Case Report

Unilateral absence of pulmonary artery: a radiographically occult cause of life-threatening hemoptysis

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ABSTRACT

Unilateral absence or agenesis of pulmonary artery (UAPA) is a rare congenital abnormality with an estimated prevelance of 1 in 2,00,000 adults. The entity occurs commonly in association with other congenital heart diseases like septal defects or patent ductus arteriosus. The condition usually runs a benign clinical course with patients usually presenting clinically in adulthood with history of recurrent respiratory tract infections. Two such patients presented with recurrent respiratory tract infections, breathlessness and hemoptysis. The chest radiograph of first patient was reported as normal in the referring hospital, while that of second patient showed volume loss in left lung. CT Pulmonary Angiography (CTPA) was then performed which demonstrated the absence of right and left pulmonary arteries respectively in the first and second patients. Pulmonary artery branches were reformed distally by multiple collaterals arising from systemic arteries. The entire spectrum, including embryology, imaging features and management of UAPA are discussed. UAPA remains a potential cause for life-threatening hemoptysis, due to extensive collateralization associated with the condition. It is important for radiologists to be aware of this uncommon entity in order to suspect it on a routine chest radiograph, diagnose it and map associated collaterals on CTPA and also embolize the bleeding collaterals.

Keywords: Computed tomography, Collaterals, Embolization, Hemoptysis, Pulmonary angiography, Unilateral agenesis of pulmonary artery

INTRODUCTION

Congenital anomalies of pulmonary arteries may range from congenital pulmonic valvular stenosis to idiopathic dilatation or anomalous origins of either right or left pulmonary artery.[1,2] Unilateral absence of pulmonary artery (UAPA) is an uncommon congenital abnormality of pulmonary artery associated with development of multiple collaterals to supply the affected lung. The condition gains significance as it predisposes to life-threatening hemoptysis, cardiac and respiratory failure in patients who may otherwise be undiagnosed and treated symptomatically owing to radiographically subtle abnormalities. The objective this article is to make the readers aware of this seemingly innocuous condition and its potential complications and management strategies.

CASE REPORT

We describe two cases of adult UAPA, with their radiological findings.

Case 1

A 20-year old male presented with complaints of three episodes of hemoptysis and recurrent respiratory tract

infections since childhood. Physical examination revealed right parasternal heaves and loud P2 sounds, suggesting pulmonary hypertension. His chest radiograph was reported as normal from the referring hospital. 2D echocardiography performed by the cardiologist revealed pulmonary arterial hypertension with moderate tricuspid regurgitation and was referred to our department for a detailed CT Pulmonary Angiography (CTPA) evaluation. On evaluation of chest radiograph, subtle features of volume loss on right side with ipsilateral tracheal deviation and mildly elevated hemidiaphragm were visualized. Mild oligemia was seen in right lung field. Right hilum appeared small with no definite identifiable right pulmonary artery (Figure 1).


Figure 1: Chest radiograph PA view of the patient showing mild volume loss in right hemithorax with ipsilateral tracheal shift and right hemidiaphragm elevation. Right hilum appears small with non visualisation of right descending pulmonary artery. Subtle difference in the vasculature of two lung fields is observed.

CTPA demonstrated the absence of right pulmonary artery from its origin with marked dilatation of main and left pulmonary artery (Figure 2).
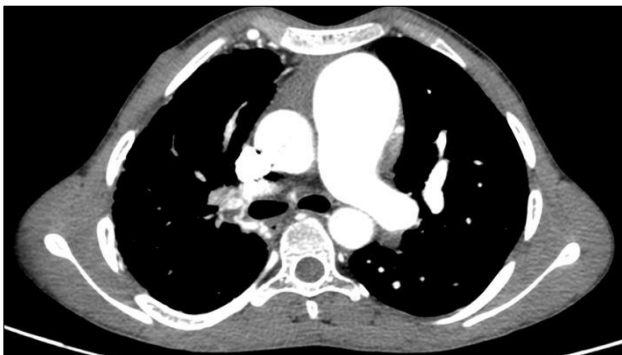
Figure 2: Axial CT Pulmonary angiography depicting absent right pulmonary artery with markedly dilated main and left pulmonary arteries. Multiple collaterals including MAPCAs are seen occupying the region of right pulmonary artery at right hilum.
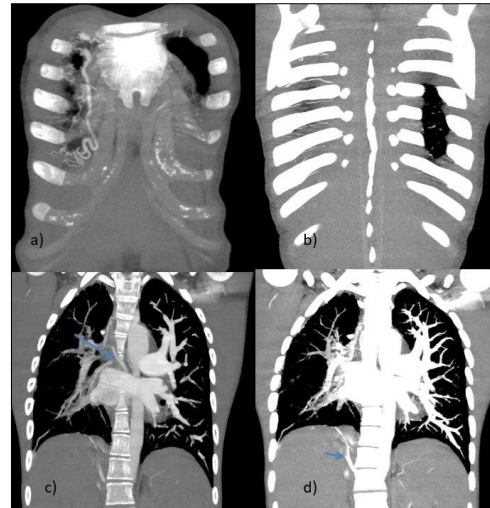

Figure 3: Coronal reconstructions of MIP images of CT Pulmonary angiography illustrate various collateral channels which had opened up to supply right lung as follows: (a) prominent right internal mammary artery; (b) right sided posterior intercostal arteries; (c) enlarged right bronchial artery (long arrow); (d) right inferior phrenic artery (short arrow).
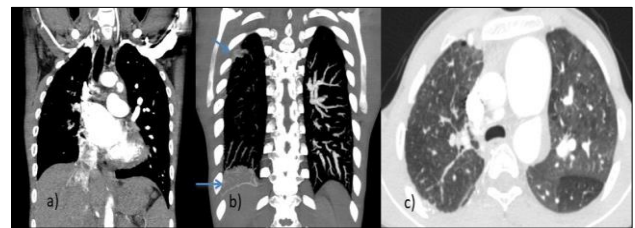

Figure 4: Coronal reconstruction of CTPA (a) revealing asymmetrical lung fields with subtle signs of volume loss on right side in the form of ipsilateral tracheal shift and right hemidiaphragm elevation; MIP images (b) clearly demonstrate the significant decrease in the size of vessels in right lung field along with focal area of 'pseudofibrosis' in apical zone (short arrow) resulting from transpleural collaterals. Subdiaphragmatic collaterals are also seen in the image (long arrow); axial section of lung window (c) reveals mosaic attenuation in both lung parenchyma.

Distal reformation of right pulmonary artery branches through collaterals from bronchial arteries, major aortopulmonary collateral arteries (MAPCAs), intercostal arteries, internal mammary, right subclavian, left gastric, and right inferior phrenic arteries was observed (Figure 3). Some collaterals were seen communicating across the pleura (transpleural collaterals) in right lung apex, giving a 'pseudofibrosis' appearance (Figure 4). Lung field appeared mildly asymmetrical with right lung being smaller than the left, with right-sided tracheal and mediastinal shift and elevation of right hemidiaphragm. Diffuse mosaic attenuation was seen in bilateral lung fields (Figure 4c).

In addition, signs of tricuspid regurgitation including right atrial dilatation with reflux of contrast in inferior vena cava and hepatic veins in arterial phase and right ventricular hypertrophy were observed (Figure 5). The patient also had associated patent ductus arteriosus and pectus carinatum (Figure 6). The patient then underwent elective embolization of bronchial and collateral arteries for management of hemoptysis at another centre.
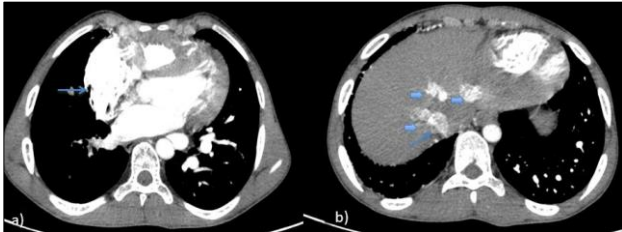
Figure 5: Axial CTPA images demonstrate indirect signs of pulmonary hypertension in the form of right ventricular hypertrophy (a) and functional tricuspid regurgitation depicted by enlarged right atrium (thin arrow) and contrast filling of hepatic veins (thick arrows) (b) and IVC (arrow) in arterial phase.
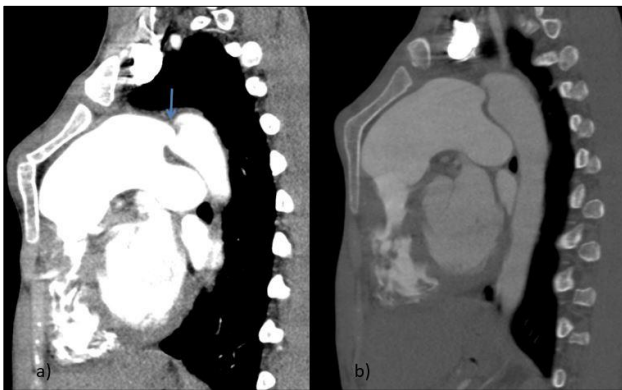
Figure 6: Sagittal reformat of CTPA (a) revealing patent ductus arteriosus (blue arrow). Note the pectus carinatum abnormality on bone window (b), attributable to long standing pulmonary hypertension.

Case 2

A 36-year old male patient presented with complaints of dyspnea since 3 years and recurrent respiratory infections since childhood. One episode of hemoptysis was also present two days before presenting to the department. Physical examination findings were not significant.

Chest radiograph (Figure 7) of the patient demonstrated moderate volume loss in left lung field with ipsilateral deviation of mediastinum and elevated left hemidiaphragm. Areas of cystic bronchiectasis were seen in left lung. Compensatory hyperinflation of right lung was noted with herniation of right lung across midline to the left. Left hilum appeared small with no definitely identifiable shadow of left pulmonary artery. Aortic

knuckle was not seen on left side with a shadow of aortic knuckle and descending thoracic aorta on right side.


Figure 7: Chest radiograph of volume loss in left lung with compensatory hyperinflation of right lung, mediastinal shift towards left side, aortic knuckle and descending thoracic aorta on right side. Multiple cystic radiolucencies representing bronchiectasis are also seen in left lung.
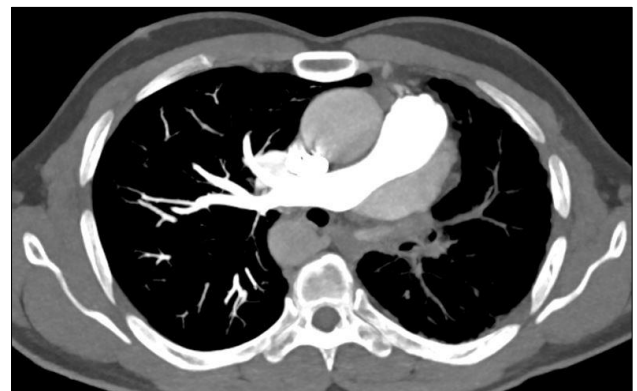

Figure 8: Axial CTPA MIP image of main pulmonary artery and right pulmonary artery with non visualisation of left pulmonary artery.

CTPA demonstrated absent left pulmonary artery from its origin (Figure 8) with attenuated segmental and sub-segmental arteries which showed faint contrast opacification - likely due to distal reformation by multiple collaterals arising from aorta and its branches. Main and right pulmonary arteries were normal in course, caliber and contrast opacification. Aortic arch was seen on right side with mirror image branching pattern (Figure 9). Multiple collaterals were seen originating from left costocervical trunk, left internal mammary artery, inferior phrenic artery and 3rd and 4th left posterior intercostal arteries and anastomosing with intrapulmonary pulmonary artery branches (Figure 9). Left lung showed reduced lung volume and increased attenuation with mediastinal and tracheal shift to left side and left sided rib

crowding. Cystic bronchiectasis was present throughout the left lung with peripheral predominance and involving lower lobe more than the upper lobe. Geographic areas of mosaic attenuation were seen throughout the right lung (Figure 10).
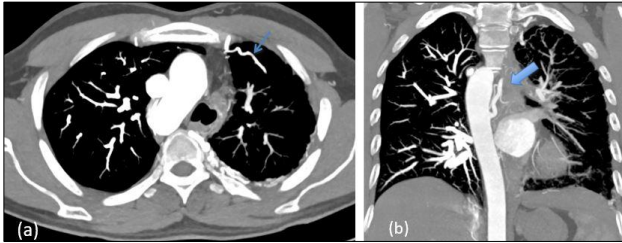
Figure 9: Axial MIP image (a) in mediastinal window showing right sided aortic arch, multiple tortuous dilated collaterals in left sided posterior intercostal spaces and and between left internal mammary arteries and subsegmental pulmonary arteries (thin arrow). Coronal MIP image (b) showing right sided descending thoracic aorta, hypertrophied bronchial artery (thick arrow), decreased contrast enhancement of segmental and subsegmental pulmonary arteries.
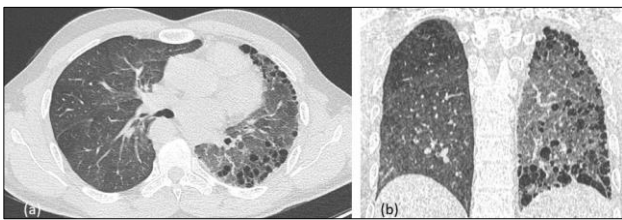
Figure 10: Axial (a) and coronal (b) CT images in lung window showing patchy areas of mosaic attenuation in right lung parenchyma with multiple areas of cystic bronchiectasis in left lung parenchyma.

This patient was managed medically for pulmonary hypertension and was advised regular follow up to look for enlarging size of collaterals or increase in episodes of hemoptysis.

DISCUSSION

UAPA is a rare radiological diagnosis with an estimated prevalence of 1 in 2,00,000 adults.[1] UAPA may occur as an isolated entity or in association with other congenital cardiac conditions like cardiac septal defects, tetrology of Fallot or patent ductus arteriosus.[1] It is commoner on right side, though associated cardiac anomalies are common when left side is affected. The condition usually occurs on the side opposite to the side of aortic arch, except in 2% cases.[3] This dictum has been followed in our two cases where pulmonary artery was absent on the side opposite to that of aortic arch.

The condition arises due to altered development of sixth aortic arch segment, resulting in origin of proximal part of pulmonary artery from ductus arteriosus, which obliterates after birth along with ductal obliteration, thus leading to interruption of pulmonary artery.[1,4]

UAPA causes increased blood flow directed to opposite pulmonary artery, which increases shear stress on endothelial lining and causes endothelin release.[1] This leads to constriction of pulmonary vasculature and resultant increased pulmonary resistance and hypertension. Decreased blood flow to the lung causes reduced delivery of inflammatory mediators, thereby increasing risk of respiratory infections.[1]

Individuals are commonly asymptomatic until adulthood, when they present with hemoptysis, exertional dyspnea, chest pain or recurrent respiratory tract infections.

Bronchoscopic findings include changes of chronic bronchitis or dilated vascular plexus around trachea or bronchi.[3]

Radiographic findings are usually subtle, including mildly asymmetrical lung fields with signs of mediastinal shift towards the smaller lung and ipsilateral elevation of hemidiaphragm. The affected lung field appears hyperlucent with pulmonary oligemia and markedly diminished or absent hilar vasculature on the ipsilateral side.[1] Bronchiectasis or patchy atelectasis may be observed. An appearance termed 'pseudofibrosis' is sometimes seen in the affected lung apex due to the formation of transpleural collateral vessels between peripheral pulmonary arterial branches and systemic arteries. Bronchiectasis was present in case 2, while pseudofibrosis was present in case 1 on the affected side. Occasionally, ipsilateral rib notching may be seen due to dilated intercostal arteries.[2]

Echocardiography aids in detecting pulmonary artery hypertension and associated cardiac anomalies. Ventilation-perfusion scan typically depicts absent perfusion with intact ventilation. However, ventilation may be decreased on affected side.[1]

CTPA is the preferred imaging modality as it clinches the diagnosis and also gives information about associated cardiac anomalies. Vascular, cardiac and parenchymal signs of pulmonary hypertension can easily be assessed. Pulmonary trunk and opposite pulmonary artery are dilated while region of absent pulmonary artery in mediastinum is seen replaced by bronchial arteries or MAPCAs. Other collaterals reforming segmental and subsegmental branches of pulmonary arteries may arise from subclavian, internal mammary, intercostal and sub-diaphragmatic arteries.[1,5] Accurate mapping of MAPCAs and collaterals is critical as they can easily rupture into trachea, bronchi, esophagus or adjacent pulmonary parenchyma and cause massive hemoptysis and death.[6] Communication between coronary and bronchial arteries may be seen in 4% of the cases.[3] Mosaic attenuation without air trapping on expiration is the commonest finding in lungs and is bilateral in 50% cases.[7] Affected

lung field shows diffusely decreased size of vessels. Similar lung findings were seen in our cases. Bronchiectasis, cystic changes, pulmonary hemorrhage and other signs of active and previous lung infections are often seen.[8]

Conventional angiography is the gold standard investigation as it helps in preoperative establishment of collaterals. However, being invasive, it is reserved for patients undergoing embolization procedures.[5]

Overall mortality rate of UAPA is approximately 7% and commonly occurs due to massive pulmonary hemorrhage or hemoptysis due to rupture of collaterals, cardiac and respiratory failure, pulmonary hypertension or high altitude pulmonary edema.[1,9]

Management strategies include medical, surgical and embolization procedures. Vasodilators constitute the medical treatment. Surgical management of MAPCAs include revascularization procedures like unifocalisation, ligation, or both, based on whether the lung is supplied singularly by MAPCAs or by both bronchial arteries and MAPCAs. Unifocalisation involves restoration of normal circulation to lung by redirecting all collaterals into a single vessel at hilum or pulmonary artery.[6] Lobectomy or pneumonectomy may be performed in patients with massive pulmonary hemorrhage.[10] Embolisation of bronchial or non- bronchial systemic arteries is usually done in patients with massive life-threatening hemoptysis, who are poor surgical candidates.[2] The salient reporting points the operating surgeon/interventional radiologist seeks in such cases include the number, origin, course and size of collaterals, with their relation to adjacent bronchial tree, pulmonary veins and esophagus along with any communication with native pulmonary artery.[6]

UAPA is a potential cause of life-threatening hemoptysis, due to extensive collateralization associated with the condition. Role of the radiologist is to suspect it on a routine chest radiograph, diagnose the same and map the different collaterals accurately on CTPA as well as embolize the bleeding collaterals. Hence, radiologists are instrumental not only in the astute diagnosis and preoperative work-up, but also in the prompt management of this potentially serious, underdiagnosed condition.

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