

A rare cause of recurrent respiratory tract infection: isolated absence of the right pulmonary artery

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SUMMARY: Özcan A, Yılmaz İ, Kahveci S, Köse M. A rare cause of recurrent respiratory tract infection: isolated absence of the right pulmonary artery. *Türk J Pediatr* 2014; 56: 186-188.

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital anomaly presenting with a wide spectrum of symptoms. UAPA is usually associated with cardiac anomalies. Patients with isolated UAPA may be asymptomatic or may present with recurrent pulmonary infections. In the present study, we report UAPA in a four-year-old boy presenting with recurrent respiratory tract infections.

Key words: child, recurrent respiratory infections, absence of pulmonary artery.

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly. The main embryologic defect is an involution of the proximal sixth aortic arch of the affected side, leading to an absence of the proximal pulmonary artery. The clinical presentation is variable and many patients may be asymptomatic for many years. Recurrent pulmonary infections, decreased exercise tolerance and shortness of breath on exertion are the most common symptoms of UAPA¹. UAPA is most frequently accompanied by cardiovascular anomalies such as tetralogy of Fallot or septal defects².

We report a case of isolated UAPA without congenital cardiac anomaly presenting with recurrent pulmonary tract infections.

Case Report

A four-year-old boy was referred to Erciyes University Children's Hospital, Pediatric Pulmonology Department because of chronic cough and recurrent respiratory tract infections. No complaints of dyspnea, hemoptysis, or exercise intolerance were detected. His physical development was normal, and there was no family history of congenital cardiovascular disease. His medical history was uneventful except for several previous hospitalizations for bronchiolitis and pneumonia.

Because of the chronic cough and recurrent respiratory tract infections, the patient was

referred to our department two years after his first symptoms. There were no clinical signs of edema, cyanosis or clubbing of fingers. On admission, his body temperature was 37°C, pulse rate 92 beats/min, blood pressure 90/60 mmHg, and respiration rate 16/min. The physical examination findings were normal.

Results of hematological, biochemical and arterial blood gas analysis were within normal ranges. Routine chest X-ray showed a loss of volume of his right lung with displacement of the mediastinum to the right (Fig. 1). Transthoracic echocardiography excluded congenital heart anomalies and secondary pulmonary hypertension. Contrast-enhanced computed tomography was recommended for further evaluation and revealed congenital absence of the right pulmonary artery with loss of lung volume from the right lung (Fig. 2). His clinical follow-up status has been stable for the one year of follow-up.

Discussion

Unilateral congenital absence of a pulmonary artery is a rare developmental anomaly, first observed by Frantzel et al. in 1868¹. Embryologically, the distal intrapulmonary pulmonary arteries arise from their respective lung buds and join the proximal portion of the sixth aortic arch (extrapulmonary pulmonary artery), whereas the main pulmonary artery is

derived from the pulmonary arterial portion of the trunco-aortic sac³. UAPA is thought to occur due to an involution of the sixth aortic arch and a persistence of the communication between the intrapulmonary artery and the ductus arteriosus⁴. There is no predilection for the right or left side, although the condition is somewhat more common on the right. However, left-sided agenesis seems to be more frequently associated with cardiac abnormalities such as tetralogy of Fallot or septal defects⁵. There was no cardiac anomaly in our patient.

Many patients with UAPA can remain asymptomatic for a long period, and the actual prevalence of UAPA is difficult to establish in these asymptomatic patients. The majority of patients are identified incidentally during routine medical evaluations performed for different reasons. Some reports state that 30% of patients are asymptomatic³. Although a majority of patients are asymptomatic, our patient experienced several recurrent respiratory infections. Patients with isolated unilateral absence of the right pulmonary artery may be asymptomatic or may present with cough, recurrent pulmonary infections, hemoptysis, or dyspnea on exertion⁶. Symptoms can sometimes be masked by factors such as pregnancy or high altitude^{7,8}. Patients may also develop pulmonary arterial hypertension with right heart failure⁹.

The pathogenesis of recurrent infections is unknown, but it is hypothesized that bronchoconstriction from alveolar hypocapnia, diminished delivery of inflammatory cells, and dysfunctional mucociliary clearance play a role. Hemoptysis is caused by excessive



Fig. 1. X-ray showed a loss of volume of the right lung with displacement of the mediastinum to the right.

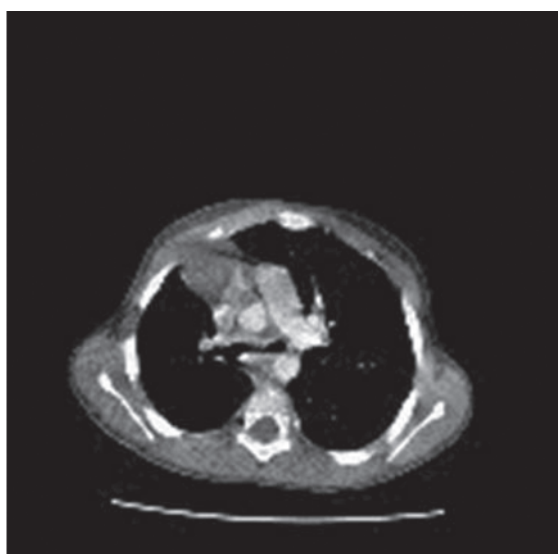


Fig. 2. Contrast-enhanced computed tomography showing absence of the right pulmonary artery with loss of lung volume from the right lung.

systemic collateral circulation, which may include bronchial, intercostal, subclavian, or subdiaphragmatic arteries¹⁰.

Typical chest radiographic findings are ipsilateral cardiac and mediastinal displacement, ipsilateral hemidiaphragm elevation with volume loss of the affected lung, absent hilar shadow, and hyperinflation of the contralateral lung. Our patient had right mediastinal displacement, right lung volume loss and diaphragm elevation in the chest radiograph. A contrast-enhanced computed tomography of the thorax can confirm the absence of the affected pulmonary artery¹¹. The principal radiologic differential diagnosis is Swyer-James, or MacLeod's, syndrome, in which the expiratory chest radiograph shows air trapping and the ventilation-perfusion scan reveals decreased ventilation and perfusion, as well as delayed washout in the ventilation scan¹².

Treatment options for the symptomatic patient are varied and controversial. Medical management of pulmonary hypertension with calcium channel blockers and prostacyclin has been proposed¹³. Surgical options include pneumonectomy and revascularization of the residual intrapulmonary pulmonary artery to a hilar artery. Embolization of the collaterals has been shown to be a safe alternative treatment in the symptomatic patient¹⁴. We did not use any medical or surgical treatment because of the patient's stable clinical status.

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