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UNILATERAL ABSENCE OF PULMONARY ARTERY – AN UNUSUAL CASE OF MASSIVE HEMOPTYSIS.

Muthu ganesh R[1], Tushar Sahasrabudhe[2], Senthamizh KR[3*], MS Barthwal[4]

[1]Junior Resident, Department of Respiratory Medicine, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, Maharashtra, India

[2]Professor, Department of Respiratory Medicine, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, Maharashtra, India

[3*]Junior Resident, Department of Respiratory Medicine, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, Maharashtra, India

[4]Professor and HOD, Department of Respiratory Medicine, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pune, Maharashtra, India

ABSTRACT:

A female in her early thirties, non-smoker, with no known comorbidities, presented to the emergency room with multiple episodes of hemoptysis and exertional dyspnea for a week. She had no history of hemoptysis or any known lung disease in the past. Chest X-Ray revealed relatively small right lung with mediastinal shift to the right. CT angiography revealed right-sided absence of pulmonary artery with collateral supply (systemic to pulmonary shunt) from hypertrophied bronchial, internal mammary and intercostal arteries. She was successfully managed with therapeutic selective embolization of bronchial arteries to control the hemoptysis.

Key-words: Unilateral absence of Pulmonary artery, Haemoptysis, Bronchial artery embolization, CT Angiography.

INTRODUCTION

Pulmonary artery agenesis or Unilateral absence of pulmonary artery (UAPA) leading to hypo-genesis of the ipsilateral lung is a rare congenital anomaly with an estimated prevalence of 1 in 200000 persons. It commonly occurs in association with congenital cardiac anomalies like patent ductus arteriosus or septal defects, which cause early symptoms requiring surgical treatment in childhood. In such cases, UAPA occurs more commonly on the left side and is an incidental diagnosis [1]. However, UAPA can also occur as an isolated congenital anomaly, more commonly on the right side. These patients may be asymptomatic in earlier years of life and survive into adulthood. The common presentation includes exertional dyspnea, exercise intolerance, recurrent respiratory infections, and hemoptysis. These patients are more prone to develop bronchiectasis due to recurrent respiratory infections [2]. Acute cardio-respiratory stress such as high altitude and pregnancy has been reported to trigger the symptoms in previously asymptomatic individuals [3, 4].

CASE HISTORY:

A lady in her thirties, non-smoker, with no known comorbidities, presented with multiple episodes of hemoptysis (each episode of approximately 100 ml) and exertional dyspnea for a week. There were no complaints of cough, fever, or chest pain, and she had no constitutional symptoms. There was no chronic history of exertional dyspnea or cough. Rather, she had two uneventful pregnancies with normal vaginal deliveries. There was no past history of tuberculosis or recurrent respiratory infections.

She was pale and breathless at rest but with normal oxygen saturation. Her respiratory system revealed right-sided volume loss with a shift of the mediastinum. She had a few crackles in the right lower chest but had no rhonchi.

All lab parameters were normal except mild anemia. ECG was normal with sinus rhythm. Chest Radiograph revealed a relatively

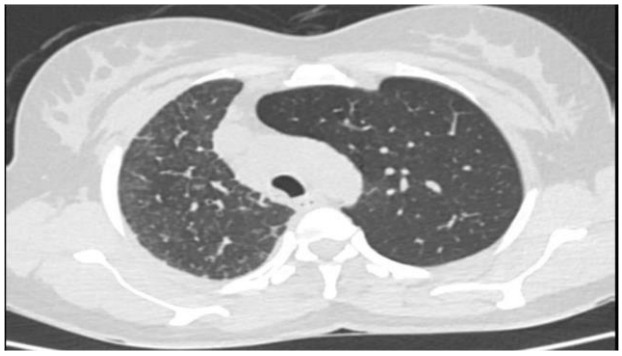

Figure 1:CT scan image showing relatively smaller right lung with increased vascular markings and compensatory left lung hyperinflation with shift of the mediastinum to right

smaller right lung, compensatory left lung hyperinflation and mediastinal shift to the right. There was no hyperlucency on either side. CT scan thorax showed a relatively smaller right lung which was normal and did not reveal any bronchial or alveolar abnormalities but showed increased vascular markings (Figure 1). Her spirometry revealed mild restriction and no obstruction. She was subjected to further workup with CT Angiography and a 2-dimensional echocardiogram (2D echo). CT angiography revealed the absence of a pulmonary artery on the right side and a normal

CT Angiography revealed the absence of a pulmonary artery on the right side and a normal pulmonary arterial tree on the left (Figure 2).

It revealed prominent and hypertrophied bronchial arteries, intercostal arteries, internal mammary and subclavian arteries with multiple collaterals from these arteries and aorta to pulmonary circulation on the right side (Figure3). There was no evidence of pulmonary thromboembolism.

2D echo revealed mild pulmonary artery hypertension and did not show any congenital cardiac septal defects. She was therefore diagnosed with a case of isolated UAPA.

During the hospital stay, she had massive hemoptysis and required a blood transfusion. Given the recurrent hemoptysis, she was subjected to direct aortography to identify all the collateral vessels. Selective therapeutic embolization of potential bleeding arteries was done using gel foam. She tolerated the procedure well and the hemoptysis was fully controlled. The patient is symptomatically and functionally better with no further episodes of hemoptysis.

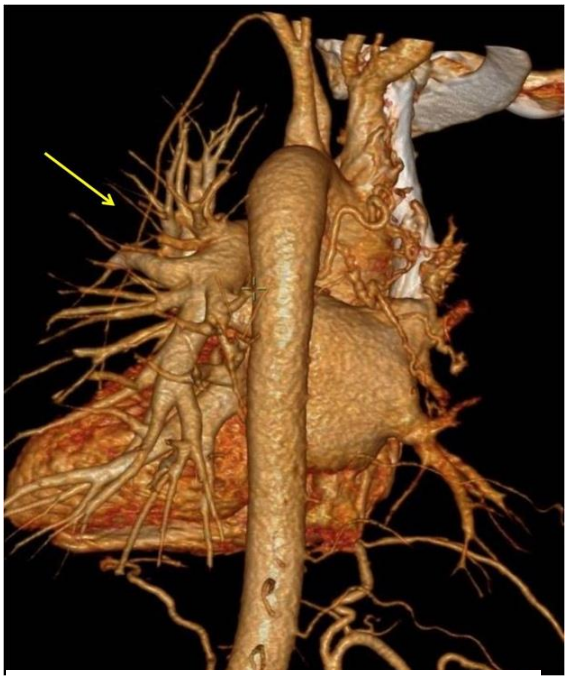

Figure 2:CT pulmonary angiogram 3D construction showing left pulmonary arterial tree (yellow arrow) with absent pulmonary artery on right side

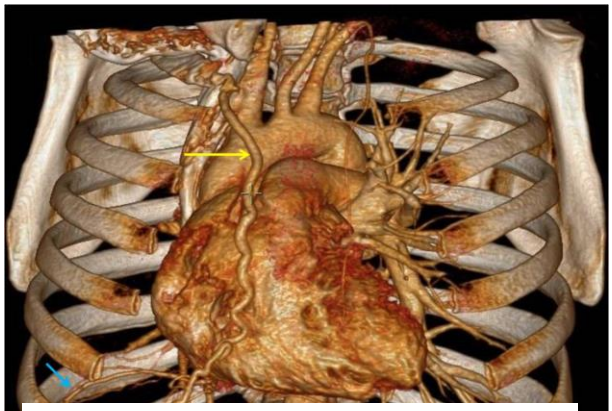

Figure 3: CT Angiography 3 D construction showing collateral supply to right lung from internal mammary artery (yellow arrow), intercostal arteries (blue arrow) and aorta (green arrow)

DISCUSSION:

UAPA is a congenital anomaly and finds its origin in embryological maldevelopment. The pulmonary arteries begin at the proximal end of the sixth aortic arch and the ductus arteriosus forms from their distal end during normal development. The absence of the pulmonary artery is due to the involution of the proximal 6th aortic arch and the persistence of the connection of the pulmonary artery to the ductus, resulting in a ductal origin of the pulmonary artery. When the ductal tissue regresses at the time of birth, this results in the proximal interruption of that vessel[7]. Transient systemic to pulmonary collateral arteries may form during the early stages of embryonic development. These temporary connections may persist as collateral arteries after birth. The absent pulmonary artery induces angiogenesis in the ipsilateral lung's systemic circulation and increases systemic to pulmonary blood flow. The affected artery's distal intrapulmonary branches often remain intact and may get collateral supply from the intercostal, bronchial, internal mammary, subclavian, sub diaphragmatic or coronary arteries. Hemoptysis frequently develops due to high pressures in the collateral circulation and hypertrophy of the intercostal, bronchial, subclavian and internal mammary arteries [1]. It also causes significant left to right shunt causing volume overload over the years.

The pathophysiology, symptomatology, diagnosis, and treatment options have been reviewed in the year 2013 [8]. A review of 108 UAPA cases, in 2002, showed average age of presentation to be 14 years. Chest pain, recurrent infections and pleural effusion was seen in 37% of cases, while exercise intolerance and shortness of

breath was seen in 40% of cases. Pulmonary hypertension was present in 44% of patients and hemoptysis in about 20% of cases [2].

Blood flow that is diverted from the affected side to the contralateral pulmonary artery may cause pulmonary hypertension. Increased shear stress on the endothelium due to increased blood flow in the contralateral artery causes the release of vasoconstrictive substances like endothelin. Pulmonary hypertension and increased resistance in the pulmonary vasculature can result from chronic vasoconstriction of the pulmonary arterioles. Exertion-induced dyspnea and chest pain are typical presentations in patients with severe pulmonary hypertension.

Swyer-James Syndrome (SJS) is one of the differential diagnoses in this case, which presents with hyperlucency of a part of or the whole lung due to poor arterial flow along with hyper-distended alveoli. It is a condition that develops because of post-infectious obliterative bronchiolitis in childhood, typically adenovirus. The diagnosis can be established by demonstrating hypoplasia of pulmonary arterial branches and post-obstructive emphysema utilizing CT angiography and ventilation-perfusion scan [5]. Post-obstructive alveolar dilatation can be differentiated from pathological emphysema using a parametric response map. UAPA, on the contrary, shows an underdeveloped lung with an absent pulmonary arterial tree and numerous collaterals can be easily demonstrated on CT angiography. Also, alveolar air trapping is absent [6]. Presentation with hemoptysis usually does not occur in SJS as there are no systemic-pulmonary collaterals unless there is secondary bronchiectasis.

The diagnosis can be made by a CT scan. Findings on CT scan apart from absent pulmonary artery include collateral circulation, intact peripheral branches of the pulmonary artery and mosaic attenuation. The transthoracic echocardiogram assesses coexisting cardiac anomalies and evaluates pulmonary hypertension. Gold standard in the diagnosis of UAPA is direct angiography but since it is invasive, it is used as a pre-op investigation for patients planned for selective therapeutic embolization of bronchial arteries and in lobectomy/Pneumonectomy.

In UAPA, there is no definite treatment consensus. In uncomplicated cases, patients are advised to have serial echocardiograms as a follow-up to assess the development of pulmonary hypertension. The management is often individualized for the patient based on the symptoms, associated conditions, and complications.

An endothelin receptor antagonist is used to treat individuals with pulmonary hypertension who have exertional chest pain and dyspnea. In cases of massive hemoptysis, radical vascular embolization and ipsilateral pneumonectomy are performed [9]. Lobectomy or pneumonectomy is carried out in cases of recurrent pulmonary infections or recurrent hemoptysis secondary to bronchiectasis.

Other infective causes of hemoptysis were ruled out by normal sputum examination and the absence of any alveolar shadows on radiological investigations.

CONCLUSION:

UAPA was well compensated allowing the patient to be completely asymptomatic before the hemoptysis episode, a rarity in this case. A high index of suspicion is necessary to diagnose such rare vascular anomalies.

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