International Journal Dental and Medical Sciences Research
Volume 3, Issue 2, Mar-Apr. 2021 pp 1028-1031 www.ijdmsrjournal.com ISSN: 2582-6018

Unilateral Pulmonary Aplasia in a 50 year old man: An incidental finding

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Submitted: 25-04-2021 Revised: 06-05-2021 Accepted: 08-05-2021

ABSTRACT:INTRODUCTION: Pulmonary aplasia, pulmonary hypoplasia, and pulmonary agenesis are rare congenital lung defects. Typical respiratory symptoms include loud breathing, rapid breathing, recurrent respiratory infections, and respiratory distress.

CASE REPORT: 50 year old man presented with chief presenting complaint of foreign body sensation throat for last 7 days. He also complained of chest pain for the past 15 days which increased on food intake. X-ray revealed complete opacification of the left hemithorax with mediastinal shift to the ipsilateral side. Right lung showed hypertrophy and herniated to the contralateral side. Patient's symptoms were diagnosed to be due to gastritis with acid reflux which resolved with a course of antacids, prokinetics and proton pump inhibitors.

DISCUSSION: Pulmonary agenesis and aplasia are very rare conditions. They are equally common in both sexes and affect both lungs equally. In asymptomatic cases, no treatment is needed. Lower respiratory tract infections require medication. If postural drainage and antibiotics fail to resolve the infection, patients with stumps may require surgical removal.

CONCLUSION: Routine investigations of any patient with chest pain should include chest X-ray, even if pain is suggestive of any non pulmonary etiology.

KEYWORDS: Aplasia, Incidental, Agenesis

I. INTRODUCTION:

Pulmonary aplasia, pulmonary hypoplasia, and pulmonary agenesis are a few rare congenital lung defects. The lung parenchyma, its vasculature, and its bronchus are all absent in pulmonary agenesis[1]. The most common variant is pulmonary aplasia, which is characterized by the absence of the distal lung[2] and the presence of a carina and the main-stem bronchial stump. Typical respiratory symptoms include loud breathing, rapid breathing, recurrent respiratory infections, and respiratory distress[2]. We present a case of a 50 year old man with unilateral aplasia of lung.

II. CASE REPORT:

A 50 year old man presented with chief presenting complaint of foreign body sensation throat for last 7 days. He also complained of chest pain for the past 15 days which increased on food intake. Chest pain was of burning nature and was located in the parasternal area on the left side. On inspection chest contour looked normal, but chest movement on left side was decreased. On auscultation, decreased breath sounds were present on left side in all auscultatory areas. Routine investigations such as electro-cardiogram (ECG) as well as chest X-Ray postero-anterior view were done. ECG showed normal sinus rhythm while chest X-ray revealed complete opacification of the left hemithorax with mediastinal shift to the ipsilateral side. Lung on the right side was hyperinflated.

Computed tomography (CT) chest was done which revealed complete opacification of left hemithorax and abrupt cut-off of left main bronchus. Ipsilateral absence of pulmonary artery and vein was appreciated. There was mediastinal shift towards the left side with left dome of diaphragm raised. Right lung showed hypertrophy and herniated to the contralateral side.

Patient's symptoms were diagnosed to be due to gastritis with acid reflux which resolved with a course of antacids, prokinetics and proton pump inhibitors.

III. DISCUSSION:

Many cases of pulmonary agenesis, aplasia, and hypoplasia have been identified prenatally, in newborns, babies, adolescents, and adults at various ages [3,4]. Anencephalic babies may develop bilateral pulmonary agenesis, an unusual malformation[2]. Unilateral agenesis, aplasia, and hypoplasia are somewhat more common, with few signs and non-specific findings, and only one-third

of cases are diagnosed during life[1]. Unilateral agenesis and aplasia are functionally identical. The sole lung is larger than average, suggesting true hypertrophy rather than emphysema[2].

Agenesis and aplasia of the lungs are caused by a halt in the development of the primitive lung during embryonic life. Obviously, the sooner the arrest happens in the development process, the more serious the defect. The bronchial tree, pulmonary parenchyma, and pulmonary vasculature do not develop in pulmonary agenesis. A rudimentary bronchial pouch is present in pulmonary aplasia. The lesion that results may affect one lobe or the entire lung; focal or bilateral defects are uncommon. Other non-pulmonary abnormalities such as microphthalmia/anophthalmia, cleft palate, heart defects, congenital diaphragmatic hernia/eventration, and limb abnormalities can be associated with pulmonary agenesis or aplasia[5]. There is a high prevalence of related cardiac, gastrointestinal, genitourinary, skeletal, and central nervous system malformations (>50.0 percent) as well as the VACTERL sequence[1–3,6].

Pulmonary agenesis and aplasia are very rare conditions with an estimated prevalence of 0.0034 percent to 0.0097 percent[7]. Agenesis, aplasia, and hypoplasia more frequently involve the right lung[8]. The aetiology could be affected by genetic, teratogenic, and mechanical factors[1,3]. Only a few records of these disorders occurring in siblings in an autosomal recessive pattern have been made. They are equally common in both sexes and affect both lungs equally[3].

When respiratory difficulty arises with tracheal deviation, a clinically symmetric lung, and a chest X-ray indicative of massive atelectasis with mediastinal change, the diagnosis should be suspected[2]. CT chest, which provides a detailed description of bronchial tree, parenchyma, and vasculature is considered to be the most definitive investigation to diagnose agenesis when chest radiograph is not diagnostic[9]. Bronchography is almost obsolete now, but bronchoscopy is useful to demonstrate rudimentary bronchus.

Pulmonary angiography or magnetic resonance imaging angiography is considered to show the absence of ipsilateral pulmonary vessel.

In asymptomatic cases, no treatment is needed[2]. Lower respiratory tract infections necessitate medication. If postural drainage and antibiotics fail to resolve the infection, patients with stumps may require surgical removal of the stump[1] .

Prognosis depends on the degree of pulmonary involvement, a history of recurrent pulmonary infections, and the presence of associated anomalies[5]. Since the tracheobronchial tree is compressed by the shifting of usually midthoracic structures into the right chest[3], patients with right lung agenesis have a higher mortality rate than those with left lung agenesis. A nearly average life span can be expected if the patient survives the first five years without significant infection.

IV. CONCLUSION:

Pulmonary aplasia is a rare congenital lung defect and even more rarer when found in adults without any associated symptoms. Routine investigations of any patient with chest pain should include chest X-ray, even if pain is suggestive of any non pulmonary etiology.

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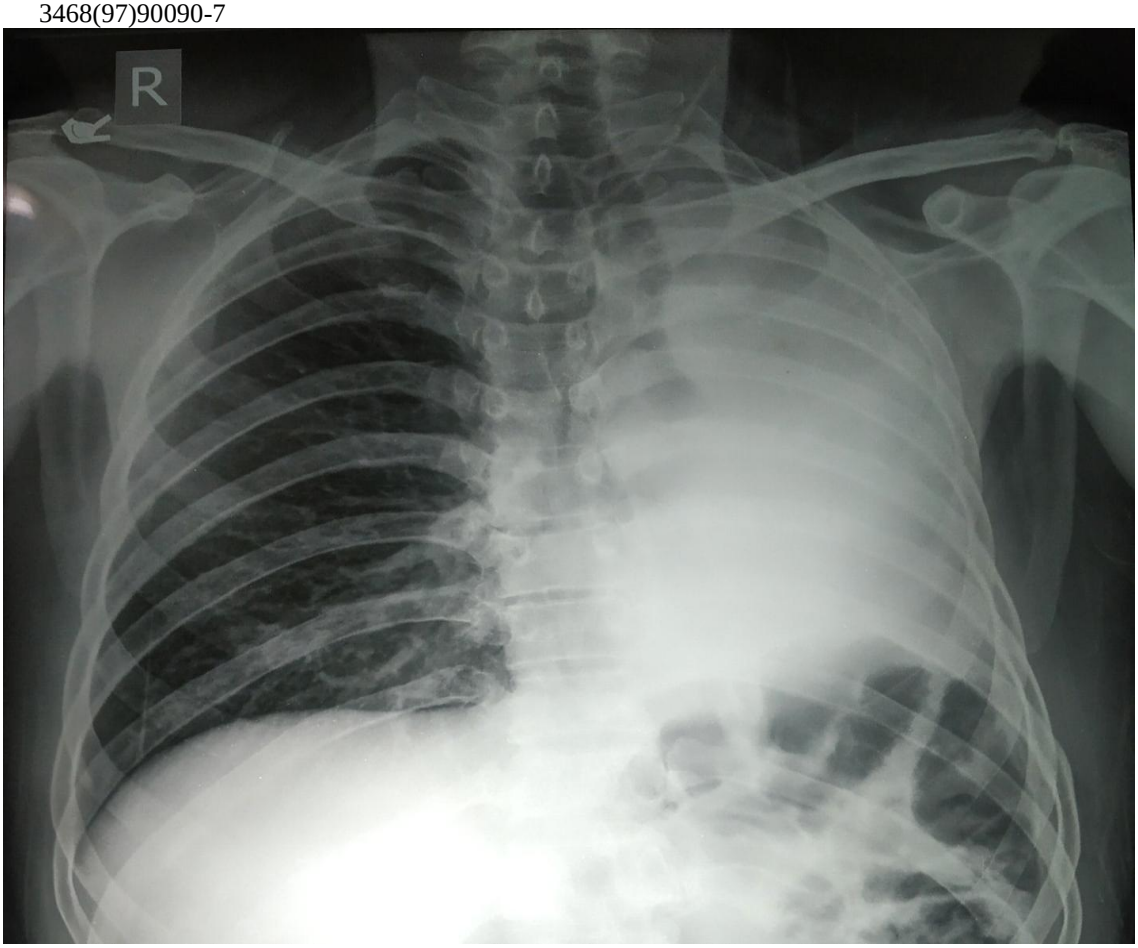
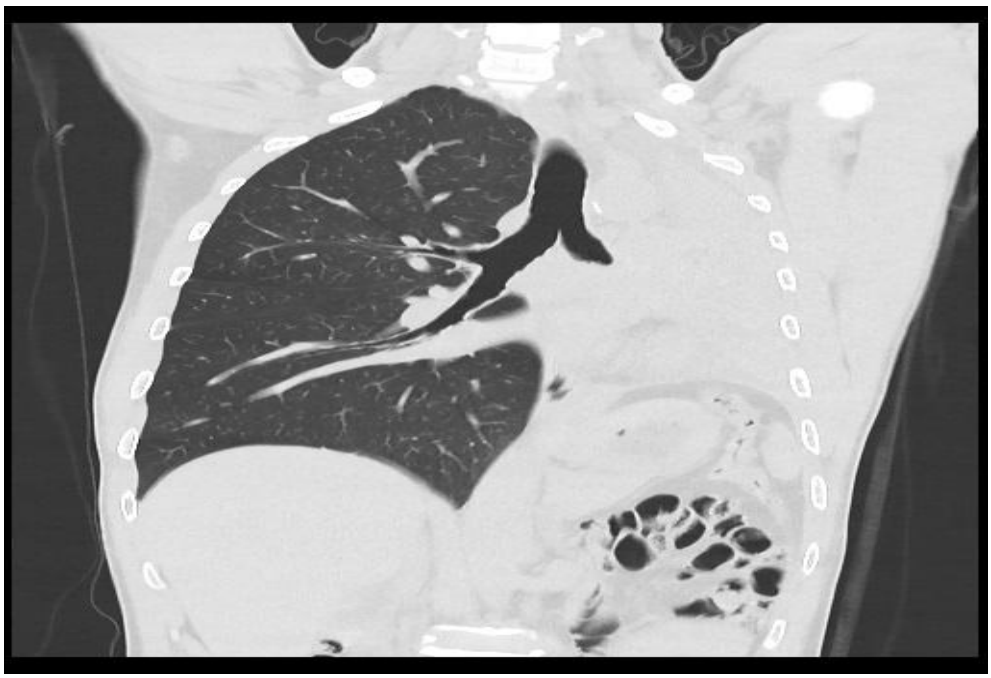


Chest X Ray PA view shows homogenous opacification of left lung with mediastinal shift to the same side



CT coronal section displaying bronchus cut off sign

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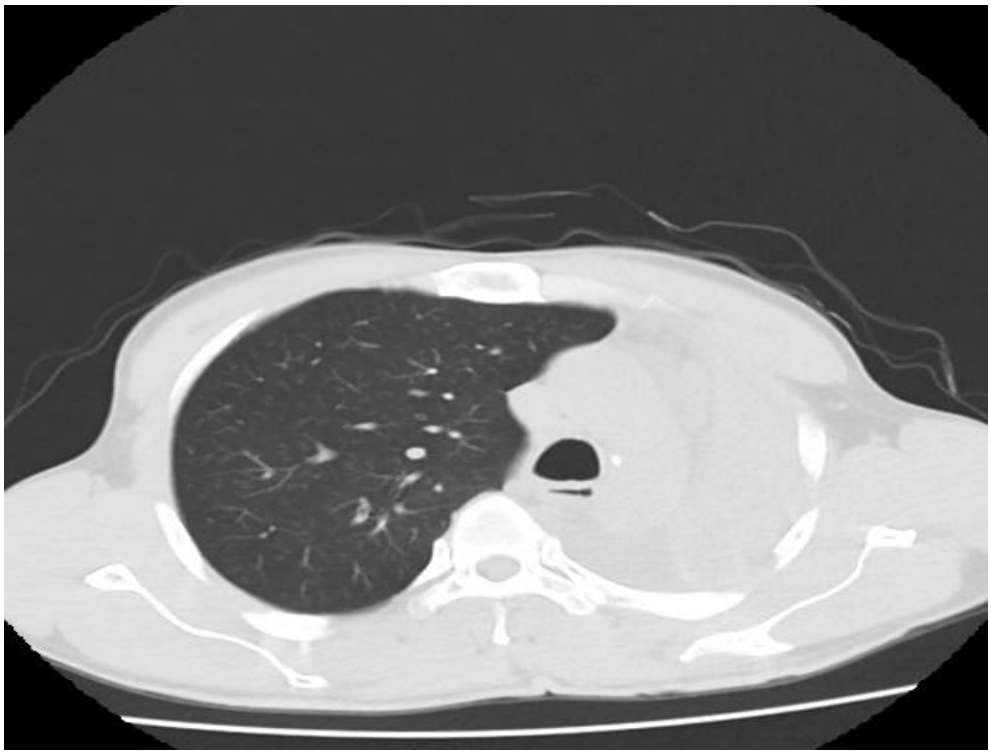

CT Transverse section showing right lung hyperinflation and left lung agenesis