Diagnosis of the case presented in the previous edition

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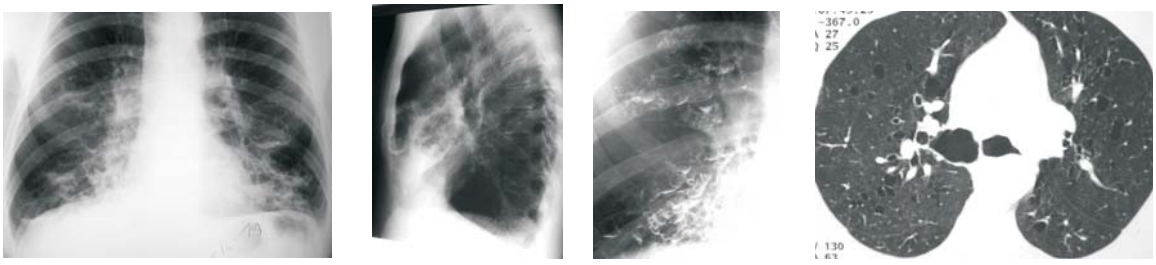
MOUNIER-KUHN SYNDROME



56 year-old male
Chronic cough and repeat infections

Mounier-Kuhn syndrome, also known as tracheobronchomegaly, is a rare disease, characterized by dilatation of the trachea and main bronchi. The disease may be accompanied by tracheal diverticulosis, bronchiectasis and recurrent infections of the upper respiratory tract.

Its etiology remains unknown. Analysis of biopsy and autopsy samples has shown that elastic and muscle fibers are atrophied, few or absent in the trachea and main bronchi. As a result, the tracheal mucosa may become herniated through the cartilaginous rings, creating abnormal outpouchings. In addition, enlargement and flaccidity of the tracheobronchial walls generate an inefficient cough mechanism, impeding proper mucociliary clearance. This, together with the accumulation of secretion in the tracheal outpouchings, favors the establishment of recurrent pneumonia, emphysema and bronchiectasis. Some cases of Mounier-Kuhn syndrome have been attributed to diffuse pulmonary fibrosis, mechanical ventilation (in neonates) and repeat infections of the lower airways. However, the majority of cases are idiopathic in origin. The syndrome is slightly more common among black men in their 30s or 40s.

It is difficult to differentiate chronic bronchitis or bronchiectasis from Mounier-Kuhn syndrome because its symptoms are nonspecific: productive cough (with or without hemoptysis) and progressive dyspnea that can lead to respiratory insufficiency. Complications, such as pneumonia, spontaneous pneumothorax or massive hemoptysis, may occur.

Diagnosis is made through imaging techniques. In radiological images, a tracheal diameter greater than 30 mm and main bronchi diameters greater than 24 mm and 23 mm on the right and left, respectively, are diagnostic of the syndrome. In computed tomography (CT) images, these same diagnostic values are 30 mm, 20 mm and 18 mm, respectively. Findings such as protrusion of musculomembranous tissue from between the cartilaginous rings, tracheal diverticulosis (outpouchings) can aid in making the diagnosis since they are present in one-third of all cases. In general, the bronchi become dilated up to the fourth segment, assuming the normal caliber thereafter. Such measurements are best taken with high-resolution CT scans. In dynamic CT imaging, tracheal dilatation can be seen in the inspiratory phase, and reduced diameter or collapse is seen upon expiration. Magnetic resonance imaging has the advantage of not emitting ionizing radiation but has low sensitivity for detecting and characterizing existing parenchymal alterations.

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