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Case Report

Unilateral pulmonary artery agenesis diagnosed in adulthood: a case report

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ABSTRACT

Isolated pulmonary artery agenesis (UPAA) diagnosed in adulthood is a rare congenital anomaly. It occurs during embryological development where there is a fusion failure between sixth aortic arch and pulmonary truck of affected side. Here we report a case of an adult patient with isolated unilateral right pulmonary artery aplasia.

Keywords: Unilateral pulmonary artery agenesis, Tuberculosis, Haemoptysis

INTRODUCTION

Isolated pulmonary artery agenesis (UPAA) diagnosed in adulthood is a rare congenital anomaly. It occurs during embryological development where there is a fusion failure between sixth aortic arch and pulmonary truck of affected side. Unilateral pulmonary artery aplasia is often diagnosed in adolescence. The estimated prevalence of single UPAA is 1/200,000 patients.[1] Here we report a case of an adult patient with isolated unilateral right pulmonary artery aplasia.

CASE REPORT

A 47-year-old male patient with no comorbidities was admitted to our department with history of acute onset dyspnea and dull aching non-exertional retrosternal chest pain with prior episodes of hemoptysis on two occasions (varying amounts of streaks of blood) 2 years ago with no current episodes. He is a nonsmoker and denied any significant occupational exposures, contact with pulmonary tuberculosis and previous treatment for pulmonary tuberculosis. He also gave a history of left below knee amputation following a road traffic accident.

General physical examination was unremarkable.

Respiratory examination revealed minimal deviation of trachea towards right and auscultation revealed decreased respiratory sounds on the right hemithorax.

Routine hematological investigations were within normal limits. Sputum for acid fast bacilli was negative. Sputum culture showed normal growth. Chest radiography showed minimally reduced right lung volume, elevation of right hemidiaphragm and cardiomegaly.

A six-minute walk test was done, and patient desaturated to 88% on room air on covering 360m. Spirometry and diffusing capacity of the lungs for carbon monoxide (DLCO) were performed and was within normal limits. Computed tomography (CT) pulmonary angiogram revealed unilateral aplasia of right pulmonary artery with no contrast opacification of right pulmonary artery and its branches in the pulmonary arterial phase. Multiple collaterals were noted arising from descending thoracic aorta, abdominal aorta, costo-cervical trunk, subclavian arteries, posterior intercoastal arteries and internal mammary arteries. Left pulmonary artery and its branches were dilated and tortuous.

Initial echocardiogram showed an ejection fraction of 65%, pulmonary artery, right atrium and right ventricle

were mildly dilated, mild tricuspid regurgitation (63 mmHg) and moderate pulmonary arterial hypertension were noted. Pulmonary angiography showed – right pulmonary artery aplasia with multiple collaterals.

Patient was treated conservatively and on discharge, was advised long term oxygen therapy.

Patient came back 2 months later with recurrent chest infections and follow up echocardiogram revealed dilated right atrium and ventricle, ejection fraction of 65%, moderate to severe tricuspid regurgitation, severe pulmonary arterial hypertension, and markedly dilated main pulmonary artery.

Magnetic resonance imaging (MRI) chest was done which revealed non visualization of right pulmonary artery with proximal dilatation of main pulmonary artery and left pulmonary artery with multiple collaterals.

Sputum for acid fast bacilli (AFB) was negative, sputum for GeneXpert showed *Mycobacterium tuberculosis* (MTB) detected, Rif resistance not detected, and he was initiated on anti-tuberculous treatment according to NTEP guidelines. The patient is currently on follow up.

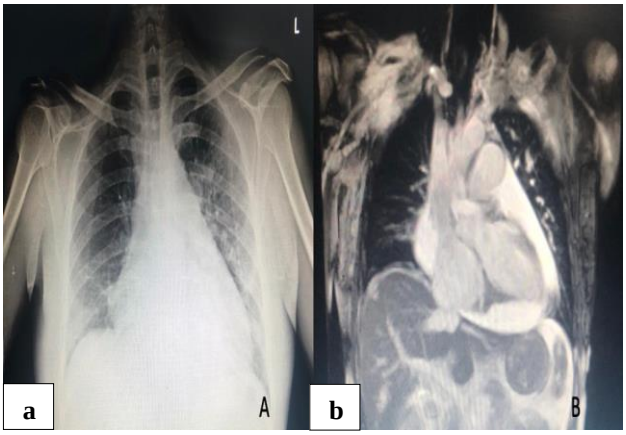

Figure 1: (a) Chest X-ray, and (b) MRI chest.

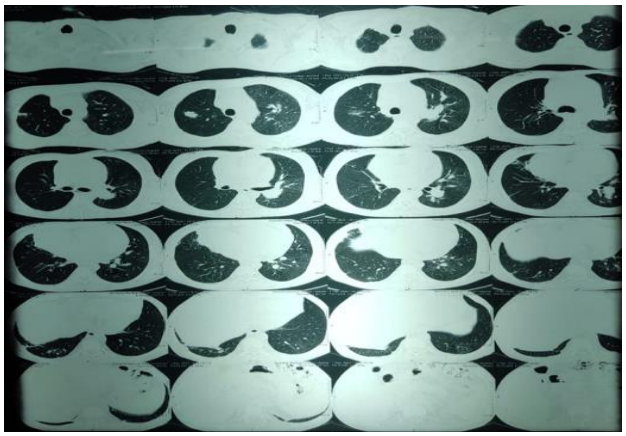

Figure 2: CT pulmonary angiogram.

DISCUSSION

The estimated prevalence of single UPAA is 1/200,000 patients.[1] While average age of diagnosis is 14 years, in our case the condition was diagnosed in adulthood.[2] Right side is affected in nearly 2/3 of cases with unilateral pulmonary artery aplasia which is similar to our case where the patient was diagnosed to have right pulmonary artery aplasia.[2] It usually presents during childhood or early adulthood and presentation in late adulthood such as in our case is a rare phenomenon.

UPAA can be accompanied by other congenital heart abnormalities, which was not noted in our patient.[2] Development of the bronchial tree takes place at about 26th to 31st day of intrauterine life. In UPAA, embryogenetic defect is a fusion failure between sixth aortic arch and pulmonary truck of affected side, causing the absence of the proximal segment of the ipsilateral pulmonary artery, which is often associated with hypo-vascularization and hypoplasia of the lung of the same side.[2,3]

Symptoms may include dyspnea on exertion, recurrent pulmonary infections, hemoptysis (in 20% of patients), chest pain, or pleural effusion.[1,2,4] Clinical examinations is usually non-diagnostic, as in our case, except the decreased breath sounds on the affected side.[1,4,6] Pulmonary function tests reveal a mild restrictive pattern with normal single breath diffusion capacity.[1,4,6] Chest X-ray in case of lung hypoplasia or aplasia may show disposition of the ipsilateral mediastinum and diaphragmatic elevation with volume loss of the affected lung, no hilar shadow and hyperinflation of the contralateral lung.[7] In our patient due to presence of multiple collaterals chest radiograph did not show any major gross abnormalities in lung volume, or significant mediastinal shift or hyperinflation of contralateral lung.

Heart echocardiography is useful in picking up the congenital cardiac defects and pulmonary artery hypertension. Congenital abnormalities were not present in our patient, but ECHO revealed severe pulmonary arterial hypertension with dilated main pulmonary artery.[2,4]

Contrast enhanced chest CT/CT chest is more than sufficient to make the suspected diagnosis of UPAA and in our case it revealed no contrast opacification of right pulmonary artery and its branches with multiple collaterals supplying the ipsilateral lung with dilatation of contralateral pulmonary artery and its branches. In our patient due to multiple collaterals supplying the affected side the ipsilateral lung showed no gross underdevelopment.

MRI chest is also a helpful modality in diagnosing UPAA. The visualization of perfusion anomalies of the lung is possible by perfusion scintigraphy and can aid in diagnosis of UPAA.[10] MRI chest done for our patient revealed

multiple collaterals supplying the ipsilateral lung with dilatation of contralateral pulmonary artery and its branches.

The gold standards for making a conclusive diagnosis and determining the collateral blood flow to the affected lung are pulmonary angiography and digital subtraction angiography.[1,4] But is reserved for cases where embolization for massive hemoptysis is indicated or for pre-operatory evaluation of patients undergoing surgical revascularization.[1,4]

Pulmonary agenesis is considered as one of the rare false positives for pulmonary embolism.

Treatment options include medical and surgical management. Asymptomatic patients must be kept on regular follow up. Early surgical repair might help in patient with heart failure by decreasing the load on heart and conserve the remaining lung function. Patients with severe complications like massive hemoptysis, recurrent pulmonary infections and severe pulmonary arterial hypertension can be treated with surgical interventions like pneumonectomy and revascularization.

Bronchial or non-bronchial systemic artery embolization and medical management is useful in patients who are not fit to undergo major surgical interventions.[2,10-13]

In our case, as there was no remnant of right pulmonary artery for surgical revascularization, patient was managed conservatively with supplemental oxygen, medical management for pulmonary arterial hypertension with endothelial receptor antagonist and discharged with advice for long term home oxygenation.

As the patient was incidentally diagnosed to have pulmonary tuberculosis, he was started on anti-tubercular drugs according to NTEP guidelines after doing a baseline liver and renal function tests, which were within normal limits.

CONCLUSION

UPAA is a rare congenital anomaly usually diagnosed in childhood or adolescence but can also be asymptomatic and be diagnosed in adults. In suspected cases thorough clinical examination and chest radiography form initial step in evaluation. Computed tomography can establish a definitive diagnosis in cases of UPAA and can pick up cardiovascular anomalies along with echocardiography for any internal cardiac malformations. Symptomatic patients can be managed with surgical management and in those where surgery is contraindicated can be managed medically.

In India due to high burden of tuberculosis, it is advisable that the patients who are susceptible for frequent lung infections, undergo screening for tuberculosis.

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