Isolated Right Pulmonary Artery Agenesis with Agenesis of Right Upper Lobe and Bronchiectasis of Right Lower Lobe with Anomalous Arterial Supply from Celiac Axis with Normal Venous Drainage

A. Chaudhry, M. Rathore and J.N. Banavaliker

Department of TB and Chest, Rajan Babu Institute for Pulmonary Medicine and Tuberculosis (RBIPMT), Delhi, India

Abstract

Isolated unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly. When detected in infancy, the condition is commonly associated with cardiovascular defects which are more frequently associated with left pulmonary artery agenesis. Patients with isolated right pulmonary artery agenesis survive into adulthood with minimal or no symptoms and are diagnosed incidentally on the chest radiographs. We report a case of a 19-year-old female patient who presented to us with recurrent haemoptysis. She was symptomatic since the age of four years. We report the rare occurrence of UAPA on right side, agenesis of right upper lobe and bronchiectasis of right lower lobe with anomalous arterial supply of right lung from coeliac axis in this patient. [Indian J Chest Dis Allied Sci 2014;56:49-52]

Key words: Pulmonary artery agenesis, Lobar agenesis, Bronchiectasis, Haemoptysis.

Introduction

Unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly with estimated prevalence of around 1 in 200,000 individuals. The first case of UAPA was reported by Fraenkel in 1868. It may occur as an isolated lesion or in combination with cardiac or vascular lesions. About 75% of the patients with left-sided pulmonary artery agenesis have associated congenital cardiac anomalies. In comparison, occurrence of associated congenital cardiac anomalies are rarely seen in patients with right-sided pulmonary artery agenesis. The most frequent accompanying cardiovascular anomalies include Tetralogy of Fallot or septal defects. Other congenital cardiac defects associated with UAPA are: coarctation of the aorta (either isolated or in combination with a ventricular septal defect), subvalvular aortic stenosis, transposition of the great arteries (either isolated or in combination with ventricular septal defect or pulmonary stenosis), Taussig-Bing malformation and coarctation, congenitally corrected transposition and pulmonary stenosis, and Scimitar syndrome. Although some patients with isolated UAPA are completely asymptomatic, others may have severe pulmonary hypertension and congestive heart failure. UAPA with lung hypoplasia is very rare. Only few cases of unilateral pulmonary artery agenesis have been reported from India. We report for the first time from India the case of a patient with right pulmonary artery agenesis with agenesis of right upper lobe and bronchiectasis of right lower lobe with anomalous arterial supply of the right lung by a branch from celiac axis, namely hepatic artery.

Case Report

A 19-year-old college girl who was a non-smoker, presented with recurrent episodes of haemoptysis starting at the age of four years, occurring almost annually with small amount (10-20 mL) of blood. But for the last three years, she had been having 2-3 episodes of haemoptysis per year and expectorating approximately 80-100 mL of blood every time. The patient also gave a history of recurrent respiratory tract infections from 1 to 4 years of age. Physical examination revealed decreased chest movement on right side, a decreased volume of right hemithorax, with accompanying decreased respiratory sounds on auscultation. Except for mild anaemia, routine haematological and biochemical parameters were within normal limits. Spirometry revealed forced vital capacity (FVC) 58% of predicted, forced expiratory volume in the first second (FEV1) 53% of predicted and FEV1/FVC ratio of 80% suggestive of restrictive pattern. Chest radiograph (postero-anterior view) showed a contracted right hemithorax with ipsilateral shift of the mediastinum, elevated right hemidiaphragm and a hyperlucent left lung (Figure 1). The left pulmonary artery appeared prominent. High resolution computed tomography (HRCT) of thorax (Figure 2) was suggestive of agenesis of right upper lobe with bronchiectasis of right lower lobe with ipsilateral shift of the mediastinum. Computed tomography (CT) angio-
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The patient was referred to a higher institute for pneumonectomy (definitive treatment), which was declined by her. Her clinical status has remained stable over the past one-and-half years of follow-up on an outpatient basis, with symptomatic and supportive treatment for recurrent haemoptysis.

Discussion

UAPA is a rare congenital anomaly that results principally due to failure of development of the ventral bud of the ipsilateral 6th aortic arch. Chromosomal defects, vitamin A deficiency, intrauterine infections, environmental factors have been held responsible for the aetiology of congenital lung malformations. When detected in infancy, the pulmonary artery agenesis is more frequent on the left side and is associated with congenital heart diseases which include Tetralogy of Fallot, septal defects and persistent ductus arteriosus. Isolated UAPA is relatively uncommon and patients present at a later age. Common presentations include shortness of breath during exercise or exercise limitation (40%), recurrent pulmonary infections (37%), haemoptysis (20%), high altitude pulmonary oedema (HAPE) (12%) and pulmonary hypertension (44%), though several patients are asymptomatic. Recurrent pulmonary infections in some patients are due to bronchiectasis.
Isolated UAPA more commonly affects the right pulmonary artery. Patients with isolated right pulmonary artery agenesis survive into adulthood with minimal or no symptoms. The diagnosis of UAPA is generally based on medical history, physical examination findings, and the results of chest radiographs. Chest radiographic findings include: ipsilateral cardiac and mediastinal displacement, absent pulmonary arterial shadow, smaller hemithorax, elevation of the hemidiaphragm, and paucity of vascular markings on the affected side. There may be hyperinflation and herniation of the unaffected lung across the midline.

Differential diagnoses include Swyer-James syndrome, lobar atelectasis, post lobectomy status, chronic pulmonary thromboembolism, pulmonary agenesis/hypoplasia complex and agenesis/hypoplasia of the pulmonary artery. A contrast enhanced CT of the thorax confirms the absence of the affected pulmonary artery. In the cases with recurrent bronchopulmonary infections, HRCT of the thorax helps in determining the presence of bronchiectasis. Magnetic resonance imaging (MRI) is helpful to evaluate the congenital cardiovascular defects. Ventilation-perfusion scan shows decreased ventilation and absent perfusion on the affected side. Absent perfusion on one side with bilateral normal ventilation may occur in unilateral absence of a pulmonary artery; thrombotic occlusion of a main pulmonary artery; or pulmonary arterial branch stenosis. These three conditions can be further differentiated by clinical history, physical examination, or angiographic studies. Echocardiography is a useful tool for exclusion/evaluation of other cardiac or major vessels abnormalities and the presence of associated pulmonary hypertension. Pulmonary angiography remains the gold standard for the diagnosis of pulmonary artery agenesis and is reserved for patients requiring embolisation or revascularisation surgery.

When revascularisation is being considered, cardiac catheterisation should be done with pulmonary venous wedge angiography to visualise hidden pulmonary artery in the hilum.

Pulmonary function test in patients with UAPA are usually unremarkable or show a mild restrictive defect with an increased ratio of physiologic dead space to tidal volume.

Recurrent haemoptysis is a less frequently found symptom in isolated UAPA (18%-20%). This haemoptysis is due to excessive systemic collateral circulation coming from the bronchial, intercostals, subclavian, subdiaphragmatic or, segmental vessels arising from the aorta, including a subphrenic contribution from the coeliac axis, as was in our case. During the embryonic development, the lung is supplied by small transitory branches of the dorsal aorta. These branches later disappear normally but, persist in cases where the pulmonary artery does not develop. These persist as enlarged supra-diaphragmatic or infra-diaphragmatic aortic branches, or as bronchial arteries. Haemoptysis may be mild and self-limiting for many years but also can be life-threatening leading to massive pulmonary haemorrhage and death.

Pulmonary hypertension may be present in 20%-44% cases of UAPA. Pulmonary hypertension in patients with UAPA may also be unmasked by predisposing factors such as HAPE and pregnancy. HAPE in such patients develops at heights less than 3,000 meters. The patient has to be brought down to a lower level as soon as possible and supplemental oxygen or nitric oxide, if available should be administered. Pulmonary hypertension adversely affects pregnant women. Patients with pulmonary hypertension have high mortality during pregnancy. The literature shows that individuals with UAPA who develop pulmonary hypertension generally do so at an early age and succumb to right-sided heart failure.
The therapeutic options in UAPA include creation of an aorto-pulmonary shunt or restoration of continuity between main and hilar pulmonary artery when diagnosed at an early age. The treatment of UAPA in adults depends upon the clinical presentation. Selective arterial embolisation or a pneumonectomy or lobectomy should be considered for recurrent or life-threatening haemoptysis or intractable pulmonary infections. In the presence of pulmonary hypertension, revascularisation of the absent artery is recommended and may improve the hypoxaemia of the patient. If revascularisation is not an option or when pulmonary hypertension is not improved, medical treatment described for patients with primary pulmonary hypertension may be helpful.

In conclusion, clinicians should consider UAPA in patients presenting with haemoptysis and recurrent respiratory infections. Chest radiograph usually suggests the diagnosis. Echocardiography is helpful for the evaluation of possible pulmonary hypertension and associated cardiac anomalies. The diagnosis can be confirmed and anatomic details can be discerned by CT and MRI. Angiography is reserved for patients requiring embolisation or revascularisation surgery. Our patient was diagnosed with right-sided UAPA after presenting with recurrent haemoptysis. She refused to undergo pneumonectomy. She was treated with an oral course of broad-spectrum antibiotics with symptomatic and supportive treatment for recurrent episodes of haemoptysis. Her clinical status has remained stable for the past one-and-half years of follow-up on an outpatient basis.

References


