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Pulmonary Aplasia: A CT Appearance

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The spectrum of pulmonary congenital anomalies due to developmental arrest of the lungs ranges from pulmonary agencies which signifies complete absence of the tracheo-bronchial tree, the pulmonary parenchyma and vasculature to pulmonary hypoplasia where the gross morphology of the lung is unremarkable but number and/or size of airways, alveoli and vessels is decreased. The spectrum is completed by pulmonary aplasia where a small rudimentary bronchus which ends in a blind pouch is present without any pulmonary parenchyma or vasculature(1). However, pulmonary agencies and aplasia have often been unfairly clubbed together because of clinical convenience and diagnostic difficulty(2). This arrangement overlooks the clinical significance of pulmonary aplasia where the blind bronchial pouch can often be the focus of repeated infections which adversely affects the prognosis of such patients(3). Recent advances in imaging have overcome the problems posed by unpleasant invasive techniques necessary for diagnosis(4). So far, probably less than a dozen reports of the two conditions have been published from the Indian subcontinent(5). The rarity of reports in Indian literature prompted the present description of a 10-year-old boy with aplasia of the left lung diagnosed with the help of CT thorax.

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Case Report

An asymtomatic 10-year-old boy was referred to our Institute for a flattened left hemithorax detected on routine school medical check-up. He was the first born of a non consanguinous marriage with uneventful antenatal and postnatal periods. He had normal milestones and had received complete immunization. There was no family history of congenital anomalies. Chest examination revealed a mediastinal shift to the left along with other features suggestive of left sided volume loss. On auscultation, normal vesicular breath sounds were audible on the right side but were reduced in intensity on the left side. Heart sounds, though normal in character, were distant.

A review of chest roentgenograms demonstrated an opaque left hemithorax with ipsilateral shift of mediastinum and associated herniation or right lung (*Fig. 1*). Fibrebronchoscopy visualised a small 1.5 cm long left main bronchus ending in a

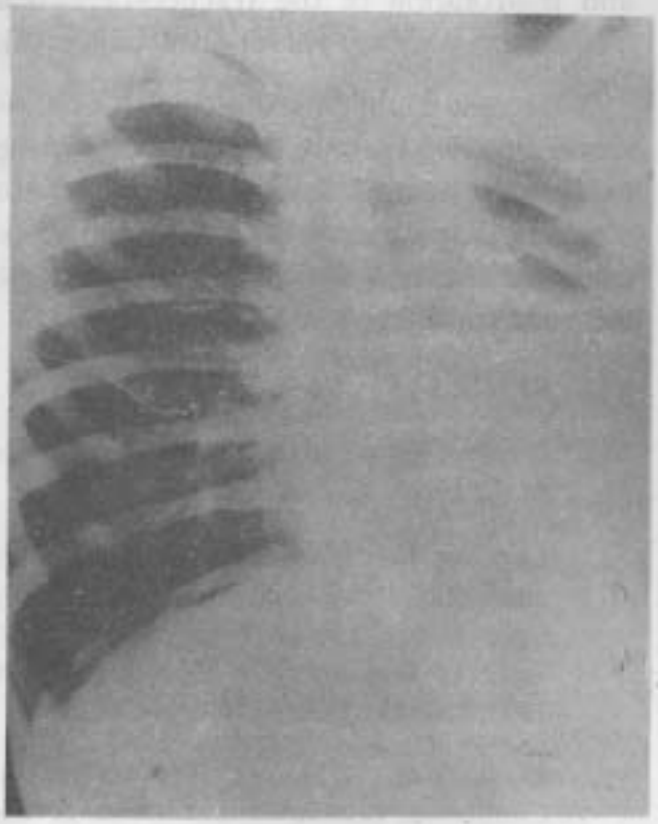

Fig. 1.Chest roentgenogram showing an opaque left hemithorax with ipsilateral shift of the mediastinum and herniated right lung.

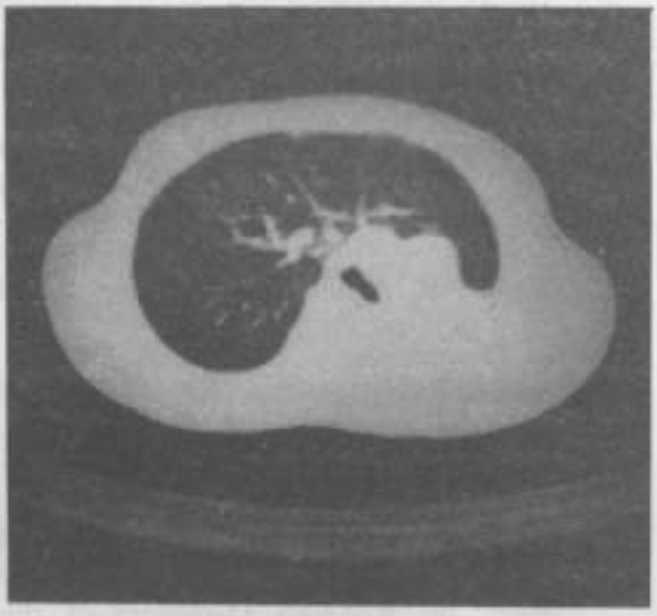

Fig. 2.CT thorax–section at level of carina: showing normal left and right main bronchi, absent left lung parenchyma and herniated right lung.

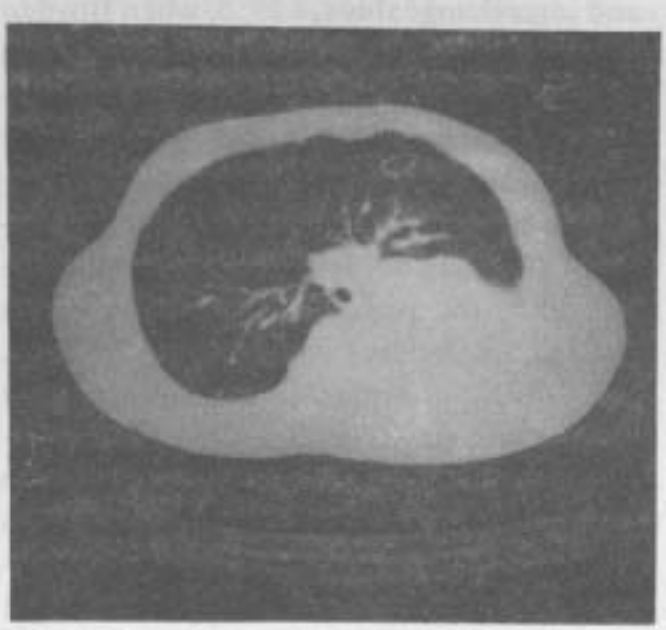

Fig. 3.CT thorax–section 1 cm below the level of carina: showing absent left main bronchus and herniated right lung with a normal right main bronchus.

blind pouch with the trachea and carina shifted to the left. The tracheo-bronchial tree on the right side was normal. A study of the serial sections of the CT thorax (*Fig. 2 & 3*) revealed an approximately 1-2 cm long left main bronchus with an absent left lung. Preaortic and precardiac herniation of the normal lung with posterior displacement of the heart was also visualised. Electrocardiogram showed low voltage normal waves with clockwise rotation of the heart. Echocardiogram demonstrated a 17.3 mm diameter main pulmonary artery, right branch of 11.5 mm and a small vestigeal left trunk of 1 cm length. Atrial and ventricular functions were essentially normal as were the skeletal survey and ultrasonographic examination of the abdomen. A diagnosis of aplasia of the left lung was made.

Discussion

The terms pulmonary agenesis, aplasia and hypoplasia were used inconsistently and interchangeably till 1955, when Boyden defined them(1). Clinically, the confusion still persisted(2) because unpleasant invasive procedures like bronchography, pulmonary angiography were necessary to establish the diagnosis. Furthermore, majority of the patients investigated usually belonged to the pediatric age groups where invasive procedures are generally disliked. The advent of advanced computerized non invasive imaging techniques like CT of thorax and echocardiogram have helped, as in our case, to establish the exact diagnosis without resorting to these invasive procedures. The short blind stump of the left main bronchus and absent left pulmonary parenchyma were easily discernible on CT thorax while the echocardiogram demonstrated a vestigeal left pulmonary artery.

Pulmonary agenesis and aplasia are said to arise as a result of embryological defect in the 5th week of intrauterine life(6). However, these two conditions should not be clubbed together because of their distinct clinical implications. In pulmonary aplasia, unlike agenesis, the bronchial stump acts as a reservoir for infected secretions. This may cause spillover of infected material to the normal lung affecting the prognosis of such patients(3). If the patient survives the first five years of life without any major infection, as in our case, an almost normal life span can be expected(7). Another favorable factor in our patient, which could explain his asymptomatic state, was the involvement of the left side as right sided lesions are reported to have a mortality twice that of left sided ones. This has been attributed to the greater mediastinal shift which occurs towards the right side leading to distortion and malrotation of the tracheo-bronchial tree alongwith great vessel disturbances(8).

Our case highlights the role of non invasive imaging techniques in distinguishing pulmonary aplasia from agenesis. It also reminds us of the distinct clinical significance of the two conditions necessitating the need to differentiate them.

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