Case Report

An Adult with Haemoptysis: A Rare Case of Congenital Anomaly

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Abstract

A non-smoker adult male presented with haemoptysis of short duration. Chest radiograph (postero-anterior view) suggested an opaque left hemithorax. Further evaluation of lung lesion pointed towards a left lung hypoplasia with absent left pulmonary artery and a right-sided aortic arch (RAA). Both kidneys were enlarged with multiple cysts and thinning of parenchyma. This case describes a unique coexistence RAA and probable autosomal dominant polycystic kidney disease.
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Key words: Pulmonary hypoplasia, RAA, ADPKD.

Introduction

Pulmonary hypoplasia manifesting in adults in association with other congenital anomalies of heart or great vessels has been described in the literature, though rarely. Unilateral absence of pulmonary artery (UAPA) has been described previously in association with pulmonary hypoplasia.[1,2] More recently it has been described as 'ductal origin of distal pulmonary artery'.[3] Here the distal pulmonary artery is supplied by ductus arteriosus in utero and the proximal pulmonary artery segment from the pulmonary trunk upto the hilum is absent. Diminished blood supply after birth due to the closure of the ductus arteriosus results in hypoplasia of lung, although the fetal lung development is normal. A right-sided aortic arch (RAA) is an uncommon congenital defect of the aorta, and is rare in the setting of an otherwise normal heart.

Pulmonary hypoplasia in association with UAPA and manifesting in adults has been reported only in one case previously, to the best of our knowledge.[1] We describe an adult with RAA and left lung hypoplasia in association with probable autosomal dominant polycystic kidney disease (ADPKD). This case is being reported due to its rarity of occurrence.

Case Report

A 42-year-old male was admitted with a history of haemoptysis of about 50 mL per day of four days duration. There was no history of dyspnoea, cough, orthopnoea or chest pain or any history of similar complaints in the past. He was a non-smoker and a teetotaller. General physical examination was unremarkable. Respiratory system examination revealed diminished breath sounds intensity over the left infra-clavicular and left mammary area. Examination of other systems did not reveal any abnormality.

Investigations revealed a normal haemogram, erythrocyte sedimentation rate, liver and renal biochemistry, bleeding and coagulation parameters. The chest radiograph (postero-anterior view) showed a veiled opacity in the left hemithorax with a shift of the mediastinum to the same side with both the diaphragms at the same level (Figure 1A). Culture of the sputum revealed no pathogen and two sputum direct smears were negative for acid-fast bacilli (AFB). Contrast enhanced computed tomography (CECT) of thorax showed changes suggestive of left lung hypoplasia (Figure 1B). Fibreoptic bronchoscopy revealed hyperaemic mucosa in the entire right endobronchial tree. On the left side, the bronchial caliber was smaller than the right throughout the lung (Figure 1C) and showed purulent secretion. The opening of lingular and left lower lobar bronchi were inflammed. Bronchoalveolar lavage fluid was negative for acid-fast bacilli (AFB) while fluid culture grew extended spectrum ß-lactamase (ESBL) producing *Escherichia coli*. Axial cuts of contrast enhanced computed tomography of thorax showed absence of the left pulmonary artery (Figure 2A) and a right-sided aortic arch with four branches arising from it (Figure 2B). Both kidneys were enlarged with multiple cysts and thinning of parenchyma (Figure 2C).

Discussion

Pulmonary hypoplasia is a rare entity that is seen in about one to two out of every 12,000 births.[4] Warburton *et al*[5] described an interaction between blood vessels and airways during the development of lung and also highlighted that development of the former controlled airway growth, particularly the formation of alveoli. The possible mechanism for recurrent pulmonary infections in pulmonary hypoplasia may be: (1)

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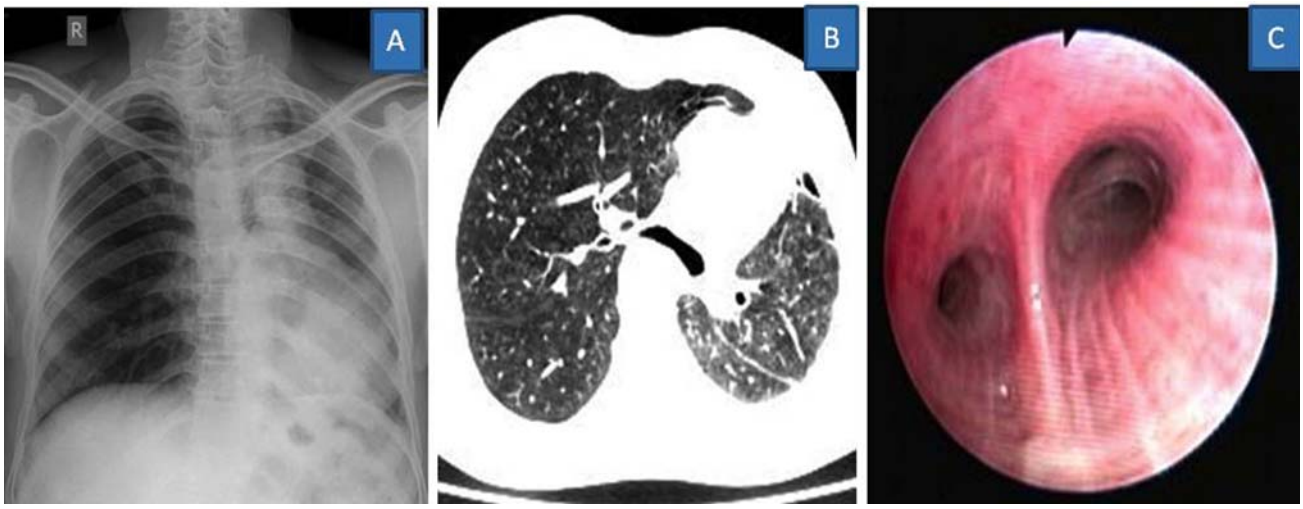

Figure 1. (A) Chest radiograph (postero-anterio view) showing veiled opacity in left hemithorax with shift of the mediastinum to the same side and both the diaphragms at the same level; (B) axial CT thorax at tracheal bifurcation level showing hypoplastic lung parenchyma on the left side; and (C) bronchoscopy image showing the small caliber opening of the left main bronchus.

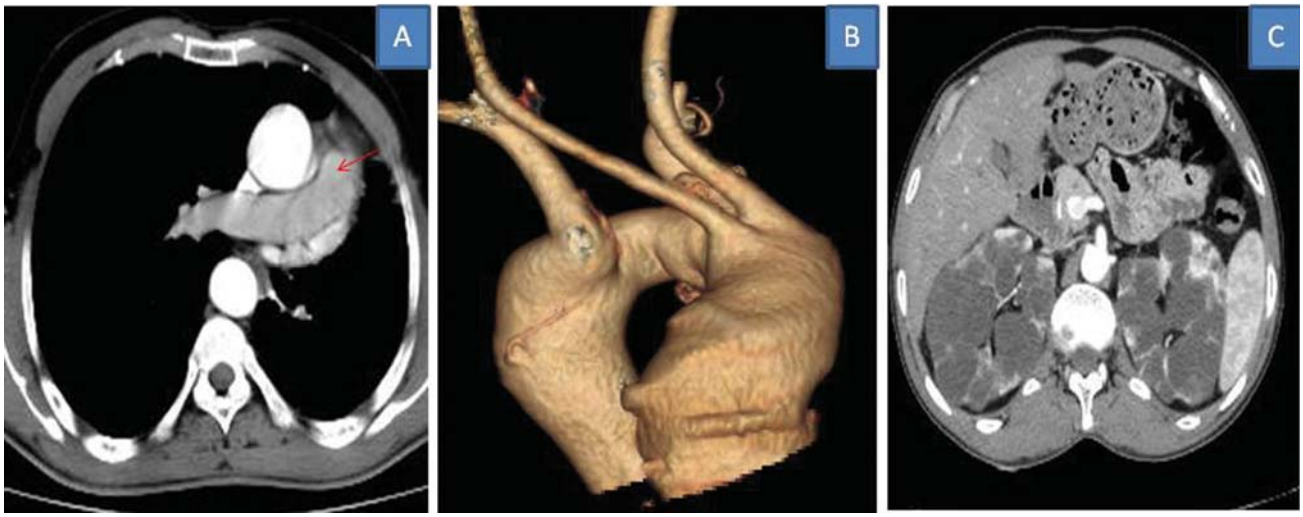

Figure 2. Axial CT thorax showing (A) absence of left pulmonary artery (arrow shows right pulmonary artery); (B) 3D-reconstruction image showing right-sided aortic arch and four branches arising from it; and (C) enlarged bilateral kidneys with multiple cysts and thinning of parenchyma.

reduced blood supply and defective ventilation of the lung parenchyma due to pulmonary artery defect;[6] (2) altered venous pulmonary circulation and (3) extrinsic compression of the large airways by the great vessels.[7] Haemoptysis may occur due to infection in the presence of above predisposing factors or may be due to increased blood flow and pulmonary hypertension in the opposite lung due to UAPA and consequent hypoplasia of the ipsilateral lung.

The RAA with aberrant origin of great vessels is a rare, but well recognised cause of tracheobronchial / oesophageal compression. The RAA is described in 0.05% to 0.1% of radiology series. Edwards[8] described three main types of RAA: type I (59%), with mirror-image branching of the major arteries; type II (39.5%), with an aberrant left subclavian artery; and type III (0.8%), with isolated subclavian artery (where the subclavian artery is connected to the pulmonary artery through the ductus arteriosus). Eighty-five percent of type 1 cases are associated with cardiac anomalies, whereas in type 2 the association is much less common (<10%). A deletion in chromosome 22q11 is known to be associated with a 24% incidence of isolated anomalies of laterality of branching of the aortic arch.[9] Though the condition can be asymptomatic, this can present at any age with features of tracheobronchial or esophageal compression.

A case of right lung hypoplasia with UAPA manifesting as haemoptysis was described in a

middle aged male.[1] Another case of pulmonary hypoplasia with RAA along with UAPA was reported in a 21-month-old child.[2] A 5-month child with RAA, right descending aorta and agenesis of the left pulmonary artery has been previously described in literature,[10] presenting with lung infection and progressive respiratory failure. In our case, the patient presented at the age of 42 years with haemoptysis. On evaluation, he was found to have left lung hypoplasia along with RAA.

The diagnosis of hypoplasia of lung manifesting in adults is based on a detailed history and physical examination, coupled with a reasonable index of suspicion. The differential diagnosis should include collapse of a lobe or of one lung, chronic tuberculous scarring, and complete obstruction of a main pulmonary artery secondary to thromboembolism or neoplasm. Though this patient did not have a family history of ADPKD, he had multiple cysts in both the kidneys which is a well-recognised genetic entity (chromosome 16p13 and 4q21) in association with various extra-renal manifestations. To the best of our knowledge, this is the first reported case with coexistence of RAA and probable ADPKD which are two separate genetic entities, the former usually manifesting in childhood and the later in adults.

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