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Clinical case

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Abnormal origin of the right pulmonary artery from the aorta. Case report and literature review

Nacimiento anómalo de la rama pulmonar derecha de la aorta. Reporte de caso y revisión de la literatura

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Palabras clave:
Nacimiento anómalo, rama pulmonar derecha, aorta, cirugía correctiva.

ABSTRACT

A 43 y/o male with a history of patent ductus arteriosus and corrective surgery of the defect performed at 11 years of age. He remained asymptomatic for a long time until he developed shortness of breath on moderate effort, associated with palpitations, productive cough and hemoptysis. On the initial investigation by Internal Medicine, he was diagnosed with a right basal pneumonia, and as part of the diagnostic approach, a chest CT scan was made, which reported an abnormal origin of the right main pulmonary artery from the ascending aorta. He was transferred to the National Institute of Cardiology, where a MRI and a cardiac angiography were performed. The diagnosis was confirmed and pulmonary hypertension was also reported. The case was discussed by the heart team, and it was decided to take him to corrective surgery. The procedure was performed two months later, with the implantation of the right pulmonary branch to the main pulmonary artery using a synthetic Dacron tube. This case is presented, since it is extremely rare to find such a pathology, making the diagnosis in adulthood, with an insidious clinical presentation, which also has an adequate clinical evolution, after surgical repair.

RESUMEN

Paciente masculino de 43 años, en el cual se detecta persistencia del conducto arterioso a los 11 años de edad, por lo cual se realiza cirugía correctiva a dicha edad. Se mantiene asintomático por largo tiempo hasta que inicia con disnea de moderados esfuerzos, acompañado de palpitaciones, tos productiva y hemoptisis. Es abordado en un principio por Medicina Interna, quien realiza el diagnóstico de Neumonía basal derecha; además, como parte del abordaje, se realiza tomografía axial computada de tórax, en la cual se encuentra el hallazgo de origen anómalo de la rama derecha de la arteria pulmonar, con procedencia de la aorta ascendente. Es trasladado al Instituto Nacional de Cardiología, donde se realizan estudios complementarios como resonancia magnética y cateterismo cardiaco derecho e izquierdo, corroborando dicho diagnóstico y determinando la presencia de hipertensión pulmonar. En sesión médico-quirúrgica, se decide hacer cirugía de corrección, la cual se lleva a cabo casi dos meses después, con reimplante de la rama anómala a la arteria pulmonar y ayuda de la colocación de un tubo de Dacrón. Posterior a la cirugía, el paciente presenta adecuada evolución clínica.

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INTRODUCTION

Hemitruncus, is a rare condition that represents the 0.05% of congenital heart anomalies.[1] One of the lungs is irrigated by the aorta, whereas the other is perfused by the main pulmonary artery, in the presence of two semilunar valves.[2]

This condition was first described by O. Fraentzel, in 1868. The anomalous origin of the right pulmonary artery from the aorta is 5-8 times more common than the anomaly of the left pulmonary artery. In 20% of the cases it is reported as an isolated malformation.[2] We present the case of this interesting pathology in an adult patient, in whom it was not possible to achieve a precise diagnosis at an early age, operated at 11 years of age for the closure of a patent ductus arteriosus.

CLINICAL CASE

A 43 y/o male patient, who was born and actually is resident in Mexico City. His past

medical history was only positive for the exposure to *Trypanosoma cruzi* in his childhood and a blood transfusion when he was 11 y/o during the surgical closure of a patent ductus arteriosus at the National Institute of Cardiology. He denies consumption of alcohol, tobacco or drugs, and has no history of chronic diseases.

The patient started his present illness complaining of shortness of breath on moderate efforts, along with palpitations, productive cough and hemoptysis. He was hospitalized in the Internal Medicine Department, where they found signs that were compatible with a right lower lobe pneumonia. A chest CT scan was made, reporting cylindrical bronchiectasis and as an incidental finding the abnormal origin of the right pulmonary artery from the ascending aorta. According to the abovementioned, it was decided to continue the investigation and treatment at the National institute of Cardiology.

On physical examination, he showed no signs of heart failure. The apex was palpated at the 5th intercostal space of the left mid-clavicular line, with a 2 cm diameter apical impulse. He also had a left lower parasternal heave. On auscultation he had a normal S1, with physiological splitting of the S2 on inspiration and an increased intensity of the pulmonary component of the second heart sound; drawing the attention the presence of a holosystolic murmur at the tricuspid valve area, with a III/VI intensity. All these signs pointing to an incipient pulmonary hypertension diagnosis.

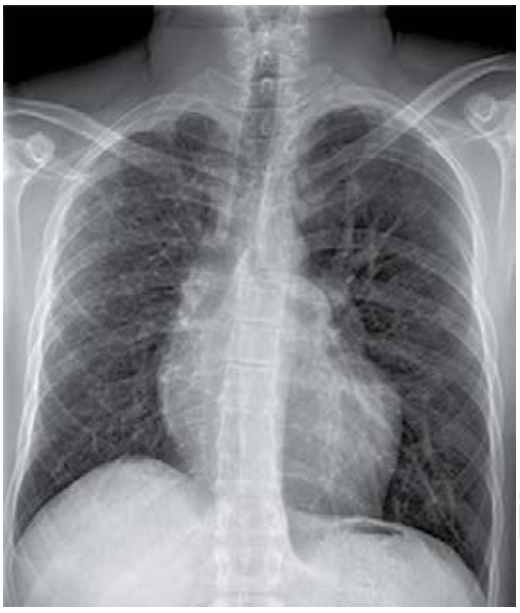

Figure 1: PA view chest X-ray, showing mild upper zone vascular flow redistribution with right lung predominance, along with enlarged pulmonary arteries with right predominance as well.

The PA (posteroanterior) view chest X-ray showed mild pulmonary flow redistribution, with a right lung predominance, along with a prominent right pulmonary artery *(Figure 1)*. A 12-lead electrocardiogram (ECG) showed an incomplete right bundle branch block and left ventricular diastolic overload *(Figure 2)*.

As part of the diagnostic approach, a transthoracic echocardiogram was performed, reporting non-dilated cardiac chambers, normal left ventricular diastolic function, a normal systolic function of both ventricles and normal valve function; it was described that the right pulmonary artery emerged from the ascending aorta.

Afterwards, to detail the vascular anatomy, a cardiac MRI was made, in which it was confirmed the anomalous origin of the right pulmonary artery from the ascending aorta as well as the following data: mild dilation of the right atrium, normal left ventricular systolic function with a left ventricular ejection fraction (LVEF) of 56%, right ventricular diameter in the upper normal limit, with a mild systolic dysfunction: ejection fraction of 45%, and with the presence of a mild tricuspid insufficiency *(Figures 3A and 3B)*.

In the inversion-recovery sequence, after the IV contrast material administration, it was observed a non-ischemic delayed enhancement pattern *(Figures 4A and 4B)*. HASTE sequence showed hyperintense areas at the apical lobe of the right lung; and as the most relevant finding, it is described that the right pulmonary artery emerges from the posteromedial side of the ascending aorta, with a diameter of 20 mm *(Figures 3C and 3D)*.

Considering the possibility of a surgical correction and specifically, to evaluate the presence of pulmonary arterial hypertension, a coronary and pulmonary angiographies were performed *(Figure 5)*, reporting the following findings:

- Normal coronary arteries.
- Oxymetric jump was not detected.
- Right pulmonary artery (RPA): vasculature with heterogeneous filling, tortuous vessels, with decrease in monopedial vessels and slow washing.
- Left pulmonary artery (LPA): vasculature with homogeneous filling and adequate amount of monopedial arteries.
- A 100% oxygen challenge was performed that showed no change in recorded pressures.

In the following table, we show the different pressure recordings in the evaluated cardiac chambers *(Table 1)*:

According to the cardiac catheterization results, it was concluded that there were signs of irreversible pulmonary arterial hypertension.

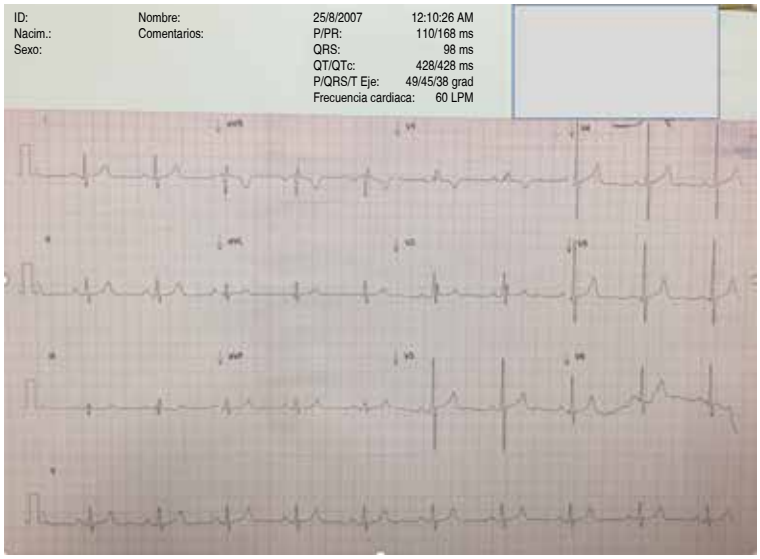

Figure 2: Twelve lead ECG, showing normal sinus rhythm, incomplete right bundle branch block and left ventricular diastolic overload.

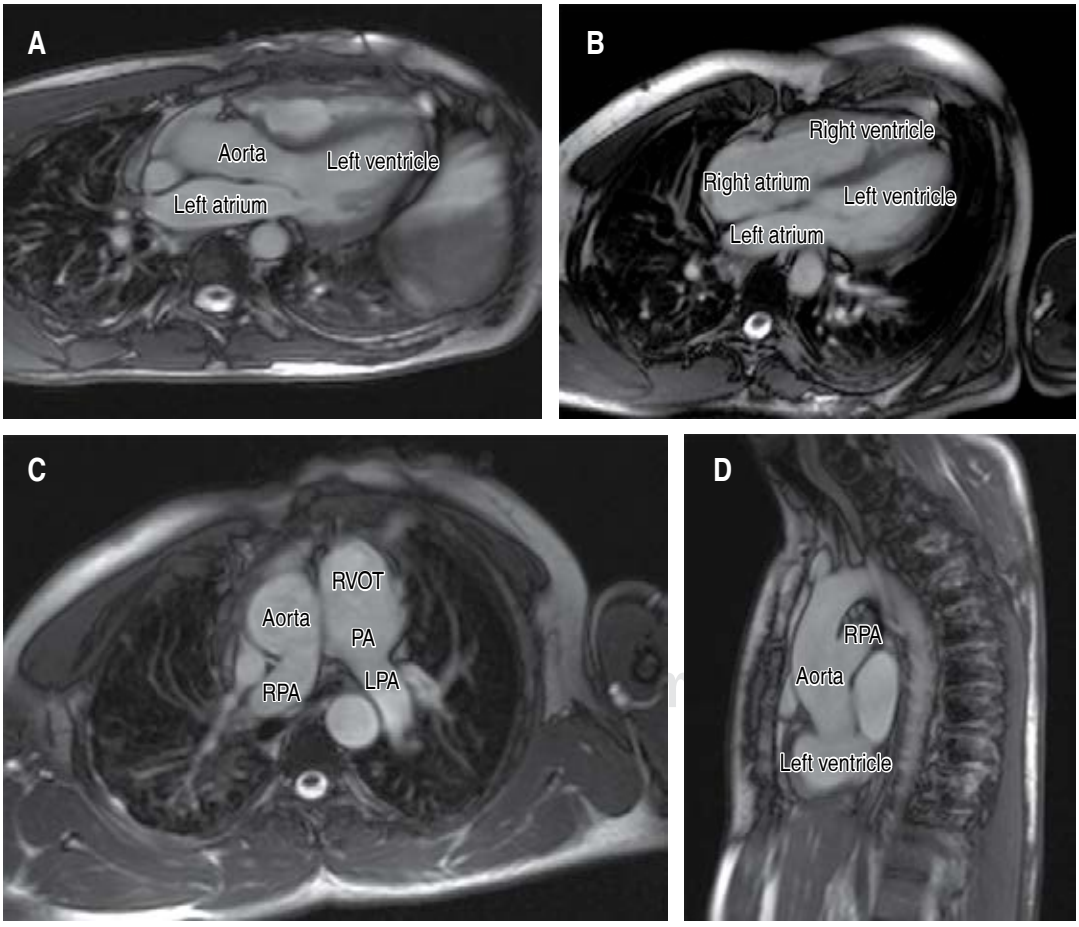

Figure 3:

Cardiac magnetic resonance (CMR) echo (Fixed) 3-chamber **(A)** and 4-chamber view **(B)**, showing normal cardiac diameters. CMR cine echo-gradient (fixed) axial **(C)** and sagittal plane **(D)**, demonstrating the origin of the right pulmonary artery from the aorta. LPA = left pulmonary artery, PA = pulmonary artery, RPA = right pulmonary artery, RVOT = right ventricle outflow tract.

However, in this particular case, the pulmonary artery pressure recording is unreliable due to the fact that the right pulmonary artery pressure is affected by its direct connection with the aorta and systemic blood pressure, that can finally impact in the pulmonary circulation.

In the background of a relatively young patient, with no other comorbidities, on a class I functional capacity of the New York Heart Association (NYHA) classification, in addition to undilated right and left ventricles, preserved right systolic function and LVEF, it was decided to take this patient to surgery.

In this particular case, the right pulmonary artery was redirected to the main pulmonary trunk, using a 16 mm woven Dacron tube and an 8 mm atrial septal defect was created as well. All these under extracorporeal circulation assistance. The patient was discharged without complications.

Two months after surgery, a chest CT scan was performed, showing post-operative modifications in the right pulmonary artery, as well as the graft patency, without evidence of stenosis or leakage. In order to evaluate the pulmonary vasculature hemodynamic evolution, a follow-up pulmonary angiogram was done. It was reported a remarkable pulmonary arterial pressure reduction, as well as a fall in pulmonary vascular resistance with 100% oxygen administration. Another oximetry run was taken but no step up was detected. The pulmonary arterial pressure recording after oxygen administration was 68/19 mean 41 mmHg and the pulmonary vascular resistance was 489 dynes, representing less than 2/3 of systemic blood pressure: 121/61 mean 83 mmHg and 677 dynes. Pulmonary angiogram showed normal size pulmonary artery branches, without filling defects, normal venous return to the left atrium and a left to right passage of contrast material through a small atrial septal defect.

Currently, the patient maintains a NYHA class I functional capacity, and is in treatment with acetylsalicylic acid 100 mg QD, sildenafil 12.5 mg BID and amlodipine 2.5 mg QD, without requiring supplemental oxygen.

DISCUSSION

Approximately, 40% of these congenital heart defects are associated to other cardiovascular anomalies,[2,3] such as: patent ductus arteriosus (75%), ventricular septal defects, aorto-pulmonary window, coarctation of the aorta, interrupted aortic arch, atrial septal defects, pulmonary vein stenosis. Accordingly, it is inferred that this condition is diagnosed at a young age; therefore, the case that we present shows a patient with this anomaly that remained relatively asymptomatic for a long time, which is unusual.

An association with CATCH 22 Syndrome has been found, including the Di George syndrome.[3] When the anomaly involves the left pulmonary artery, the congenital defect may be associated with tetralogy of Fallot.[4]

This condition must be diagnosed at an early stage, in order to perform a surgical correction at the appropriate time due to the high risk of developing an irreversible pulmonary vascular

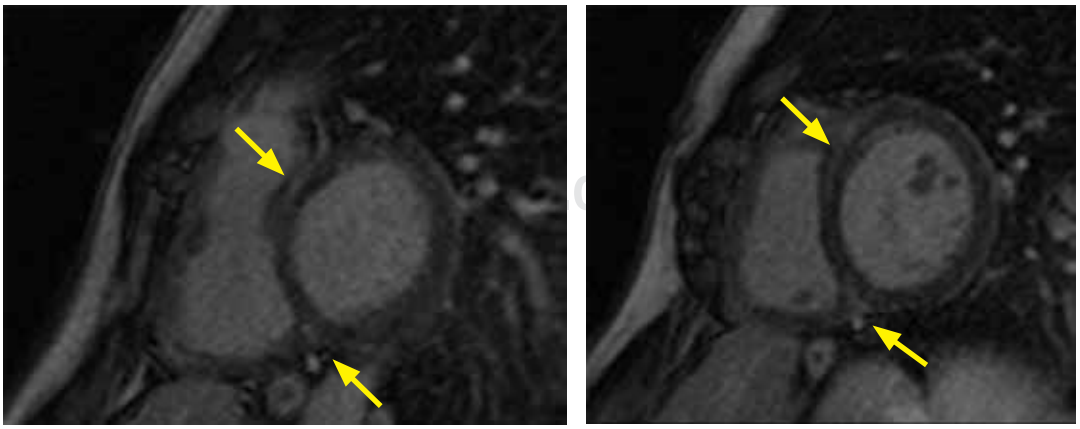

Figure 4:

Cardiac magnetic resonance (CMR) short axis view. Base **(A)** and mid-cavity **(B)** sections, showing a septal intra-myocardial late enhancement pattern, in relation to a non-ischemic pattern (yellow arrows).

disease. If it is not corrected at an early age, the survival rate reported could be as low as 30% at one year.[2]

Embryological pathology

The defect is caused by an incomplete migration of the right sixth aortic arch to the left. It can be the consequence of the lack of septation of the truncus arteriosus. The septation process starts with the emergence of two appendages in the primitive truncus, which will spread in a cephalic direction through the base of the conus and join with the conus septum. This septum will divide the aorta from the pulmonary trunk.[2] It is proposed that the anomalous origin of the pulmonary arteries could be related to a malposition of the Tandler's aorto-pulmonary septum, constituted by neural crest-derived mesenchymal cells. Normally, this septum has a right to left and dorsal to ventral orientation and separates the fourth aortic arch (located anterior and to the right and giving origin to the aortic arch) from the sixth aortic arch (located posteriorly and to the left, giving origin to the proximal portions of the pulmonary artery branches). The emergence of the right pulmonary artery from the aorta is caused by a leftward malposition of the aorto-pulmonary septum, while the anomalous origin of a left pulmonary artery from the aorta is caused by an abnormal position of this same septum to the right.[5]

Table 1: Intracavitary pressure record.

Anatomic site	Pressure in mmHg (millimetres of mercury)
LPA	50/25 mean 33
RV	50/4 end-diastolic 28
RA	Mean 4
RPA	120/80 mean 80
LV	120/ 80 D2 10

LPA = left pulmonary artery, RV = right ventricle, RA = right atrium, RPA = right pulmonary artery, LV = left ventricle, D2 = left ventricular end-diastolic pressure.

Classification

These types of defects are classified among the type III group of aorto-pulmonary septation defects. Usually originated at the posterolateral portion of the ascending aorta, in contrast with the presentation of our patient in whom it was

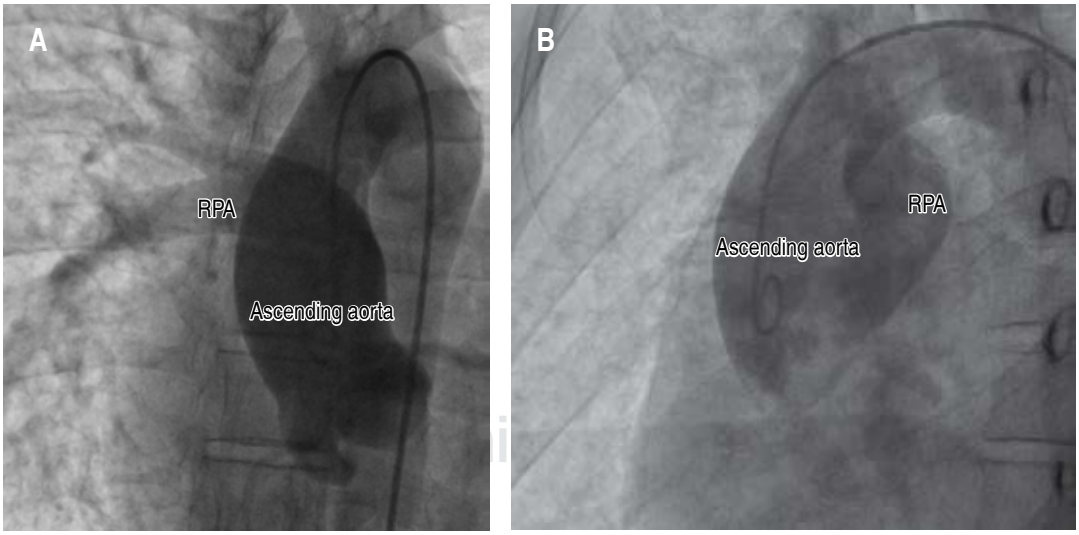

Figure 5: A. Aortography. Anteroposterior projection with caudal angulation. It is observed the presence of a 5fr pigtail catheter in the ascending aorta from a femoral access. Calling the attention the anomalous origin of the right pulmonary artery. **B.** Aortography. Left lateral projection with 90º angulation, where it is confirmed the anomalous origin of the right pulmonary artery from the posteromedial border of the ascending aorta.

originated at the posteromedial portion of the aorta. The origin of the right pulmonary artery can be located at different sites: proximal, with the origin at the ascending aorta, close to the aortic valve annulus; and distal, originated from the ductus arteriosus. Some other authors accept a third origin location close to the innominate artery.[3]

Pathophysiology

The abnormally connected lung is perfused at a systemic blood pressure, while the other is exposed to all the stroke volume coming from the right ventricle.[4] If a short-circuit is added from left to right, the hemodynamic situation gets even worse.[5]

After birth, with the fall in pulmonary vascular resistances, the flow to the anomalous pulmonary artery increases, causing a pulmonary overflow, leading to pulmonary hypertension.[4] Pulmonary hypertension developed in the lung irrigated by the main pulmonary artery could be caused by circulating vasoconstrictive substances or crossed neurogenic factors.[6]

CLINICAL PRESENTATION

Clinical manifestations include heart failure, cardiac murmurs, shortness of breath, tachypnea and frequent respiratory tract infections. Being heart failure the most prominent manifestation, and especially worst when associated with coarctation of the aorta or interrupted aortic arch.[7] Cyanosis may be present when pulmonary hypertension and vascular resistances are elevated.[2]

Diagnostic studies

The electrocardiogram can show signs of right ventricular hypertrophy.[2] Chest X-ray usually shows cardiomegaly and pulmonary flow redistribution, being more prominent on the side of the anomalous pulmonary artery.[2,8] An echocardiogram may show a posterior vessel that grows from the ascending aorta and perfuse the lung in the parasternal and suprasternal views.[2,9] Subcostal views could be useful if the pulmonary artery is originated from the lateral portion of the ascending aorta, but the most common origin is at the posterior portion of the aorta.[2,9] The anomaly may go unnoticed in up to 15% of the cases.[7]

Cardiac catheterization is only indicated in the evaluation of pulmonary vascular resistances in patients over six months of age.[6] A CT angiography and magnetic resonance angiogram (MRA) could also be used as complement studies when there is a high clinical suspicion and also to define the surgical treatment strategy.[10] Both cardiac catheterization and imaging studies such as CT angiogram and MRA represent helpful tools in the preparation of a corrective surgery and to asses prognosis, because they can contribute with important and precise anatomic details.

Prognosis and treatment

Due to the tendency to develop congestive heart failure on an early stage and irreversible pulmonary vascular disease, the prognosis is poor, often with a fatal outcome.[2] In the case of our patient, it draws the attention the fact that he became symptomatic late in his life.

The most commonly used surgical correction procedure is a direct anastomosis of the anomalous pulmonary artery to the main pulmonary trunk. A terminus-terminal anastomosis with the use of a synthetic graft can also be done, using a homograph patch or an autologous pericardial patch with the purpose of increasing the diameter of the anomalous pulmonary artery and avoid stenosis.[2] The most common postsurgical complication is the anastomosis stenosis (10.6%), a complication that could develop even months after surgery.[11] In these particular cases, re-operation is required, being the balloon dilation or a stent angioplasty the usual techniques.[11,12]

Without an early surgical correction, it is usually fatal, although, there are reports of late stage corrections.[13] Peng, et al. report a mortality close to 0%.[14] Among late complications after surgery, they described a stenosis of the anastomosis site (12.5%) and the need of a patch growth (12.5%).[14] Prifti, et al. mention a trans-operatory mortality around 20%, and 100% reoperation-free survival on late follow up.[15]

CONCLUSIONS

The congenital anomaly of the pulmonary artery is a rare condition, generally diagnosed at an early age as it can be associated with other cardiac defects, producing a more evident clinical presentation, although if isolated, it may remain unnoticed until adolescence and become an incidental finding on imaging studies.

In the case of our patient, we found the presence of a patent ductus arteriosus at a late age associated with the presence of the anomalous origin of the right pulmonary artery. Calling our attention is the fact that he remained with only a few and vague symptoms until a late time of life for this pathology. As part of the diagnostic approach, it is important to perform an echocardiogram, where these types of defects can be identified, and even more, to describe the hemodynamic impact on all cardiac chambers. Finally, a chest CT scan can be a useful diagnostic tool, and in order to describe in detail the anatomy of the pulmonary arteries, a MRA and cardiac catheterization with a pulmonary angiogram can be performed. This former procedure apart from a diagnosis confirmation, can be helpful to define the presence of pulmonary hypertension and also for hemodynamic and vasoreactivity assessment, with the purpose of a planned corrective surgery and long term prognosis. Due to its excellent outcomes, it is mandatory to perform an early surgical correction in order to avoid the progression of pulmonary vasculature disease to an irreversible condition, where it has an ominous prognosis and surgery is usually unsuitable.

In this particular case, the right pulmonary artery was redirected to the main pulmonary trunk, using a 16 mm woven Dacron tube and also, an 8 mm atrial septal defect was created with the purpose of reducing pressure on the right side of the heart due to the previous presence of pulmonary arterial hypertension. The intervention was an absolute success and the patient remains asymptomatic to date.

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