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PULMONARY APLASIA – A CASE REPORT OF THIS RARE PATHOLOGY

Radiodiagnosis

Dr. Chaitnya D. Patel* Resident Doctor, Department of Radiodiagnosis and Imaging, Govt Medical College and New Civil Hospital, Surat *Corresponding Author

Dr. Viral D. Panchal Senior Resident, Department of Radiodiagnosis and Imaging, Govt Medical College and New Civil Hospital, Surat

Dr. Bhagvati V. Ukani Associate Professor, Department of Radiodiagnosis and Imaging, Govt Medical College and New Civil Hospital, Surat

Dr. Rajat D. Arora Resident Doctor, Department of Radiodiagnosis and Imaging, Govt Medical College and New Civil Hospital, Surat

ABSTRACT

Chest Xray findings of unilateral opacification of hemithorax with mediastinal shift towards the affected side may prompt the differential diagnoses of complete lung atelectasis, suspected foreign body aspiration, bronchial mass lesions. We must also consider the rare congenital condition of pulmonary underdevelopment, which includes pulmonary agenesis, aplasia and hypoplasia. We report a case of 20 years old female with left sided pulmonary aplasia.

KEYWORDS

Pulmonary agenesis, Pulmonary aplasia, congenital condition

INTRODUCTION

Schneider classified agenesis into three groups, which has been subsequently modified by Boyden. Depending upon the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories:

Type 1 (Agenesis) – Complete absence of lung and bronchus and no vascular supply to the affected side.

Type 2 (Aplasia) – Rudimentary bronchus with complete absence of pulmonary parenchyma..

Type 3 (Hypoplasia) – Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

CASE REPORT

A 20 years old female presented to Medicine OPD at Government medical college and new civil hospital, Surat with chief complaints of prolonged breathlessness . On clinical examination , findings were as follows:

Tachycardia (HR=120/min), Tachypnea (RR=28/min) , with mild subcostal recession.Chest was normal in shape with slightly reduced movements on left side. Trachea was deviated towards left side.Apex beat was palpable on the left side at 5th Intercostal space in anterior axillary line. There was dull percussion note on the whole left side of chest. Breath sounds were reduced on left side of chest.

Xray chest showed complete opacification of left hemithorax with ipsilateral mediastinal shift. Lung on the right side was hypertrophied with gross herniation towards left.

Multislice CT-Chest with non-ionic IV contrast was advised.

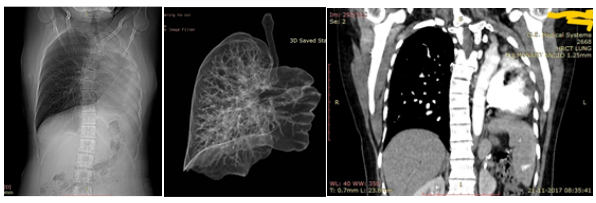

Figure 1: Coronal CT image and its volume rendered image showing left lung aplasia.

On CT-scan , left lung parenchyma not visualized . Only rudimentary left main bronchus noted. Left pulmonary artery and left superior and inferior pulmonary veins not visualized. Resultant shifting of mediastinal structures and trachea towards left side with overcrowding of ribs noted on left side with retrosternal and prevertebral herniation of right lung parenchyma . Left dome of diaphragm appears elevated. Right superior and inferior pulmonary veins appear dilated.

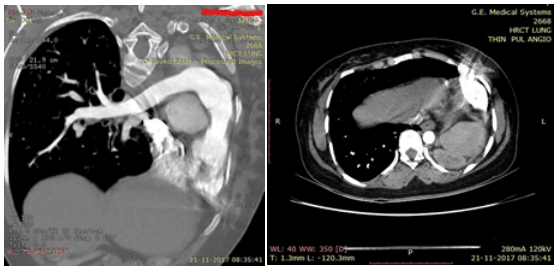

Figure 2: CT image showing retrosternal and prevertebral herniation of right lung

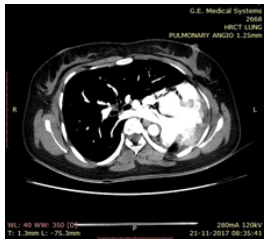

Figure 3: Axial CT-image showing left lung aplasia with absent left pulmonary artery . Only right pulmonary artery visualized arising from main pulmomary artery (arrow)

DISCUSSION

Our patient would classify as Type 2 (Aplasia).

The onset of symptoms in pulmonary aplasia is remarkably variable. In many cases, presence of this anomaly usually comes to light during infancy because of recurrent chest Infections, cardiopulmonary insufficiency or due to associated congenital anomalies.

Nearly 50% cases of pulmonary agenesis have associated congenital defects, involving cardiovascular, skeletal, gastroint estinal and genitourinary system.

The exact aetiology of this condition is unknown although genetic factors, viral agents and dietary deficiency of Vitamin A during pregnancy have been implicated.

Left sided agenesis is more common and these subjects have a longer life expectancy than those with right sided agenesis. This is probably due to excessive mediastinal shift and malrotation of carina in right sided agenesis which hinders proper drainage of the functioning lung and increases chances of respiratory infections.

Abnormal blood flow in the dorsal aortic arch during the 4th week of gestation had been hypothesized to cause pulmonary agenesis . The contralateral lung may develop as much as twice more alveoli in response to pulmonary aplasia / agenesis .

Differential diagnosis includes Complete lung atelectasis , thickened pleura, destroyed lung and pneumonectomy , pulmonary hypoplasia Final diagnosis can be established after bronchoscopy , bronchography but, in some cases, angiography is also needed.

Diagnosis should be suspected when respiratory difficulty occurs with tracheal deviation, in the presence of a clinically symmetric chest and Chest X-ray suggestive of massive atelectasis with mediastinal shift .

No treatment is required in asymptomatic cases .

Treatment is necessary for lower respiratory tract infections.

Prognosis depends on two factors. Firstly , the severity of associated congenital anomalies and secondly , involvement of the normal lung in any disease process.

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