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Unilateral Agenesis of Pulmonary Artery: A Rare Entity to be Known

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Abstract

Unilateral Agenesis of Pulmonary Artery (UAPA) is rare, with a prevalence of 1: 200,000 in young adults and usually occurs in conjunction with cardiovascular anomalies. We present a case of a 31 year old man with no underlying co-morbidity who presented with a 7 month history of dyspnea and recurrent chest infections and was later diagnosed with isolated unilateral agenesis of pulmonary artery. Our case alerts the physicians and radiologists about this rare entity as a cause of recurrent infections in young adults.

Introduction

Agenesis of pulmonary artery is an uncommon congenital abnormality and results from an abnormality of the 6th aortic arch [1,2]. Unilateral Absence of Pulmonary Artery (UAPA) can have varied presentations. The age of presentation is usually teen age with a median of 14 years as reviewed in 108 cases in a previous study [3]. We present a brief review of the symptomatology, diagnosis and treatment of isolated UAPA.

Case report

A 31-year old male with no underlying comorbidity was referred to us with a history of exercise intolerance for the past seven months which had progressed to breathlessness at rest. The patient had previous admissions to the hospital with lower respiratory tract infections. The vitals and laboratory investigations were normal. On auscultation loud P2 and systolic murmur heard all over the chest more on the right side. Postero-anterior

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chest X-ray revealed hyperlucent left lung field with decreased volume. Left hilum was less appreciable with slightly elevated left hemi diaphragm (Figure 1). Echocardiography revealed hypertrophied right ventricle with tricuspid regurgitation with non-visualization of Left Pulmonary Artery (LPA). CT Pulmonary Angiography (CTPA) done on CT SOMATOM SENSATION 64 revealed absence of LPA with evidence of paucity of vessels on the left side compared to right (Figure 2) with hypertrophied left bronchial (Figure 3) and left posterior intercostal arteries (Figure 4). Systemic supply to the left lung was also seen arising from the left subclavian artery (Figure 5). Right Pulmonary Artery (RPA) just proximal to bifurcation at hilum showed 50% stenosis (Figure 2). However, RPA and its segmental and sub segmental branches revealed normal contrast opacification. Catheter pulmonary angiography revealed absent LPA with moderate proximal RPA stenosis. Descending Thoracic Aorta angiography revealed hypertrophied bronchial and intercostals arteries supplying the left lung. The patient was put on medical management (endothelin receptor antagonist) for Pulmonary Arterial Hypertension (PAH) after cardiac catheterization revealed raised pulmonary artery pressure and is presently doing well.

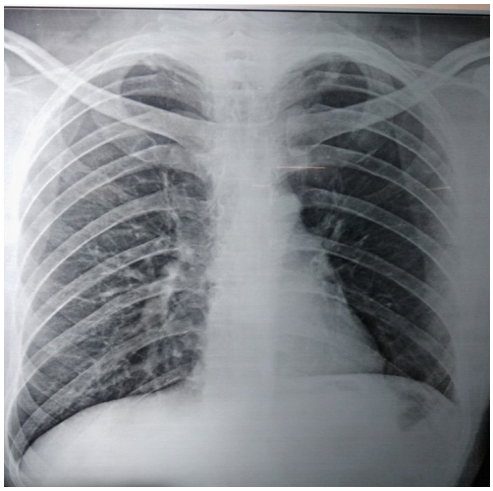

Figure 1: Postero-Anterior chest X-ray revealing hyperlucent left lung field with decreased volume. Left hilum was less appreciable with slightly elevated left hemi diaphragm.

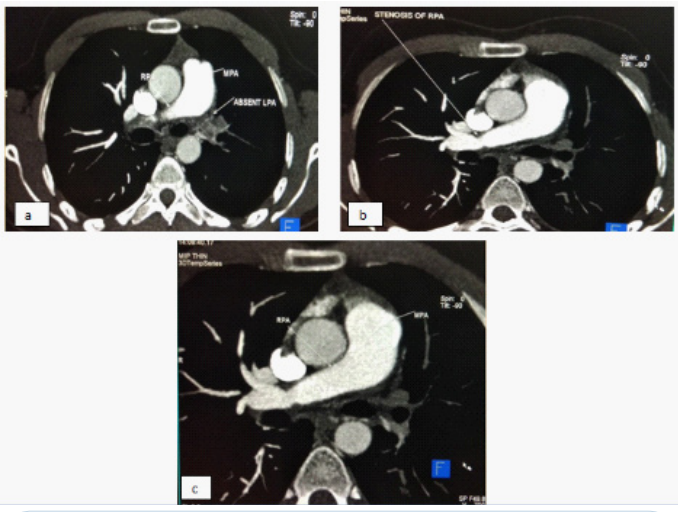

Figure 2: CT Pulmonary Angiography (CTPA) axial images at the level of pulmonary bifurcation revealing absence of LPA (arrow in a). RPA is seen and is normal at origin with a stenosis just distal to its origin (b and c).

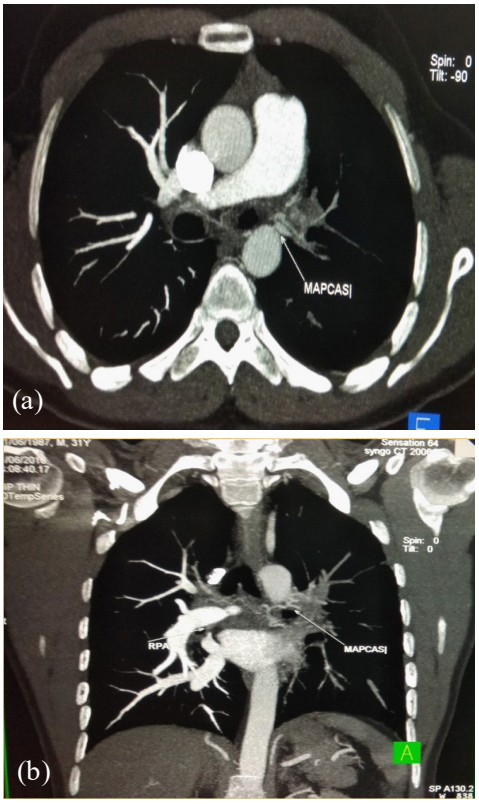

Figure 3: CT Pulmonary Angiography (CTPA) MIP axial **(a)** and coronal **(b)** images revealing absence of LPA and multiple MAPCAS entering the left hilum and supplying the left lung. MAPCAS - Major Aorto Pulmonary Collateral Arteries.

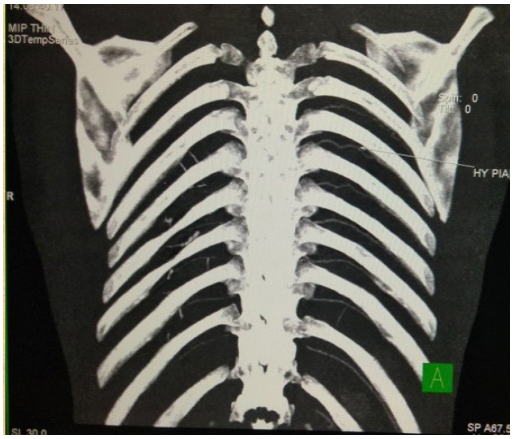

Figure 4: CT Pulmonary Angiography (CTPA) MIP coronal images revealing hypertrophied left posterior intercostal arteries in comparison to the right side serving as systemic supply to the left lung.

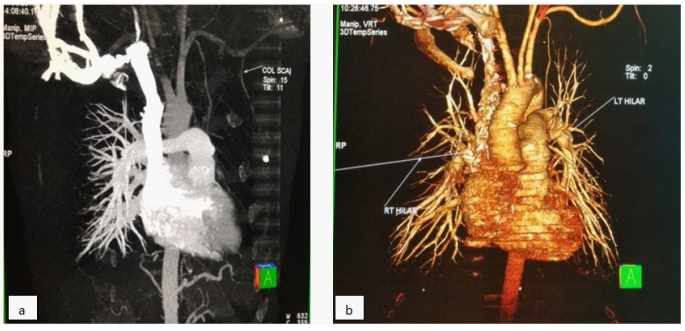

Figure 5: Coronal MIP **(a)** and VRT **(b)** images showing asymmetry of hila secondary to absence of left pulmonary artery. Also systemic supply to the left lung from left subclavian artery is also noted (arrow in a).

Declarations

Acknowledgement

Department of General Medicine, Sheri Kashmir institute of medical sciences.

Ethics approval and consent to participate

Our study was an observational study with no requirement for ethical clearance in our institution. The consent from the patients was however taken in all the cases.

Discussion

UAPA is an entity known from the late 1800's and reported subsequently in many patients [4,5]. These patients may have an asymptomatic course into adulthood but some may present with asthma or recurrent chest infections requiring medical management. Congestive heart failure and breathlessness with activity are some other common presenting features [3,6]. The diagnosis of UAPA on clinical grounds is very difficult; however a routine chest radiograph may provide some information guiding us towards the diagnosis. The chest radiograph usually shows asymmetrical lung zones on the two sides with the affected side being small and hyperlucent owing to lack of sufficient vasculature [6,7]. The diagnosis can be confirmed by cross sectional imaging in the form of Computed Tomography, Magnetic Resonance Imaging or Transthoracic Echocardiography. Absence of pulmonary artery on these modalities is diagnosed when the vessel is not visualized or terminates within 1 cm of its origin from MPA [6].

The management of this entity is debatable but in most cases a combination of surgery in the form of lung resection (total or partial), surgical or endovascular closure of Major Aorto Pulmonary Collaterals (MAPCAS) except when they are the only vessels supplying the lung. In cases were distal vasculature is well developed and anastomosis is possible, a one time or staged anastomosis can also be done [8]. In our case option of RPA stenting was considered; however it was thought that RPA stenting may lead to right lung overflow and consequently right lung hypertension. RPA stenosis was therefore protective to the distal right lung and hence not stented.

On follow up, with compliant medical treatment, the patient is presently doing well with some tolerance to exertion. Breathlessness during routine work has settled and quality of life is somewhat better.

Conclusion

UAPA is frequently associated with cardiac anomalies. Cross sectional imaging is a sine qua- non in the diagnosis of UAPA and help identify associated anomalies. Recurrent chest infections should prompt a physician to consider UAPA as a differential especially if chest radiograph is suggestive. UAPA can go un-noticed till adulthood with no associated anomalies. The mortality associated with UAPA is mostly secondary to development of pulmonary artery hypertension and its effects.

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