Unilateral Pulmonary Artery Agenesis in an Elderly Female

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Abstract

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly due to a malformation of the sixth aortic arch of the affected side during embryogenesis. It can occur in isolation or may be associated with other congenital cardiovascular malformations. The diagnosis is usually set at adolescence, however it can remain asymptomatic and late diagnosis is possible. Recurrent pulmonary infections, decreased exercise tolerance, and shortness of breath on exertion are the most common symptoms.

Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly due to a malformation of the sixth aortic arch of the affected side during embryogenesis.

The estimated prevalence of isolated UPAA without associated cardiac anomalies ranges from 1/200,000 to 1/300,000 adults.² Isolated UPAA is twice as common on the right side.³ There is no sex predilection. Approximately 75% of the patients with left-sided pulmonary artery agenesis have associated congenital cardiac anomalies. In comparison, occurrence of associated congenital cardiac anomalies is rarely seen in patients with right-sided pulmonary artery agenesis.³

The most frequent accompanying cardiovascular anomalies include tetralogy of Fallot, septal defects, patent ductus arteriosus, coarctation of the aorta and transposition of great vessels and Scimitar syndrome.³ The patients with isolated UPAA without other cardio-vascular abnormalities are often asymptomatic and therefore remain undiagnosed. They may present with exercise intolerance, haemoptysis or are incidentally detected during chest radiography.

Case Report

A 60 year-old female was admitted for evaluation of progressively worsening shortness of breath. Her exercise tolerance was reduced to walking few feet. Her past medical records revealed that she had hypertension which was well controlled. There was no history of coronary artery disease. She had three uneventful pregnancies, and all her children were healthy.

On physical examination she was an overweight elderly woman who had mild respiratory distress. Cardiovascular examination revealed a left parasternal heave, prominent pulmonary artery pulsation and loud P2. Examination of the respiratory system revealed tracheal shift to the right, reduced right chest movements and reduced breath sounds over the right hemithorax. There were no added sounds. Routine laboratory tests were normal. Her chest radiograph (Figure 1) showed evidence of volume loss in the right hemithorax, a mediastinal shift to the right and crowding of the right ribs. Pulmonary function tests revealed a mixed restrictive and obstructive pattern. A contrast enhanced computerised tomography (CECT) scan (Figure 2), revealed the absence of right pulmonary artery and a dilated main pulmonary artery with evidence of collaterals at the right hilum and apex. These findings led to the diagnosis of right pulmonary artery agenesis.

Discussion

UPAA is a rare anomaly that can occur in isolation or is accompanied by other cardiovascular malformations. It results from the involution of the proximal sixth aortic arch of the affected side, leading to an absence of the proximal pulmonary artery. Intrapulmonary vessels and distal portion of the affected pulmonary artery trunk can develop normally, and blood supply is achieved by systemic collaterals from bronchial, major aortopulmonary collaterals and other systemic arteries.³ Two types of presentations have been described based upon the timing of presentation. The first one is seen in infants, who present with congestive cardiac failure and pulmonary hypertension. The other presentation is in older patients, who are often asymptomatic; they may present with exercise intolerance, haemoptysis or recurrent infections. Recurrent infection can lead to bronchiectasis in some.¹ Recurrent infections observed in patients with UPAA results from lack of arterial blood flow to the affected lung. This results in poor delivery of inflammatory cells to sites of inflammation, impaired ciliary function and alveolar hypocapnia. Alveolar hypocapnia promotes secondary bronchoconstriction and mucous
trans-thoracic echocardiography. On magnetic resonance imaging (MRI) or by computed tomography (CT), complaints. had three term deliveries, she had no or high altitude. Although our patient unmasked by factors such as pregnancy as asymptomatic unless the symptoms are missed while in adults it may be totally signs can be subtle and can be easily to make the diagnosis. In infancy, the high index of suspicion is required Diagnosis of UPAA is difficult and a of PA agenesis. Currently, with the remnant the gold standard for diagnosis remain asymptomatic and undiagnosed in childhood but some subjects may remain asymptomatic and undiagnosed until adult age. A number of imaging techniques are available to aid the diagnosis. Physicians should bear in mind the possibility of undiagnosed UPAA in adults also.

References


Fig. 2: Coronal CT chest showing main pulmonary artery continues as left pulmonary artery and complete absence of right pulmonary artery - agenesis of right pulmonary artery. (A) Right internal mammary artery, (B) Collaterals, (C) Prominent intercostal arteries, (D) Collateral from inferior phrenic artery

trapping. Haemoptysis in patients with UPAA is from collateral circulation. Pulmonary hypertension (PHT) results from increased blood flow in the contralateral pulmonary artery which leads to shear stress, with subsequent release of vasoconstrictive compounds. Chronic vasoconstriction leads to pulmonary vasculature remodeling and increased resistance and PHT. Diagnosis of UPAA is difficult and a high index of suspicion is required to make the diagnosis. In infancy, the signs can be subtle and can be easily missed while in adults it may be totally asymptomatic unless the symptoms are unmasked by factors such as pregnancy or high altitude. Although our patient had three term deliveries, she had no complaints.

When there is suspicion of UPAA, definitive diagnosis should be pursued by computed tomography (CT), magnetic resonance imaging (MRI) or transthoracic echocardiography. On cross-sectional imaging (CT or MRI), the absent PA typically terminates within 1 cm of its origin from the main PA. Other findings include intact peripheral branches of the PA, variable collateral circulation, mosaic parenchymal changes and bronchiectasis. Transthoracic echocardiography is a good tool for establishing the diagnosis, excluding any other cardiovascular abnormalities and evaluating the presence of associated PHT. Magnetic resonance angiography (MRA) with contrast provides real-time assessment of the haemodynamic status. Angiography remains the gold standard for diagnosis of PA agenesis. Currently, with the development of CT, MRI and MRA techniques, it is rarely performed unless embolization is indicated for massive haemoptysis. Pulmonary venous wedge angiography is particularly useful in delineating the presence of an ipsilateral hilar PA and intrapulmonary vessels, which is required before revascularization.

There is no consensus regarding the treatment for isolated absence of PA. The therapeutic approach for isolated absence of PA should be based on symptoms of the patient, PA anatomy, associated cardiovascular anomalies and aortopulmonary collaterals and PHT. No treatment is required in patients without any evidence of cardiopulmonary dysfunction (as seen in adults with incidental detection); they should be followed up on a regular basis. Pneumonectomy and surgical revascularization are considered in cases of recurrent hemoptysis, pulmonary infections and PHT. Selective embolization of bronchial or non-bronchial systemic arteries is an alternative for patients with massive hemoptysis not eligible for surgery. Pharmacological treatment for PHT is recommended for patients unable to undergo surgical revascularization or in cases not improved after surgery.

In conclusion, UPAA is a rare congenital anomaly usually diagnosed in childhood but some subjects may remain asymptomatic and undiagnosed until adult age. A number of imaging techniques are available to aid the diagnosis. Physicians should bear in mind the possibility of undiagnosed UPAA in adults also.