Congenital bronchial atresia: a fortuitous diagnosis

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A 25-year-old woman is admitted in the Radiology Department to evaluate by echography a tumefaction in the right axillary region. Multiple lymph nodes are found and the examination is completed by neck computed tomography. The presence of inflammatory lymph nodes associated with a positive serology for Bartonella Henselae led to the diagnosis of «Cat-scratch disease». Fortuitously, we discovered a hypodense left upper lung. A chest CT in breathing in and out revealed bronchial discontinuity of the apicoposterior left upper lobe bronchus. Associated radiological signs were: air trapping in the culmen while breathing out (Fig. A), local hypovascularization, small bronchoceles (Fig. B, arrow) and the presence of two bronchogenic cysts in the upper (Fig. C) and lower mediastinum.

The patient is asymptomatic and shows satisfying respiratory volumes and flow rates. No treatment is necessary as long as the patient stays asymptomatic; she will be followed in pneumology consultation annually.

Comment

Bronchial atresia is a congenital anomaly of the tracheobronchial tree. It has been described for the first time in 1953. In most case it affects the upper left lobe. It is estimated that 60% of the patients displaying this anomaly are asymptomatic. The atresia is generally discovered as a fortuitous radiologic event. Symptomatology may be persistent cough, recurrent respiratory infections and dyspnea.

The pathophysiological mechanism is not clearly known yet. The most probable assumption is an ischemic phenomenon by obstruction of a bronchial artery during the intra-uterine life. This causes a segmental interruption of the bronchial tree which is totally formed at this time. The distal bronchial tree is normal but alveolar number is diminished in the concerned segment, alveoli are distended rather than destructed. This hyperinflation can be explained by collateral ventilation through the interalveolar pores of Kohn, bronchoalveolar channels of Lambert and interbronchiolar channels. Bronchoceles are typically associated.

Bronchial atresia has to be mentioned when a one-sided lung hyperclarity is discovered. The differential diagnosis includes bronchial tumor, Swyer James syndrome, allergic bronchopulmonary aspergillosis, cystic fibrosis or inhalation of a foreign body (1).

The best diagnostic test is chest CT. The exploration must be completed with a bronchoscopy which is generally negative and enables the elimination of other bronchial obstructive causes.

Associated malformations are possible: bronchogenic cysts (as in this case), sequestration, interauricular communication, patent left superior vena cava, pericardial defect, partial abnormal pulmonary venous drainage.

Management of the patients with bronchial atresia is variable. If the malformation is discovered during childhood, a surgical excision of the concerned lobe can be performed to allow a normal development of the remaining lung. During adulthood, the surgical treatment is meant for patients who present repeated infections. Lobectomy is preferred to segmentectomy.

Reference