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Two Cases of Incidentally Picked Up Adult Unilateral Pulmonary Artery Atresia with Variable Imaging Features

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Abstract: *Unilateral absence of pulmonary artery is a rare congenital disorder, a variant of pulmonary atresia, can either be isolated or associated with other congenital cardiac anomalies. Adult patients with isolated UAPAA are usually asymptomatic and incidentally diagnosed when presenting with other thoracic complaints such as chest pain, dyspnea, etc…Here we present two cases of isolated unilateral right pulmonary artery atresia who presented with chest pain, dyspnea referred for CT scan suspecting infective causes.*

Keywords: *Unilateral pulmonary artery atresia, UPAA, Unilateral absence of the pulmonary artery, UAPA, Major aortopulmonary collateral arteries*

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I. Introduction

Unilateral pulmonary artery atresia or unilateral absence of the pulmonary artery is a rare congenital anomaly, which is actually a variant of pulmonary artery atresia. Frentzel first described it in 1868. It can either be isolated or sometimes even associated with septal defects or other cardiac anomalies such as tetralogy of fallot, right sided aortic arch, coarctation of aorta etc.

II. Case Report

Case 1 is a 43-year-old male patient, presented with complaints of dyspnea and chest pain, initially evaluated by a pulmonologist, further referred for a CT Thorax plain study. On identifying the narrow caliber and tapering of right pulmonary artery we suggested CT pulmonary angiogram and eventually diagnosed isolated UPAA.

2.1 Imaging Findings:

Only a stump of the proximal right pulmonary artery was visualized and appeared extremely narrow in caliber (7mm) with no contrast opacification in the pulmonary arterial phase. In the aortographic phase, multiple aorto-pulmonary collaterals were seen arising from the aorta, subclavian arteries, intercostal arteries and lumbar arteries partially reconstituting the right pulmonary artery, the lobar and segmental branches.

Case 2 is another 34-year-old male who presented with fever and occasional dyspnea was referred for HRCT and on noticing the unilateral absence of right main pulmonary artery, we further recommended CECT but due to preexisting renal insufficiency, the contrast examination could not be performed.

2.2 Imaging Findings:

In this case, the right pulmonary artery was absent right from the origin and the collaterals were mainly from intercostal arteries suggestive of proximal interruption of right pulmonary artery.Peribronchovascular interstitial thickening was seen in the right lung with nodular fissural thickening and diffuse pleural thickening. There was also volume loss in the right lung with compensatory hyperinflation and transthoracic herniation of left lung.

III. Discussion

Unilateral pulmonary artery atresia or unilateral absence of the pulmonary artery is a rare congenital anomaly, which is actually a variant of pulmonary artery atresia. Frentzel first described it in 1868. It can either be isolated or sometimes even associated with septal defects or other cardiac anomalies such as tetralogy of fallot, right sided aortic arch, coarctation of aorta etc.

3.1 Etiology

The prevalence of isolated UAPA without associated cardiac anomalies ranged from 1 in 200,000 to 1 in 300,000 adults.

3.2 Embryology

The main pulmonary artery is derived from the trunco-aortic sac while the intrapulmonary pulmonary arteries arise from the lung buds, the extrapulmonary pulmonary arteries arise from the proximal portion of the sixth aortic arch. Involution of the proximal sixth aortic arch along with persistent connection of the intrapulmonary PA to the distal sixth aortic arch leads to the absence of pulmonary artery. It is also been noted that every reported case of absent pulmonary artery had a ductus arteriosus or ligamentum arteriosus ipsilateral to the absent PA.The affected lung usually receives collaterals from bronchial arteries but have also been documented to arise from aortal, intercostal, subdiaphragmatic, subclavian and even coronary arteries.

3.3Clinical symptoms and their Pathophysiology

Infants usually present with congestive cardiac failure and PHT. Adult patients with UAPA are often asymptomatic and they may present with exercise intolerance (18–40%), haemoptysis (20%) or are incidentally detected during chest radiography [2]. Another study by Ten Harkel et al. [3] found that PHT was present in 44%, haemoptysis in 20%, recurrent pulmonary infections in 37% and limited exercise tolerance in 40% of patients with isolated UAPA. Recurrent pulmonary infections, decreased exercise tolerance and mild dyspnoea during exertion are the most common symptoms.

The aetiology of recurrent infections in patients with UAPA is likely to be due to the reduced arterial blood flow to the affected lung, which may result in affected lungs receiving reduced inflammatory cells.Poor blood flow to the affected lung may also result in alveolar hypocapnia, leading to secondary bronchoconstriction and mucous trapping.Haemoptysis in patients with UAPA is due to excessive collateral circulation.

PHT may result from blood flow directed away from the absent PA to the remaining PA. Increased blood flow in the contralateral PA leads to shear stress within the endothelium, with subsequent release of vasoconstrictive compounds, such as endothelin. Chronic vasoconstriction of the pulmonary arterioles may lead to remodelling, resulting in increased resistance of the pulmonary vasculature and PHT.

IV. Treatment

These patients are usually treated based on the presenting symptoms, associated cardiovascular anomalies and pulmonary hypertension (PHT).

Figures

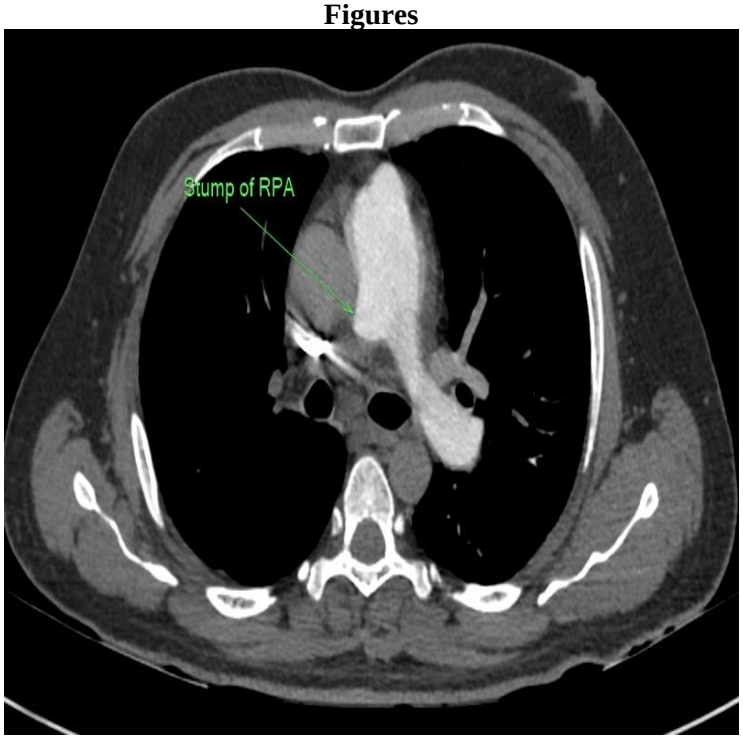

Figure 1: 43 year old male with unilateral absence of right pulmonary artery. Axial CT section of pulmonary angiogram demonstrating extremely narrow caliber of right pulmonary artery.

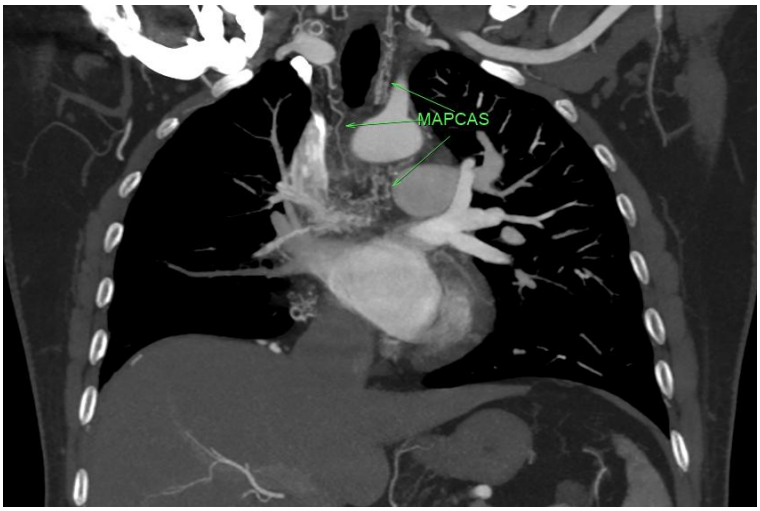

Figure 2: 43 year old male with unilateral absence of right pulmonary artery. Coronal MIP reformat of the pulmonary angiogram demonstrating collaterals.

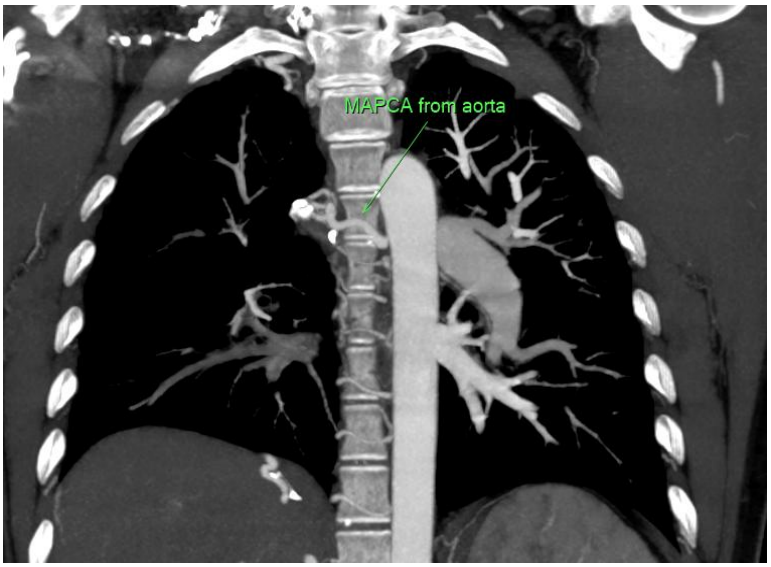

Figure 3: 43 year old male with unilateral absence of right pulmonary artery. Coronal MIP reformat of the pulmonary angiogram demonstrating collaterals.

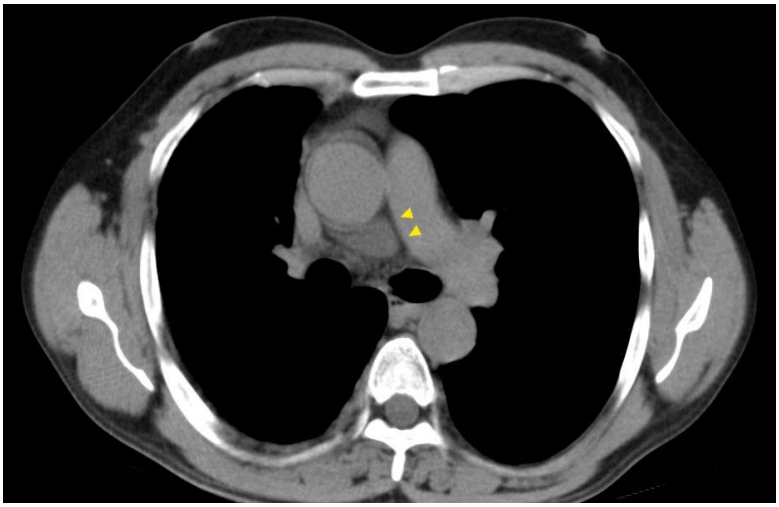

Figure 4: 34 year old male with proximal interruption of right pulmonary artery Axial plain CT of the thorax demonstrates absence of right pulmonary artery.

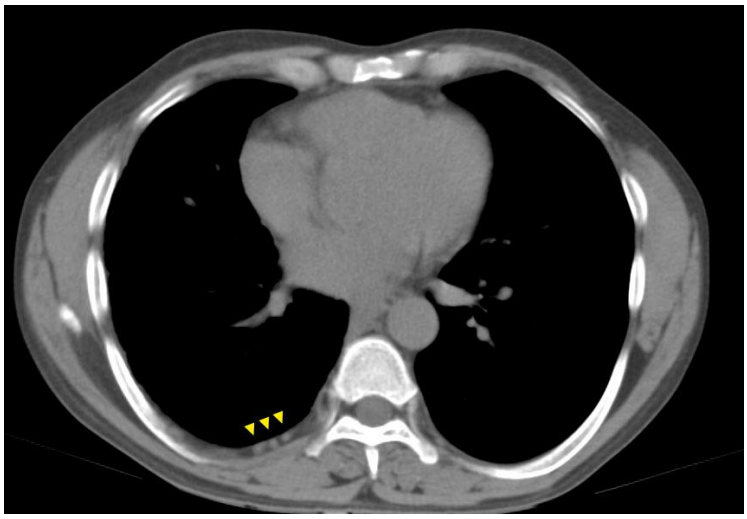

Figure 5: 34 year old male with proximal interruption of right pulmonary artery Axial plain CT of the thorax demonstrates right posterior pleural thickening, with few prominent vessels suggestive of collaterals from intercostal arteries.

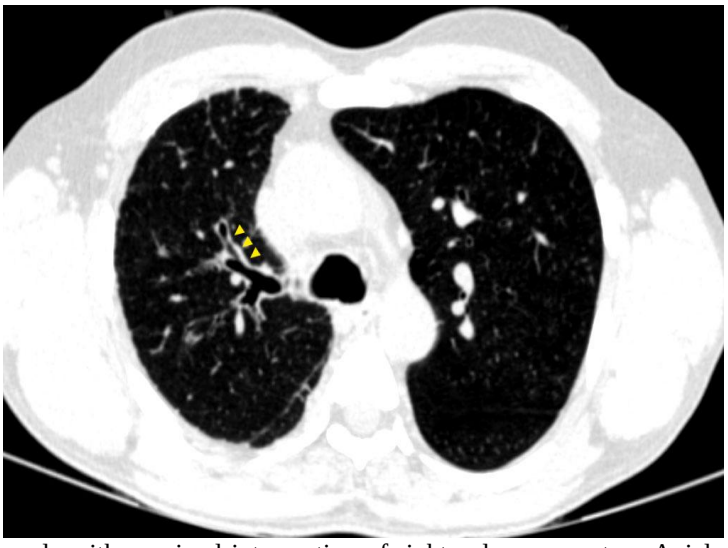

Figure 6: 34 year old male with proximal interruption of right pulmonary artery Axial plain CT of the thorax showing mild peribronchial cuffing.

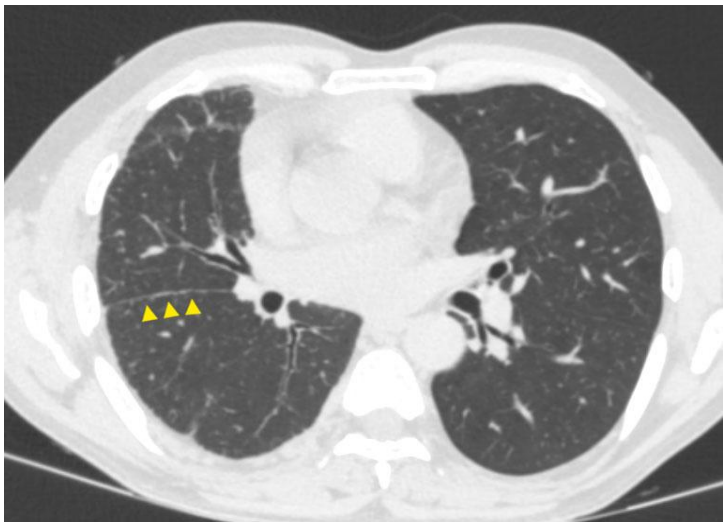

Figure 7: 34 year old male with proximal interruption of right pulmonary artery

Axial plain CT of the thorax showing nodular fissural thickening in the right lung. There is volume loss in the right lung with Subpleural fibrotic strands and compensatory hyperinflation of left lung.

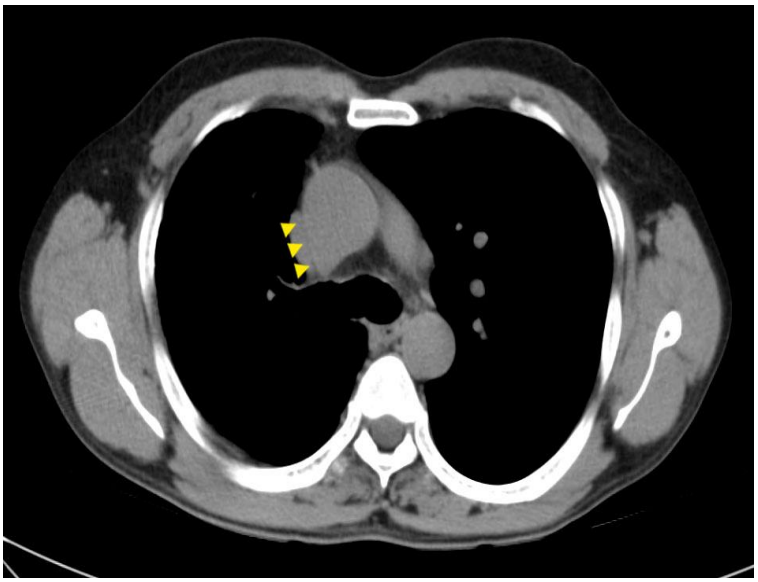

Figure 8: 34 year old male with proximal interruption of right pulmonary artery Axial plain CT of the thorax showing irregular contour of ascending thoracic aorta.

V. Conclusion

Incidental congenital unilateral anomalies of pulmonary arteries with variable clinical and radiological presentations in the absence of associated cardiac manifestations in middle-aged population is not uncommon.

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