An unusual cause of recurrent respiratory tract infection: unilateral pulmonary agenesis

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Abstract

Unilateral pulmonary agenesis may be asymptomatic or present with recurrent respiratory symptoms like respiratory distress or respiratory tract infections. The computed tomography is found to be most invaluable non-invasive imaging modality in the patient management. We report two cases of unilateral pulmonary agenesis, one presented with recurrent respiratory tract infections and the other was asymptomatic.

Keywords: Pulmonary agenesis; Respiratory infection; Infections

1. Introduction

Unilateral pulmonary agenesis is an extremely rare congenital malformation resulting from genetic, teratogenic and mechanical factors [1]. Unilateral pulmonary agenesis may be compatible with life, provided it is not associated with other congenital malformations or respiratory complications [2]. We describe two cases of unilateral pulmonary agenesis, one patient presented with recurrent respiratory tract infections and the other patient was asymptomatic.

2. Case reports

2.1. Case 1

A 2-month-old male baby admitted to our hospital with a history of recurrent respiratory infections and respiratory distress. The boy was born at term following spontaneous delivery. The baby started to show symptoms like mild fever, wheezing and respiratory distress from 15th day after birth. Physical examination revealed increased respiratory rate and absence of breath sounds on right of chest. There was no cyanosis. A chest radiograph revealed opaque right hemithorax with dextrocardia and hyperinflated left lung crossing the midline. Contrast enhanced CT scan of the chest revealed the absence of right main bronchus and right lung parenchyma (Fig. 1). The right hemithorax occupied by the heart and hyperinflated left lung was herniated to the right anterior hemithorax. Bronchoscopy confirmed the absence of right bronchus.

2.2. Case 2

A 6-year-old female child was admitted to the hospital for adenotonsillectomy. Physical examination revealed dullness in left hemithorax and absent air entry on left side of chest. A chest radiograph revealed opaque left hemithorax with ipsilateral mediastinal shift. Contrast enhanced CT scan revealed the absence of left bronchus and left lung parenchyma (Fig. 2). There was crowding...
of intercostal spaces of left hemithorax. Subsequently, bronchoscopy confirmed the absence of left bronchus.

3. Discussion

De Pozze, who discovered it accidentally during autopsy of a female adult in 1673, first described the condition of pulmonary agenesis [1]. Although there are no definite etiological factors that might be responsible for the development of this malformation, various genetic, teratogenic and mechanical factors have been proposed as the possible causes of pulmonary agenesis. The pulmonary agenesis may occur as an isolated anomaly, however, usually associated with other congenital malformations involving skeletal, gastrointestinal tract and genitourinary systems [3].

Depending upon the extent to which bronchopulmonary tissue is absent, the pulmonary agenesis divided into [4]

1) Bilateral complete agenesis (aplasia)
2) Unilateral agenesis with
   a) complete absence of bronchi, alveolar tissue and blood supply
   b) rudimentary bronchus present but no pulmonary tissue
   c) rudimentary bronchus invested by ill defined pulmonary tissue
3) Lobar agenesis and other abnormalities.

Bilateral pulmonary agenesis is incompatible with life. Whereas unilateral agenesis may be compatible with life, but has a high mortality rate if associated with other congenital malformations. The unilateral pulmonary agenesis is a rare congenital malformation and may present with recurrent respiratory symptoms or it may be asymptomatic [5]. The former is amenable to surgical treatment and symptoms are relieved by pneumonectomy [5]. The differential diagnosis of opaque hemithorax in children with respiratory distress includes pulmonary agenesis, diaphragmatic hernia, congenital adenomatoid malformation and sequestration [6,7].

In conclusion, in children with respiratory symptoms if chest radiograph shows opaque hemithorax with ipsilateral mediastinal shift, pulmonary agenesis should be considered in differential diagnosis and contrast enhanced CT will be more informative in such cases.
References


Fig. 2. (a) Axial CT section of chest shows absence of left lung parenchyma and herniation of right lung across the midline to left side anteriorly. (b) Lung window of the same section shows the absence of left main bronchus.