Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Congenital Isolated Unilateral Hypoplasia of the Right Pulmonary Artery: A Rare Incidental Anomaly in an Elderly Female

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DOI: https://doi.org/10.36347/sjmcr.2024.v12i10.023 | **Received:** 26.08.2024 | **Accepted:** 01.10.2024 | **Published:** 09.10.2024

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Abstract **Case Report**

Unilateral pulmonary artery hypoplasia is a rare congenital malformation. The diagnosis is usually set at adolescence, however it can remain asymptomatic and late diagnosis is possible. Radiologically, this anomaly may be suggested on a standard chest X-ray but must be confirmed by angiography. We report the case of a young patient, 51 years old, presenting with NYHA stage II dyspnea. An angiogram was performed and revealed hypoplasia of the right pulmonary artery. The management of these patients is not yet consensual and depends on clinical evolution. However, it most often consists of regular clinical and paraclinical monitoring to detect potential complications.

Keywords: Pulmonary Artery Agenesis, Malformation, Hemoptysis, Pulmonary Hypertension.

INTRODUCTION

Congenital isolated unilateral hypoplasia of the pulmonary artery (CIUPAH) or agenesis is a rare congenital anomaly that accounts for 1% of congenital heart diseases. It is defined by the total absence of parenchymal and bronchial vascular structures in the lung. Its prevalence is approximately 1 in 200,000 patients and also represents 1% of congenital heart diseases [1].

It can be unilateral or bilateral. Unilateral involvement is often well tolerated clinically and is frequently discovered incidentally or in the context of respiratory symptoms. The diagnosis is confirmed by a thoracic CT angiography. Therapeutic management is not yet consensual and depends on the clinical progression [2].

CASE REPORT

This is a 51-year-old female with a history of rheumatic purpura, who has experienced NYHA stage II dyspnea for the past six months. Clinical examination revealed blood pressure asymmetry, a heart rate of 120 beats per minute, a respiratory rate of 22 cycles per minute, and an oxygen saturation of 97% in ambient air. Cardiac auscultation revealed a systolic murmur at the tricuspid area. An electrocardiogram was performed, showing a right atrial hypertrophy. Doppler echocardiography indicated a severely dilated right ventricle, a dilated pulmonary artery trunk, and a paradoxical septum. The chest X-ray showed an asymmetry in the size of the two lung fields, distension of the left lung, with attraction of the mediastinal structures and interstitial and bronchial syndrome (figure 1).

Given these anomalies, a CT angiogram of the chest was performed, revealing a filiform and hypoplastic appearance of the right branch of the pulmonary artery. In contrast, there was significant left systemic collateral circulation provided by the intercostal and bronchial arteries, along with dilation of the pulmonary artery trunk and its left lobar and segmental branches, as well as compensatory bilateral pulmonary veins (figure 2). Additionally, it also showed a reduction in the volume of the right lung with a shift of mediastinal structures (figure 3). A pulmonary ventilation-perfusion scintigraphy was performed (figure 4), showing a non-visualized right lung that was almost non-perfused, with no tracer uptake.

The diagnosis of hypoplasia of the right pulmonary artery was established. During subsequent follow-ups, the patient remained asymptomatic, and regular radiological monitoring was conducted.

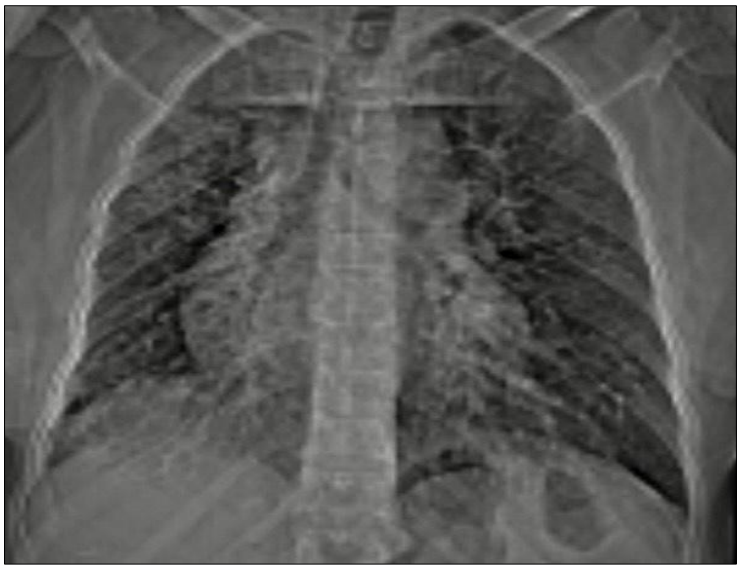

Figure 1: A chest X-ray: showed an asymmetry in the size of the two lung fields, distension of the left lung, with attraction of the mediastinal structures and interstitial and bronchial syndrome.

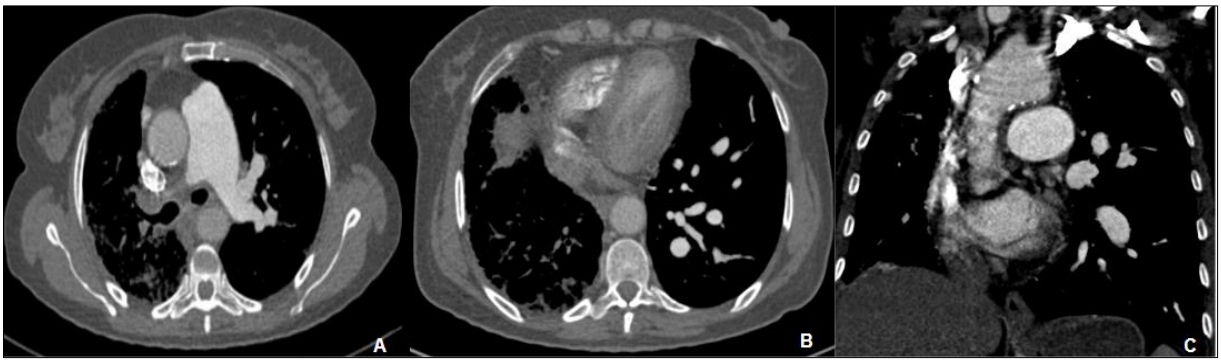

Figure 2: Chest angiogram in axial sections (a, b) and coronal (c): showing hypoplasia of the right pulmonary artery (a) and dilation of the left pulmonary veins with devascularization of the right lung field (b,c).

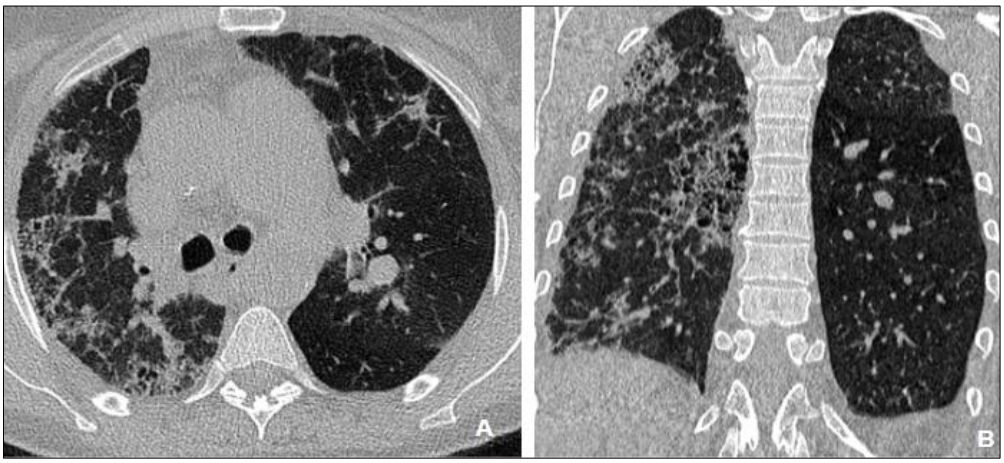

Figure 3: Axial and coronal sections (a, b) in a parenchymal window showing: a reduction in the volume of the right lung with a shift of mediastinal structures and multiple triangular bronchiectasis visible in the left upper lobe and right lower lobe.

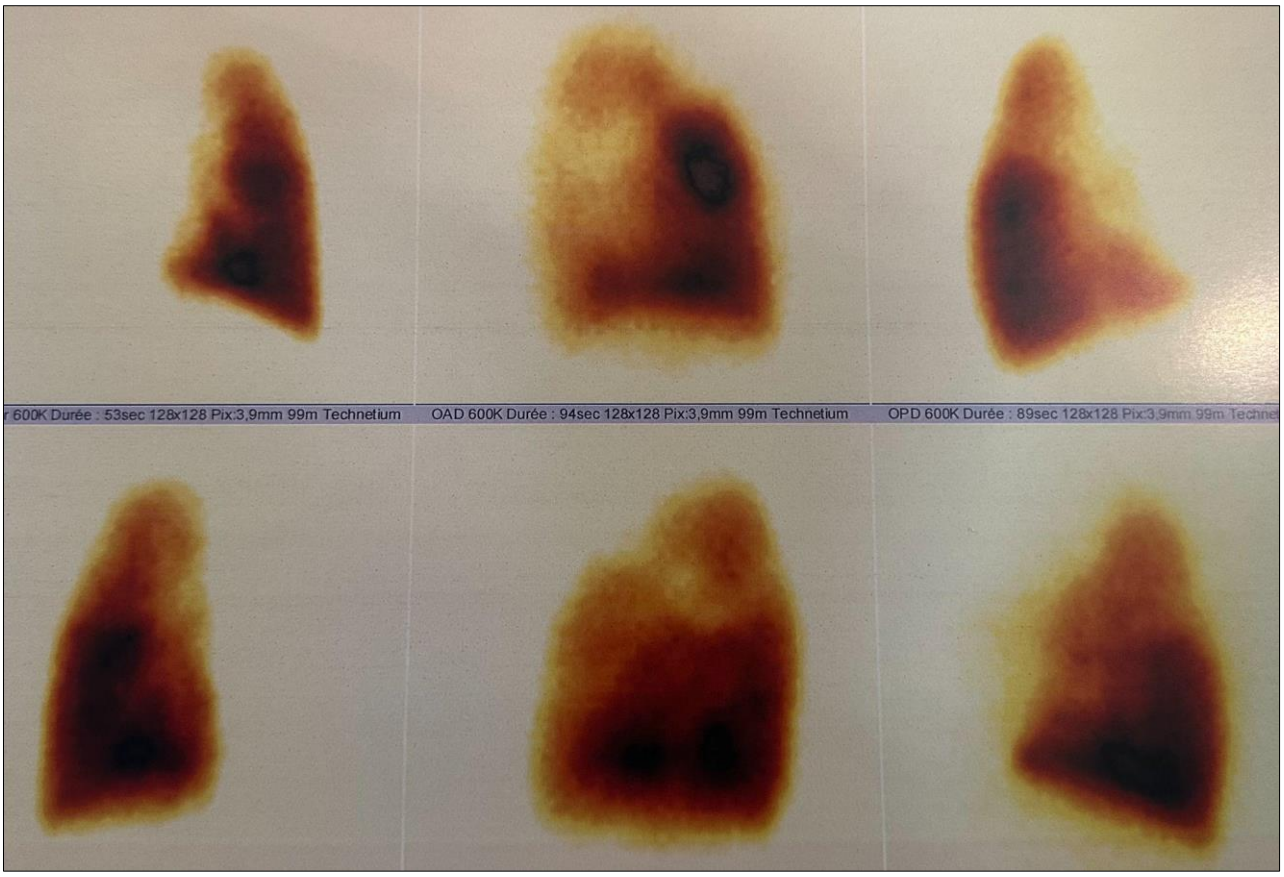

Figure 4: A ventilation pulmonary scintigraphy showing a non-visualized right lung that was almost non-perfused, with no tracer uptake.

DISCUSSION

The hypoplasia of a pulmonary artery is a rare congenital malformation, first described in 1868 by Fraentzel and visualized via angiography by Madoff *et al*., in 1952 [3, 4]. It most commonly affects the right pulmonary artery and can be isolated or associated with other congenital anomalies. If the malformation occurs on the left side, it is frequently associated with congenital heart defects such as tetralogy of Fallot, right aortic arch, or atrial septal defect. In cases of bilateral agenesis, it is considered incompatible with extrauterine life.

From an embryological perspective, it is an accident that likely occurs during the involution of the proximal branches of the sixth aortic arch, which normally determines the formation of the pulmonary arteries and the ductus arteriosus [5]. The associated ipsilateral pulmonary hypoplasia can be explained by the parallelism between vascular development and alveolar growth. The cessation of growth of the pulmonary artery leads to a deficiency in peripheral alveolar growth, resulting in diffuse, harmonious pulmonary hypoplasia.

The clinical manifestations of CIUPAH can vary widely, from asymptomatic cases to severe respiratory distress. It is usually discovered in childhood due to complications or as part of a vascular malformative syndrome such as tetralogy of Fallot, which includes pulmonary valve stenosis, aortic coarctation over the interventricular septum, a large ventricular septal defect, and right ventricular hypertrophy [6]. Other anomalies, such as a ventricular septal defect, right aortic arch, transposition of the great vessels, abnormal venous return, and persistence of the ductus arteriosus, have also been described. Adult diagnosis is especially challenging due to nonspecific symptoms, which are similar to more ominously acquired diseases. In many instances, patients may present with recurrent respiratory infections, cough, and exercise intolerance due to the compromised lung function associated with hypoplasia [7]. The severity of symptoms often correlates with the degree of hypoplasia and the presence of associated congenital heart defects. The complications include recurrent respiratory infections (37%), dyspnea or exertional discomfort (40%), hemoptysis (20%), which may be a consequence of systemic treatment, pulmonary hypertension (25%), and acute cardiogenic pulmonary edema[8-11].

Because of the nonspecific symptoms and imaging findings, it is easy to misdiagnose parenchymal change induced by unilateral pulmonary artery hypoplasia as other lung parenchymal diseases or pneumonia. Imaging studies, including chest X-rays can reveal characteristic findings such as vascular asymmetry, a small hyperlucent lung with mediastinal shift, absence of hilar shadow, and sometimes compensatory emphysema of the contralateral lung.

Based solely on standard radiology, other diagnoses may also be considered: thrombosis or embolism of a pulmonary artery, unilateral emphysema, Mac-Leod syndrome, coarctation or stenosis of a pulmonary artery, and pulmonary agenesis [9].

Diagnosis was traditionally performed through angiography. However, thoracic CT angiogram has proven to be highly effective and minimally invasive: it demonstrates the absence of vascular structures along the course of the pulmonary artery, with a smooth and regular wall between the main pulmonary artery and the remaining right or left pulmonary artery. CT angiography immediately rules out pulmonary embolism, which is the main differential diagnosis in adults [10, 11]. Additionally, it allows for the analysis of pulmonary veins, the exclusion of Felson's venolobar syndrome, and assessment of pulmonary parenchyma: complete pulmonary agenesis, aerated or non-aerated hypoplastic lung, and other complications affecting prognosis [12-15]. CT findings of chronic lung infarction may include parenchymal bands, wedge-shaped opacities or irregular peripheral linear opacities. In the study by Sakai *et al.*, [13], in eight patients with unilateral pulmonary artery agenesis, CT findings such as serrated pleural thickening were observed in six patients (75%), subpleural parenchymal bands in five (63%), and mosaic attenuation in three (38%) on the affected lung [14]. However, Magnetic resonance imaging is useful in cases of diagnostic uncertainty to differentiate between pulmonary hypoplasia and unilateral agenesis of the pulmonary artery. Echocardiography is valuable for screening associated cardiac malformations and is essential for follow-up, allowing for verification of the absence of pulmonary hypertension [15]. Pulmonary ventilation-perfusion scintigraphy is helpful for assessing the functionality of the affected lung and can guide potential treatment; it typically reveals a total absence of perfusion on the affected side, with normal or reduced ventilation [16].

Management of HRPA typically focuses on symptomatic treatment and addressing any associated complications. There is no general consensus on the treatment of unilateral pulmonary artery hypoplasia. However, there are several case reports of unilateral pulmonary artery hypoplasia with hemoptysis that were successfully treated with variable therapeutic options, such as pneumonectomy, vasodilator therapy, and embolization of collateral arteries, although the decision to operate is often complex and depends on the overall clinical picture. The long-term prognosis for individuals with HRPA varies, with some patients experiencing significant morbidity due to chronic respiratory issues, while others may lead relatively normal lives if the condition is managed appropriately [17].

CONCLUSION

In conclusion, hypoplasia of the right pulmonary artery is a rare but clinically significant condition that can lead to a spectrum of respiratory complications. Understanding its associations with other congenital anomalies, diagnostic imaging findings, and management options is crucial for optimizing patient outcomes.

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