

Case report:

**Left pulmonary artery agenesis with pulmonary hypoplasia in an elderly patient:
a rare case report**

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Abstract

Proximal interruption of the unilateral pulmonary artery is a rare congenital anomaly, which is often associated with other cardiovascular abnormalities. It is usually diagnosed in children but rarely discovered in adulthood as an isolated phenomenon, occurring more frequently on the right side and is often associated with a contralateral aortic arch. We are presenting a rare case of a sixty year old male who was diagnosed with left lung hypoplasia due to proximal interruption of left pulmonary artery with left sided aortic arch without any associated cardiovascular anomalies.

Key words:

unilateral pulmonary artery agenesis, pulmonary hypoplasia, pulmonary angiography.

Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare entity with an incidence of about one in 200,000 individuals¹ and it is often associated with other developmental anomalies of cardiovascular system. UPAA is commoner on right side¹ and is usually associated with contralateral aortic arch². Although several hypotheses like chromosomal anomaly, vitamin A deficiency, intrauterine infection and environmental factors have been proposed³, no definite aetiology has been detected till now. Diagnosis of this condition is usually made during childhood, but it may be delayed till late adulthood, especially in asymptomatic patients. We hereby describe a case of left sided pulmonary artery agenesis with hypoplasia of left lung without any cardiological abnormality, which was diagnosed late in his life and had a same-sided aortic arch.

Case Presentation

A thin built sixty year old male patient presented to our pulmonary medicine OPD with complaints of

occasional left sided chest pain over last one year. He had no history of breathlessness, palpitation, cough, fever or haemoptysis. He did not have any significant past history except for occasional respiratory tract infection. He was a non-smoker, non-alcoholic and did not have any co-morbidity. On clinical examination, he was afebrile and vital signs were normal. Chest examination revealed reduced movement of left hemithorax. Trachea was central in position, but apex beat was localized at one inch lateral to mid-clavicular line in left fifth intercostal space. There was localized crepitation over left infrascapular area. Cardiac examination was within normal limit except for location of apex beat. Routine haematological and biochemical profiles revealed no abnormality.

Chest X-ray in postero-anterior view (Fig.1) revealed substantial volume loss in left hemithorax with shifting of lower mediastinum to left. Right lung appeared to be hyperinflated and there was a laterally displaced left anterior junction line.

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Left pulmonary artery agenesis with hypoplastic left lung



Figure 1: Chest X-ray (PA) showing reduced left lung volume with mediastinum shifted to left. Left anterior junction line displaced laterally (indicated by arrow)

Contrast enhanced CT scan of thorax was done for further evaluation and it showed proximal interruption of left pulmonary artery (Fig.2) along with prominent main and right pulmonary artery. There was marked hyperinflation of right lung associated with herniation to the left and bronchiectatic changes were detected in lower lobe of hypoplastic left lung (Fig.3). Aortic arch was located on the left side (Fig.4).



Fig.2: CECT thorax (mediastinal window) showing proximal interruption of left pulmonary artery (indicated by arrow). MPA-main pulmonary artery, RPA-right pulmonary artery

12-lead electrocardiogram showed features of left axis deviation, probably due to shifting of lower mediastinum towards left. Echocardiogram (Fig.5) showed prominent main and right pulmonary artery with absent left pulmonary artery. There was no evidence of any congenital heart disease and mean pulmonary artery pressure was 23mm of Hg.

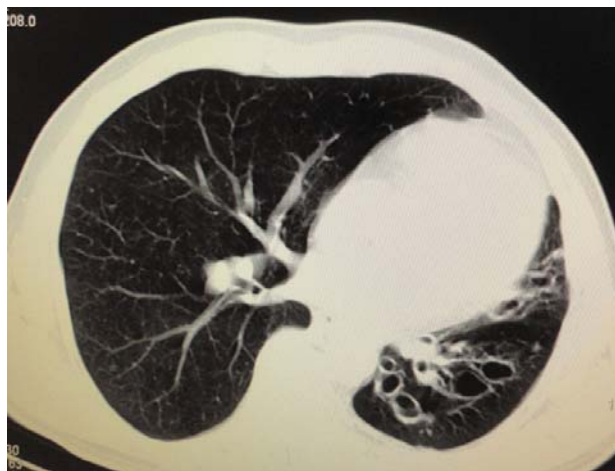


Fig.3: Lung window of CT thorax showing markedly hyperinflated right lung with herniation to left. Left lung is hypoplastic with bronchiectatic changes in lower lobe.



Fig.4: Mediastinal window of CECT thorax showing a left sided aortic arch

Bronchoscopy was done to rule out any obstruction or developmental anomaly of trachea-bronchial tree and it revealed normal carina and bronchi. Ventilation-perfusion scan could not be done due to lack of facility. Spirometry showed a restrictive pattern with FEV1/FVC of 77.3 with FVC of 65% and FEV1 of 70% of predicted value. Subsequent pulmonary angiography (Fig.6) was carried out which confirmed absence of left pulmonary artery.

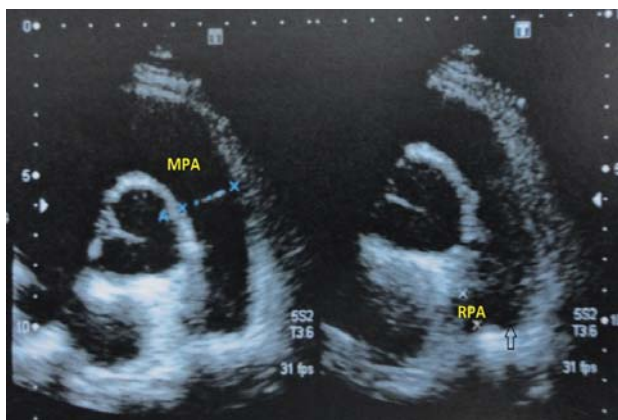


Fig. 5: Echocardiography showing MPA (main pulmonary artery) and RPA (right pulmonary artery). Arrow denoting proximal interruption of left pulmonary artery.

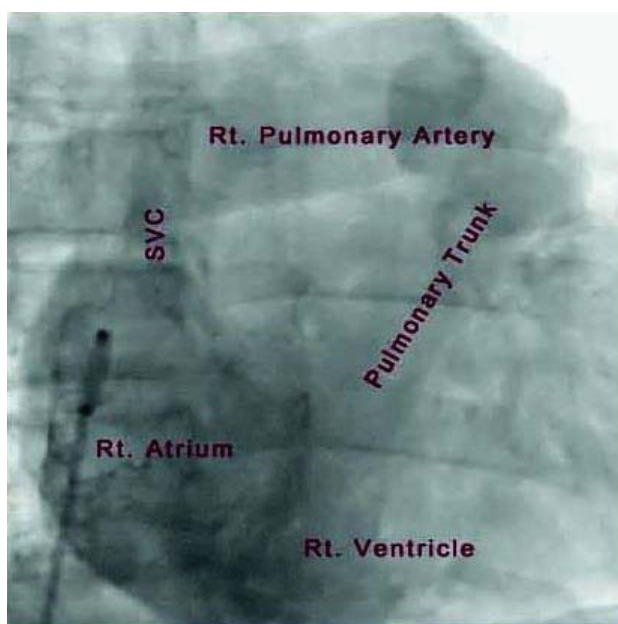


Fig. 6: Pulmonary angiography showing pulmonary trunk and dilated right pulmonary artery. No left pulmonary artery is noted.

Thereby, a final diagnosis of isolated left pulmonary artery agenesis with left pulmonary hypoplasia with left sided aortic arch and left lower lobe bronchiectasis was made. Patient was given symptomatic management and antibiotic, with which he symptomatically improved. He was counseled about the nature of his disease and had been kept under observation.

Discussion

Although unilateral pulmonary artery agenesis was first described by Fraentzel⁴ in 1868, very limited number of cases was detected worldwide since then. It occurs as a result of failure in the connection of sixth aortic arch with pulmonary trunk during embryologic development¹. In a recent review of isolated UPAA, absence of right pulmonary artery was found to be commoner than left (63% vs. 37%)¹ although our patient had an absent left pulmonary artery. Left sided UPAA is more commonly associated with cardiologic abnormalities (like Fallot's tetralogy, septal defect, patent ductus arteriosus etc) and therefore, often requires early diagnosis and surgical repair during initial years of life⁵. On the other hand, isolated UPAA is commoner on right side and such patients often present later because of lack of symptoms. And in most cases, the affected pulmonary artery is on the opposite side of the aortic arch⁶. But above mentioned patient did not have any cardiac anomaly in spite of having a left UPAA and had an ipsilateral aortic arch.

Clinical presentation is widely variable. According to some reports, 30% of such patients are asymptomatic⁷, though Ten Harkel et al¹ found only few asymptomatic patients in their review. Recurrent respiratory tract infection, breathlessness on exertion and decreased exercise tolerance are the commonest mode of presentation. But infrequently patient may present with haemoptysis, necrotizing bronchopneumonia, congestive heart failure or features of pulmonary hypertension. Median age of presentation is 14 years¹. Diagnosis of UPAA becomes difficult with increasing age and a high index of suspicion, coupled with appropriate investigation is required to detect this condition in adulthood. On examination, patient may have features of reduced hemithoracic volume with ipsilateral mediastinal shift⁷.

Chest X-ray in PA and lateral view usually show decreased size of affected hemithorax, elevation of ipsilateral diaphragm, absent ipsilateral hilum and an ipsilateral mediastinal shift. Ventilation-perfusion scan, CT angiography, MRI and digital subtraction angiography (DSA) are usually sufficient to make the diagnosis. In our case, CECT clearly showed absence of left pulmonary artery which was later confirmed by pulmonary angiography. Bronchiectatic changes are often detectable in

affected lung explaining recurrent respiratory infection. Pulmonary angiography is considered to be gold standard for diagnosis of UPAA, but it is often not required unless surgical approach has been considered. Echocardiography should be done routinely to evaluate pulmonary hypertension and to rule out cardiac abnormality.

Radiologically, Swyer-James syndrome (or McLeod's syndrome) is the principal differential diagnosis in which affected hemithorax may not be small and there is a classical delay in the wash out phase of ventilation-perfusion scan. Other differential diagnoses like pulmonary artery obstruction due to thromboembolism, unilateral pulmonary vein atresia, fibrosing mediastinitis and primary neoplasm of pulmonary artery should be kept in mind.

Asymptomatic patients with isolated UPAA require close monitoring to detect early development of pulmonary hypertension. They should be informed about predisposing factors like high altitude pulmonary oedema (HAPE) and pregnancy which may unmask pulmonary hypertension¹. But sometimes selective vascular Embolization or even pneumonec-

tomy is required to manage massive haemoptysis from excessive collateral circulation through bronchial, intercostal, subclavian or subdiaphragmatic arteries⁸. Correction of congenital cardiac anomaly should be done early in life. Revascularization can be an effective prevention against haemoptysis or pulmonary hypertension in patients who have reconstructable pulmonary artery and reparable lung⁹⁻¹⁰. Heart-lung transplantation may be an option to simultaneously improve blood oxygenation¹¹. Medical therapy for pulmonary hypertension with calcium channel blocker or intravenous prostaglandin infusion may be tried if surgery cannot be undertaken.

Conclusion

UPAA should be suspected in presence of recurrent respiratory infection, haemoptysis or pulmonary hypertension even in older population when no other definite aetiology is detected. In childhood, early diagnosis of this entity can avert serious complication. A high index of suspicion and judicious use of a battery of imaging techniques can lead to the appropriate diagnosis.

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