A 21 Years Old Woman, with Chest Pain and Cardiac Dextroposition Pulmonary Hypoplasia or Scimitar Syndrome?

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A B S T R A C T

Background: Physicians seldom encounter chest x-ray of adult patients diagnosed with dextrocardia.
Case presentation: Here, we present the case of a 21-year-old woman who referred to the hospital with the clinical diagnosis of dextrocardia but, she has pulmonary hypoplasia and this rare disease in adults may be a component of scimitar syndrome and scimitar sign is not pathognomonic to define the syndrome. MR angiography was done, but finally the diagnosis was hypoplasia of right upper lobe and aplasia of middle lobe.
Conclusion: As lung hypoplasia is rare in adults, some cases are misdiagnosed as dextrocardia on chest x-ray.

►*Implication for health policy/practice/research/medical education:* As lung hypoplasia is rare in adults, some cases are misdiagnosed as dextrocardia on chest x-ray.

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1. Introduction:

Congenital pulmonary hypoplasia is a rare developmental abnormality of the lung with an incidence rate of around 1/5000000. As a fatal condition associated with respiratory insufficiency after birth, this disease is rare in adults (1).

Thoracic congenital malformations may go unnoticed and unsuspected until adulthood. They should be kept in mind when reviewing unusual CT scans of the chest in adults (2).

The respiratory system begin to develop at 3 week of gestation, and aberration in developmental process may give rise to a group to structural abnormalities collectively referred to as broncho-pulmonary foregut malformation (BPFMs) (3).

According to Boyden, there are three degrees of mal development: a) agenesis, in which there is complete absence of lung tissue, b) aplasia, in which rudimentary bronchus present but no lung tissue is present and c) hypoplasia, in which all the normal pulmonary tissue are present but under-developed (4). Congenital lung anomalies are increasingly diagnosed in adults, because they are missed in childhood. They have unique

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manifestation, often mimic other thoracic pathology, and can present acutely and necessitate emergent evaluation and management (5).

Congenital anomalies of the chest are an important cause of morbidity in infants, children and even adults. The evaluation of affected patients requires multiple imaging modalities to diagnosis the anomaly and plan surgical correction (6).

Encounterig a developmental lung anomaly in adult can be a challenge, as the abnormally may be mistaken for something more sinister (7). This article introduces a case of lung hypoplasia, which was a misdiagnosed as dextrocardia.

2. Case Report:

A 21 year old woman, college student presented to our hospital with chest pain in right hemithorax radiating into the right axilla since 2 years ago which was aggravated from 2 weeks before presentation. It was usually a dull aching pain with no relationship to the respiration. She was able to perform her daily living activities and had no dyspnea on exertion.

She did not report cough, night sweat, and palpitation.

Her past medical history was unremarkable. Her brother has von Recklinghausen disease. Standard Chest X-Ray - past-anterior revealed shifting of heart and mediastinum to right side.

A spiral CT scan of lung and mediastinum was performed which revealed volume decrease in the right side with cardiac and mediastinal shift to right. Diaphragmatic elevation in the right side and vessels in the base of right lung had been decreased. Abdominal sonography was normal. Echocardiography revealed good left ventricular systolic function.

In bronchoscopy by Olympus CLE-10 (made in japan) the right upper bronchus had only one segment was at aberrant site and no evidence of right middle lobe bronchus was seen.

According to CT scan and brochoscopy, congenital lobar aplasia and hypoplasia of the Lung was suggested.

To confirm the diagnosis, CT angiography was performed. In CT angiography, congenital right airway system anomaly, absence of bronchus intermedius, middle lobe, and hypoplasia of right upper lobe bronches was revealed. There was also associated anomaly of arterial branches plus ipsilateral mediastinal shift and elevation of right hemidiaphragm.

The triad of respiratory distress, right lung hypoplasia and edextro of the heart should alert the clinician to think of scimitar syndrome. MR angiography was performed and anomalous venous connection was excluded.

3. Discussion:

This case may be isolated Lung hypoplasia-Aplasia or may be part of scimitar syndrome. Scimitar syndrome is a rare congenital disease (9).

This is a clear bimodal presentation of this syndrome with either as infantile manifestation or a pediatric/adult form. The patient with the adult form is less severely affected and maybe asymptomatic on diagnosis (10).

The syndrome is characterized by partial or complete anomalous venous drainage of the right or left lung into inferior vena cava and is commonly associated with hypoplasia of the right lung, pulmonary sequestration, persistent left superior vena cava and dextroposition of the heart (11). As the different components of this malformation can be present in various combinations and the sign of scimitar is not pathognomonic to define the syndrome (12) MR angiography was done and scimitar syndrome in this patient was rolled out.

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