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Case Report

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Pulmonary Artery Aneurysm in an Adult with Pulmonary Hypertension and Unrepaired Congenital Heart Disease

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Abstract

Congenital heart disease is the most frequent malformation. It is estimated that 10% are diagnosed in adulthood. In adult population the true prevalence is six cases per 1,000 adults. The main complication in unrepair congenital heart disease is pulmonary arterial hypertension. With serious heart defects pulmonary artery aneurysms can be a complication. The hemodynamic stress caused by increased left to right flow and left pressures transmitted to the pulmonary artery fosters the pulmonary artery aneurysms formation. I present a case of a young adult with severe unrepair congenital heart disease, pulmonary arterial hypertension and secondary pulmonary artery aneurysm.

Case

A 26 year old female patient with a late childhood diagnosis of a double outlet right ventricle (DORV), ventricular septal defect (VSD), and pulmonary arterial hypertension (PAH). Due to the severity of the PAH and high surgical risk, the congenital heart disease (CHD) was not repaired. She did not receive medical treatment for more than two decades. A year ago she was hospitalized for an episode of hemoptysis. On a recent assessment in the adult congenital heart disease unit (ACHDU) and pulmonary hypertension department she had an NYHA functional class III, 72% saturation on room air, precordial pain, increased right sternal activity, a right infraclavicular thrill, a systolic ejection murmur and a short right parasternal diastolic murmur, with an intense, palpable second heart sound. The echocardiogram showed DORV, unrelated VSD, RV hypertrophy and dilation, 15 mm TAPSE, right atrial dilation, mild pericardial effusion (Figure 1), and pulmonary artery aneurysm (PAA) (Figure 2). The diagnosis was PAH and Eisenmenger syndrome (ES). Magnetic cardiac resonance was performed, showing the same diagnosis. Dissection and thrombosis were excluded (Figure 3). She was treated with pulmonary vasodilators (sildenafil, bosentan). On follow-up she had 80% saturation on room air, an NYHA functional class II, resolved pericardial effusion, 17 mm TAPSE and no chest pain.

Discussion

CHD is the most frequent malformation, sixty percent of CHDs are diagnosed and treated in the first year of life, and 30% throughout childhood and adolescence up to 16 years of age. It is estimated that 10% are diagnosed in adulthood [1]. Nowadays CHD has a true prevalence is thirteen cases per 1,000 children and six cases per 1,000 adults [2]. The adult congenital heart disease population (ACHD) is growing at a rate of 5-6% per year, and it is estimated that there will be 11% more per year in 2030 [3]. There are great differences between European and American ACHD populations, especially the Latin American population. An overview in developing countries the ACHD population

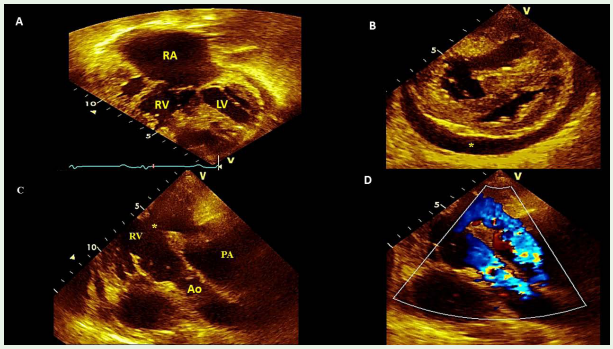

Figure 1: A: apical 4 chamber view shows dilated RA, dilated and hypetrophy RV. Due to the large dilatation and anterior position of RA, do es not possible to see left atrium in this view. **RA:** right atrium, **RV:** right ventricle, **LV:** left ventricle. **B:** short axis view shows mild pericardial effusion (*). **C:** apical modified RV inflow and outflow tracts view, shows DORV and unrelated VSD (*). **D:** comparative doppler color shows the same in C.

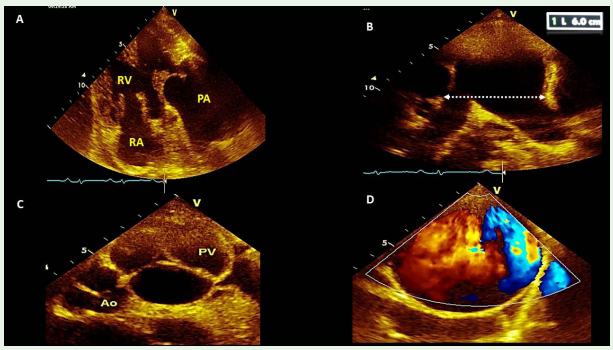

Figure 2: A: apical modifield RV outflow tract view shows dilated pulmonary artery. **PA**: pulmonary artery. **B**: pulmonary artery aneurysm, in this view was measured in 60 mm. **C**: short axis view of the great vessels, in this view due to the PAA, the PV is placed in a more anterior position and it is possible to see the 3 veils (these are not usually seen in routine echocardiographic studies). **PA**: pulmonary artery, **Ao**: Aortic valve. **D**: short axis view doppler color during systole, just above the PV, at the PT level. This view shows PAA, the systolic flow into the PT has a circular motion (red and blue color at the same time), due to the PAA and this was the explanation about the thrill in the patient.

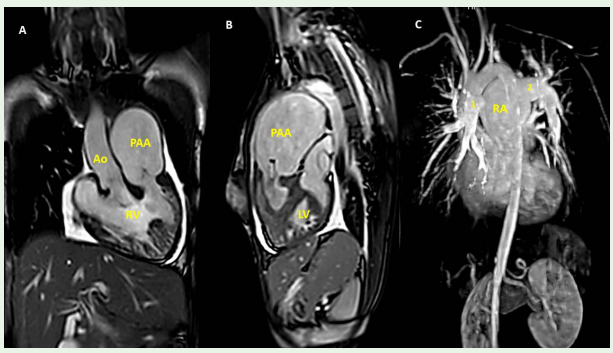

Figure 3: A: coronal view shows PAA and DORV and unrelated VSD. **B**: longitudinal view shows PAA. **C**: angiography shows PPA central, Left (1) and right (2) branch are not dilated.

is still young, their health status oscillates between relapses and hospitalizations due to CHD decompensation. The most frequent hospitalization emergencies in ACHD include: arrhythmia, heart failure, syncope, aortic dissection, endocarditis, thromboembolism, bleeding (noncerebral), pulmonary hypertension, aortic aneurysm/ dissection, and sudden cardiac death [4]. PAH is defined as pulmonary medium pressure (PMP) by right heart catheterization ≥ 25mm Hg, pulmonary capillary wedge pressure ≤15 mm Hg and pulmonary vascular resistance ≥3 Wood units.

PAA have a low prevalence of 0.057% in 109,571 consecutive autopsies [5]. Fifty-six percent are caused by CHD, with the three most frequent being persistent ductus arteriosus, VSD, and atrial septal defect. PAH causes 1.25% of aneurysms, and ES 6.5%. In unrepaired CHD, persistent increased pulmonary flow leads to PAH, endothelial hyperplasia and hypertrophy of the medial layer of the arterioles, and increased pulmonary vascular resistance (PVR), at the same time the hemodynamic stress caused by increased left to right flow and left pressures transmitted to the pulmonary artery (PA) fosters the formation of PAA. The currently expert guidelines recommendations about when a CHD may be repaired if the following criteria are met:

1. under baseline conditions:

- PVR ≤ 2.3 wood units (WU) or PVRi (index) < 4 WU/m2
- PVR/ systemic vascular resistance (SVR) ratio is ≤ 0.3.

2. in the event that the PVR is ≥ 2.3-4.6 WU or 4-8 WU/m2 and the PVR/SVR ratio is 0.3-0.5, a pharmacological challenge with nitric oxide is required to determine pulmonary vascular reactivity. The challenge is considered to be positive if the PVR or PMP decrease by 20%, the PVR/SVR ratio decreases by 20% (< 0.33), or the PVR is ≤ 4-5 WU/m2. Finally when PVR > 4.6 WU or PVRi > 8 WU/m2, CHDs should not be repaired [6].

The international clinical classification of PAH associated with CHD includes 4 groups [7]:

1. Eisenmenger syndrome

Includes all large intra and extra cardiac defects which begin assystemic to pulmonary shunts and progress with time to severe elevation of PVR and to reversal (pulmonary to systemic) or bidirectional shunting; cyanosis, secondary erythrocytosis and multiple organ involvement are usually present.

2. Left-to-right shunts

- Correctable
- Noncorrectable

Include moderate to larged efects; PVR is mildly to moderately increased systemic to pulmonary shunting is still prevalent, whereas cyanosis is not a feature.

3. PAH with coincidental CHD

Marked elevation in PVR in the presence of small cardiac defects, which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. To close the defects in contraindicated.

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4. Post-operative PAH

CHD is repaired but PAH either persists immediately after surgery or recurs/develops months or years after surgery in the absence of significant postoperative hemodynamic lesions. The clinical phenotype is often aggressive.

The present case is from clinical group 1 (ES) commonly manifested in chronic hypoxemia with multiple systemic complications, just like in this case. The evolution time, severity of CHD and clinical signs are sufficient for the diagnosis and cardiac catheterization is not necessary. Patients with ES are at high risk for procedures that require general anesthesia.

PA diameter > 40 mm is considered to be a PAA. It is central if it is found in the pulmonary trunk and the main right and left PAs. The case presented was diagnosed in childhood and cardiac catheterization was performed with measures of PVR (no data available), due to the severity of the PAH, was not possible to repair the CHD, however in the past decades there were no available pulmonary vasodilators, and maybe that was one reason why special medical therapy was never administered in this patient, in addition to poor medical control by a specialist in the field. Medical therapy with pulmonary vasodilators was enough to improve the patient's clinical condition, demonstrated by increased saturation, better functional class and resolving the pericardial effusion.

Today ACHD population must be concentrating in specialized centers of management in CHD and PAH. They must be managed by a sub specialized cardiologist in ACHD. Mortality in ACHD is reduced when their management adheres to the directives in the currently available guidelines [7]. PAA can be complicated with dissection, rupture, thrombosis, hemoptysis, and compression of adjacent structures [8]. Pulmonary artery aneurysms related to unrepaired CHD, PAH and ES should only receive medical treatment; they cannot be repaired [9]. That is the best option recommended by experts in adults with CHD.

Conclusions

PAA have a low prevalence in general population and when they are associate with CHD and PAH serious complications are described. Due to the severity of PAH, especially in the group with SE, the best alternative is medical treatment with pulmonary vasodilators. ACHD populations are chronic patients; they should never be discharged from cardiology follow-up, which should be done in an ACHDU and by an expert in PAH.

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