Case Report

DOI: https://dx.doi.org/10.21088/ijmhs.2347.9981.5118.9

Hypoplastic left Pulmonary Artery Presenting as Pulmonary Hypertension in an Adult: A Case Report

A.K. Badrinath[1], Suresh Babu S.[2], T. Vishwa Teja[3]

IJMHS (January-June 2018) 05 (1): 45-477 / ©Red Flower Publication Pvt. Ltd.

Abstract

Unilateral hypoplasia or absence of a branch of the pulmonary artery is a rare anomaly usually associated with other congenital cardiac disorders. Isolated hypoplasia of unilateral pulmonary artery is still rare. Patients present with varied clinical manifestations with varied age at presentation, younger age or in adulthood with pulmonary hypertension (PAHT). Early diagnosis with definitive treatment prevents development of PAHT in adulthood. Here we present an elderly female who presented with PAHT with right heart failure secondary to chronic pulmonary thromboembolism was diagnosed to have hypoplastic left pulmonary artery.

Keywords: Pulmonary Artery; Pulmonary Hypertension; Pulmonary Angiography.

Case Report

A 50 year old female patient presented with complaints of breathlessness on exertion for 2 months duration relieved on rest associated with swelling of legs. History of giddiness present on and off, no chest pain, hemoptysis or palpitations. Patient was a known case of type 2 diabetes for 3 years on oral hypoglycemic drugs. No history of similar complaints in the past and no other comorbidities present. On examination patient was conscious and oriented, tachypneic (RR- 28/ min), saturation 78% in room air which improved to 90% with 6L of O_2, no pallor, bilateral pitting pedal edema was present, pulse rate- 98/ min, regular, low volume; blood pressure- 100/70 mm Hg, JVP elevated. Systemic examination revealed CVS – S1, S2 present, pan systolic murmur present in tricuspid area with a loud P2 in pulmonary area. RS – reduced air entry on the left hemithorax with bilateral fine basal crepitations (R>L). Per abdomen soft, no organomegaly, epigastric pulsations were present and CNS was intact. Patient was admitted with provisional diagnosis of heart failure and evaluated.

Her complete hemogram revealed Hb; total count and platelet. Her renal and liver function tests were normal. ECG showed sinus tachycardia with right axis deviation with right ventricular hypertrophy with right bundle branch block. Chest X ray showed cardiomegaly with hypoplastic left lung with absent vascularity with compensatory emphysema right lung. A 2D echocardiography revealed right atrium and ventricle dilated with severe tricuspid regurgitation and pulmonary hypertension (PAHT) with RVSP of 89 mm Hg with mild RV dysfunction. The patient was diagnosed as a case of chronic pulmonary hypertension with right heart failure and further evaluated.

There was no family history of similar symptoms or disease. No history or clinical finding of respiratory disorders, chronic drug intake, no history suggestive of collagen vascular diseases and serum ANA by IF method was negative. Viral markers (HIV, hepatitis B & C virus) were negative. 2D echocardiography did not reveal any cardiac pathology. At this juncture the etiology

Author's Affiliation: [1]Professor [2]Assistant Professor [3]Senior Resident, Department of General Medicine, Sri Manakula Vinayagar Medical college and hospital, Puducherry 605107, India.

Corresponding Author: A.K. Badrinath, Professor, Department of General Medicine, Sri Manakula Vinayagar Medical college and hospital, Puducherry 605107, India.

Email- akbsts@yahoo.co.in

Received on 10 February 2018
Accepted on 23 April 2018

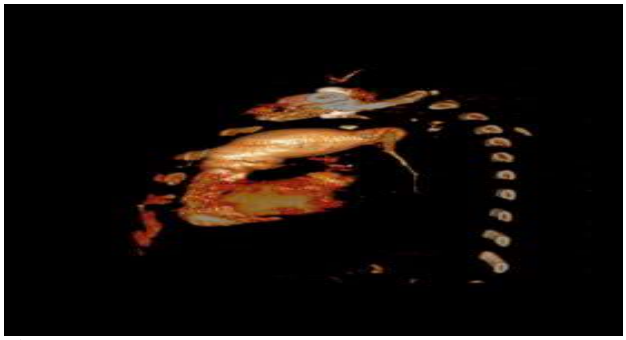

Fig. 1:

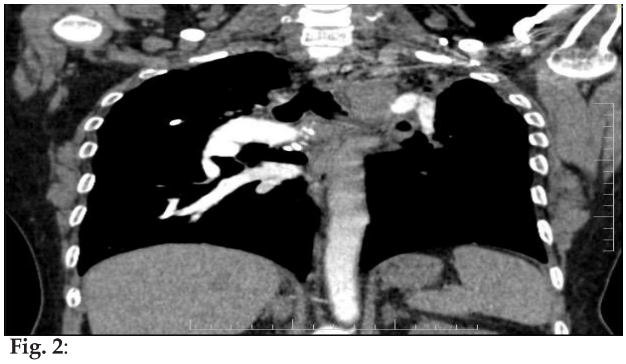

Fig. 2:

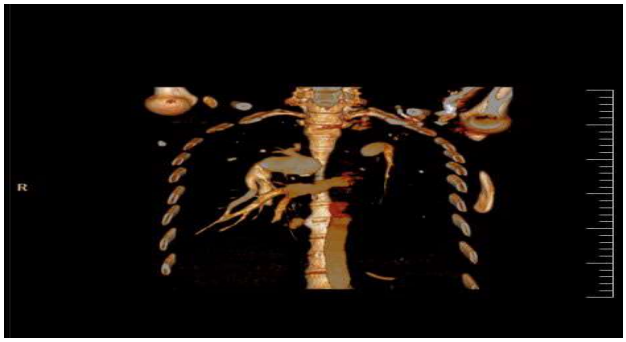

Fig. 3:

Fig. 4:

was towards PAHT due to chronic pulmonary thromboembolism. Patient was started on oxygen supplementation, diuretics and oral anticoagulants. CT Pulmonary angiography revealed normal course of the right pulmonary artery with a hypoplastic left pulmonary artery. Patient diagnosis was revised with final diagnosis of Congenital Hypoplastic Left Pulmonary Artery with Severe Pulmonary Arterial Hypertension with Right Heart Failure. Oral anticoagulant was stopped. Patient started on tab. Sildenafil 10 mg tds and tab. Bosentan 62.5 mg bd, salt restricted diet, bed rest and discharged with home oxygen therapy.

Discussion

Pulmonary hypertension (PAHT) results from left heart failure, pulmonary parenchymal or vascular disease and thromboembolism or combination of them. Patients presents with exertionaldyspnea. Diagnosis is made by ECG, chest X ray and echocardiographic studies. Further identifying the cause of PAHT needs detailed evaluation. In our case the cause of pulmonary hypertension was secondary to congenital absence of the left pulmonary artery.

Unilateral absence of the pulmonary artery (UAPA) is a very rare congenital abnormality seen in 1/ 2 lakh population. Pulmonary artery malformations are seen due to malformations in the sixth aortic arch during embryogenesis. Most of the cases were associated with other congenital cardiac defects. The common associated cardiac defects were tetralogy of fallot, atrial septal defect, coarctation of aorta, truncusarteriosus, right sided aortic arch and pulmonary atresia. Isolated unilateral absence of pulmonary artery (UAPA) is rare and in about 2/3 of cases the right pulmonary artery is commonly involved but in our case the left pulmonary artery was absent.

The age at presentation is varied with patients presenting at younger age with recurrent lower respiratory tract infections or have remain asymptomatic presenting later in life as in our patient with pulmonary hypertension with exercise intolerance. Hemoptysis is also common in these patients. Clinically these patients have an ejection systolic murmur in pulmonary area with reduced chest movements and reduced to absent air entry on the affected side. ECG may be normal to features of right ventricular hypertrophy from various case reports. Our patient had RBBB with right ventricular hypertrophy. Chest X ray is one of the important investigation and gives a clue for further evaluation. Our patient had similar finding in chest X ray as in other case reports with small hemithorax on the affected side with hyperlucency. A transthoracic echocardiography is necessary to find associated cardiac abnormalities and development of pulmonary hypertension. The definitive diagnosis of UAPA is made by computed

tomography – pulmonary angiography. The other investigations done in such patients are ventilation perfusion scan which shows reduced to absent ventilation and perfusion. Finally a high index of suspicion is necessary to diagnose UAPA.

The development of pulmonary hypertension (PAHT) in patients with UAPA is an important prognostic indicator. PAHT occur due to various mechanisms, the persistentfetal structure of the unaffected pulmonary artery was the predominant mechanismpostulated for pulmonary hypertension. The other possibility was a reduction in the elastic property of the vasculature of thenormal lung which is exposed to the total cardiac output fromthe right ventricle. Also the increased blood flow to the normal lung leadingto sheer stress on the endothelium, which results in the releaseof vasoconstrictors. Chronic vasoconstrictionof the pulmonary arterioles further lead to remodelling resulting inincreased resistance in the pulmonary vasculature and PAHT.

One of the important considerations in these patients presenting in adult life is chronic pulmonary thromboembolism. In our case the initial diagnosis was towards chronic pulmonary thromboembolism. But patient had no evidence or risk factors for hypercoagulable state. Management of these patients are based on clinical manifestations of individual patients. Asymptomatic patients can be monitored periodically for the development of PAHT. In patients with recurrent serious pulmonary infections and hemoptysis can be managed therapeutically with lobectomy or arterial embolization. Patients with PAHT should be started on vasodilator therapy as in our case.

To conclude unilateral absence of pulmonary artery is a rare disease and a high degree of suspicion is necessary for diagnosis. Treatment is dependent on individual patient's condition.

References

1. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. Chest. 1995;108(3):670-76.
2. Presbitero P, Bull C, Haworth SG, de Leval MR. Absent or occult pulmonary artery. Br Heart J. 1984;52(2):178-85.
3. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: A case report and review of the literature. Chest. 2002;122(4):1471-77.
4. Wyman SM. Congenital absence of a pulmonary artery. Its demonstration by roent-genography. Radiology.1954;62:321.
5. Yiu MWC, Le DV, Leung Y, et al. Radiological features of isolated unilateral absence of pulmonary artery. J HK CollRadiol. 2001;4: 277-80.
6. Welch K, Hanley F, Johnston T, Cailes C, Shah MJ. Isolated unilateral absence of right proximal pulmonary artery: surgical repair and follow-up. Ann Thorac Surg. 2005;79(4):1399-402.