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Absent Unilateral pulmonary artery in an adult. A rare entity

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Abstract: *Unilateral absent pulmonary artery is an uncommon condition and may be associated with other cardiac diseases. Patients without associated cardiac abnormality usually present in adulthood with symptoms of dyspnea, hemoptysis etc. We encountered an adult with absent right pulmonary artery who presented with hemoptysis and on investigations turn out to have absent right pulmonary artery. Considering its rarity, being reported.*
Key word: *Hemoptysis, Unilateral absent pulmonary artery*

I. Introduction

Absence of one or other main pulmonary artery is rare, which may be seen as an isolated condition or in association with other congenital heart defects.[1] This abnormality is a consequence of failure of development of either branch of sixth branchial arch in the embryo[2]. Although this situation occurs equally on both the sides but right pulmonary artery agenesis is more often seen than left. This is because absent left pulmonary artery is often associated with severe congenital cardiac defects. Therefore, patients with absent left pulmonary artery do not survive long enough to report late in life. On the contrary right pulmonary artery agenesis has no strong relationship with congenital cardiac defect; therefore these individuals survive long and are diagnosed late in the life, often without symptoms[3]. We encountered a patient with absent right pulmonary artery at the age of 19. Considering its rarity, hence reported.

II. Case Report

A nineteen year old male presented with history of hemoptysis since four days. There was no cough, expectoration, chest pain, breathlessness, palpitation or syncope. Family history was negative for any congenital heart disease. Patient denied any history of smoking, alcohol or drug abuse. Past medical history was unrewarding.

Physical examination revealed a fair built adult. His vitals were with in normal limits. Clubbing, cyanosis, hepatomegaly & edema feet were absent. Jugular venous pressure was also not raised. Respiratory and cardiovascular examinations were normal except shift of apex beat to medially in fifth intercostal space just lateral to left sternal border.

Electrocardiogram, two dimensional echocardiography & resting arterial blood gas analysis were within normal limits. Spirometry was not performed as patient was having hemoptysis. Chest X ray posteroanterior view (figure 1) showed reduced right lung volume with shift of mediastinum toward same side. Right lung field appeared hazy due to diffuse reticulations in it.

Contrast enhanced computed tomography of thorax in mediastinal window (figure 2a) showed absent right pulmonary artery. Rest of the major mediastinal vessels and trachea appeared normal. Lung window (figure 2b) at level of main bronchi & just above the diaphragm showed increase vascular marking in right lung field giving appearance of diffuse reticulations. Left lung field appeared normal. Diagnosis of absent right pulmonary artery was made and symptomatic treatment was given to the patient. Hemoptysis subsided in next 24 hours

III. Discussion

Pulmonary vasculature is derived from three principle sources. Main pulmonary artery is derived from the arterial portion of truncoaortic sac. Extra pulmonary portion of pulmonary arteries develops from respective 6^{th} branchial arch. Intrapulmonary pulmonary vasculature develops from their respective mesenchymal tissue surrounding the lung buds. Agenesis of pulmonary artery is due to the involution of 6^{th} brachial arch. Since, intrapulmonary vasculature which develops from respiratory mesenchyme is normal in these patients. In these patients lung receive blood retrogradely from collateral arteries & from persistent ductus arteriosus.[4] The term interruption is more preferred to absent pulmonary artery by some authors.[5] Common collaterals supplying to lung include bronchial arteries, transpleural branches of intercostals, internal mammary, subclavian & innominate artey[6]. Rarely collaterals from coronary artery also supply of lung with absent pulmonary artery.[7] During first half of 20^{th} century, all recorded cases of absent pulmonary artery were from the autopsy series.[8] Subsequently with the availability of cardiac surgery in early seventies for cyanotic heart diseases, many more

cases were identified at the time of operations.[9,10] After the development of angiographic techniques, diagnosis of this entity was possible without operation or autopsy.[11]

Advent of computed tomography in last quarter of 20th century made the diagnosis of this entity easy. Patients were diagnosed to have this abnormality when computed tomography was performed for the evaluation of some or other unrelated chest problems or during the workup of symptoms related to this abnormality e.g. dyspnea / hemoptysis.

Occasionally, this entity can also be diagnosed when skiagram chest is performed as a part of routine check up, or during pre employment examination. Small hemi thorax with diffuse reticulations due to collateral blood supply from systemic circulation to lung points toward the possibility of absent pulmonary artery and need of further workup. Although smaller hemi thorax may also be seen in scimitar syndrome & hypoplastic lung. Congenital hypoplastic lung and absent/hypo plastic pulmonary artery disease can be excluded from scimitar syndrome by normal venous drainage to the left atrium.

On computed tomography affected part of pulmonary artery gets terminated with in 1 cm of its origin. Direct anastomosis of transpleural collateral vessels with peripheral branches of the pulmonary artery appears as serrated thickening of pleura and sub pleural parenchymal bands on CT film. Such CT appearance may mimic interlobular septal thickening as seen in IPF.[12] Contrast enhanced computed tomography of chest not only diagnose absent pulmonary artery but also provide information about mediastinal structures and lung parenchyma. Although invasive angiography is better in certain aspects as it provides better delineation and haemodynamic data, but considering the availability, expense and risk, pulmonary angiography is to be reserved only for those having structural cardiac anomaly on echocardiography.[13]

Subject with isolated unilateral absent pulmonary artery may experience recurrent respiratory infections, dyspnea on exertion, high altitude pulmonary edema, pulmonary hypertension in contra lateral lung or hemoptysis.[1,14] Hemoptysis occurs in about 20% of cases and usually self limiting. Occasional case report of massive hemoptysis and death is available in literatures.[6] Rupture of an aneurysm between systemic to pulmonary collateral is said to be responsible for hemoptysis.

There is no consensus regarding treatment of this condition. However, those who are asymptomatic or have minor symptoms should be observed closely for any development of pulmonary arterial hypertension. Medical treatment of pulmonary hypertension in patients with unilateral absent pulmonary artery without associated congenital heart disease is not available yet.[13] Surgical option to correct pulmonary hypertension when present include anastomosis of the hilar arteries of affected lung to the main pulmonary artery.[15] Pneumonectomy or lobectomy is to be considered in cases with massive hemoptysis.[13] In patients having poor pulmonary reserve and or co morbid conditions, selective embolization of the systemic artery is indicated for the management of hemoptysis[16].

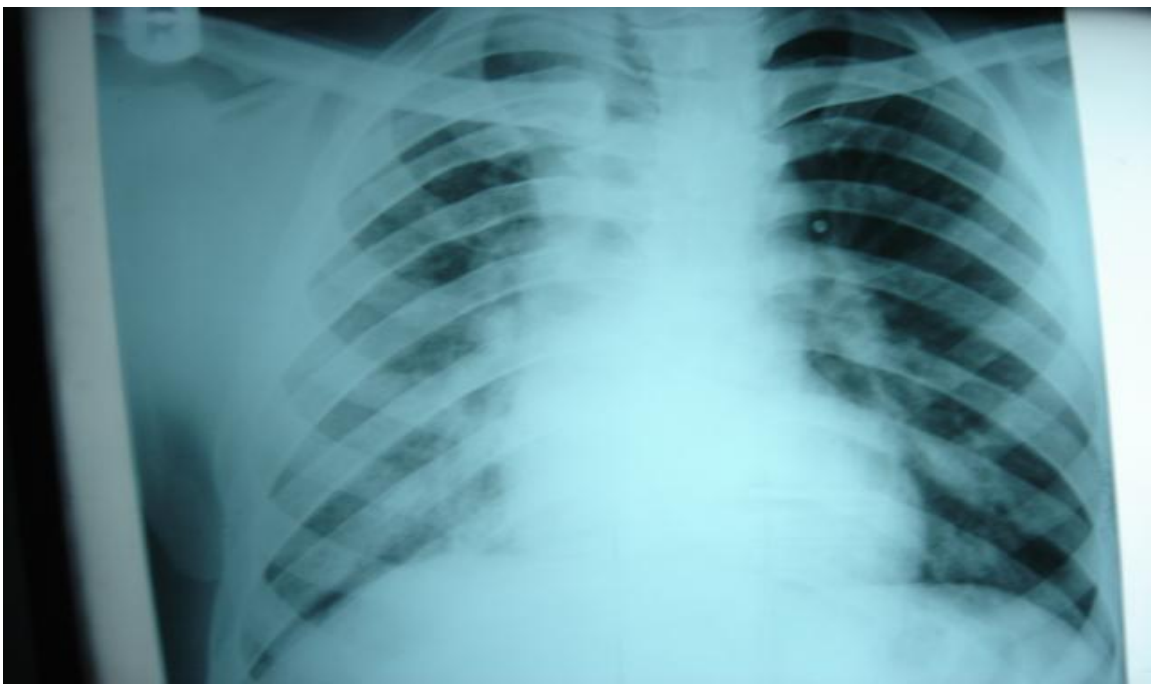

Figure 1

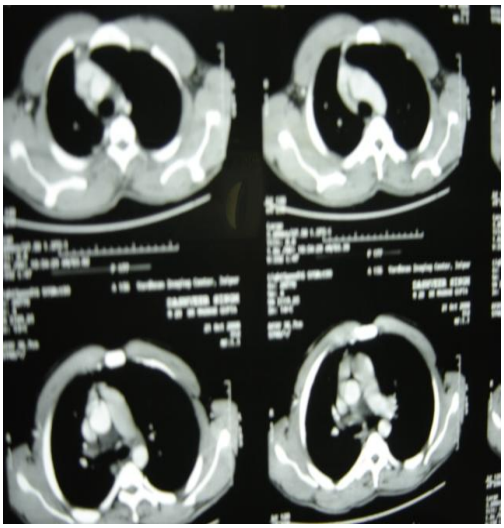

Fig 2a

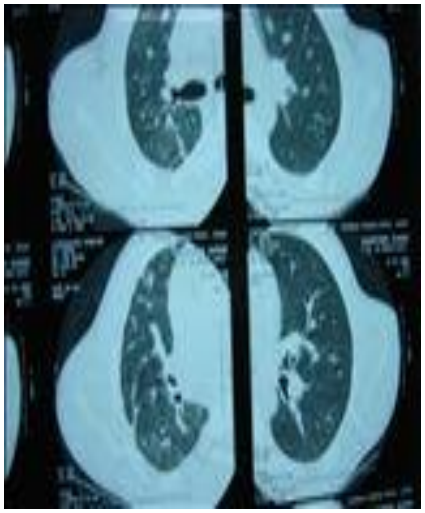

Fig 2b

IV. Legends

Figure 1. X ray posteroanterior view showing small right hemithorax with shift of mediastinum toward right side with diffuse parenchymal haziness.

Figure 2. (a) Contrast enhanced computed tomography of thorax showing absent right pulmonary artery. Rest other mediastinal structures including left pulmonary artery appears normal. (b) High resolution section at the level of carina & just above the diaphragm showing diffuse reticulations attributable to direct anastomosis of transpleural collaterals with peripheral branches of pulmonary artery.

Figure 3 Perfusion scan thorax with Tc99m macroaggregated albumin showing complete absence of perfusion to right lung

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