Exertional dyspnoea and haemoptysis in an adolescent: is it tuberculosis only?

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Abstract

Unilateral pulmonary artery atresia (UPAA), commonly detected in childhood, can occasionally present in adulthood with typical symptoms of recurrent pulmonary infections, dyspnoea on exertion and haemoptysis. An 18-year-old girl presented with complaints of cough, dyspnoea on exertion and haemoptysis. Chest radiograph revealed dilated pulmonary trunk, signs of left sided volume loss and cavitating lesion in left lower zone. Pulmonary CT angiography found left pulmonary artery atresia. Endobronchial lung biopsy revealed granulomatous inflammation. Diagnosis of left sided UPAA with pulmonary tuberculosis was established. She responded well to the anti-tubercular therapy. This case highlights the importance of awareness about UPAA as a possible differential for exertional dyspnoea, recurrent chest infections, haemoptysis and pulmonary hypertension in adults.

Introduction

Unilateral pulmonary artery atresia (UPAA) is a defect that occurs due to involution of proximal part of embryonic sixth aortic arch and its failure to fuse with pulmonary trunk [1]. It is mostly associated with other cardiovascular malformations but rarely can occur in isolation, with right sided pulmonary artery atresia being more common [2]. Most commonly it is detected in childhood with only a few cases remaining asymptomatic till adulthood and these usually present when they develop complications [3]. In adults, it presents most commonly with haemoptysis, dyspnoea on exertion and recurrent chest infections [4]. Few cases of pulmonary tuberculosis (TB) in a patient with unilateral pulmonary artery atresia have been reported globally, with no such case being reported from India.

Case Report

An 18-year-old non-smoker female presented with complaints of cough, dyspnea on exertion and left sided chest pain for 1.5 years, anorexia and weight loss for past 1 month and haemoptysis (< ½ tsp per day) for 5 days. On physical examination only significant finding was tachycardia. On auscultation, bilateral normal vesicular breath sounds were audible. Chest X-ray (Figure 1a) revealed dilated pulmonary trunk, left sided oligemia with signs of left sided volume loss, a cavity in left lower zone and increased bronchovascular markings in right upper and mid zone. Pulmonary CT angiography was suggestive of left pulmonary artery atresia with dilatation of main and right pulmonary artery (Figure 2) and evidence of increased right upper lobe vasculature. CT thorax also showed bilateral lower lobe cavitating lesions and mosaic perfusion and centrilobular nodules (Figure 3). Echocardiography revealed severe pulmonary artery hypertension with dilatation of right atria and right ventricle.

Mantoux test was positive with 15 mm induration. Sputum smear and GeneXpert were negative. On bronchoscopy, bronchoalveolar lavage analysis was inconclusive. Endobronchial biopsy revealed granulomatous inflammation. Hence, based on above a course of anti-tubercular treatment was given for a period of six months.

After completing the treatment, patient’s symptoms improved, and chest X-ray (Figure 1b) showed improvement with evidence of resolution of left lower zone cavitating lesion.

Diagnosis: Pulmonary tuberculosis with left sided isolated unilateral pulmonary artery atresia (UPAA) with pulmonary hypertension.

Discussion

Isolated unilateral pulmonary artery atresia (UPAA) is a very rare developmental anomaly [5]. In a study, conducted in India incidence of pulmonary atresia was found to be 0.47% (46 cases out of 9,728 cases of congenital heart diseases [6]). It was first described by Frantzel in 1868 [7]. In two third of cases the right
sided pulmonary artery is affected [8]. But in our case left side was involved. Most of the times, it is associated with other congenital cardiac defects [9]. It usually presents in childhood. Rarely it remains asymptomatic till adulthood and presents with recurrent chest infections. Our patient had remained asymptomatic in childhood and presented in adolescence. She was diagnosed as a case of left pulmonary artery atresia and severe pulmonary artery hypertension with no other cardiac defects.

Isolated UPAA assumes significance in pulmonology practice because of the following reasons:

i. **Differential diagnosis of dyspnoea/exertional breathlessness in adolescent age group.** Patients of UPAA manifest dyspnoea because of large amount of dead space in the affected lung having pulmonary artery atresia. This is one of the causes of exertional dyspnoea in these cases with pulmonary hypertension also contributing to the same.

ii. **Pulmonary hypertension.** Evidence of pulmonary artery hypertension has been reported in up to 25% of patients with unilateral pulmonary artery atresia and has been reported to be related to long term survival in such patients [10]. Absence of pulmonary artery on one side in cases of UPAA leads to diversion of blood to the other remaining pulmonary artery. This results in endothelium intimal shear stress injury and release of mediators like endothelin causing vasoconstriction. This further leads to remodelling in pulmonary vasculature. All these factors contribute to development of pulmonary hypertension which clinically presents as dyspnoea and poor exercise tolerance [11]. Hence, it is important to be aware of UPAA as one of the causes of pulmonary hypertension in adults.

iii. **UPAA can also present with haemoptysis in young patients** because of large collateral circulations in the pulmonary venous system [4]. The haemoptysis in these patients can be mild and self-limiting or sometimes even massive. In developing countries, like India, with high burden of tuberculosis (TB), the most common diagnosis for haemoptysis in young patient remains TB. Based on radiology findings, UPAA can also be a differential diagnosis for haemoptysis in young patient. However, in the present case, we had both UPAA and Pulmonary TB co-existing and presenting as haemoptysis.

iv. **UPAA can present with recurrent chest infections:** The mechanisms for recurrent infections in these patients is not well established. However, it is postulated that alveolar hypoxia and impaired mucociliary clearance mechanisms leading to the above. Also, decreased blood supply results in decreased delivery of the appropriate inflammatory cells. The affected side lung parenchyma can also develop bronchiectasis and bullae [2]. Further, it has been postulated that the patients suffering from congenital heart diseases associated with shunting of pulmonary blood flow (left to right shunt), develop an increased blood flow to the upper regions of the lung. This creates an aerobic environment, hence favouring the growth of tubercle bacillus [12]. In our case, due to atresia of left pulmonary artery, right pulmonary artery got dilated, thereby increasing blood flow to the right lung and probably making it more prone to TB.

v. **Radiology:** Chest x-ray in unilateral pulmonary artery atresia can show decreased size of the affected hemithorax, elevation of the same sided hemidiaphragm, ipsilateral hyperlucent hemithorax, absent ipsilateral hilar shadow, features suggestive of absent affected pulmonary artery with enlarged opposite side pulmonary artery shadow and displacement of the mediastinum to the affected side [13]. The differential for these radiology findings could include massive pulmonary embolism, Swyer-James syndrome and Scimitar syndrome. However, when these suggestive findings are present the transthoracic echocardiography, computed tomography (CT) thorax or MRI will clinch the diagnosis of UPAA.

Bronchoscopy findings in UPAA can range from a completely normal study to dilated blood vessels (mesh like vascularisation) which may also be the cause of haemoptysis [5,14,15]. In our case bronchoscopy was grossly normal. The treatment of UPAA in adults is still a therapeutic challenge and depends on clinical presentation.

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**Figure 1.** a) Chest X-ray (posteroanterior view) shows dilated pulmonary trunk, signs of left sided volume loss and a cavity in left lower zone, increased bronchovascular markings in right upper and mid zone. b) Chest X-ray (posteroanterior view) – after completion of Anti-Tubercular therapy (ATT) showing resolution of cavity.
Case Report

Figure 2. Pulmonary CT Angiography showing left pulmonary artery atresia with dilatation of main and right pulmonary artery. MAPCAS, major aorto-pulmonary collateral arteries; LPA, left pulmonary artery.

Figure 3. CT thorax showing bilateral lower lobe cavitating lesions and mosaic perfusion and centrilobular nodules.
The treatment options may vary from chronic pulmonary vasodilator therapy to selective embolization in cases of massive haemoptysis and even lobectomy or pneumonectomy [11].

This case emphasizes that we should consider a diagnosis of unilateral pulmonary artery atresia in an adult patient presenting with exertional dyspnoea, haemoptysis or recurrent respiratory infections with asymmetric aeration of lung on radiography.

Conclusions

UPAA is one of the disease entities to be considered while dealing with cases of dyspnoea, haemoptysis and recurrent pulmonary infections, especially in presence of unilateral pulmonary oligemia and asymmetric lung field lucency. Further, presence of pulmonary tuberculosis in a patient with unilateral pulmonary artery atresia is a very rare clinical entity. Since both pulmonary TB and isolated UPA have haemoptysis as the common presenting feature, we need to be aware of both the disease entities to be able to diagnose the culprit pathology.

Similarly, the present case also emphasizes the fact that the physicians managing the cases of unilateral pulmonary atresia should have high index of suspicion for pulmonary tuberculosis also, especially in high burden countries like India.

References