

Bronchiectasis with tracheobronchial dilation

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A 27-year-old man complained of irritating cough and recurrent respiratory infection. He reported progressive dyspnea for 3 years. Chest CT showed diffuse bronchiectasis, with marked dilation of the trachea and main bronchi (Figure 1).

Bronchiectasis, by definition, is a permanent and irreversible dilation of the airways. Numerous etiologies can result in bronchiectasis. They include airway obstruction (tumors, foreign body aspiration, etc.), cystic fibrosis, immunological disorders, congenital alterations, lung infections (tuberculosis and allergic bronchopulmonary aspergillosis), among others.(1-3)

Bronchiectasis can be classified in several ways. Clinically, the current tendency is to classify them as fibrocystic or nonfibrocystic. Morphologically, they are classified as tubular (cylindrical), varicose, or cystic (saccular). The distribution of bronchiectasis can be important for diagnosis. They may, according to distribution, be divided into focal or diffuse, or may predominate in certain regions of the lungs. When they predominate in upper fields, cystic fibrosis, allergic bronchopulmonary aspergillosis, tuberculosis, and sarcoidosis must be remembered. When they predominate in anterior regions symmetrically and especially affecting the middle lobe and lingula, they suggest atypical mycobacteriosis. The predominance in

lower fields is more often seen when they are secondary to aspiration or when associated with fibrosing diseases, such as usual interstitial pneumonia or nonspecific interstitial pneumonia. Some imaging findings are characteristic of certain etiologies, such as branched tubular opacities with high density, corresponding to dilated bronchi containing hyperdense mucus, as seen in allergic bronchopulmonary aspergillosis; bronchiectasis associated with situs inversus totalis and sinusitis, corresponding to the immotile cilia syndrome (Kartagener's syndrome); or bronchiectasis associated with marked dilation of the trachea and main bronchi, as observed in our patient, suggesting cartilage atrophy and characterizing tracheobronchomegaly, or Mounier-Kuhn syndrome.(1-3)

Mounier-Kuhn syndrome is a congenital condition characterized by the absence or marked atrophy of elastic fibers and smooth muscles of the walls of the trachea and main bronchi. Patients generally present with cough and recurrent respiratory infections, and imaging tests show a marked increase in the caliber of the large airways, in addition to bronchiectasis. These abnormalities can be seen on chest X-rays but are better identified on CT. The main alteration observed in the respiratory physiology of these patients is the total collapse of the airways during expiration. Small diverticula can be observed on the walls of the upper airways, also related to parietal fragility.⁽¹⁻³⁾



Figure 1. In A, chest CT during inspiration showing bilateral bronchiectasis, in addition to marked dilation of the main bronchi. In B, coronal reconstruction in minimal intensity projection showing, in addition to bronchiectasis, dilation of both the main bronchi and the trachea. These alterations characterized Mounier-Kuhn syndrome.

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