

Isolated Unilateral Absence of a Pulmonary Artery: An Unusual Cause of Recurrent Pulmonary Infection

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Isolated unilateral absence of a pulmonary artery (UAPA) is a rare anomaly. Some patients with isolated UAPA are completely asymptomatic and may be undetected until an incidental finding on routine chest x-ray, or the appearance of complications such as hemoptysis or recurrent pulmonary infections.

We report a 17-year old male patient with agenesis of the right pulmonary artery who presented with cough, sputum and dyspnea on exertion.

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Case Report

A 17-year old male patient was admitted to hospital with exertional dyspnea, productive cough and sputum. The patient stated that exertional dyspnea has first occurred three months ago. There was a past history of recurrent pulmonary infections.

Physical examination revealed a well-developed male in no distress. His blood pressure 110/60 mmHg, pulse 82 beats/min and respiratory rate 24 breathes/min. Coarse crackles over the right posterior lung field were audible on auscultation. Heart sounds were regular, without murmurs. No clubbing, cyanosis, telangiectasia or oedema were noted.

The chest x-ray on admission revealed loss of volume of the right lung and ipsilateral shift of the heart and mediastinum. Reticular, linear infiltrative shadows on middle and lower zones of the right hemithorax were noted (Fig.1). The sputum was negative for acid-fast bacilli. An electrocardiogram was normal. Computed tomographic (CT) angiography revealed the absence of the right pulmonary artery. No other cardiovascular abnormality was noted (Fig.2). Hypertrophied intercostals arteries were attributed to the collateral blood flow of the right lung (Fig 3). High resolution

computerized tomography (HRCT) demonstrated peribronchial thickening and bronchiectasis especially on basal segments of the right lung (Fig. 4). Echocardiography showed normal cardiac size and good left ventricular contractility, with no evidence of congenital cardiac lesions or pulmonary hypertension.

Bronchoscopy revealed normal bronchial structure. A perfusion scintigram showed the absence of perfusion in the right lung. Pulmonary angiog-

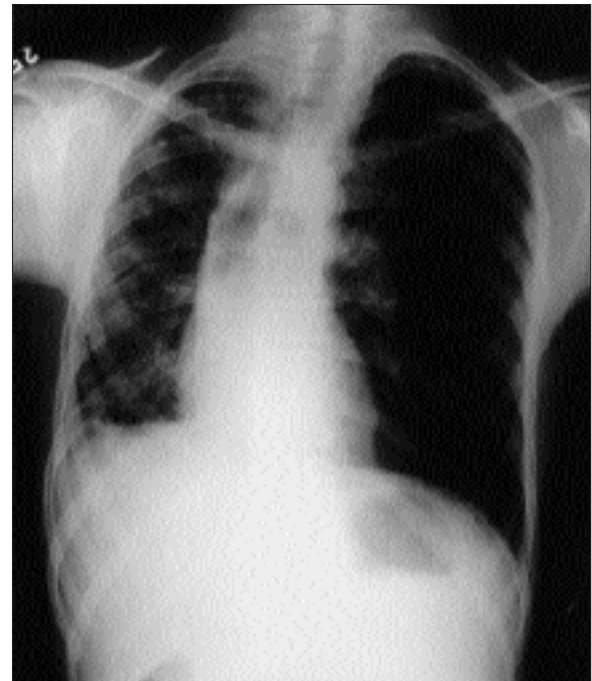


Fig 1: Posteroanterior chest radiograph.

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raphy confirmed the diagnosis of an absent right pulmonary artery (Fig.5). Arterial blood gas analyses was in normal range. Respiratory function test and carbonmonoxide diffusing capacity were found normal.

Since there was no pulmonary hypertension and any symptom of pulmonary hemorrhage, we decided to carefully follow this patient and at that time not perform any intervention.

Discussion

Unilateral absence of a pulmonary artery is a rare congenital anomaly that results principally from the failure of development of the ventral bud of the ipsilateral sixth aortic arch (1,2). Mo-

re frequently UAPA is combined with severe cardiovascular anomalies such as tetralogy of Fallot or septal defects (3-6). Since many patients with UAPA can remain asymptomatic for a long period, the actual prevalence of UAPA is difficult to establish. In Bouros and colleagues study, the prevalence of isolated UAPA has been found 1 among 200.000 men (7).

Absence of the left pulmonary artery is more commonly seen in patients with tetralogy of Fallot or truncus arteriosus, whilst absence of the right pulmonary artery is more often an isolated finding as in our case (8).The clinical presentation of these patients is variable, about 30% of patients remain asymptomatic. Most patients had symptoms such as dyspnea or limited exercise tolerance (40%), recurrent pulmonary infections (37%), or hemoptysis (20%) (7,9). Our patient



Fig 2: CT angiography of the patient at the level of aorticopulmonary window revealing the absence of the right pulmonary artery , arrows are showing dilated intercostals arteries.



Fig 3: Hypertrophied intercostal arteries due to collateral circulation on the affected lung is shown (arrowhead).



Fig 4: HRCT of the patient showing bronchiectasis on the affected lung.

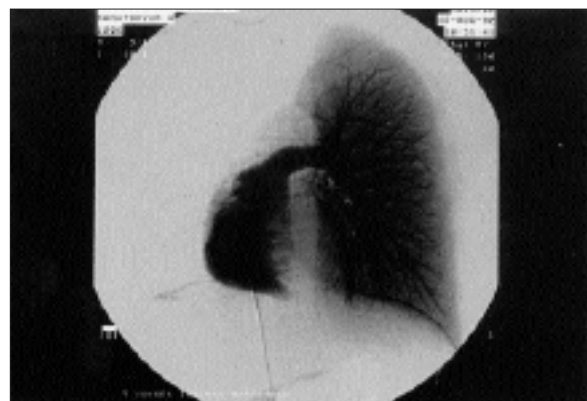


Fig 5: Pulmonary angiogram in the anterior view showing the absence of the right pulmonary artery.

stated dyspnea on exertion. His pulmonary function tests and arterial blood gas analysis were all in the normal range. The mechanism of dyspnea on exertion in patients with isolated UAPA remains unclear (7). Our case was also giving the history of recurrent pulmonary infections from the childhood. HRCT scans of the patient revealed peribronchovascular thickening and bronchiectasis in the affected lung. The pathogenesis of the recurrent infections and bronchiectasis is unclear. It has been suggested that bronchoconstriction secondary to alveolar hypocapnia is one possibility. Impaired mucociliary clearance and the diminished delivery of appropriate inflammatory cells may also contribute to the high incidence of infections in patients with UAPA (7). UAPA is an unusual cause of hemoptysis that results from hypertrophied collateral vessels (7,8). The systemic collaterals to the affected lung in patients with isolated UAPA come from the bronchial, intercostal, subclavian or subdiaphragmatic arteries (10). In our case the hypertrophied intercostal arteries on CT angiography suggested that the systemic collaterals came from the intercostal arteries. Various diagnostic possibilities are available for patients with UAPA. Physical findings can include an asymmetrical chest with decreased breath sounds in the affected lung, occasional systolic murmur across the pulmonary outflow tract because of turbulent blood flow into the remaining pulmonary artery. Cardiac examination and the electrocardiogram are usually within normal ranges when there is no associated congenital heart disease and pulmonary hypertension. Some patients may have digital clubbing (2,8). The chest radiograph can be very valuable, typically revealing an ipsilateral small, contracted lung, and a contralateral enlarged, hyperinflated, well-vascularized lung that frequently herniates across the midline (8). In addition, the absence of the left or right pulmonary artery ipsilateral cardiac and mediastinal displacement, ipsilateral hemidiaphragm elevation may be present (11). An echocardiogram may confirm the diagnosis and exclude other cardiovascular abnormalities. Pulmonary hypertension also can be diagnosed (3). Scintigraphic studies will demonstrate complete absence of perfusion to one whole lung with diminished ventilation (12). Bronchoscopy and

bronchograms generally reveal normal bronchial structure, although it is not unusual to find crowding or irregular constrictions on the affected side (8). CT scanning and MRI should be performed to assess UAPA. HRCT scanning has been found valuable in the evaluation of congenital heart defects and distal pulmonary arteries in UAPA patients. Both of the three techniques enabled congenital UAPA to be distinguished from acquired obstruction of the pulmonary artery (7,9,13). Digital subtraction angiography (DSA) and pulmonary angiography are usually definitive in establishing the diagnosis (7).

Other conditions mimicking UAPA on plain chest radiograph are; collapse of a lobe or of one lung, chronic tuberculous scarring, unilateral emphysema, hypoplasia of one lung, Swyer-James syndrome or hyperlucent lung, and complete obstruction of a main pulmonary artery secondary to thromboembolism, neoplasm or fibrosing mediastinitis (9,13).

Therapeutic approach should be based on symptomatology and severity of disability. When pulmonary hypertension is present in a patient with UAPA, revascularization of affected side may improve the condition. If revascularization is not possible, long-term vasodilator therapy may improve survival. Massive hemoptysis can be treated by selective embolization of the systemic collaterals or even pneumonectomy of the affected side (3,14).

References

1. Thomas P, Gaubert MR, Bartoli JM, et al. Exsanguinating hemoptysis revealing the absence of left pulmonary artery in an adult. *Ann Thorac Surg* 2001; 72: 1748-50.
2. Kuo PH, WU HD, Lee LN. A patient with hemoptysis and a smaller right lung. *Eur Respir J* 1996; 9: 847-49.
3. Harkel ADJ, Blom NA, Ottenkamp J. Isolated Unilateral Absence of a Pulmonary Artery. *Chest* 2002; 122: 1471-77.
4. Pac A, Özme S, Çeliker A, et al. Absent pulmonary valve syndrome with agenesis of the left pulmonary artery. *Turk J Pediatr* 1994;36:249-53.
5. Presbitero P, Bull C, Hawort SG, et al. Absent or occult pulmonary artery. *Br Heart J* 1984; 52:178-85.
6. Whitman G, Stephenson LW, Weinberg P. Vascular

- ring: left cervical aortic arch, right descending aorta, and right ligamentum arteriosum. *Thorac Cardiovasc Surg* 1982;83: 311-15.
7. Bouros D, Pare P, Panagou P, et al. The varied manifestation of pulmonary artery agenesis in adulthood. *Chest* 1995; 108:670-76.
 8. Pool PE, Vogel JH, Blount SG. Congenital unilateral absence of a pulmonary artery. *Am J Cardiol* 1962;10: 706-32.
 9. Shakibi JG, Rastan H, Nazaraian I, et al. Isolated unilateral absence of the pulmonary artery: review of the world literature and guidelines for surgical repair. *Jpn Heart J* 1978; 19: 439-51.
 10. Oakley C, Glick G, McCredie RM. Congenital absence of a pulmonary artery: a report of a case, with special reference to bronchial circulation and review of the literature. *Am J Med* 1962; 34:264-70.
 11. Krall WR, Ploy-Song-Sang Y. Unilateral pulmonary artery aplasia presenting with chest pain and pleural effusion. *South Med J* 1980;73:233-36.
 12. Bouros D, Panagou P. Pulmonary agenesis. *Eur Respir J* 1990; 3:379-80.
 13. Link KM, Lesco NM. Magnetic resonance imaging in the evaluation of congenital heart disease. *Magn Reson Q* 1991;7:173-90.
 14. Taugchi T, Ikeda K, Kume K, et al. Isolated unilateral absence of left pulmonary artery with peribronchial arteriovenous malformation showing recurrent hemoptysis. *Pediatr Radiol* 1987; 17: 316-18.