SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Agenesis of the Right Pulmonary Artery Associated with Patent Ductus Arteriosus: A Case Report

Gourram Kenza[1*], S. Jabri[1], H. Belgadir[1], A. Merzem[1], O. Amriss[1], N. Moussali[1], N. El Benna[1]

[1]Radiology Department, University Hospital Center 20 Aout 1953, Casablanca, Morocco; Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco

DOI: 10.36347/sasjm.2023.v09i06.023 | **Received:** 27.04.2023 | **Accepted:** 22.05.2023 | **Published:** 16.06.2023

***Corresponding author:** Gourram Kenza
Radiology Department, University Hospital Center 20 Aout 1953, Casablanca, Morocco; Faculty of Medicine and Pharmacy, Hassan II University, Casablanca, Morocco

Abstract **Case Report**

***Introduction*:** Unilateral pulmonary artery agenesis is a rare congenital malformation that can be the source of serious complications. Its diagnosis must be considered in the presence of recurrent respiratory infections. Adult patients are often asymptomatic, which makes the diagnosis difficult. Chest CT angiography is the reference examination. We report a case of agenesis of the right pulmonary artery associated with patent ductus arteriosus revealed at the age of 50 years on a thoracic CT angiography. ***Observation*:** A 50-year-old female patient, treated for pulmonary tuberculosis in childhood, presented with exertional dyspnea. Thoracic CT angiography showed agenesis of the right pulmonary artery with patent ductus arteriosus and right pulmonary hypoplasia. There was significant systemic vascularity with evidence of pulmonary hypertension and right heart failure. ***Discussion*:** Unilateral pulmonary artery agenesis has been rarely reported in association with a patent ductus arteriosus. Right pulmonary artery agenesis often presents as an isolated, asymptomatic finding without associated congenital heart defects. CT angiography is used to make the diagnosis. Management is often limited to surveillance. ***Conclusion*:** Isolated pulmonary artery agenesis is a rare congenital malformation but its true frequency is not well known because of the inherent difficulties in its diagnosis. The main problem remains the risk of serious complications which requires regular follow-up.

Keywords: Agenesis of the pulmonary artery, pulmonary hypoplasia, patent ductus arteriosus, CT angiography.

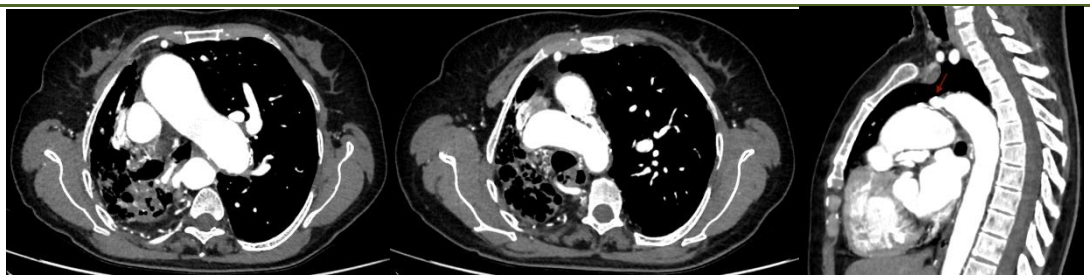
INTRODUCTION

Unilateral pulmonary artery agenesis is a rare congenital malformation. It occurs in only about 1% of congenital heart defects. It is often discovered in childhood as part of a cardiovascular malformation syndrome or following severe complications. Very few patients remain asymptomatic until adulthood. Its diagnosis is established by CT angiography. Management is not yet consensual and depends on the clinical course [1]. The overall mortality rate is 7% [2]. Because of the often delicate and therefore sometimes unrecognized diagnosis, we thought it would be interesting to report a case of agenesis of the right pulmonary artery associated with patent ductus arteriosus in a 50-year-old patient.

CLINICAL OBSERVATION

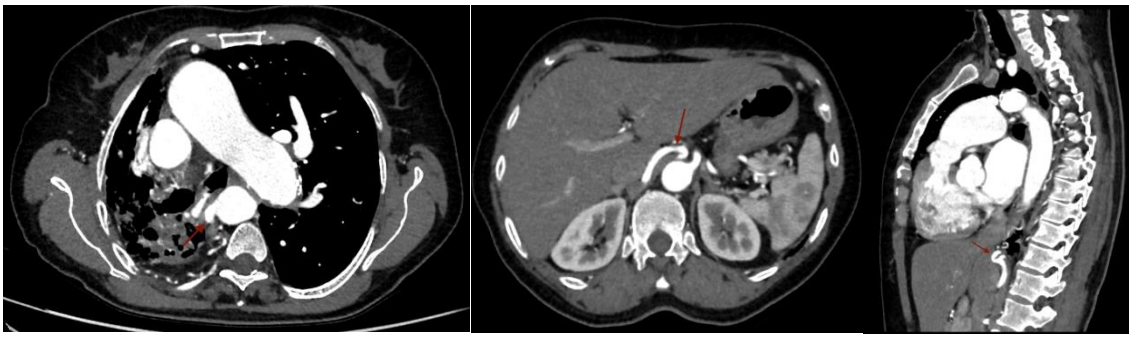
A 50-year-old female patient with a history of pulmonary tuberculosis in childhood presented with exertional dyspnea in a setting of apyrexia. The clinical examination was unremarkable. Respiratory function tests showed a mixed ventilatory disorder. A thoracic CT scan was performed, which showed dilatation of the trunk of the pulmonary artery with doubt about the absence of the right pulmonary artery.

For those, we had completed by a thoracic CT angiography. It showed an agenesis of the right pulmonary artery with patent ductus arteriosus (fig.1). On the other hand, there was significant systemic vascularization of the right lung by the right bronchial and inferior diaphragmatic arteries (Fig. 2).



A B C

Figure 1: Thoracic CT angiogram, mediastinal window in axial and coronal sections shows the absence of visualization of the right pulmonary artery, with dilatation of the pulmonary artery trunk (a). There is a patent ductus arteriosus with calcifications connecting the trunk of the pulmonary artery to the aortic arch (b, c)

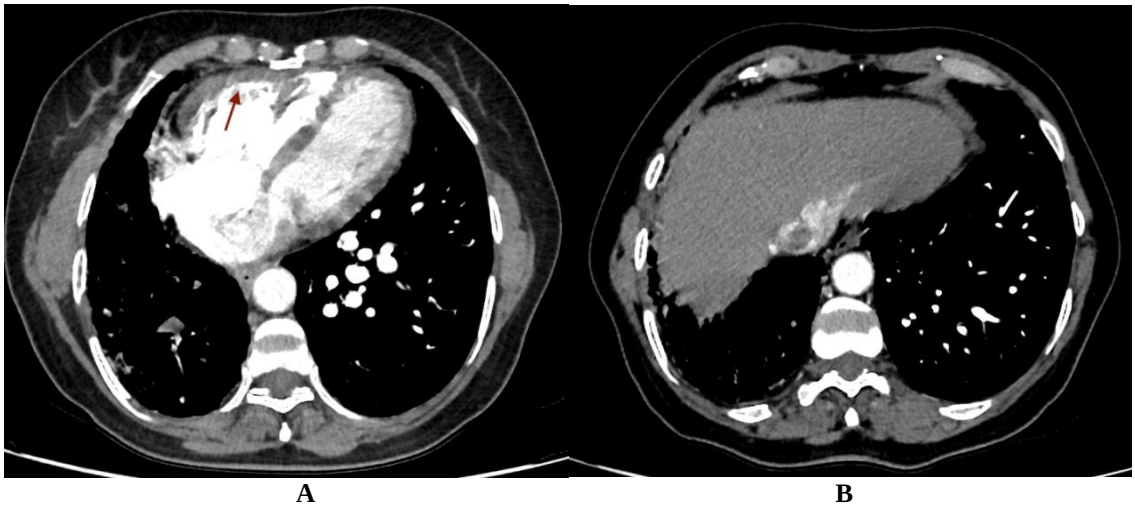


A B C

Figure 2: Thoracic CT angiogram showing systemic vascularization of the right lung, with dilated bronchial arteries emanating from the descending aorta (a), and a network originating from the right inferior diaphragmatic artery (b, c)

There were signs of pulmonary hypertension, notably a ratio of the diameter of the pulmonary artery to that of the aorta greater than 1.

CT angiography also showed signs of right heart failure including right ventricular myocardial hypertrophy, paradoxical septum, dilatation of the inferior vena cava and hepato-cava reflux (fig. 3).



A B

Figure 3: Thoracic CT angio in axial slices objectifying myocardial hypertrophy of the right ventricle with a paradoxical interventricular septum (a), as well as dilatation of the inferior vena cava with hepato-cava reflux (b)

On the parenchymal window, CT showed right pulmonary hypoplasia with deviation of the mediastinum to the homolateral side. It also showed cystic retractile bronchiectasis in the right upper lobe (fig. 4).

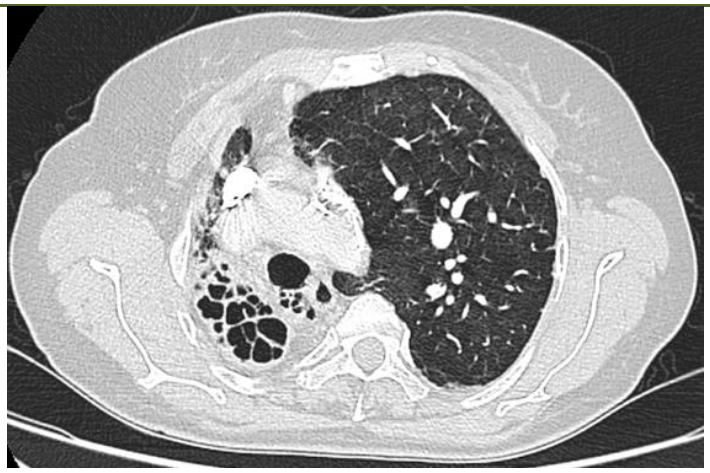


Figure 4: Thoracic CT angiogram in parenchymal window, objective hypoplasia of the right lung with dilatation of the right upper lobar cystic retractile branches with peribronchovascular thickening without mucoid impaction

The patient was treated with vasodilator-based medical therapy for pulmonary hypertension with regular clinical and echocardiographic monitoring.

During the course of the disease, the patient did not present any episode of hemoptysis.

DISCUSSION

Pulmonary artery agenesis is found more frequently on the right than on the left. Moreover, 58% of right pulmonary artery agenesis is isolated compared to only 19% of left pulmonary artery agenesis [3]. Associated cardiac malformations are tetralogy of Fallot, coarctation of the aorta, right aortic arch, transposition of the great vessels and more rarely with patent ductus arteriosus [4].

Our patient was diagnosed with asymptomatic right pulmonary artery agenesis in association with patent ductus arteriosus on the basis of a thoracic CT angio.

Embryologically, the branches of the pulmonary artery are derived from the proximal portion of the primitive sixth aortic arches. The disappearance of the proximal portion of the right or left arch results in the absence of pulmonary artery formation. Homolateral pulmonary hypoplasia is explained by the parallelism between vascular development and alveolar growth [3].

The possible complications are recurrent respiratory infections (37%), dyspnea or discomfort on exertion (40%), and hemoptysis (20%), which would be the consequence of pulmonary hypertension and systemic vascularization [1].

Pulmonary hypertension is present in 44% of patients. The systemic vascularization comes from the bronchial, subclavian, intercostal and subdiaphragmatic arteries [2].

Isolated patent ductus arteriosus is characterized by a left-to-right shunt from the aorta to the pulmonary artery. On the other hand, the patent ductus arteriosus associated with agenesis of the pulmonary artery leads to a right-left shunt by pulmonary hypertension. This causes a dilatation of the right cavities [5]. This is the case found in our patient.

The diagnosis is based primarily on CT angiography. It shows the absence of vascular structure in the path of the pulmonary artery with a smooth and regular wall between the primary pulmonary artery and the remaining right or left pulmonary artery. It eliminates pulmonary embolism which is the main differential diagnosis in adults. It allows the assessment of the pulmonary parenchyma in search of complete pulmonary agenesis, hypoplastic lung with or without ventilation. It also highlights associated congenital heart disease and complications likely to worsen the prognosis. Finally, CT angiography allows a study of the systemic vascularization, which is particularly interesting in the case of hemoptysis since it guides embolization [1].

Standard radiology can find different signs that will help to evoke the diagnosis but they are not specific. The pulmonary scintigraphy allows to evaluate the pulmonary function. It shows the absence of perfusion of a pulmonary field with persistence of normal or slightly decreased ventilation. Angiography is used to evaluate the systemic vascular network. It is performed if embolization is considered. Cardiac ultrasound is essential for follow-up. It allows to verify the absence of pulmonary arterial hypertension. Respiratory function tests are usually normal or show a discrete restrictive syndrome [2-4].

If diagnosed early, treatment is based on regeneration of the affected side or formation of a systemic-pulmonary shunt to support intra pulmonary vessel growth [5]. If the diagnosis is late, management must be adapted on a case-by- case basis depending on

the complications. Pneumonectomy is performed in case of failure of endovascular treatments of hemoptysis [4].

CONCLUSION

The diagnosis of pulmonary artery agenesis requires cross-sectional imaging based on thoracic CT angiography. It allows simultaneous analysis of the vessels and the pulmonary parenchyma. It is important to emphasize the frequent association with other embryological anomalies. The main problem remains the risk of serious complications. This imposes a regular follow-up of the patients even in the absence of any pre-existing symptomatology.

DECLARATION OF INTEREST

The authors declare that they have no ties of interest.

REFERENCES

1. Ukkola-Pons, E., Weber-Donat, G., Potet, J., Minvielle, F., Baccialone, J., & Bonardel, G. (2010). Agénésie unilatérale d'une artère pulmonaire, à propos d'un cas. *Journal de radiologie (Paris)*, *91*(6), 723-725. https://doi.org/10.1016/S0221-0363(10)70105-0.
2. Ten Harkel, A. D. J., Blom, N. A., & Ottenkamp, J. (2002). Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. *Chest*, *122*(4), 1471-1477. https://doi.org/10.1378/chest.122.4.1471.
3. Boudard, I., Mely, L., Labbé, A., Bellon, G., Chabrol, B., & Dubus, JC (2004). Isolated agenesis of the pulmonary artery. About eight observations. *Archives of Pediatrics*, *11*(9), 1078-1082. https://doi.org/10.1016/j.arcped.2004.04.026.
4. Laaraje, A., El Hafidi, N., & Mahraoui, C. (2017). Rare cause of a dry cough on exertion: agenesis of the left pulmonary artery with pulmonary hypoplasia. *The Pan African Medical Journal*, *27*, 146. https://doi.org/10.11604/pamj.2017.27.146.11399.
5. Beker-Acay, M., Ozkececi, G., Unlu, E., Hocaoglu, E., Kacar, E., & Onrat, E. (2014). Right pulmonary artery agenesis with patent ductus arteriosus and Eisenmenger syndrome: A rare case diagnosed during the postpartum period. *Heart & Lung*, *43*(4), 363-366. https://doi.org/10.1016/j.hrtlng.2014.04.009.