Case Reports

Isolated Unilateral Pulmonary Agenesis

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Two cases of isolated pulmonary agenesis with no other associated congenital abnormality are reported. First case was 13-year-old girl who presented for the first time with chest infection and was found to have isolated unilateral pulmonary agenesis and responded well to antibiotics, Second case was a 10-year-old girl who had presented with total duration of illness of one year with clinical features suggestive of chronic chest infection with family history of pulmonary tuberculosis and later found to have agenesis of upper lobe of left lung in addition to having gastric aspirate positive for Mycobacterium tuberculosis. She responded to anti tubercular therapy.

Keywords: *Pulmonary agenesis.*

Pulmonary agenesis is a rare condition as a result of embryological defects, usually unilateral and is associated with skeletal, cardiovascular and other anomalies(1). Isolated pulmonary agenesis without any other associated anomaly except absence of right pulmonary artery is being reported for its rarity.

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Case Report

Case 1

A 13-year-old girl presented with main complaints of poorly localized chest pain, fever, cough and expectoration for the last two months. There was no past history of similar complaints. Her two siblings were asymptomatic. The perinatal and developmental history was normal. No parental consanguinity was present. She was moderately built and nourished with mild pallor. There was no jaundice, edema, significant lymphadenopathy or any obvious congenital abnormality. Chest examination showed diminished movements of chest on the right side with trachea deviated to the right side. No breath sounds could be heard over the right hemithorax. Normal heart sounds were heard on the right side. Rest of the examination was unremarkable.

X-ray chest (*Fig. 1*) showed complete opacification of right hemi-thorax with hyperinflation of left lung and herniation to the contralateral side with shifting of mediastinum to right side. Pulmonary function tests showed moderate restriction with no airway obstruction. Three consecutive sputum examinations for AFB were negative. Sputum pyogenic cultures showed *S. pneumoniae*.

HRCECT scan chest (*Fig. 2*) showed no evidence of any lung parenchyma on the right side. Apical region showed a small soft tissue with focal calcification. Pulmonary trunk was

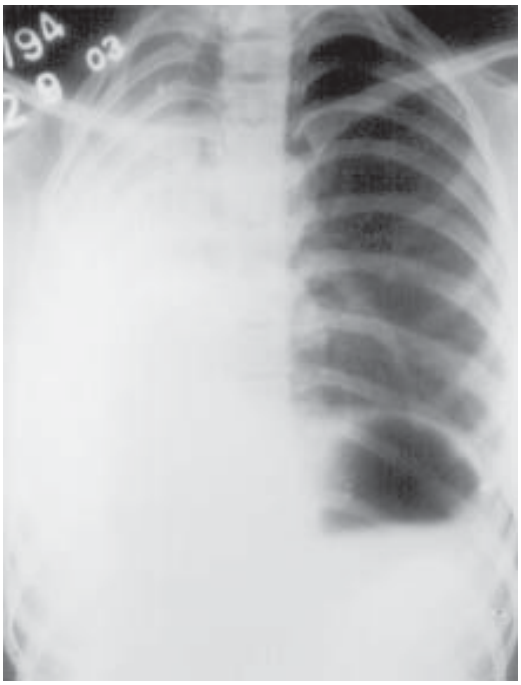

Fig. 1. Showing complete opacification of right hemithorax with hyperinflation of left lung.

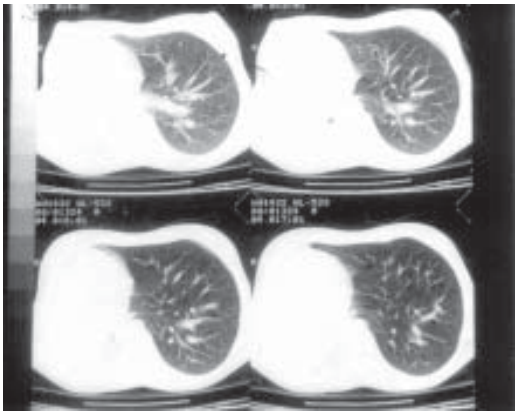

Fig. 2. Showing HRCECT of chest with absent lung parenchyma on right side, absence of right branch of pulomonary artery.

located anterolateral to ascending aorta with absent right branch of pulmonary artery. Echocardiographic examination showed dextroposition of the heart with no other cardiac anomaly. Ultrasound abdomen, *X*-rays of hands, face, spine, pelvis and lower limbs were normal. Bronchoscopy confirmed the diagnosis.

The pneumonia responded to a course of amoxycillin and is on follow-up regularly.

Case 2

Second case was a 10-year-old girl born of non-consanguineous parents. She presented with fever, cough and expectoration of about one year duration. She also complained of progressive, non-paroxysmal dyspnea. There was no past history of similar complaints, tuberculosis, measles, diabetes and chicken pox. Both parents had sputum positive pulmonary tuberculosis and had completed anti tubercular therapy and were cured. She was moderately built and nourished. Chest examination revealed diminished movements over left side of chest with trachea and apex beat shifted to the left side. Auscultation showed no breath sound over interscapular and left supra-scapular region with coarse crackles over left infra-scapular region. Cardiovascular examination was normal.

X-ray chest PA view showed a homogeneous shadow over left upper zone and a non- homogeneous shadow over left lower zone with trachea and heart shifted to the same side, Left lateral film showed a homogenous shadow over posterior mediastinum.

HRCECT scan chest (*Fig. 3*) showed reduction in volume of left hemithorax, left upper zone had low attenuating soft tissue opacity with no airways and communication with the bronchial tree. Left upper lobe bronchus was not visualized, Left lower lobe showed parenchymal destruction with cavitary changes. Right lung was hyper inflated with herniation. Pleura was normal. These features were suggestive of complete

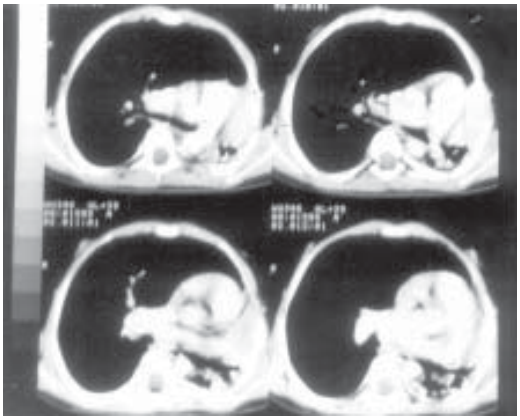

Fig. 3. HRCECT chest shows reduction in lung volume of left hemithorax with no airways and communication with the bronchial tree. Left lower lobe showed cavitory changes.

bronchial atresia of left upper lobe with destroyed left lower lobe with compensatory over inflation of right lung, There was agenesis of upper lobe of left lung.

Bronchoscopy under general anesthesia showed normal right bronchial tree. Left upper lobe bronchus was absent and left lower lobe bronchial tree showed inflammation with mucoid secretions. Acid fast bacilli were isolated in bronchial washings and also in gastric aspirate suggesting co-infection with pulmonary tuberculosis. Category I anti tubercular therapy was started.

Discussion

Pulmonary agenesis usually presents in childhood but may present in later life. If not complicated by other anomalies it is quite compatible with normal living. Pulmonary agenesis needs to be differentiated from pulmonary hypoplasia by the absence of bronchial tree on bronchoscopy and the absence of lung tissue on radiological investigations(2). Diagnosis is usually made by chest *X*-ray and CT scan, and invasive procedures like bronchography, bronchoscopy and angiography can be avoided(3). The typical CT findings are opaque hemithorax with mediastinal shift towards the affected side with absence of lung parenchyma; pulmonary and bronchial tree as seen in our Case 1(4,5).

The condition is asymptomatic unless compromised by infection, which can be bacterial (as in Case 1) or even mycobacterial (as in Case 2). Differential diagnosis is from atelectesis, diaphragmatic hernia which can be differentiated with the help of tomography (HRCT), bronchography, and angiography (6). Asymptomatic cases do not require any treatment especially when no other anomalies are present. Chest infections are to be treated energetically. These cases are high-risk cases for any surgery because of low respiratory reserve and anesthetist and surgeon should take appropriate precautions(6).

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