Atrésie bronchique congénitale révélée par une toux chronique : A propos d'un cas
Congenital bronchial atresia presenting as a chronic cough: a case report

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Résumé :

L'atrésie bronchique représente une malformation congénitale pulmonaire rare, de découverte le plus souvent fortuite. Nous rapportons l'observation d'un nourrisson de 15 mois qui présente une toux chronique révélant une atrésie bronchique congénitale.
L'examen physique a montré un murmure vésiculaire diminué à gauche. La tomodensitométrie thoracique a montré un emphysème lobaire en amont d'une bronche lobaire gauche rétrécie sans obstruction intraluminale ni compression extrinsèque. L'endoscopie a permis d'écarter les autres diagnostics différentiels et de retenir le diagnostic d'atrésie bronchique gauche. L'aspect radiologique de l'atrésie bronchique peut mimer d'autres pathologies respiratoires. L'exploration initiale pour le diagnostic est le scanner thoracique. L'échographie prénatale reste un moyen de dépistage.

Abstract

Congenital bronchial atresia is a rare congenital anomaly of the lung, which usually presents as an incidental finding on routine examination. In this report, we report the case of a 15-month–old boy presenting congenital bronchial atresia revealed by a chronic cough. The physical examination showed decreased breath sound on the left lung. Computed tomography (CT) of the chest revealed a giant lobar emphysema upstream a narrowed left lobe bronchus without intraluminal obstruction or compression. Further examination including bronchoscopy excluded other disorders and the diagnosis of congenital bronchial atresia was made. The radiological presentation of congenital bronchial atresia may occasionally mimic serious lung diseases. The main procedure for diagnosis is CT of the chest. Ultrasound during the prenatal period is useful for the diagnosis.

Mots clés : atrésie bronchique, emphysème, toux, tomodensitométrie, fibroscopie
Key words : bronchial atresia; emphysema;cough;computed tomography; bronchoscopy

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Introduction :

Congenital Bronchial Atresia (CBA) is a rare congenital anomaly of the lung. It has variable appearance and can mimic some acquired lung diseases. It results from focal interruption of lobar, segmental or sub segmental bronchus and leads to distal mucus impaction forming a bronchocele or mucocele and is often associated with regional lung hyperinflation. We report the case of a 15-month –old boy presenting congenital bronchial atresia.

Case Presentation :

A 15 month-old male infant weighting 10Kg, with the history of chronic cough, was referred to our department for an abnormal chest X-ray. He was born by spontaneous vaginal delivery after an uncomplicated full-term pregnancy and has a normal physical examination at birth. He had no history of previous disease.

Physical examination showed decreased breath sounds in the left hemithorax. The cardiac auscultation was normal.

The chest X-ray showed hyperinflation of the upper left lung suggestive of a proximal bronchial obstruction. The routine laboratory tests were within normal range. Bronchoscopy showed a mild degree of extrinsic obstruction at the entrance of the lingual bronchus and no foreign body. The Microbiological and cytological examination of the broncho-alveolar lavage were negative. A computed tomography scan of the chest showed a giant lobar emphysema (figure 1)

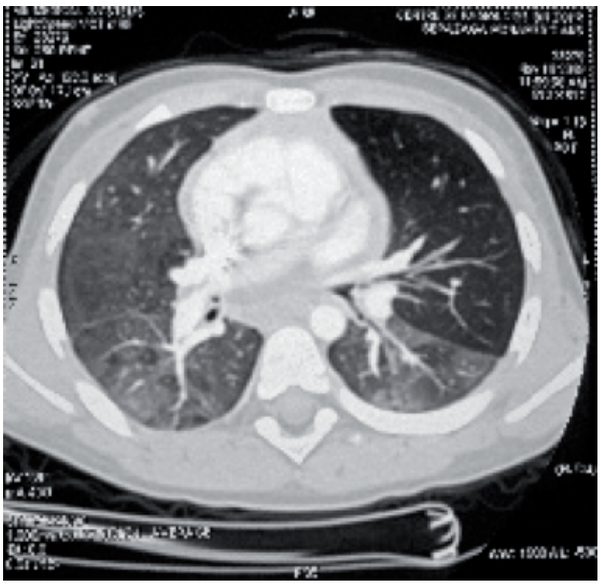

Figure 1 : CT scan showing giant left lobar emphysema

upstream a narrowed left lobe bronchus without intraluminal obstruction or compression (figure 2).

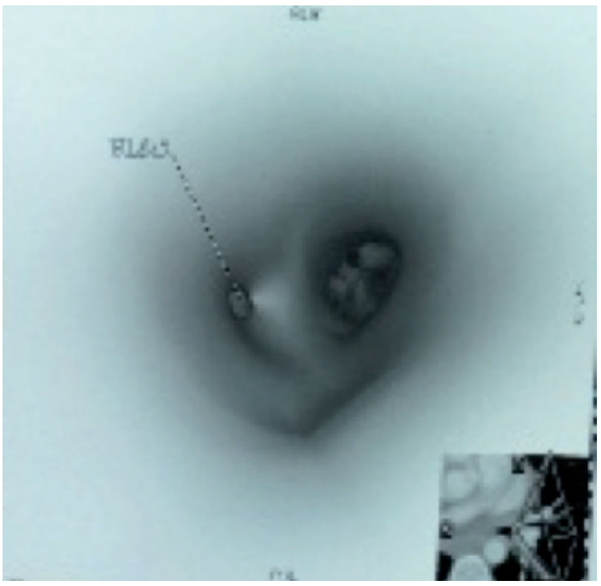

Figure 2: CT scan showing a narrowed left lobe bronchus without intraluminal obstruction or extraluminal compression

The diagnosis of congenital bronchial atresia was made and the patient was discharged without further investigation.

The patient presented on the follow-up several episodes of wheezing and dyspnea. He was treated with inhaled corticosteroids and bronchodilators.

Discussion :

Congenital bronchial atresia is characterized by a mucocele or bronchocele resulting from a mucus-filled, blind-terminating sub-segmental, segmental or lobar bronchus, at or near its origin, and hyperinflation of the isolated lung parenchyma [1]. The first case was reported by Ramsay and Byron in 1953 and since this date more than 150 cases have been reported in the literature [1]. A male predominance in the incidence has been reported with sex-ratio of 2/1 [2].

The exact cause of bronchial atresia is unknown and many mechanisms are proposed to explain the pathogenesis. Bronchial atresia is hypothesized to occur as a focal interruption before birth. One proposal is that bronchial atresia is caused by an intrauterine ischemia after 16th week of gestation. Congenital bronchial atresia has been described with several congenital anomalies (congenital cystic adenomatoid malformation, unilateral renal agenesis, pericardial defect ...) which are known to develop earlier in embryogenesis and suggested the possibility that the lesion develops much earlier, during weeks 4-6 of intrauterine development. The findings of Louw and Barnard support vascular occlusion rather than a failure of growth as the

cause [2].
Patients are often asymptomatic; however they may present cough, dyspnea, and recurrent infections.
Chest radiographs showed an area of pulmonary hyperlucency in 67%, a hilar mass-like shadow in 89%, and both findings in 67% [3]. The Accumulation of secretions and mucoid impaction distal to the bronchial atresia results in ovoid, round and branching opacities near the hilum in most patients. Our patient presented hyperlucency of the upper left lobe.
Chest CT is the most sensitive tool for the diagnosis. It showed mucoid impaction, segmental overinflation, and hypovascularity [3]. Mucoid impaction is readily recognized by the presence of branching soft tissue densities in a bronchial distribution, usually associated with bronchial dilatation. The findings in our patient are: mucocele, occlusion of segmental bronchi, and emphysematous change of the upper left lung lobe.
Flexible-bronchoscopy has a limited role for the diagnosis of CBA. It may be normal up to 50% cases; However in some cases it may reveal a blind-ending bronchus [4]. Flexible-bronchoscopy is interesting to exclude differential diagnosis and to demonstrate the patency of the central bronchi, especially in doubtful cases.
The management of CBA remains debatable. For some authors, surgery with resection of the affected lobe is indicated for all patients; however others believe in conservative approach in asymptomatic and mildly symptomatic patients [5].
Antenatal ultrasonography may be useful for the diagnosis showing hyperechogenic mass at the hilum of the lung which corresponds to a mucocele [6, 7].

Conclusion :

Congenital bronchial atresia is a rare and benign entity, which might mimic serious diseases on radiographic examination. The chest CT scan is the tool of choice for the diagnosis and bronchoscopy had supplementary role especially for excluding other diagnosis.

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