

Isolated Congenital Unilateral Agenesis of the Left Pulmonary Artery with Left Lung Hypoplasia in an Asymptomatic Adult Patient

Sadık Volkan Emren,¹ Selcen Yakar Tülüce² and Kamil Tülüce³

The congenital unilateral agenesis of pulmonary artery is a congenital cardiovascular anomaly rarely seen in adulthood. A 21-year-old asymptomatic male was admitted to our hospital to obtain a routine health report to accompany a job application. Posteroanterior chest radiograph revealed a mediastinal shift to the left, with increased radiopacity in the left lung and increased radiolucency in the right lung. Thoracoabdominal computed tomography revealed hypoplasia of the left pulmonary artery. Transthoracic echocardiography excluded any accompanying cardiac abnormalities. Pulmonary angiography was undertaken and confirmed diffuse hypoplasia of the left pulmonary artery while right pulmonary artery was significantly enlarged. The patient's pulmonary artery pressure was within the normal limits, after which he decided to be carefully followed-up.

Key Words: Cardiovascular anomaly • Hypoplasia • Pulmonary artery

INTRODUCTION

Congenital unilateral agenesis, unilateral absence of the pulmonary artery (UAPA) and hypoplasia of a pulmonary artery (UHPA) are rare congenital anomalies.¹ Agenesis might occur either in the left or right pulmonary artery (PA). Unilateral agenesis of pulmonary artery is most frequently accompanied by congenital cardiovascular anomalies; less frequently, UHPA may also occur as an isolated finding. In the great majority of the cases in the literature, patients were diagnosed in infancy or childhood. In fact, only a limited number of patients appear to remain asymptomatic until adulthood.

In this paper, we report a 21-year-old male with a

left pulmonary artery agenesis and left pulmonary hypoplasia diagnosed in adulthood which was not accompanied by other congenital cardiovascular abnormalities.

CASE REPORT

A 21-year-old male patient was admitted to our hospital to obtain a health report for a job application. He was asymptomatic until one year prior to admission when he suffered sudden shortness of breath and chest pain which were resolved spontaneously without the need to be admitted to a hospital. The patient had no history of any significant diseases, and his family lacked any prior history of congenital cardiovascular disease. On physical examination, his pulse rate was 70 beats/min, respiration rate was 16 breaths/min, and blood pressure was 110/70 mmHg in his arms bilaterally. A grade 3/6 systolic murmur was heard at the second left intercostal rib, and it was difficult to hear breathing sounds in the left lung area. Otherwise, his biochemistry parameters were normal. Arterial blood gas analyses revealed pH: 7.35; PO₂: 92 mmHg; PCO₂: 36 mmHg. Electrocardiographic testing revealed sinus rhythm with 70

Received: September 24, 2014 Accepted: May 11, 2015

¹Department of Cardiology, Afyonkarahisar State Hospital; ²Izmir Katip Celebi University Atatürk Research and Department of Cardiology, Training Hospital; ³Department of Cardiology, Izmir Karsiyaka State Hospital, Turkey.

Address correspondence and reprint requests to: Dr. Sadık Volkan Emren, Department of Cardiology, Afyonkarahisar State Hospital, Afyonkarahisar 03500, Turkey. Tel: +90 05052644578; E-mail: vemren@hotmail.com

beats per minute with an incomplete right bundle branch block. No other typical right heart overload signs were detected, such as pulmonary P waves or increase of R wave in V1, V2. A posteroanterior chest radiograph revealed a mediastinal shift to the left, increased radiopacity in the left lung, and increased radiolucency in the right lung (Figure 1A). Initially, we performed diagnostic examinations to rule out malignancy or tuberculosis that might cause atelectasis of the left lung. Laboratory analyses and culture tests for infection were all negative. Bronchoscopic examination also revealed no endobronchial lesion. However, it was observed that the carina was displaced to the left, and concentric narrowing of the left main bronchus and the left upper lobe bronchus were noted. No evidence of malignancy was detected in cytological preparations, which had been collected during the bronchoscopic procedure. Thoracoabdominal computed tomography (CT) showed agenesis of the left PA (Figure 1B) with hypoplasia of the parenchyma of the left lung, and atelectasis in the upper lobe of the left lung. Thereafter, a nuclear ventilation/perfusion scan showed a complete absence of uptake in perfusion scans of the left lung; ventilatory scans were diminished in the left lung area beside a part of the left upper lobe where no ventilation was observed. To exclude congenital cardiac anomalies, the patient was evaluated with transthoracic echocardiography (TTE). However, no additional congenital cardiac anomalies besides UAPA were detected (Figure 1C). Soon thereafter, the patient underwent right heart catheterization. The mean PA pressure was 20 mmHg; the mean right PA pressure was 25 mmHg; and the mean left PA pressure was 17 mmHg. Pulmonary angiography confirmed the agenesis of the left PA and displayed significant enlargement of the right PA (Figure 1D). The patient was evaluated by the council of cardiology and cardiovascular surgery, ultimately choosing a non-invasive course of treatment of careful follow-up because he had no current symptoms. His clinical status has remained stable since that time, and he was followed-up in our outpatient clinics for one year.

DISCUSSION

Congenital agenesis of PA or hypoplasia is a rare

anomaly. It has a customary prevalence in the general population of 1/200,000. The first report of UAPA was published in 1868. Unilateral hypoplasia of a PA is caused by a malformation of the sixth aortic arch of the affected side during embryogenesis. While proximal interruption of pulmonary arteries was seen in UAPA, these remained rudimentary in UHPA.

Unilateral hypoplasia of a PA occurs twice as frequently as compared to the right side. Although UHPA and UAPA are less frequently seen on the left side, they can be accompanied by other congenital heart abnormalities such as tetralogy of Fallot, atrial septal defect, coarctation of the aorta, right aortic arch, truncus arteriosus, patent ductus arteriosus, and pulmonary atresia.² Interestingly, this patient had a left-sided pulmonary artery agenesis without the accompanying congenital cardiac anomalies which could contribute to the maintenance of an asymptomatic state.

The most common symptoms in PA agenesis are re-

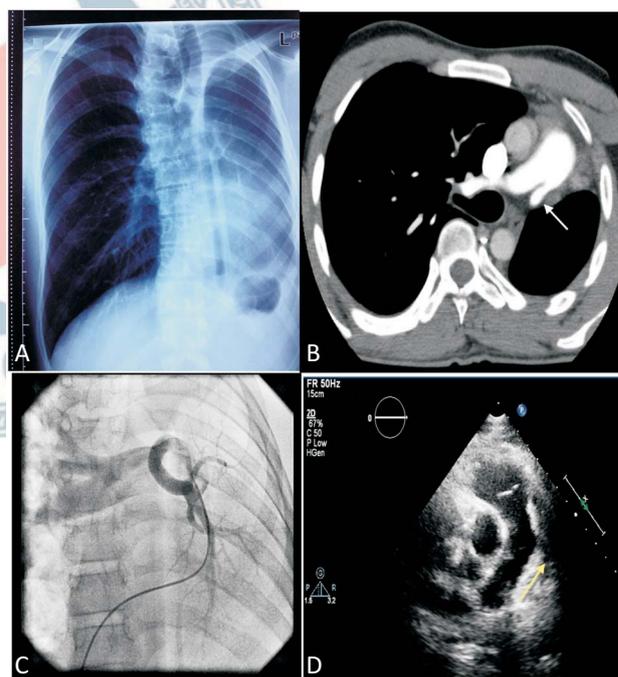


Figure 1. (A) Posteroanterior chest radiograph showing mediastinal shift to the left, increased radiopacity in the left lung, and increased radiolucency in the right lung; (B) Computed tomography angiography of the patient (Arrowhead – Left pulmonary artery hypoplasia); (C) Pulmonary angiography showing diffuse hypoplasia of the left pulmonary artery and significant enlargement of the right pulmonary artery; (d) Transthoracic echocardiography in a short axis view (Arrowhead – Rudimentary left pulmonary artery).

current pulmonary infections, exercise intolerance, and mild dyspnea during exertion; hemoptysis, however, is rarely seen. Due to the absence of disease specific symptoms, some patients may be belatedly diagnosed almost 30 years after disease onset.¹ This patient suffered from shortness of breath only once during the previous year.

The proper diagnosis of UAPA or UHPA can be a challenging undertaking, and an elevated level of suspicion often helps to facilitate a correct diagnosis. In general, a complete medical history, physical examination, and imaging examinations are required. Physical examination may reveal an asymmetrical chest with abnormal breath sounds on the affected side.³ Additionally, there may be systolic ejection murmur across the pulmonary outflow tract. In our case, this patient was asymptomatic; however, his physical examination revealed decreased breath sounds in the left lung area and a systolic ejection murmur at the left second intercostal space during auscultation. We concluded that these findings warranted further investigation.

The electrocardiogram is usually normal in patients with uncomplicated isolated absence or hypoplastic PA (without pulmonary hypertension), whereas it presents right ventricular dominance in cases associated with pulmonary hypertension (PHT). The chest radiograph of patients with UHPA typically show asymmetric lung fields with an ipsilateral small hemithorax holding a hyperlucent lung. The mediastinum shifts towards the affected side and the hilar vasculature on that side is greatly diminished. When suspect findings are noted on a chest radiograph, the diagnosis of UAPA can be made definitively by CT, magnetic resonance imaging (MRI) or TTE. In this case, we further performed bronchoscopy in order to rule out the possibility of pathologies that might be the cause of unilateral atelectasis, such as a malignancy or an infection.

Transthoracic echocardiography was also performed in this patient to establish the diagnosis, to exclude any other cardiac or major vessel abnormalities, and to evaluate the presence or development of PHT. Thereafter, pulmonary angiogram and right heart catheterization were performed to confirm the diagnosis, to evaluate the entire anatomy of the pulmonary arteries and to make hemo-dynamic calculations; some authors suggest performing these invasive procedures in cases where-in

embolization is indicated for massive hemoptysis.⁴

Among the most important differential diagnoses of congenital agenesis of the left PA is left pulmonary hypoplasia. Similar clinical findings and radiographic changes may also be observed in simple pulmonary hypoplasia with a markedly hypoplastic PA. Pulmonary hypoplasia is also rarely seen (1-2/12,000) in adulthood.⁵ The majority of cases exhibit other congenital abnormalities as well.⁶ The value of ventilation-perfusion scintigraphy is conflicting in the case of differential diagnosis. In cases with a good cross-sectional imaging, it appears unnecessary to perform a ventilation-perfusion study to confirm UPAA.⁷ Generally, in patients with UPAA, diminished or total absence of ipsilateral perfusion is demonstrable by perfusion scintigraphy, since ventilation scan of these patients are typically normal or rarely diminished in patients with agenesis of PA.⁸ In patients with pulmonary hypoplasia, ventilation-perfusion scan of the chest most often shows complete absence or diminished ventilation and perfusion in the affected lobe and segment of the lung.⁹ In this case, ventilation was diminished and perfusion was absent in the left lung, favoring a diagnosis of a UAPA with a left pulmonary hypoplasia. The cause of lung hypoplasia in this case is secondary hypoplasia attributed to the pulmonary artery agenesis. This view is supported by the report indicating that reduction in pulmonary blood flow prevents lung development and causes lung hypoplasia.¹⁰

No treatment is required in patients who lack any manifestation of cardiopulmonary dysfunction. However, as a cautionary measure, these patients should be followed-up on a regular basis. Treatment is warranted for a small number of patients with hemoptysis, recurrent lower respiratory tract infection or PHT. Accordingly, since this patient has remained asymptomatic, and his systolic PA pressure has remained within normal limits, it was decided to follow up subsequent to discharge. He has been examined in our outpatient clinic regularly for one year, and a TTE is performed to calculate systolic pulmonary arterial pressure.

Left PA agenesis or hypoplasia without accompanying cardiovascular abnormalities is a rarely occurring anomaly, especially in adulthood, which can complicate the process of making an accurate diagnosis. However, it should be included in the differential diagnosis of a chest radiograph with a contracted hyperlucent hemi-

thorax. Physicians must be properly cautious regarding the possibility of an undiagnosed UHPA in adults. By presenting this case, we intended to remind physicians about the often differential signs, findings, and imaging modalities that need to be carefully reviewed in the correct diagnosis of UAPA with left pulmonary hypoplasia.

REFERENCES

1. 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.
2. Vergauwen S, Bracke P, De Schepper A. Unilateral absence of a pulmonary artery. *J Belge Radiol* 1998;81:254.
3. Kruzliak P, Syamasundar RP, Novak M, et al. Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. *Arch Cardiovasc Dis* 2013;106:448-54.
4. Yiu MWC, Le DV, Leung Y, et al. Radiological features of isolated unilateral absence of the pulmonary artery. *J HK Coll Radiol* 2001;4:277-80.
5. Kant S. Unilateral pulmonary hypoplasia - a case report. *Lung India* 2007;24:69-71.
6. WIER JA. Congenital anomalies of the lung. *Ann Intern Med* 1960;52:330-48.
7. Griffin N, Mansfield L, Redmond KC, et al. Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: a review of four cases. *Clin Radiol* 2007;62:238-44.
8. Gluck MC, Moser KM. Pulmonary artery agenesis; diagnosis with ventilation and perfusion scintiphotography. *Circulation* 1970;41:859-67.
9. Katsenos S, Antonogiannaki EM, Tsintiris K. Unilateral primary lung hypoplasia diagnosed in adulthood. *Respir Care* 2014;59:47-50.
10. Sunam G, Ceran S. Pulmonary artery agenesis and lung hypoplasia. *Eur J Gen Med* 2009;6:265-7.

