Proximal Interruption of Isolated Right Pulmonary Artery with Ipsilateral Pulmonary Hypoplasia: Case Report

Interrupción proximal de la arteria pulmonar derecha aislada e hipoplasia pulmonar ipsilateral: Presentación de caso

Maria Carolina Pérez1
Miguel Ronedores Dumit 2
Juan Manuel Pérez1
Javier Castaño3
William Andrés Prada4

Summary
The complex congenital heart disease may present with a variety of malformations in the main pulmonary artery. However, it is not common to find in isolation the absence of one of the pulmonary arteries. In this case report, we explain the diagnostic imaging aid that led to the diagnosis of proximal interruption of the right pulmonary artery without association to other congenital malformation in a patient in whom a syndrome of anomalous venous drainage was initially thought.

Resumen
Las cardiopatías congénitas complejas se presentan con una gran variedad de malformaciones que pueden afectar el tronco de la arteria pulmonar y las arterias pulmonares. Es infrecuente encontrar de manera aislada la interrupción de una de las mismas. En este caso se explican las ayudas diagnósticas imaginológicas que llevaron al diagnóstico de interrupción proximal de la arteria pulmonar derecha sin asociación con otra malformación congénita, en una paciente a quien se le diagnosticó, inicialmente, síndrome de drenaje venoso anómalo.

Case
This is a 13-year-old female who visits the emergency department for a syncopal episode at rest, preceded by a sensation of dizziness and subsequent mild cranial trauma in the occipital region. Subsequently, the patient has two emetic episodes. As a relevant antecedent, pneumothorax refers to two days after birth. Tonsillectomy at age 6 due to recurrent episodes of tonsillitis. In the systems review, the mother reports symptoms of dyspnea on moderate physical activity and sensation of spontaneous and self-limiting palpitations, without cyanosis.

On physical examination, there are no signs of neurological deterioration. Cardiovascular examination does not identify blows or alterations in the pulse. The diagnostic impression was a syncopal episode of origin to be established. Paraclinics were performed; chest x-ray (Figure 1); electrocardiogram, with sinus rhythm without the presence of a pathological tract and glycemia, normal.

The chest x-ray with AP and lateral projection show findings suggestive of proximal interruption of the right pulmonary artery and pulmonary hypoplasia. The cardiomediatinal silhouette and bony structures of the rib cage were normal. Congenital malformations were considered: pulmonary hypoplasia, focal stenosis of the right pulmonary artery, congenital venolobar syndrome (total or partial anomalous venous drainage of the pulmonary venous flow) (1), bronchial atresia (distal to the atrophic bronchus presenting cystic degeneration) (2) and lung sequestration with cystic masses (1).
The pediatric cardiology service diagnosed anomalous venous drainage, scimitar syndrome. Cardiac catheterization was ordered (Figure 2a and b).

Cardiac catheterization demonstrates disconnection of the right pulmonary branch and severe pulmonary hypertension; there are also several rudimentary branches in the upper right lobe that originate from the intercostal arteries and neck vessels, the morphology and drainage of the pulmonary veins are normal.

Figure 1. Chest X-ray PA. In the right hemithorax, the inferior interlobar artery (arrow) is missing; there is a marked decrease in the distal vascular weave, mainly in the upper lobe (circle); the lung volume is decreased compared to the contralateral hemithorax and the cardiomiadiastinum, displaced to the right side.

Figure 2. a) Angiography of the pulmonary trunk: Absence of proximal filling of the right pulmonary artery. b) Angiography of the thoracic aorta: intercostal vessels (oval), internal mammary (vertical line) and bronchioles (arrow) perfusing the right lung.

Figure 3. Angio CT of the thorax, axial, window to the mediastinum. Absence of the right pulmonary artery (black arrow), dilation of bronchial arteries (white date). Asymmetry in the diameter of the internal mammary arteries (circles).

Figure 4. Angio CT, axial, pulmonary window. Decreased right lung volume with deviation of the anterior junction line and visceral space of the mediastinum.

Figure 5. Coronal CT angiography. Absence of the right pulmonary artery (proximal disconnection of the right pulmonary artery), hypoplasia of the right lung.
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Given these findings, an angiotomography (angio CT) of the thorax is performed (Figures 3-5), in which it is evident that the right pulmonary artery is not present and there is an important decrease in the right lung volume due to pulmonary hypoplasia. There was also an increase in the size of the right internal mammary (3.5 mm), intercostal (3.5 mm) and bronchial (2.6 mm) arteries (figure 6); without an abnormal venous drainage, with adequate concordance of the cardiac chambers.

Discussion
Proximal interruption of the right pulmonary artery is a rare congenital anomaly. Its incidence is 1 per 200,000 live births (3-5). It is more common the anomaly of the right side than of the left, but when it is of the left side, it is frequently associated with dextrocardia (2, 6).

This anomaly is usually associated with congenital cardiovascular malformations, such as tetralogy of Fallot and interauricular and interventricular communication (7, 8); however, it can also be found in isolation and clinically asymptomatic at the time of diagnosis (5, 9). In the case of this patient, the main symptom was decreased functional capacity (Figure 7).
The etiology of this condition is of embryonic origin, due to the abnormal involution of some of the central pulmonary branches. From week 8 to the end of the first trimester, from the sixth brachial arch the pulmonary arteries develop; of the left pulmonary artery develops the ductus arteriosus that communicates with the descending aorta and is fundamental in the fetal circulation (10-12). However, the trunk of the pulmonary artery originates from the truncus arteriosus by week 5 and connects with the pulmonary arteries for definitive vascular development; therefore, if the formation of the pulmonary arteries is interrupted, only the trunk of the pulmonary artery develops (12). The physiological effect of the absence of any of the pulmonary arteries is the hypoplasia of the lung on the affected side, dilation and tortuosity of the collateral arteries (bronchial arteries, mammary arteries, intercostal arteries) (10).

Pulmonary irrigation is given by the bronchial arteries. There are two left bronchial branches that emerge from the caudal aspect of the aortic arch. The right bronchial artery usually arises from the third intercostal artery (6, 12).

In the case of this patient, the irrigation of the right lung described in the angiography was done by the intercostal arteries and the right internal mammary artery. Collateral vessels of the neck, mainly of the right common carotid artery, were also identified.

The clinical manifestations in patients with this entity are mainly respiratory tract infections (37%), haemoptysis (20%), pulmonary hypertension (44%), dyspnoea and functional impairment (40%), patient (13-15).

Clinical suspicion and findings on chest radiography establish the most important initial criteria for suspecting this pathology. Despite this, it is necessary to perform extension studies and establish all the anatomical and functional conditions to define the treatment of these patients. Extension studies include angio CT and cardiac catheterization in asymptomatic patients, as was done in this case. However, when patients consult with respiratory symptomatology, differential diagnoses, such as the Swyer James syndrome, should be ruled out in addition to those initially described (16). For this, high resolution tomography offers an important diagnostic performance, since it allows to define clearly the subpleural spaces, the venous dilation, thickening of the centrilobular septum and the development of pulmonary emphysema (16, 17).

After the first reported case more than a century ago (9), many of those that have been studied are associated with another cardiovascular malformation; however, this isolated anomaly is uncommon. In this case, the patient presented association with pulmonary hypoplasia without any other anomaly.

In conclusion, discontinuation of the isolated right pulmonary artery is an uncommon diagnosis; however, it should be considered in the context of pathologies related to pulmonary hypoplasia without other associated anomalies. These patients have a poor long-term prognosis, due to the hemodynamic imbalance in the pulmonary circulation that can lead to recurrent hemoptysis, the development of cystic lung lesions and even pulmonary vascular malformations; for this reason, early diagnosis in these patients allows a therapeutic intervention that improves the quality of life in the long term (6, 7, 13).

References

Correspondence
William Andrés Prada Mancilla
Fundación Cardio Infantil
Calle 163A # 13B-60
wpradamancilla@gmail.com

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