Case Report

Access this article online
Quick Response Code:

Website: www.eurasianjpulmonol.com
DOI: 10.4103/ejop.ejop_62_19

Unilateral absence of pulmonary artery: A rare cause of dyspnea in a 77-year-old woman

RK Chopra, Mohanakrishnan Jayamani, Ganesh Pratap

ORCID:
RK Chopra: https://orchid.org/0000-0001-6240-528X
Mohanakrishnan Jayamani: https://orchid.org/0000-0001-6975-7587
Ganesh Pratap: https://orcid.org/0000-0001-9364-665X

Abstract:

Isolated unilateral absence of pulmonary artery is quite rare and usually associated with congenital cardiac defects. It is usually diagnosed in childhood with recurrent respiratory tract infections and signs of pulmonary hypertension. It rarely remains asymptomatic until adulthood and may present with dyspnea, hemoptysis, and recurrent respiratory infections later, while it can rarely be an incidental finding of other imaging. Surgical management in early days of life will prevent pulmonary hypertension while in adults it is aimed to treat the complications. Here, we report the case of a 77-year-old female, a mother of five children. She came with worsening of dyspnea for a week, and her computed tomography pulmonary angiography showed absence of left pulmonary artery accompanying hypoplastic lung. Ventilation perfusion imaging showed absence of perfusion of the left lung. She had severe pulmonary hypertension and was managed with long-term oxygen therapy, diuretics, and vasodilators.

Keywords:

Absent pulmonary artery, dyspnea, hypoplastic lung, pulmonary hypertension

Introduction

Unilateral absence of pulmonary artery (UAPA) is a very rare congenital entity with the incidence of 1 in 200,000 people.[1] The first case was reported by Fraentzal in 1868 and up to now, 400 cases have been reported in literature.[2,3] UAPA of the right lung is seen in 65% of cases, whereas that of left side is seen 35% of the reported cases.[4] It is usually associated with cardiac anomalies such as tetralogy of Fallot, septal defects, and transposition of great arteries.[5] Absence of pulmonary artery is thought to be the involution of the sixth branchial arch. The average age of clinical presentation was reported as 14 years of age.[4] We reported the case of a 77-year-old female with absence of the left pulmonary artery without any cardiac defects. As far we know, our report is the oldest case of UAPA.

Case Report

A 77-year-old woman, a mother of five with no comorbidities, came to our hospital with worsening dyspnea for 5 days. She never had any prior hospitalization in medical history. On physical examination, she had tachycardia, tachypnea, and hypotension. Her arterial oxygen saturation was 78% and jugular venous pressure was high, and she had bilateral pretibial edema. Pulmonary examination revealed the signs of volume loss of the left hemithorax together with dull note on percussion and

Department of Chest Medicine, Ruby Hall Clinic, Pune, Maharashtra, India

Address for correspondence:
Dr. Mohanakrishnan Jayamani, Department of Chest Medicine, Ruby Hall Clinic, 40, Sasoon Rd, Sangamvadi, Pune - 411 001, Maharashtra, India.
E-mail: drmkpulmo@gmail.com

Received: 18-07-2019
Revised: 07-09-2019
Accepted: 24-12-2019
Published: 30-04-2020

How to cite this article: Chopra RK, Jayamani M, Pratap G. Unilateral absence of pulmonary artery: A rare cause of dyspnea in a 77-year-old woman. Eurasian J Pulmonol 2020;22:64-6.

stony dull note over the right lower hemithorax. Breath sounds reduced on the left and fine crackles were heard over the right with loud P2. Arterial blood gas was consistent with Type 1 respiratory failure, and chest X-ray revealed shift of mediastinum to the left side and left lung hypoplasia [Figure 1]. Two-dimensional (2D) transthoracic echocardiography showed that pulmonary artery pressure was 92 mmHg with grossly dilated right heart chambers and left ventricular ejection fraction of 50%. With a suspicion of massive pulmonary embolism, the patient underwent computed tomography pulmonary angiography (CTPA) and Doppler ultrasonography of the lower limbs. Doppler was normal; however, CTPA showed absence of left pulmonary artery without any evidence of pulmonary thromboembolism. There was a gross mediastinal shift to the left, hypoplasia of the left lung, patchy infiltrates in the right upper lobe, and bilateral pleural effusion with right basal atelectasis [Figure 2]. During the course of hospitalization, the patient had desaturation, and she underwent intubation and needed mechanical ventilation support. Pleural effusion was consistent with transudate and attributed to right heart failure, and post diuretic therapy, there was reduction in the pleural fluid volume. No organisms were isolated in the sputum and body fluids and here, infective markers were within normal limits. Ventilation perfusion scintigraphy showed the absence of perfusion on the left lung [Figure 3]. The patient was taken to the ward and then, she was discharged with long-term oxygen therapy, vasodilators, and prophylactic oral anticoagulants. The patient's follow-up continues in the outpatient clinic.

Discussion

Congenital UAPA is a very rare anomaly with the incidence of 1 in 200,000 population.[1] The first case was reported by Fraentzel in 1868.[2] Nearly 400 cases had been reported in literature so far.[3] UAPA of right lung seems to be 65%, whereas that of left lung seems to be involved in 35% of the reported cases.[4] In our patient, left pulmonary artery was absent.

The most common clinical presentation of UAPA is dyspnea (40%) followed by recurrent respiratory tract infections (37%) due to bronchiectasis, and hemoptysis (20%) because of systemic collaterals from bronchial, intercostal, subclavian, subdiaphragmatic, and rarely coronary vessels. Rarely, it can be recognized as an incidental finding of asymptomatic person.[4,6,7] In our case, the patient presented with dyspnea as her major symptom. Only very few cases had been reported and as far we know, our report was of the oldest female in literature.

Chest X-ray of patients with UAPA shows hypoplasia of lung with mediastinal shift similar to our case, and others findings can be ipsilateral elevation of diaphragm, absence of hilar shadow, and contralateral compensatory hyperinflation of the hemithorax.[3,8]

CT thorax and also CTPA help the diagnosis as similar to our case, and they also identify parenchymal abnormalities. Ventilation perfusion scintigraphy shows the absence of perfusion in the affected side as in our patient.[9] Pulmonary

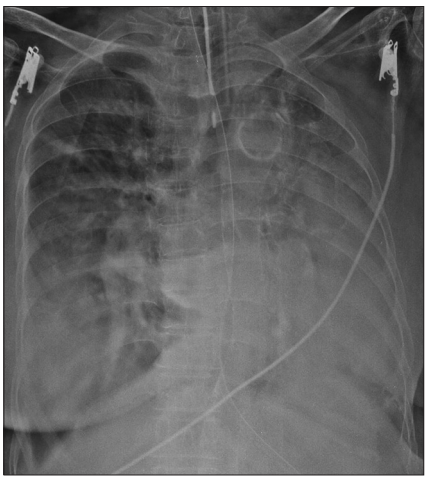

Figure 1: Chest X-ray showing mediastinal shift toward the left side and left lung hypoplasia and prominent right pulmonary artery with opacities in the right mid and lower zone

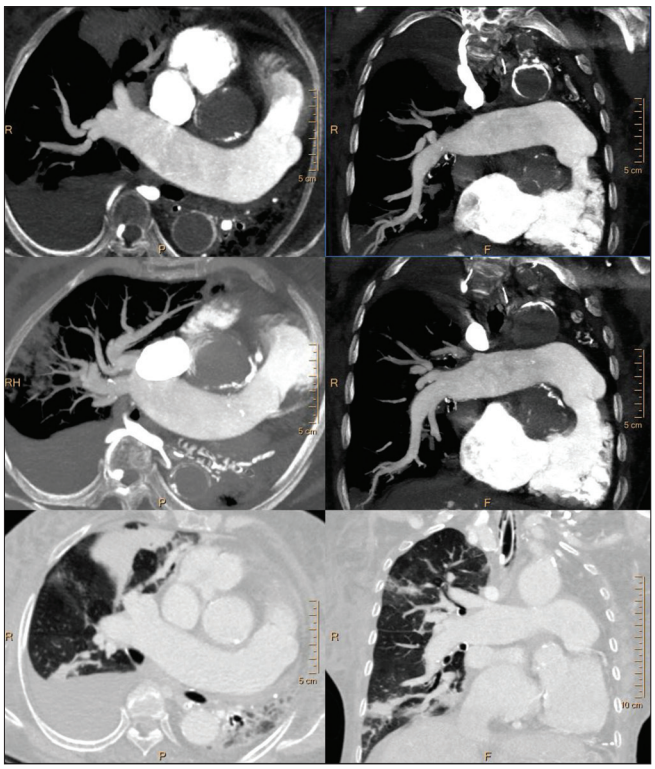

Figure 2: CTPA showed absence of left pulmonary artery without any evidence of pulmonary thromboembolism. There was a gross mediastinal shift to the left, hypoplasia of the left lung, patchy infiltrates in the right upper lobe, and bilateral pleural effusion with right basal atelectasis

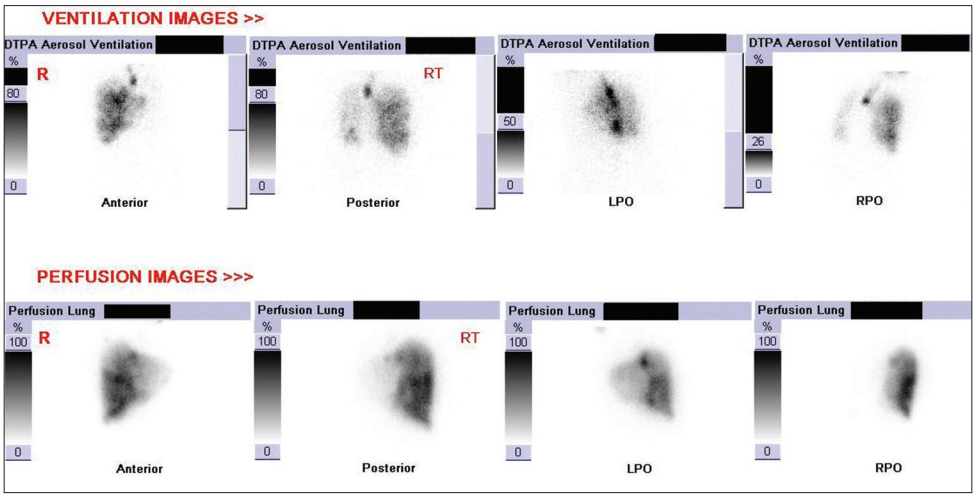

Figure 3: Ventilation perfusion scintigraphy showed the absence of perfusion on the left lung

angiography is the gold standard of diagnosis of UAPA, and it also identifies collateral circulation.[8]

2D transthoracic echocardiography helps the identification of cardiac defects which are usually seen together with UAPA. In our case, there were no cardiac defects, and the patient had severe pulmonary hypertension.

Medical management of UAPA cases includes treatment of pulmonary hypertension with long-term oxygen therapy, calcium channel blockers, and vasodilators.

Surgical options include creation of aortopulmonary shunt or restoration of continuity between main and hilar pulmonary arteries when diagnosed at an early age.[10] Pneumonectomy or lobectomy in the case of massive hemoptysis and recurrent respiratory infections should be needed. Selective arterial embolization can be considered in some patients who are not fit for surgery. The mortality rate is about 7% in patients with UAPA.

Conclusion

Thus, UAPA can have various spectra of symptoms and can also rarely be found in older patients. It is rarely thought in the differential diagnosis in patients presenting with dyspnea. Imaging techniques can help the diagnosis and various medical and surgical management options are available for this condition depending on the age of patient. In this case report, we tried to emphasize the importance of this very rare congenital anomaly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. Chest 1995;108:670-6.
2. Fraentzel O. An case of abnormal communication with the aorta the pulmonary artery.Virchow's Arch Pathol Anat 1868;43:420.
3. Bockeria LA, Makhachev OA, Khiriev TK, Abramyan MA. 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.
4. 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.
5. Pfefferkorn JR, Löser H, Pech G, Toussaint R, Hilgenberg F. Absent pulmonary artery. A hint to its embryogenesis. Pediatr Cardiol 1982;3:283-6.
6. Aypak C, Yıkılkan H, Uysal Z, Görpelioğlu S. Unilateral absence of the pulmonary artery incidentally found in adulthood. Case Rep Med 2012;2012:942074.
7. Mohan V, Mohan B, Tandon R, Kumbkarni S, Chhabra ST, Aslam N, *et al*. Case report of isolated congenital absence of right pulmonary artery with collaterals from coronary circulation. Indian Heart J 2014;66:220-2.
8. Griffin N, Mansfield L, Redmond KC, Dusmet M, Goldstraw P, Mittal TK, *et al*. Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: A review of four cases. Clin Radiol 2007;62:238-44.
9. Gluck MC, Moser KM. Pulmonary artery agenesis diagnosis with ventilation and perfusion scintiphotography. Circulation 1970;41:859-67.
10. Toews WH, Pappas G. Surgical management of absent right pulmonary artery with associated pulmonary hypertension. Chest 1983;84:497-9.