Unsuspected case of congenital bronchial atresia in workup for supposed pulmonary AVM

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A 58-year-old man was send to our hospital for embolisation of an arteriovenous (AV) malformation of the right lung. This diagnosis was based on a CT made in the referring hospital. Diagnostic pulmonary angiography with catheter placed in the right main pulmonary artery (Fig. A) did not show an AV-malformation. Nevertheless striking was the diminished number of pulmonary vessels in the superior and middle part of the right lung (arrows). When revising the CT scan a hypodense mass was seen in the right hilum (Fig. B, arrow). This lesion was not vascularised and was located near the right upper lobe bronchus (Fig. C, arrow). The lung segment distal to this mass contained little vessels. These findings are compatible with bronchial atresia. Retrospectively this congenital anomaly could already be suggested based on the chest X-ray (Fig. D), which showed a perihilar mass (short arrow) and decreased vascular marking of the right peripheral lung zones (arrows).

Comment

Bronchial atresia is defined as congenital atresia of a segmental bronchus with normal distal architecture. Etiology is supposed to be related to bronchial ischemia in utero between 5th and 15th week of gestation. The bronchus loses communication with the bronchial bud. Mucus accumulates and a bronchocele develops. Due to hypoventilation oligemia ensues. Hyperinflation of the alveoli of the effected segment with airtrapping is possible from collateral air drift. The best diagnostic clue is a round, sharply defined perihilar mass with distal hyperinflation. CT is the best diagnostic tool to characterize the bronchocele, airway anatomy, distal hyperinflated lung and other associated anomalies. Because this congenital anomaly in most cases does not cause symptoms, the lesion is only incidently discovered, frequently just in adulthood. Treatment is usually not necessary. Surgical resection is reserved for those with recurrent infection or encroachment of normal pulmonary structures. The differential diagnosis consists of other lesions with mucoid impaction and hyperinflation as intralobar sequestration and intrapulmonary bronchogenic cyst. In an AV-malformation a contrast study can differentiate this lesion from a bronchocele, which does not enhance after intravenous contrast administration. Congenital lobar emphysema is also associated with hyperinflation but contains no bronchocele and is usually more extensive and diagnosed in infants with respiratory distress. Allergic bronchopulmonary aspergillosis may have mucoid impaction in central bronchiectasis, but is usually bilateral and patients have asthma. In Swyer-James syndrome patients have a history of recurrent infection and bronchiolitis obliterans in childhood. Imaging can look similar due to involvement of only one lung segment, hyperinflation and hypoplasia of the pulmonary artery.

Reference