

Congenital Right Lung Agenesis with Dysplastic Sternum - A Constellation of Two Rare Cases

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Abstract

Congenital lung agenesis is a rare disease spectrum which includes complete agenesis, lung aplasia (with rudimentary bronchus) and partial agenesis. It is embryological anomaly arising from failure of unilateral lung bud to develop. Sternal dysplasias are very rare entities with only few reported cases^[1]. We present case report of a 1 year old female with complete right lung agenesis associated with dysplastic sternum and vertebral segmentation anomaly.

Key words: Lung agenesis, sternal dysplasia, opacified hemithorax

INTRODUCTION

Congenital lung agenesis is a rare disease spectrum which includes complete agenesis, lung aplasia (with rudimentary bronchus), and partial agenesis. Sternal dysplasias are very rare entities with only few reported cases.^[1] We present the case report of a 1-year-old female with complete right lung agenesis associated with dysplastic sternum and vertebral segmentation anomaly.

A 1-year-old female infant with clinical complaints of recurrent pneumonia was referred to the department of radiodiagnosis for chest evaluation. On examination, the patient had apex beat on the right side. The breath sounds were absent on the right side and the right chest wall motion was reduced.

Scout computed tomography (CT) film showed completely opacified right hemithorax with mediastinal shift toward the right side. Cardiac silhouette and cardiac apex were absent from the left side. The left lung was

hyperinflated and herniated across anterior junctional line [Figure 1].

CT chest lung window showed no lung tissue on the right side. The right main bronchus was absent. Trachea continued into the left main bronchus which further divided into two lobar bronchi. The left lung was hyperinflated and herniated toward the right side across anterior and posterior junctional lines [Figure 2].



Figure 1: Scout computed tomography film showing completely opacified right hemithorax with mediastinal shift toward the right side. Cardiac silhouette and cardiac apex were absent from the left side. The left lung was hyperinflated

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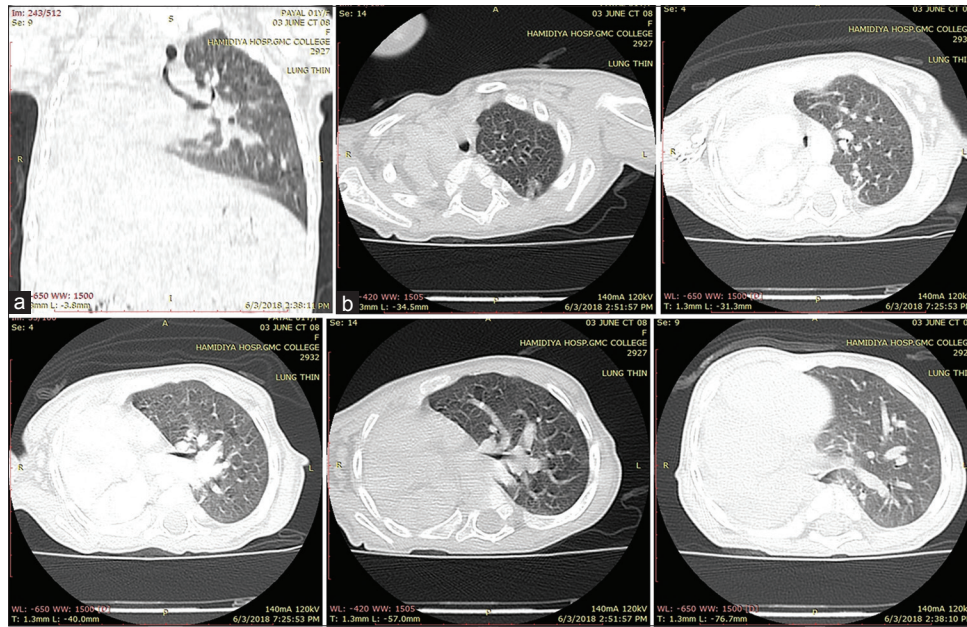


Figure 2: (a): Coronal reformatted computed tomography chest image in lung window showing absent right main bronchus, (b): Absent right lung parenchyma with herniation of the left lung across anterior and posterior junctional lines

Contrast-enhanced CT chest mediastinal window showed heart occupying the right hemithorax with cardiac apex pointing toward the right side; however, aortic arch was the left sided. The right pulmonary artery was not visualized. Few prominent superficial veins were noted over the right hemithorax which drained into the right brachiocephalic vein; however, no venous thrombosis was seen [Figure 3].

CT chest bone window depicted dysplastic sternum with abnormal development of the manubrium, the presence of a hypoplastic body, and the absence of the xiphoid process. There is also an alteration of the sternoclavicular joint [Figure 4].

Body and spinous process of D8 vertebra were unfused in midline [Figure 5]. The left lung parenchyma was normal.

POINTS TO PONDER

- Congenital lung agenesis can be diagnosed antenatally by target scan.
- It should be considered in differentials of patients presenting with absent or reduced breath sounds, completely opacified hemithorax, and ipsilateral mediastinal shift in any age group. Case report of congenital lung agenesis diagnosed in 24-year-old patient does exist!^[2]
- Although it appears to be very morbid condition scientifically, case reports of asymptomatic patients

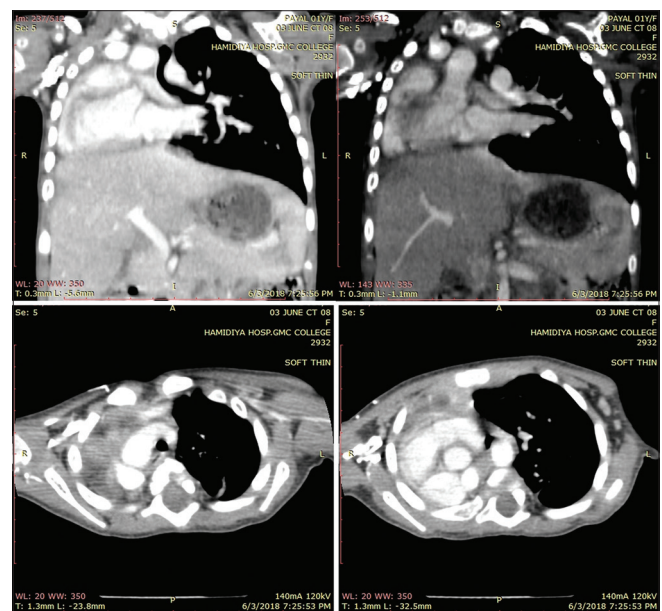


Figure 3: Contrast-enhanced computed tomography chest mediastinal window showing shifting of mediastinum toward the right side with heart occupying the right hemithorax. Cardiac apex is toward right; however, aortic arch is toward left

have been published. Oldest patient of lung agenesis reported at autopsy was 72 years old.

- In literature, the right lung agenesis is considered to have a poorer prognosis and is often associated with cardiac anomalies. It can be attributed to the fact that both lung bud development and cardiac migration occur around 4–5 weeks.^[3]

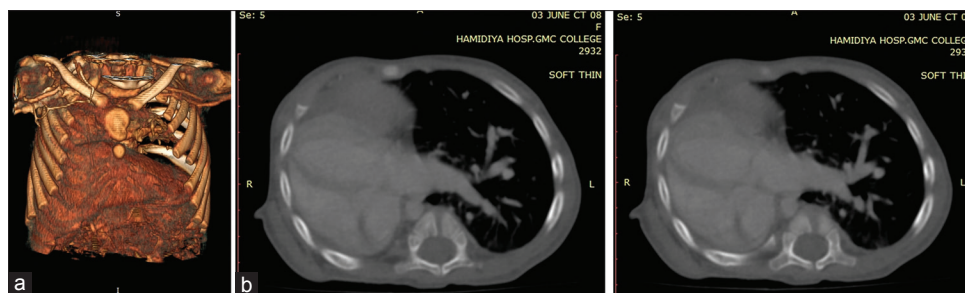


Figure 4: (a): Dysplastic sternum with abnormal development of the manubrium, the presence of a hypoplastic body, and the absence of the xiphoid process, (b): Unfused D8 vertebral body and spinous process

- In almost half of the cases, lung agenesis is associated with some other systems anomaly; like in this patient, it is associated with skeletal system anomaly. So be on lookout for other anomalies to catch!
- Difference between lung agenesis and aplasia is that rudimentary bronchus is found in lung aplasia, while it is completely absent in lung agenesis.^[4]
- Dysplastic sternum is a very rare anomaly and is usually associated with other chest wall abnormalities.

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