Unilateral left Pulmonary Artery Agenesis with Pulmonary Hypertension: Unusual First Presentation

Vikas Dogra¹, Manu Bhardwaj², Lakshay Beriwal³

ABSTRACT

Introduction: Unilateral absence of pulmonary artery is a rare congenital disorder. We present here the case of Unilateral left pulmonary artery agenesis with pulmonary hypertension. Case Report: A 33-year young male patient presented to our tertiary care hospital in the out-patient department with complaints of shortness of breath on exertion for 7 months and bilateral feet swelling for 6 months. Routine investigations were within normal limits except 2 dimensional ECHO showed right atrium and right ventricle dilatation with severe pulmonary artery hypertension and 60% left ventricular ejection fraction. Patients with isolated right pulmonary artery agenesis commonly survive into adulthood with minimal or no symptoms, which makes their identification challenging. But our patient had left pulmonary artery agenesis which has significant association of life threatening cardiovascular malformations with left-sided UAPA and surgical repair is often required during the first year of life. None of which is present in our patient which is unsusual.

Conclusion: Clinicians should be aware that recurrent respiratory infections may be presenting feature of UAPA. Initial investigation is usually a chest radiograph. Echocardiography is required for evaluation of possible pulmonary hypertension. Confirmation of the diagnosis and anatomic details can be discerned by CT scanning and MRI. Angiography is reserved for patients requiring embolization or revascularization surgery.

Present case demonstrates a rare presentation of UAPA as right heart failure in first visit.

Keywords: Recurrent Infections, Unilateral Artery Pulmonary Agenesis

INTRODUCTION

Unilateral absence of pulmonary artery (UAPA) or pulmonary artery agenesis is a rare congenital disorder presenting with wide spectrum of symptoms. The first case was reported in 1868. UAPA is a rare disease with prevalence of around around 1 in 200,000 individuals.¹ The clinical presentation is variable and patients may be asymptomatic for many years and even throughout their lives. Recurrent pulmonary infections, decreased exercise intolerance and shortness of breath on exertion are the most common symptoms.

CASE REPORT

A 33-year young male patient presented to our tertiary care hospital in the out-patient department with complaints of shortness of breath on exertion for 7 months and bilateral feet swelling for 6 months. There was no history of leg pain, cough, sputum, hemoptysis, fever. He has been smoking 2-3 cigarettes per day for 3 years. There is a history of

sputum positive pulmonary tuberculosis for which he took ATT for 6 months. Patient is a chronic alcoholic and he has been taking around 250ml/week alcohol for last 10 years.



Figure-1: ECG showing ST depression in V2-V6 leads.



Figure-2:

¹Assistant Professor, Department of Pulmonary Medicine, Rajiv Gandhi Super Speciality Hospital, Taharpur Road, Taharpur, Taharpur Village, Dilshad Garden, New Delhi, Delhi, ²Senior Resident, Department of Critical Care Medicine, Rajiv Gandhi Super Speciality Hospital, Taharpur Road, Taharpur, Taharpur Village, Dilshad Garden, New Delhi, Delhi, ³Junior Resident, Department of Pulmonary Medicine Rajiv Gandhi Super Speciality Hospital, Taharpur, Taharpur Village, Dilshad Garden, New Delhi, India

Corresponding author: Dr. Manu Bhardwaj, Senior Resident, Department of Critical Care Medicine, Rajiv Gandhi Super Speciality Hospital, Taharpur Road, Taharpur, Taharpur Village, Dilshad Garden, New Delhi, Delhi, India

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Figure-4A, B, C & D:

J18

There is no history of hypertension, bronchial asthma and diabetes mellitus. On presentation his pulse rate was 98/min, respiratory rate was 16/min., blood pressure 132/94 mm Hg,

temperature 98.8 ° F, SpO_2 of 96% at room air. His general physical examination was unremarkable except bilateral pitting pedal edema.

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Figure-4E, F:

Routine investigations were within normal limits. Cardiac enzymes (CK-MB, hs-troponin I) were negative. Electrocardiogram showed tall R wave and ST depression and T wave inversion in leads V2-V6 (Fig 1). 2 dimensional ECHO showed right atrium and right ventricle dilatation with severe pulmonary artery hypertension and 60% left ventricular ejection fraction.

A plain chest radiograph showed a loss of volume of his left lung with absence of pulmonary artery shadow on left side and dilated on right side.(Fig 2)

For further evaluation CT Pulmonary Angiography was done and it showed reduced volume of left hemithorax, patchy pleural thickening and few peripheral reticular opacities with multiple trans-pleural collaterals with mild dilated right pulmonary artery suggestive of left pulmonary artery agenesis (Fig3 a and b). 3D CT reconstruction images are shown in Fig 4. He was treated with Bosentan (endothelin receptor antagonist) and torsemide and spirinolactone (diuretics). On follow up his symptoms improved and is doing well.

DISCUSSION

Congenital UAPA is a rare anomaly that may occur in isolation but most frequently is accompanied by cardiovascular malformations such as Tetralogy of Fallot, Septal defects, Patent ductus arteriosus, Coarctation of the aorta and transposition of great vessels.⁵ Dyring embryogenesis there is an involution of the proximal sixth aortic arch of the affected side, which leads to an absence of the proximal pulmonary artery. There is normal development of intrapulmonary vessels. In some cases distal portion of the affected pulmonary artery trunk can develop normally. Blood supply is achieved by systemic collaterals from bronchial, major aorto-pulmonary collaterals and other systemic arteries.¹

UAPA is twice as common on the right side. There is significant association of life threatening cardiovascular malformations with left-sided UAPA and surgical repair is often required during the first year of life.^{6,7} Patients with

isolated right pulmonary artery agenesis commonly survive into adulthood with minimal or no symptoms, which makes their identification challenging.

Multiple conditions like Swyer-James-MacLeod's syndrome (SJMS), compensatory emphysema and pulmonary thrombo-embolic disease can have similar radiographic appearance. A study has shown that around 30% patients were asymptomatic.⁸ Their detection was incidental during routine medical examination.² Sometimes disease can be unmasked by pregnancy or high altitude.⁹ The symptoms are commonly nonspecific and due to this reason diagnosis has been delayed in some cases by up to 30 years after onset of symptoms.²

Chest radiographic findings typical of UAPA include ipsilateral displacement of heart and mediastinum, absent hilar shadow, volume loss of affected lung with hyperinflation of contralateral lung.¹⁰ For confirming absence of pulmonary artery, a contrast-enhanced CT of the thorax can be performed. High resolution CT scanning can also evaluate the presence of bronchiectasis in cases of recurrent bronchopulmonary infections. Magnetic resonance imaging (MRI) is helpful in the evaluation of congenital cardiovascular defects.¹¹

Echocardiography is necessary to exclude any other cardiac abnormalities and to evaluate the presence of associated pulmonary hypertension. Ventilation-perfusion scintigraphy can be useful in distinguishing UAPA from SJMS.¹⁰ The gold standard for the diagnosis of pulmonary artery agenesis is angiography. Cardiac catheterization with pulmonary venous wedge angiography is necessary to visualize hidden pulmonary arteries in the hilum.² This is done when revascularization is considered.

Multiple therapeutic approaches have been described for treatment of UAPA depending upon the clinical presentation. 8% of the patients underwent either a pneumonectomy or lobectomy for recurrent hemoptysis or intractable pulmonary infections.² Revascularization of the absent artery is recommended in presence of pulmonary hypertension. If revascularization is not an option or it fails to improve pulmonary hypertension is, medical treatment described for

Section: Medicine

CONCLUSION

Clinicians should be aware that recurrent respiratory infections may be presenting feature of UAPA. Initial investigation is usually a chest radiograph. Echocardiography is required for evaluation of possible pulmonary hypertension. Confirmation of the diagnosis and anatomic details can be discerned by CT scanning and MRI. Angiography is reserved for patients requiring embolization or revascularization surgery. Present case demonstrates a rare presentation of UAPA as right heart failure in first visit. He is doing well on endothelin receptor antagonist and diuretics .

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