotomy was performed under general anesthesia. Cardiopulmonary bypass was established by cannulating the ascending aorta and the

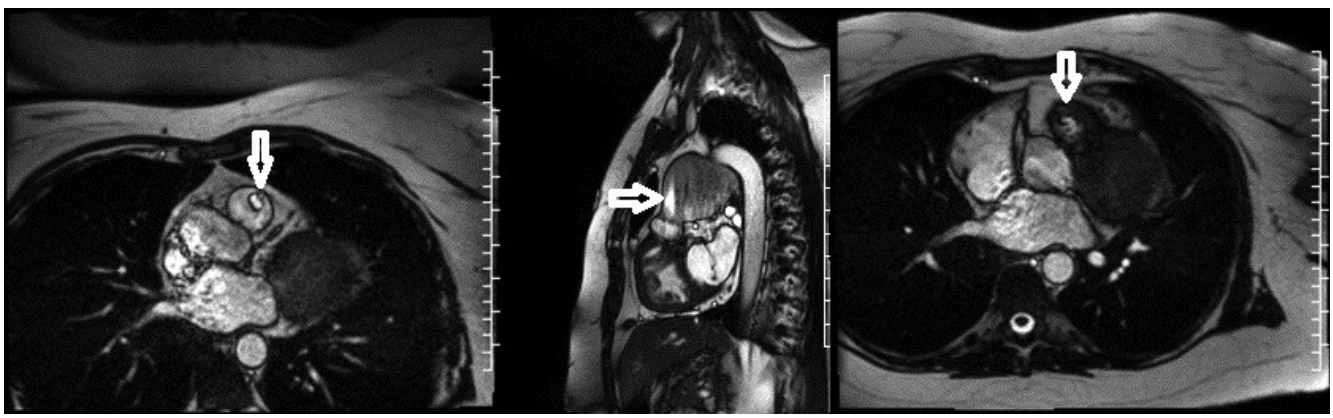

Figure 5. Preoperatory cardiac magnetic resonance, True FISP Cine- Sequences. Left: transverse image- reduced pulmonary valve opening (arrow); Middle: sagittal image- gigantic main pulmonary artery aneurysm (arrow), significant RVOT narrowing; Right: transverse image- significant RVOT narrowing (arrow). See text for details.

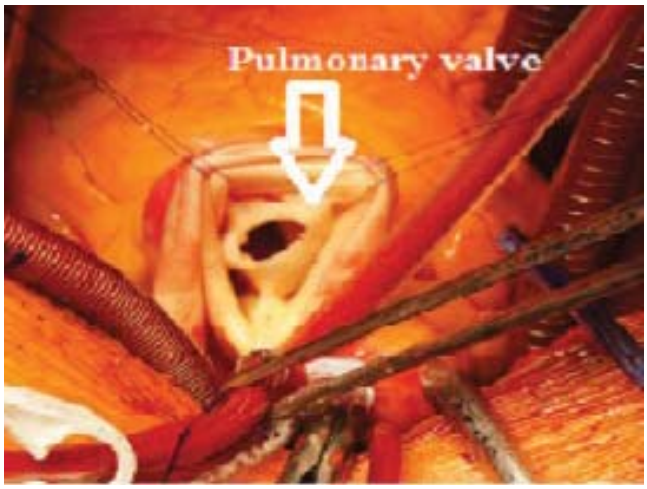

Figure 6. Sclerotic pulmonary valve with sutured commissures and impaired opening. Intraoperatory view.

superior and inferior vena cava. The ascending aorta was clamped and anterograde cardioplegy was started. The pulmonary valve could be inspected after opening the pulmonary artery. It was severely fibrotic and sclerotic so the decision was to replace the valve instead of repairing it (Figure 6). The excision of the pulmonary valve revealed a marked RV hypertrophy with a fibrotic aspect and severe RVOT stenosis. The RVOT was reconstructed using a pericardial patch, the pulmonary valve was replaced with a homograft, which was designated for the valve and the proximal part of the PA, the PAA was resected and replaced with a Y-vascular prosthesis, and the atrial shunt was sutured. The intraoperative TEE validated a good result with no pulmonary regurgitation. The surgical intervention lasted for 208 minutes. Postoperatively the patient was transferred to the intensive care unit, where she was extubated after 5 hours.

Histopathological examination of the valve and artery revealed myxoid degenerescence with a high quantity of fibrotic tissue especially in the valvular segment. Postoperative evolution was favorable, without complications, except for transient pleural and pericardial effusion that resolved with medical therapy. TTE, TEE and CMR reevaluation along with the 3-year symptom-free evolution of the patient demonstrated the success of the intervention (Figure 7), without alteration of the RV systolic function. Her long- term medication included an antiplatelet drug, an angiotensin converting enzyme blocker, a beta-blocker, a loop diuretic and an aldosterone receptor blocker.

DISCUSSIONS

Pulmonary artery aneurysms are frequently associated with pulmonary stenosis or left to right shunts including atrial septal defects. When both pulmonary valve stenosis and atrial septal defects are present, significant left-to-right shunt is often prevented by the RV outflow obstruction, which protects the pulmonary bed until adulthood[7]. In our case, the cause of the aneurysm development was, most probably, the high pressure through the pulmonary stenosis acting on a pathologically modified pulmonary artery wall structure. The atrial shunt in this case was the result of the high pressure in the right cavities, secondary to the outflow obstruction, acting on the interatrial septum and resulting in a significant IAS aneurysm and opening the foramen ovale as an adapting mechanism to reduce the pressure. This might have slowed the evolution to heart failure, explaining the late symptomatology, but

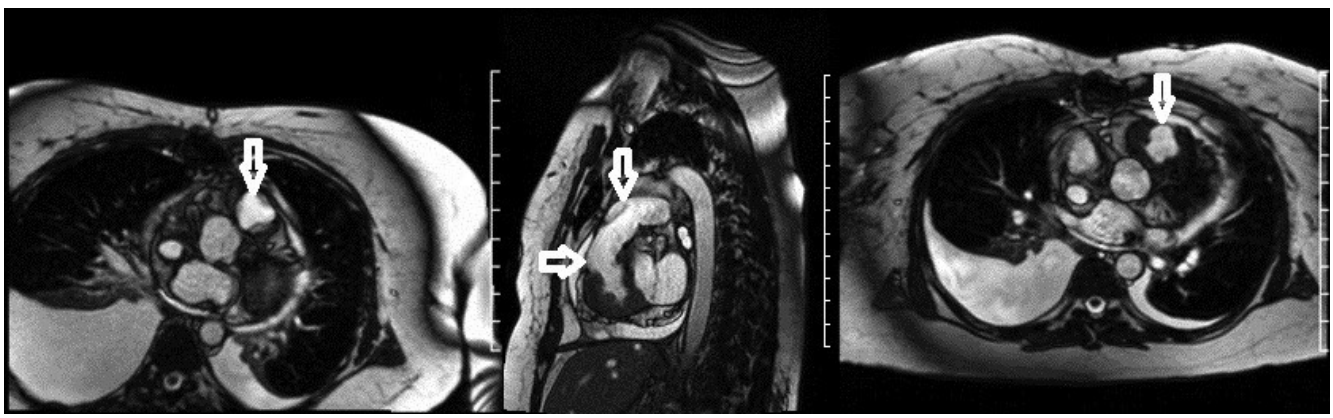

Figure 7. Postoperatory cardiac magnetic resonance, True FISP Cine- Sequences. Left: transverse image- normal pulmonary valve homograft opening (arrow); Middle: sagittal image- normal aspect of the main pulmonary artery prosthesis and of the RVOT plasty (arrows); Right: transverse image- normal aspect of the RVOT plasty (arrow). See text for details.

it was not enough to prevent the PAA development. The spontaneous closure was uncertain after resolving the obstruction, and for this reason the decision was to close it during the same surgical intervention.

A debatable aspect is the RVOT (subvalvular) stenosis. First, there is the discrepancy between the anatomical aspect of the RVOT visualized by echo, but better by CMR and direct inspection during surgery, which proved subvalvular stenosis, and the uncertain/ indefinite aspect of the PW Doppler flow anvelope at this level. This problem was also described in patients with coexisting aortic stenosis and obstructive hypertrophic cardiomyopathy. The left ventricle outflow tract obstruction is often masked in this situation by the extreme afterload of the LV induced by the valvular stenosis, consecutively minimizing the subvalvular obstruction in an artificial manner[8]. This may also be a valid explanation for our situation. Another problem is the possible RVOT obstruction worsening immediately after PS afterload relief with refractory heart failure worsening. Cases of right ventricular failure because of exacerbated RVOT gradient after pulmonary balloon valvuloplasty are described in the literature. Beta- blockers proved to be efficient in this situation and the RV hypertrophy is reported to regress after several weeks-months even in adults[9]. Data is poor in the literature, though, and given the fibrotic aspect of the RVOT in our patient and the risk for this to impede reverse remodeling, RVOT plasty was considered to be a safer approach[9,10]. The significant improvement in the afterload after surgery, without pulmonary regurgitation, contributed to the preservation of RV function by decreasing intraventricular pressures, wall stress and oxygen demand[11].

Most probably the PAA developed secondary to the high velocity and pressure of the flow through the stenotic pulmonary valve, acting as a long-term and continuous stress on a pathologically modified PA wall structure. The CMR aspect was concordant with the observations of Fenster et al; they demonstrated a supravalvular systolic separated flow, with a significant region of counterclockwise recirculation, by 4D CMR, in a 33 year old woman with bicuspid pulmonary valve and moderate pulmonary stenosis. They hypothesize that this flow may be responsible for pulmonary artery aneurysm formation in these patients4. The best therapeutic option is still debated. Some authors reported cases without complications and even regression of the aneurysm after valvuloplasty for pulmonary stenosis[12]. Other authors recommend the use of the same criteria as for aortic aneurysms to decide surgical intervention, consisting in a diameter of more than 5,5cm, symptoms, more than 0,5cm diameter increase in 6 months, complications, etc[1]. A careful management is generally recommended, because even if the rupture risk is low, there are also other possible fatal complications that may occur, like pulmonary embolism, heart failure and compression of the surrounding structures including the coronary arteries[13]. Duijinhouwer et al. concluded in a recent review and analysis that pulmonary hypertension in congenital heart disease (CHD), a fast diameter growth (>2 mm/ year), tissue weakness mostly due to infection are the most frequent predisposing factors for dissection, and in the rest of the cases, the risk is low[3]. On the other hand, Adodo et al consider PA dissection to be underestimated because it is most often a fatal condition, as it has the tendency to rupture rather than extend[14]. The surgical outcomes for PAA are satisfactory according to data in the literature. There are two main strategies: aneurysmorrhaphy with excision of the excessive arterial and complete replacement with arterial grafts, including homografts and Y-vascular grafts. In our case, complete replacement was preferred, as cases of PAA recurrence after aneurysmorrhaphy were reported. The Y-vascular graft was used along with the homograft because of the gigantic dimensions of the aneurysm. Also, in adults with PAA secondary to congenital heart disease, the PA wall is weak[15,16], and may increase the risk of such complications on the long term.

Interventional treatment in this case would have been incomplete and most probably with a poor outcome, even if the surgical risk was high in this patient, given all the pathologies that needed to be resolved and the associated comorbidities.

CONCLUSIONS

The entity associating PAA and congenital PVS in adulthood is rare, but a complete imaging of the heart and great vessels is always mandatory with this congenital disorder as it has important treatment implications. Surgical intervention in this setting represented the only appropriate and complete curative treatment. The case is worth mentioning as it reports an unusual presentation of a rare congenital disease in an adult patient, describing its complete particular phenotype, management and long-term evolution.

Conflicts of interes: none declared.

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