A 38-Year-Old Man With Tracheomegaly, Tracheal Diverticulosis, and Bronchiectasis*

Luiz Claudio Lazzarini-de-Oliveira, MD; Carlos Alberto Costa de Barros Franco, MD; Cristiane Linhares Gomes de Salles, MD; and Amarino C. de Oliveira, Jr, MD

(A CHEST 2001; 120:1018–1020)

A 38-year-old man was referred to our University Hospital for evaluation of chronic cough with sputum production. During the last 15 years, he had experienced an increased expectoration of mucoid sputum that became purulent during infectious exacerbations, sometimes with bloody sputum. He denied fever, wheezes, chest pain, dyspnea, or weight loss. No other respiratory illness was present. His parents and siblings were normal. He works as a security officer. He has never smoked and denies regular alcohol intake.

Physical examination revealed a thin but well-nourished patient. Vital signs were normal. Clinical respiratory examination disclosed decreased breath sounds over both lungs and inspiratory crackles at the lower third of both lung fields. Finger clubbing was present. The results of blood analysis were within normal limits. A chest radiograph and helical CT scan (Fig 1, 2) were performed, showing tracheomegaly with transversal diameters of the trachea and right-main bronchus of 36 mm and 26 mm, respectively. Many diverticular out-

pouchings were present from the trachea to the main bronchi. There was also cystic bronchiectasis involving both lower lobes (Fig 3).

A bronchoscopy was performed and disclosed several openings in the posterior and lateral wall of trachea and main bronchus; some of the tracheal diverticula are identified surrounding the trachea and main bronchus.

The rest of the examination was normal. Sputum and tracheal aspirate results were negative for mycobacteria.

What is the diagnosis?
Figure 2. Chest CT scan in axial plane at the level of mid-trachea, above the aortic arch, showing tracheomegaly and adjacent diverticula that are visible in Figure 1.

Figure 3. Chest CT scan at the level of heart. Note the presence of cystic bronchiectasis in both lower lobes.
Diagnosis: Mounier-Kuhn syndrome

Mounier-Kuhn syndrome is a rare disorder characterized by marked dilatation of the trachea and main bronchi, sometimes with tracheal diverticulosis, bronchiectasis, and recurrent lower respiratory tract infection. The clinical and endoscopic features of tracheobronchomegaly were described by Mounier-Kuhn in 1932. The etiology is uncertain. Autopsy studies suggest a congenital defect or atrophy of the elastic and smooth muscle tissue of the trachea and main bronchi. Because of this weakened trachea, some patients also develop mucosal herniations between the tracheal rings, leading to tracheal diverticulosis and retention of secretions in them. The airways distal to the fourth-order and fifth-order division are usually normal in diameter. A familial form has been described with a possible recessive inheritance and acquired forms as a complication of pulmonary fibrosis in adults and of mechanical ventilation in preterm neonates. Secondary tracheobronchomegaly was also described in association with Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome, connective tissue diseases, ataxia-telangiectasia, Bruton-type agammaglobulinemia, ankylosing spondylitis, cutis laxa, and light chain deposition disease. Nevertheless, the majority of cases appear to be sporadic. The disease predominantly occurs in men in their third and fourth decades of life.

The symptoms of tracheobronchomegaly are non-specific, with sputum production secondary to bronchiectasis and lower respiratory tract infection. The grossly enlarged but weakened airways and inefficient cough mechanisms block mucociliary clearance leading to mucus retention with resultant recurrent pneumonia, bronchiectasis, and fibrosis. Excessive sputum production with occasional hemoptysis occurs and patients may develop dyspnea and respiratory failure as the lungs become progressively damaged. In addition, spontaneous pneumothorax, hemoptysis, pneumonia, and finger clubbing may develop.

On a plain chest radiograph, the increased caliber of the central airways may be visible. This is usually best seen in the lateral projection. For an adult, any diameter of the trachea, right main bronchus, and left main bronchus that exceeds 3.0 cm, 2.4 cm, and 2.3 cm, respectively, on a standard chest radiograph or bronchogram is diagnostic of tracheobronchomegaly because these are the upper limits of the means plus three standard deviations. For chest CT, these values are 3.0, 2.0, and 1.8, respectively. Tracheal diverticulosis is seen in approximately one third of patients and most commonly originate from the right posterolateral wall. The experience with MRI is still limited to one case report in the literature. Pulmonary function tests may reveal an obstructive pattern and increased residual volume.

Treatment is limited to physiotherapy to assist in clearing secretions and appropriate antibiotics during infectious exacerbations. There is no role for surgery because of the diffuse nature of the disease. There is one case report of tracheal stenosis requiring surgical correction secondary to tracheal intubation for 15 days with a high-volume, low-pressure cuff. It is recommended that Mounier-Kuhn syndrome patients who require mechanical intubation should use an uncuffed tube. Tracheal stenting has been shown to be useful in advanced cases.

References