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Case Report

A Case Report of Right Pulmonary Artery Agenesis Presentation in an Adult Female

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ABSTRACT
Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly resulting from maldevelopment of the sixth aortic arch. Its presentation in adulthood, particularly in the absence of associated congenital cardiac anomalies, is uncommon and often poses a diagnostic challenge.

INTRODUCTION

Unilateral absence of the pulmonary artery or unilateral pulmonary artery agenesis (UPAA) is a rare congenital disorder presenting with a wide spectrum of symptoms. The prevalence of isolated UAPA is estimated to be around 1 in 2,00,000 individuals.(1) Only a few cases of unilateral pulmonary artery agenesis have been reported from India. Due to its rarity, it remains a diagnostic and therapeutic challenge in adults. The clinical presentation is variable ranging from infants, where they usually present with congestive cardiac failure and pulmonary hypertension to(2) Often in association with other cardiac defects, such as tetralogy of Fallot or septal defects, but it can occur in isolation. Other presentation in adults may be asymptomatic for many years and even throughout their lives with the disease being detected by chance when chest radiography is performed or may have symptoms like recurrent pulmonary infections, decreased exercise tolerance and shortness of breath on exertion, hemoptysis. And a few patients may present with respiratory distress and heart failure.(3) The final diagnosis is based on imaging. Chest X-rays show an ipsilateral contracted lung with ipsilateral mediastinal shift and hyperinflation of the contralateral lung due to compensatory emphysema that may be accompanied by herniation towards the affected side.(4) This is a case report of the rare disease of pulmonary agenesis presenting as pneumonia in an adult in our institute.

Case Report

A 60 year old woman, farmer by occupation, with history of systemic hypertension on medication from 7 years presented with complaints of cough for 20 days aggravated during night and on lying down on left side and reduced on lying towards right side, cough was assosiated with scanty yellowish sputum non blood tinged and non-foul smelling, cough partially relieved on medication, (details of medication unknown) associated with shortness of breath for 20 days gradually progressive from MMRC grade 1 to 2, not assosiated with wheeze or orthopnea, aggravated on walking up and down the stairs and on walking distances, relieved with rest. For these symptoms patient took medications prescribed by a local doctor but presented to us as she developed occasional episodes of high grade fever with chills and rigor for 2 days with aggravation of respiratory complaints. Patient had history of pneumonia 2 years ago and had no other hospitalization history, no significant family, birth history. On arrival vitals were Bp 130/80 mmHg, Pulse rate of 160 bpm, irregularly irregular pulse felt, RR 18 cpm, spo2 95 % on room air temp 98 F. general examination was normal. On respiratory system examination showed trachea deviation to right side, with drooping of right shoulder, right supra and infraclavicular hollowing and decreased movements of right hemithorax. On measurements right spino scapular distance and right spino acromian distance was reduced, right hemithorax measurement was reduced compared to left side, vocal fremitus was increased in right infra axillary, infra scapular, inter scapular areas, on auscultation right sides coarse crepitations heard in infra axillary, axillary, infra scapular and inter scapular areas, vocal resonance increased in corresponding areas and right infraclavicular area. A provisional diagnosis of right lung collapse

consolidation was made and investigations were done.

Ecg showed irregularly irregular rhythm with no p waves atrial fibrillation was present and which was reverted on antiarrythmics. Chest X ray showed deviation of trachea to right, opacities notes in right upper and lower zones which suggested infective etiology. HRCT chest was done and reported as multifocal areas of pathy ground glass opacities and nodules seen in right upper and right lower lobes, volume loss collapse consolidation with cystic bronchiectatic changes in apical segment of right upper lobe, fairly defined subpleural /peri fissural/ intraparenchymal soft tissue attenuation with speculated margin and suggested for further evaluation. CECT chest was done which was reported as congenital right pulmonary artery agenesis with hypertrophied intercostal and bronchial arteries supplying the right lung. ECHO showed dilated left atrium and moderate MR with normal ejection fraction. Other blood investigations were normal.

Patient was treated as community acquired pneumonia and symptoms relieved. Planned for further work up of solitary pulmonary nodule with PET-CT scan, but patient was not willing hence was discharged.

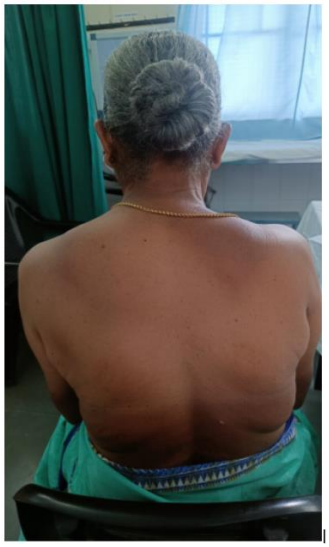

Figure 1. Right Drooping Of Shoulder and Volume Loss of Lung on Right Side

DISCUSSION

Normally During embryonic development, there are 6 Aortic arches each arch develops to give rise to various arteries.

Arch	Derivatives
1st Aortic Arch	Maxillary Arteries
2nd Aortic Arch	Hyoid and stapedial arteries
3rd Aortic Arch	Common carotid and first part of internal carotid arteries
4th Aortic Arch -left side	Arch of aorta from the left common carotid to left subclavian arteries
4th Aortic Arch - right side	Right subclavian artery proximal portion
5th Aortic Arch	Regresses
6th Aortic Arch left side	Left pulmonary artery and ductus arteriosus
6th Aortic Arch on right side	Right pulmonary artery

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 sixth aortic arch forms on each side of the heart. The proximal portion of this arch normally develops into the main pulmonary artery, while the distal portion contributes to the ductus arteriosus. In unilateral pulmonary artery agenesis, the proximal portion of the sixth aortic arch on the affected side fails to connect with the pulmonary trunk. This failure results in the absence of the main pulmonary artery on that side. Embryologically, the branch pulmonary arteries arise from the ventral aspect of the sixth aortic arches, and the intraparenchymal

arterial system originates from the primitive lung buds. If maldevelopment of the sixth aortic arch occurs on the affected side, it will lead to total absence of the branch of pulmonary artery giving rise to UPAA. Normally the lung buds are supplied by arterial branches from the dorsal aorta, however in UPAA, as the pulmonary artery fails to develop, these arteries persist as collaterals from aorta, subclavian arteries and intercostal arteries. The foetal development of the lungs and the associated broncho vascular anatomy are usually normal, since the hilar and distal pulmonary arteries and their branches are supplied by an ipsilateral patent ductus arteriosus, although they are not connected to the pulmonary trunk. After birth, closure of the ductus arteriosus leads to a reduction in blood flow which in turn results in pulmonary hypoplasia and the development of aortopulmonary collateral vessels. These are the reasons why no anomalies will be detected in prenatal ultrasounds and pulmonary and vascular manifestations developed after birth.[5] Left-sided pulmonary artery agenesis is more likely to develop with a concomitant congenital cardiac defect such as Tetralogy of Fallot, Atrial septal defect, Coarctation of aorta and Truncus arteriosus. This explains why left sided UPAA patients present with cardiovascular symptoms during childhood. Conversely, right sided UPAA without cardiovascular anomalies presents later in adulthood with a milder clinical course.[6] Clinical features are classified into three groups, depending on the clinical presentation in group I, a left-to-right shunt develops and is diagnosed in childhood. In group II, severe pulmonary hypertension occurs and these patients generally die in the first few months of life. Group III consists of adults with few manifestations as our patient.[7] Absence of pulmonary artery leads to excessive collateral circulation. Blood supply to the affected lung comes from bronchial arteries, aortopulmonary arteries and other systemic arteries such as the intercostal and trans pleural vessels. These vessels produce numerous vascular channels in the submucosa of the bronchial walls, which may enlarge and rupture. As a result, hemoptysis can be severe and persistent.[8]

Radiology

Chest X ray shows findings of cardiac and mediastinal displacement, absence of the pulmonary artery shadow on the affected side, an ipsilateral elevation of the diaphragm and mediastinum shift, a contralateral compensatory hyperinflation of the lung and herniation across the midline.[9] **Ventilation-perfusion scintigraphy** elucidates the anomaly by showing absence or reduction in perfusion on the affected side with normal ventilation.[10] **Contrast enhanced chest CT** is adequate for UPAA diagnosis, limiting the use of more invasive techniques. Parenchymal findings include bronchiectasis and mosaic attenuation pattern in both lungs, possibly caused by an increased perfusion of the unaffected lung, by the development of PH or by a compensatory over-inflation of the unaffected lung. In addition, **Chest CT** provides useful information regarding PH or congenital heart defects and perfusion of lung parenchyma.[11] **Pulmonary angiography and digital subtraction angiography** are the gold standard to establish a definitive diagnosis and identify the collateral blood flow to the affected lung[1] Conventional angiography is reserved for patients requiring embolization or revascularization surgery

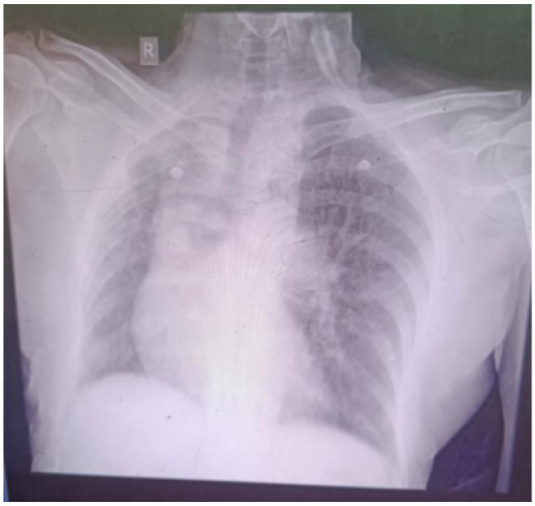

Figure 2. Chest X Ray Showing Tracheal Shift, Right Hemi-Diaphragm Elevation

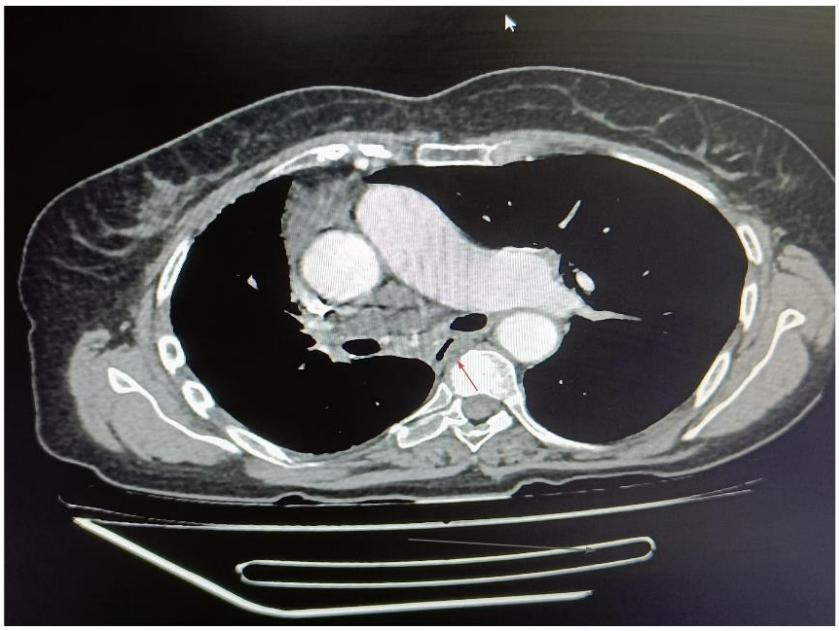

Figure 3. CECT Image Showing Absent Right Pulmonary Artery, Arrow Mark Showing Presence Of Left Pulmonary Artery

Management

Pneumonectomy and surgical revascularization, selective embolization of systemic arteries, and pharmacological treatment for pulmonary hypertension are optional treatments for UPAA.[12] Surgical treatment for UPAA is considered for patients in whom associated anomalies are present or for those who are symptomatic. However, there are currently no established guidelines for the treatment approach in these patients.

Complications

1. There is a possibility of the occurrence of lung cancer with UPAA that chronic hypoxia may cause DNA damages. Chronic hypoxia may result in the release of reactive oxygen species (ROS), induction of hypoxia-inducible factor-1(HIF-1) and p53, and induction of cell proliferation, leading to cancer.(13)
2. Massive hemoptysis is an another complication. Although many patients with UPAA can be asymptomatic for a considerable time. The development of pulmonary hemorrhage or pulmonary hypertension can impact their long-term survival.
3. The hypoplasia of lung tissue can be secondary to decreased lung perfusion which is classified as secondary lung hypoplasia. It exhibits various pathologies such as bronchiectasis, interstitial fibrosis, cystic changes resulting in multiple bullae formation, especially in the later part of life, resulting in recurrent respiratory infections. The basic pathology is reduced lung perfusion leading to decreased pulmonary parenchymal development, impaired ciliary function, mucus trapping, and chronic bronchitis.(14)
4. The causes of death comprise sudden cardiac death, right heart failure, respiratory failure, severe pulmonary hemorrhage, and high-altitude pulmonary edema. Massive hemoptysis is also one of the main causes of death in patients with UPAA.

Differential diagnosis

Various clinical conditions such as Swyer-James syndrome, unilateral emphysema, lobar atelectasis, bronchiectasis, post-lobectomy status, chronic pulmonary thromboembolism, and agenesis of the lung resemble UPAA.(15)

1. **Swyer-James Syndrome**: It is a rare abnormality characterized by a hyperlucent lobe or lung and by air trapping during expiration. There is substantial evidence of viral bronchiolitis causing the condition. It shows air trapping on radiographs or CT scans at the end of maximum expiration.(16)
2. **Chronic Pulmonary Thromboembolism**: History of deep vein thrombosis and CT pulmonary angiography shows an obvious depiction of thrombotic occlusion from the clinical and radiological perspectives, although it can often resemble UPAA in adulthood.(17)

CONCLUSION

Not all congenital heart diseases manifests with symptoms and signs in childhood or infancy some defects may go unnoticed in prenatal scans and can present in adults masquerading as respiratory complaints and few cases may go undiagnosed in life. This case highlights the presentation of a cardiac defect such as right pulmonary artery agenesis presenting with recurrant respiratory symptoms in an adult and the treating physician may likely miss the underlying anomaly. The importance of clinical examination and radiological investigations should be recognised in diagnosing such diseases and helps guiding an appropriate treatment to reduce mortality.

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