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Computed Tomography / Tomodensitométrie

Imaging Appearances of Congenital Thoracic Lesions Presenting in Adulthood

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Abstract

Many congenital lesions of the thorax are detected for the first time in adulthood when they can simulate a wide range of pathologies, including infection and neoplasia. They can be broadly classified into tracheobronchial, parenchymal, vascular, and combined parenchymal/vascular abnormalities. An awareness of their typical imaging features enables a confident diagnosis and helps direct appropriate patient management.

Abrégé

Bon nombre de lésions congénitales du thorax sont décelées pour la première fois à l'âge adulte, alors qu'elles peuvent simuler toutes sortes de pathologies, notamment des infections et des néoplasies. On peut en gros les considérer comme des anomalies trachéobronchiques, parenchymateuses, vasculaires ou parenchymateuses-vasculaires combinées. Une bonne connaissance de leurs caractéristiques d'imagerie type permet de poser un diagnostic fiable et favorise une prise en charge adéquate du patient.

Key Words: Congenital; Thoracic; Tracheal bronchus; Tracheobronchomegaly; Cystic adenoid malformation; Bronchogenic cyst; Pulmonary hypoplasia; Scimitar syndrome; Pulmonary sequestration

Many congenital lesions of the thorax are detected for the first time in adulthood when they can simulate a wide range of pathologies, including infection and neoplasia. Although some of these conditions are simply incidental findings that require no specific management, others potentially can be life threatening. This review illustrates the plain film and cross-sectional imaging appearances of these conditions, which can be broadly classified into tracheobronchial, parenchymal, vascular, and combined parenchymal/vascular abnormalities (Table 1).

Abnormalities of the Tracheobronchial Tree

Tracheal Bronchus

Tracheal bronchus is an anatomical variant whereby an anomalous airway arises from the lateral wall of the trachea. The majority of tracheal bronchi are right sided and arise within 2 cm of the carina; however, left tracheal bronchi have also been described. Tracheal bronchus has a prevalence of 0.1%−2% from bronchoscopic studies [1] and is more common in patients with Down syndrome. There are many possible anatomical configurations, including a displaced right upper-lobe bronchus, giving rise to all 3 upper-lobe bronchopulmonary segments, or, more commonly, it may just supply the right apical segment [2].

Tracheal bronchus is most often an incidental finding, although, in the critical care setting, attention is drawn to its possibility when persisting lobar atelectasis is noted in

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Table 1
Developmental thoracic lesions that may present in adulthood

Abnormalities of the tracheobronchial tree
Tracheal bronchus
Tracheobronchomegaly
Bronchial atresia
Bronchogenic cyst
Abnormalities of the lung parenchyma
Pulmonary agenesis/aplasia
Pulmonary hypoplasia
CAM
Abnormalities of the vasculature
Vascular rings
Arteriovenous malformation
PAPVR
Combined parenchymal/vascular abnormalities
Scimitar syndrome
Pulmonary sequestration

spite of an adequately positioned endotracheal tube (Figure 1). It may also be associated with recurrent pneumonia, chronic bronchitis, and bronchiectasis. Computed tomography (CT) can eloquently demonstrate the aberrant bronchus and its precise anatomical configuration (Figure 2). Surgical resection is considered in severely symptomatic cases [3].

Tracheobronchomegaly

Tracheobronchomegaly (Mounier-Kuhn syndrome) is a rare condition characterized by atrophy of elastic and smooth muscle fibers in the trachea and central bronchi, which become markedly dilated. An abrupt change to normal calibre occurs at the fourth or fifth order bronchial divisions [4]. It has been reported as a familial trait with an autosomal recessive pattern of inheritance [5] and also in association with Ehlers-Danlos and Marfan syndrome; however, in many cases, it is thought to be an acquired condition. Inhalation of chemical irritants and barotrauma from prolonged mechanical ventilation are recognized associations.

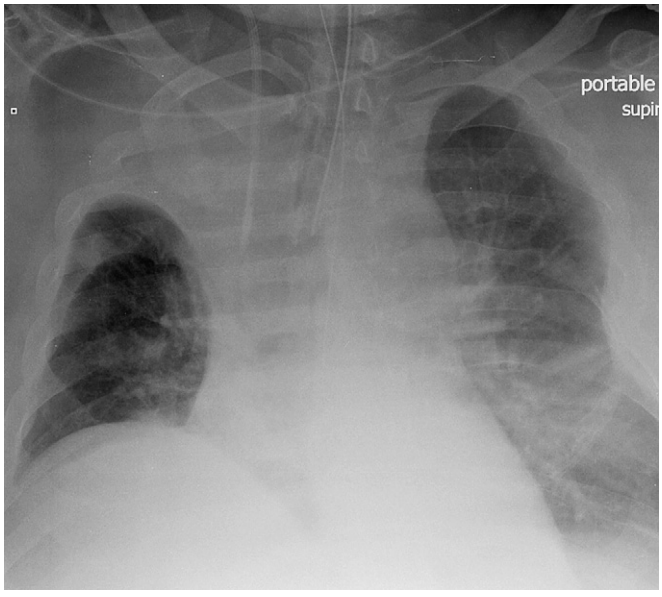

Figure 1. Chest radiograph of a 52-year-old man admitted to intensive care after a road traffic accident. An endotracheal tube is appropriately positioned, but there is right upper-lobe collapse. These changes persisted, and a subsequent CT scan showed a right tracheal bronchus as the cause.

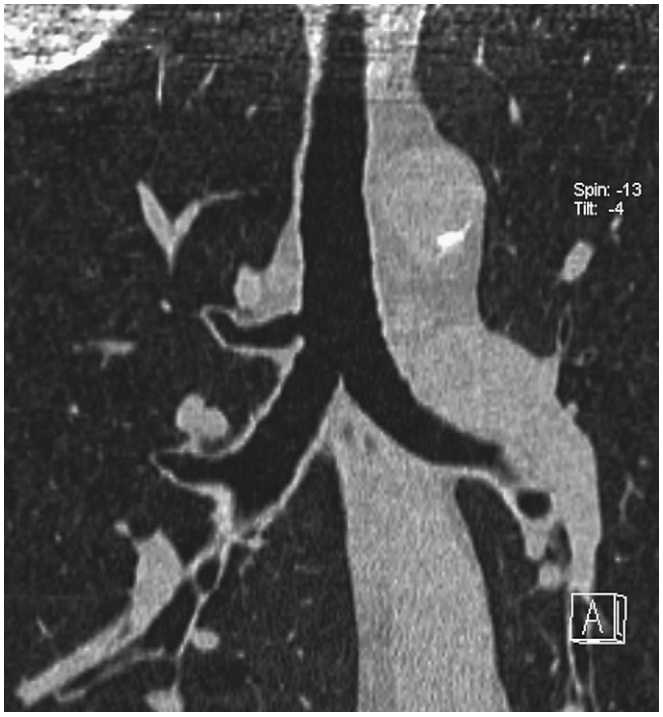

Figure 2. Coronal reformatted CT image, showing a right-sided tracheal bronchus.

Most cases present in adult life with chronic cough, excessive sputum production, and recurrent chest infections as a consequence of impaired clearance of secretions [6]. Diagnostic criteria based on radiologic findings are tracheal and main-stem bronchial diameters that exceed more than 3 standard deviations from the mean. A tracheal diameter larger than 26 mm in men and 23 mm in women is considered diagnostic. CT enables a more comprehensive assessment of the central airways and may show supportive features, such as tracheobronchial diverticulosis, cystic bronchiectasis, and complete collapse of the central airways on expiration (Figure 3).

Bronchial Atresia

Bronchial atresia is a rare condition caused by congenital occlusion of a proximal bronchus, most likely the consequence of a vascular insult in early fetal life [7]. The bronchus immediately distal to the occlusion fills with mucoid secretions, but the distal lung parenchyma develops normally and becomes hyperaerated via collateral air-drift pathways. The left upper-lobe bronchus is most frequently involved [8]. Most cases are incidental findings on chest radiographs;