Int. J Med. Pharm. Res. (P-ISSN: 2958-3675 | E-ISSN: 2958-3683)
Available on: https://ijmpr.in/

International Journal
of Medical and Pharmaceutical Research

CASE REPORT **OPEN ACCESS**

Unilateral Bronchiectasis With Absent Right Pulmonary Artery presenting as Obstructive Airway Disease

Dr. Priyajeeta Mohapatra[1]*, Dr.Rekha Manjhi[2], Dr.Aurobindo Behera[3], Dr.Prabir Kumar Prusti[4], Dr.Bikash Panda[5]

[1]Post Graduate Trainee, Department of Pulmonary Medicine, VIMSAR, Burla, 768017, Odisha, India
[2]Professor and HOD, Department of Pulmonary Medicine, VIMSAR, Burla, Odisha, 768017, India
[3]Associate Professor, Department of Pulmonary Medicine, VIMSAR, Burla, Odisha, 768017, India
[4]Assistant Professor, Department of Pulmonary Medicine, VIMSAR, Burla, Odisha, 768017, India
[5]Senior Resident, Department of Pulmonary Medicine, VIMSAR, Burla, Odisha, 768017, India

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***Corresponding Author**
Dr. Priyajeeta Mohapatra

Post Graduate Trainee, Department of Pulmonary Medicine, VIMSAR, Burla, 768017, Odisha, India

Received: 05-09-2024
Accepted: 02-11-2024
Available online: 07-11-2024

A B S T R A C T

The pulmonary arteries arise from the 6th aortic arch. The involution of the ipsilateral proximal 6th aortic arch may result in unilateral absent pulmonary artery (UAPA). Unilateral absent pulmonary artery is a rare disorder with an estimated prevalence of 1 in 200,000 adults.We report a case of absent right pulmonary artery with bronchiectasis presenting as obstructive airway disease (OAD).A 40yearold female presented with acute onset of fever, shortness of breath, cough and mucoid sputum production for 4 days. She had a past history of OAD since 15years.On investigation, Chest X ray showed trachea shifted to right, smaller right lung with a small hilar shadow on right side. CECT Thorax showed right side bronchiectasis with small right lung with absent right pulmonary artery, cardiomegaly & Pulmonary hypertension. Patient was treated with IV antibiotics, bronchodilators, mucolytics, moist O2, antihypertensive. Patient clinically improved and was discharged as obstructive airway disease.

Keywords: Unilateral absent pulmonary artery, obstructive airway disease, bronchiectasis.

INTRODUCTION

Unilateral absent pulmonary artery(UAPA) is a rare disorder with an estimated prevalence of 1 in 200,000 adults. The pulmonary trunk and the main pulmonary arteries arise from the 6th aortic arch. The involution of the ipsilateral proximal 6th aortic arch may result in Unilateral absent pulmonary artery. The lung that receives the abnormal blood supply is frequently hypoplastic and may contain bronchiectatic changes [1].

Here we present a case of unilateral absent right pulmonary artery with bronchiectasis presenting as obstructive airway disease.

Case Description

A 40 year female presented with chief complaints of fever, shortness of breath, cough with mucoid sputum production for 4 days. The patient was symptomatic with intermittent exacerbations since 15years. Dyspnea was intermittent with seasonal variation and with dust/smoke allergy.

She is a known case of hypertension since 5years and on medication. No past history of PTB or recurrent childhood respiratory infection. There is history of biomass exposure for 10years.

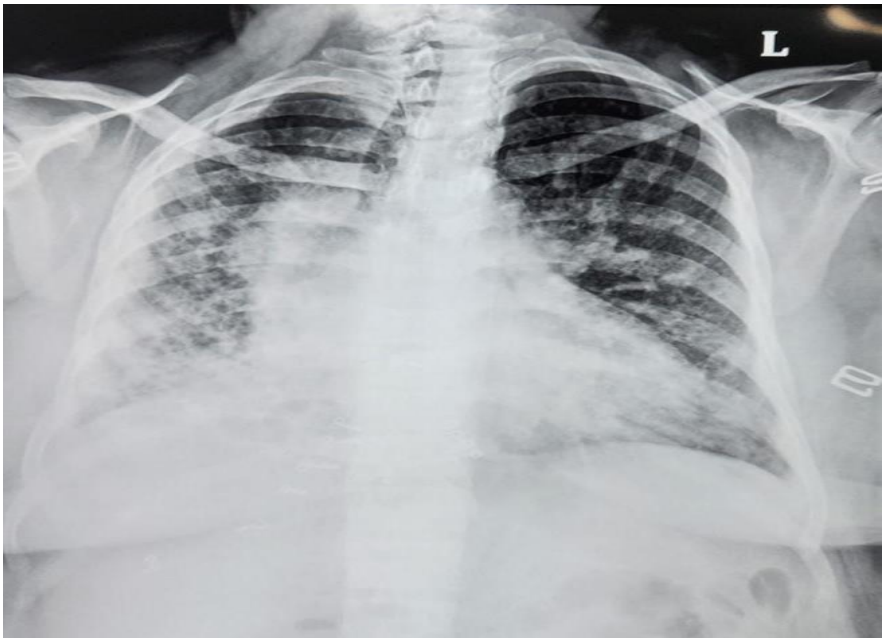
The patient had similar episodes of dyspnea and cough in past for which she was treated as obstructive airway disease.

EXAMINATION & INVESTIGATIONS

On examination patient was dyspneic with pulse rate 90/min, respiratory rate of 30/min. SP02 was 84% in room air. Blood pressure was 180/90 mmHg with bilateral pitting pedal edema.

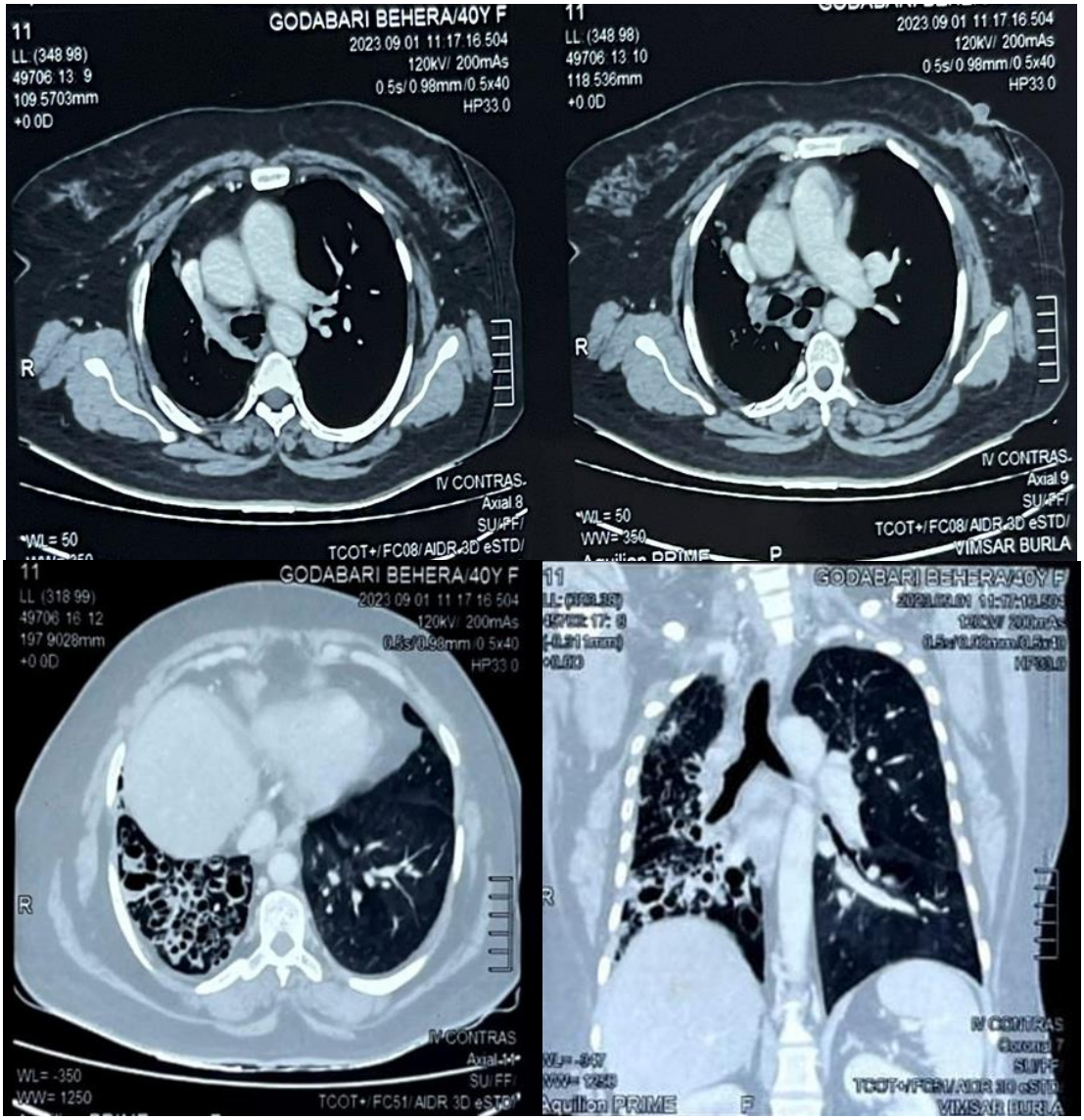
On auscultation of chest patient had bilateral ronchi in all areas and coarse biphasic leathery crepitation over right hemithorax.

Chest X ray-There is presence of Loss of lung volume on right side, smaller right lung with upper mediastinal displacement to right and right hilar prominence. A left cervical rib was found incidentally.



2D Echo- Grade 3 diastolic dysfunction with heart failure with preserved ejection fraction.

CECT Thorax and CT pulmonary angiography- right side bronchiectasis with small right lung with absent right pulmonary artery, cardiomegaly &Pulmonary hypertension.



MANAGEMENT & CONCLUSION

Patient was treated with IV antibiotics, bronchodilators, mucolytics, moist O2, antihypertensive &chest physiotherapy.

The lung that receives the abnormal blood supply is frequently hypoplastic and may contain cystic and bronchiectatic changes [2].

After managing bronchiectasis and hypertension patient improved clinically. Bronchiectasis is usually seen post infectious or post tuberculosis in India. But in this case there is a smaller right lung associated with unilateral absent pulmonary artery which was undiagnosed previously, a predisposing factor for the right lung bronchiectasis.

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