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Unilateral Absence of the Pulmonary Artery: A Case Report and Literature Review

S. Ouassil*, F. Jaafari, B. Boutakioute, M. Ouali Idrissi, N, Cherif Idrissi Elganouni

Department of Radiology, ARRAZI hospital, UHC Mohamed VI, Cadi Ayad University, Marrakech, Morocco

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***Corresponding author:** S. Ouassil

Abstract **Case Report**

Unilateral agenesis of the pulmonary artery is a rare congenital cardiovascular malformation (1% of congenital heart diseases)**,** it can be isolated or associated with different congenital heart diseases. We report the case of a young patient aged 18, operated on in childhood for congenital heart disease without document, admitted to resuscitation unit for heart failure. After stabilization of the patient's hemodynamic status, a thoracic CT angiography (done as part of the etiological assessment) was performed showing agenesis of the left pulmonary artery, left brachiocephalic artery and a permeable blalock-tausing shunt between the brachiocephalic artery and the trunk of the pulmonary artery which is reduced in size with direct emergence of the arch of the aorta the right subclavian artery and the right common carotid. The patient remained hemodynamically stable with 60% ambient air saturation.

Key words: unilateral agenesis, pulmonary artery, chest CT scan.

INTRODUCTION

Unilateral absence of the pulmonary artery (UAPA) is a very rare congenital cardiovascular malformation [1, 2]. It was first described by Frantzel in 1868 [3], the prevalence of isolated UAPA is estimated to range from 1 in 200,000 to 1 in 300,000 young adults, but the actual prevalence of this malformation in children is still unknown.

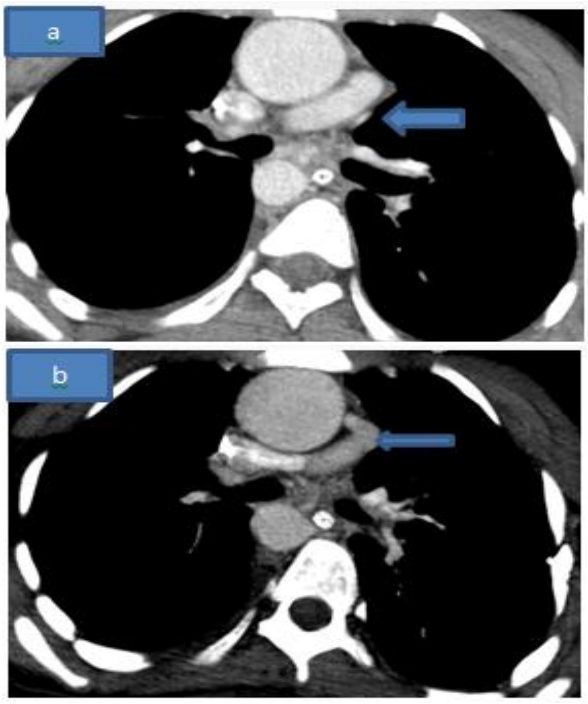
CASE REPORT

A 18 years old young patient operated in childhood for congenital heart disease without document, was admitted to the resuscitation unit for heart failure.

After stabilization of the patient's hemodynamic status, a thoracic CT angiography (done as part of the etiological assessment) was performed showing agenesis of the left pulmonary artery, left brachiocephalic artery and a permeable blalock-tausing shunt between the brachiocephalic artery and the trunk of the pulmonary artery which is reduced in size with direct emergence of the arch of the artery aorta the right subclavian artery and the right common carotid. Biological assessment revealed a discreet inflammatory syndrome.

The patient subsequently underwent angiography through catheterization of the left radial artery, which also showed the absence of the left pulmonary artery, without clearly identifying the blalock-tausing shunt.

The patient remained hemodynamically stable with 60% ambient air saturation.



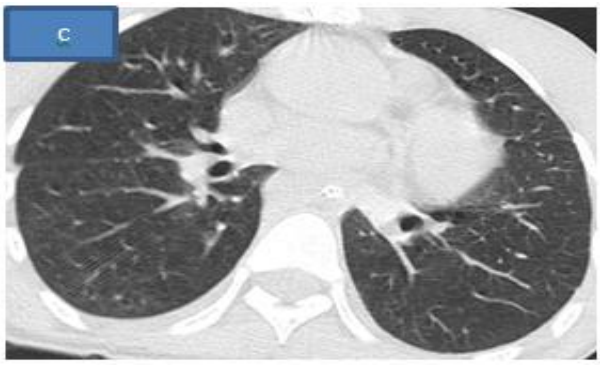


Fig-1: Chest angiography Absence of the left pulmonary artery (a) with hypoplasia of the pulmonary artery trunk (b). (C): normal appearance of lung parenchyma

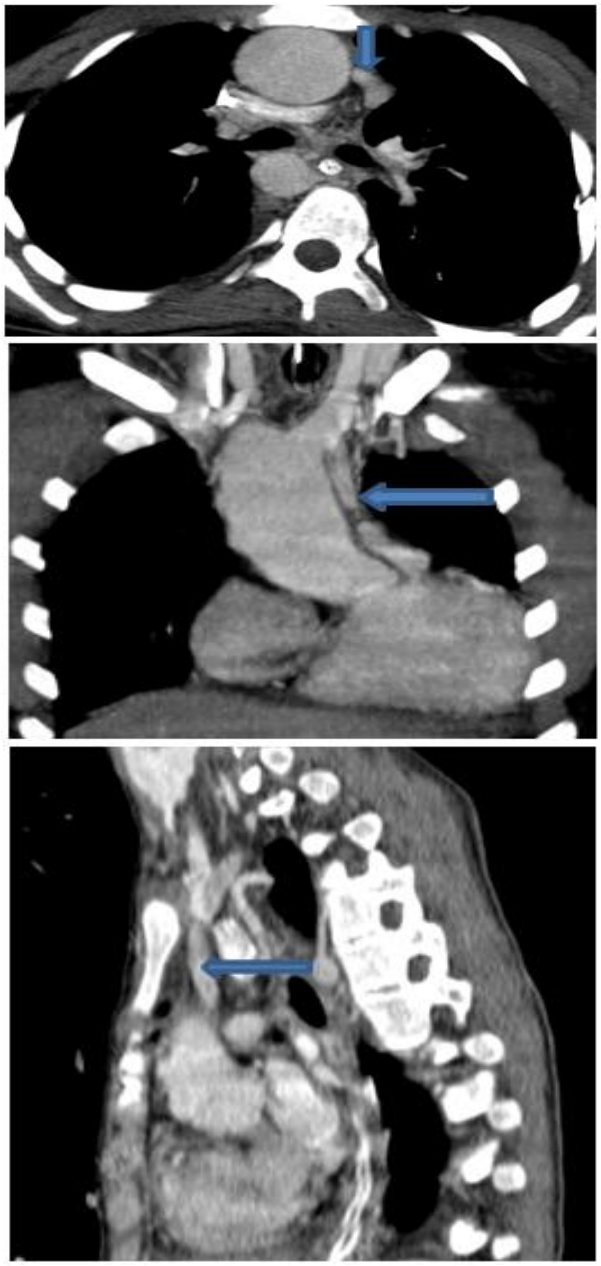


Fig-2: Permeable blalock-tausing shunts between the brachiocephalic artery and the trunk of the pulmonary artery (blue arrow): axial section (a), coronal (b) et sagittal (c)

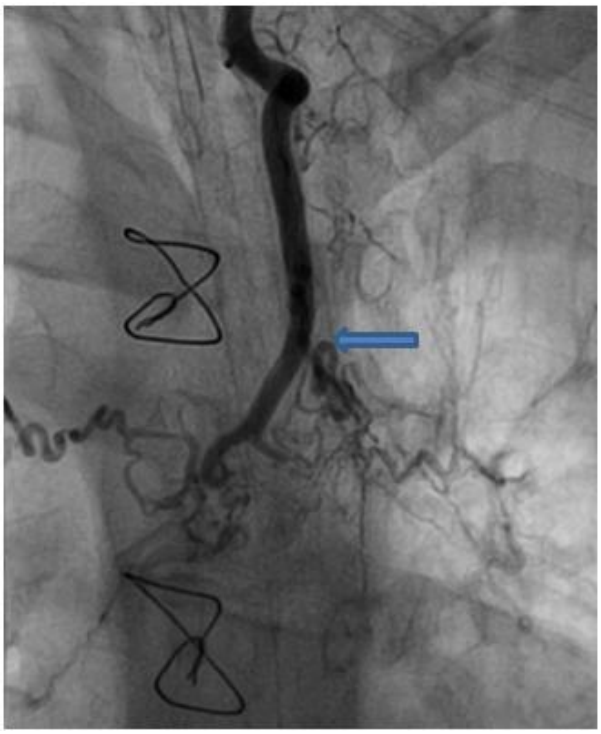


Fig-3: Angiography through catheterization of the left radial artery showing the absence of the left pulmonary artery (blue arrow) with evidence of a collateral circulation probably resulting from the right pulmonary artery and which ensures the vascularization of the left lung

DISCUSSION

Unilateral absence or the pulmonary artery (UAPA) is a very rare congenital cardiovascular malformation. It was first described by Frantzel in 1868 [3], the prevalence of isolates UAPA is estimated to range from 1 in 200,000 to 1 in 300,000 young adults, but the actual prevalence of this malformation in children is still unknown.

Unilateral absence of pulmonary artery (UAPA) is the congenital absence of the left or right pulmonary artery caused by involution of the proximal sixth aortic arch with persistence of intrapulmonary part to the distal sixth aortic arch. As a result, "proximal interruption of pulmonary artery" has been suggested as a preferred term [4, 5]. It's often associated with other cardiac anomalies; Bockeria et al. reviewed 352 cases of UAPA reported in literature, of whom 237 (67%) were associated with other congenital heart defects [6]. Among the cardiac malformations possibly associated with UAPA, tetralogy of Fallot, atrial septal defect, coarctation of the aorta, right aortic arch, truncus arteriosus and pulmonary atresia are the most frequent ones [7]. However it may seldom occur as an isolated lesion, which mostly concerns the right pulmonary artery [8, 9]. Patients with isolated UAPA are usually asymptomatic at birth; thereafter they may develop a progression of symptoms such as exercise intolerance,

dyspnea, chest pain, hemoptysis and recurrent pulmonary infections [10].

In a retrospective review of 108 patients reported between 1978 and 2000, 14 patients (median age 14 years; range 0.1-58 years) were asymptomatic.

Diagnosis of UAPA can be very difficult and should be considered on the basis of medical history, physical examination and imaging studies, including echocardiogram, chest roentgenograms, CT scans perfusion scintigraphy and angiography [11, 12].

The typical findings on CT scans or roentgenograms may include elevation of the ipsilateral hemidiaphragm, a diminished hemithorax, a mosaic attenuation pattern, and displacement of the mediastinum, as well as interruption in the pulmonary artery and loss of normal pulmonary vascular markings [11, 13, 14]. However, it should be noted that the affected lung is supplied by the bronchial, diaphragmatic, intercostal, or aortopulmonary collateral arteries that may be visualized on angiogram or echocardiogram [15].

If the diagnosis of UAPA is suspected on an abnormal chest radiograph, the authors would advocate the use of transthoracic echocardiography and cross-sectional imaging as second – investigation. Transthoracic echocardiography is non-invasive test that is widely available and reproducible [1], but it is more difficult in adults, and so transoesophageal echocardiography could be considered. However given the more invasive nature of this test, cross-sectional imaging is preferred [1].

Contrast enhanced CT and MRI studies [16] are both helpful in establishing the diagnosis. The UAPA is demonstrated along with the presence of collateral arteries. Intracardiac and great vessel anomalies can be identified. Both techniques offer a fast and non-invasive approach, and can be used to diagnose and evaluate pulmonary hypertension [17]. The advantage of combined three-dimensional cine gradient echo MRI and MR angiography (using gadolinium as a contrast agent) is that there is no irradiation. It has also been shown to have excellent correlation with findings on cardiac catheterization, echocardiogram and surgery [16].

CONCLUSION

UAPA is a rare malformation that requires an early diagnosis in order to avoid serious complications, namely hemoptysis and PAH, the thoracic angiography CT makes the diagnosis and allows a simultaneous study of the vessels and the pulmonary parenchyma.

REFERENCES

1. Griffin N, Mansfield L, Redmond KC, Dusmet M, Goldstraw P, Mittal TK, Padley S. Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: a review of four cases. Clinical radiology. 2007 Mar 1;62(3):238-44.
2. Kastler B, Clair C, Livoisi A, Narboux Y, Delabrousse E, Sarlième P. Malformation de l'artère pulmonaire. Cardiopathies cyanogènes: aspect IRM. Encycl Méd Chir, Radiodiagnostic-Cœur-poumon. 2002; 32-015-F-70.
3. Frentzel O. Ein fall von anormer communication der aorta mit der arteria Pulmonalis. Virchow's Arch Path Anat. 1868;43:420–2.
4. Kruzliak P, Syamasundar RP, Novak M, Pechanova O, Kovacova G. Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. Arch Cardiovasc Dis. 2013;106(8–9):448–454.
5. Steiropoulos P, Archontogeorgis K, Tzouvelekis A, Ntolios P, Chatzistefanou A, Bouros D. Unilateral pulmonary artery agenesis: a case series. Hippokratia. 2013;17(1):73–76.
6. ockeria LA, Makhachev OA, TKh K. Congenital isolated unilateral absence of pulmonary artery and variants of collateral blood supply of the ipsilateral lung. Interact Cardiovasc Thorac Surg. 2011;12:509–10
7. Shakibi AG, Rastan H, Nazarian I. Isolated unilateral absence of the pulmonary artery. Review of the world literature and guidelines for surgical repair. Jpn Heart J. 1978;19:439–51
8. Welch K, Hanley F, Johnston T. Isolated unilateral absence of right proximal pulmonary artery: surgical repair and follow up. Ann Thorac Surg. 2005;79:1399–402
9. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. Chest. 1995;108:670-6.
10. Raymond A, Pedretti E, Privitera G, Cicero C, Biasucci G. Neonatal diagnosis of isolated absence of the right pulmonary artery: a case report and review of the literature. Italian journal of pediatrics. 2018 Dec;44(1):1-4.
11. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest. 2002;122:1471-7.
12. Zhang LZ, Ma WG, Gao SG, He J. Unilateral absence of pulmonary artery associated with contralateral lung cancer. Journal of thoracic disease. 2016 Sep;8(9):E942.
13. Sakai S, Murayama S, Soeda H, Furuya A, Ono M, Ro T, Akamine T, Masuda K. Unilateral proximal interruption of the pulmonary artery in adults: CT findings in eight patients. Journal of computer assisted tomography. 2002 Sep 1;26(5):777-83.
14. Camera L, Fusari M, Calabrese M, Sirignano C, Catalano L, Santoro A, Galderisi M, Salvatore M. Isolated unilateral absence of pulmonary artery mimicking chronic pulmonary embolism at chest X-ray: multidetector-row CT angiographic

findings. Clinical imaging. 2012 Nov 1;36(6):845-9.

15. Welch K, Hanley F, Johnston T, Cailes C, Shah MJ. Isolated unilateral absence of right proximal pulmonary artery: surgical repair and follow-up. The Annals of thoracic surgery. 2005 Apr 1;79(4):1399-402.
16. Prasad S, Soukias N, Hornug T. Role of the magnetic resonance angiography in the diagnosis of major aorto-pulmonary collateral arteries and partial anomalous venous drainage. Circulation. 2004; 109: 207-14.
17. Di Guglielmo L, Dore R, Vespro V. Pulmonary hypertension: role of the computed tomography and magnetic resonance. Ital heart J. 2005; 6: 846-51.