Mounier-Kuhn Syndrome Diagnosed in an Adult

Síndrome de Mounier-Kuhn diagnosticado en edad adulta

To the Editor:

This syndrome, also known as tracheobronchomegaly, was first described in 1932 by Mounier-Kuhn and now bears his name.[1] It consists of an anomalous and diffuse dilation of the trachea and main bronchi, generally accompanied by bronchiectasis. Very few cases have been described in the literature, and most patients are men between the ages of 20 and 50 years[2,3]; diagnosis is rare in patients over 65 years of age.

We report the case of a woman with a history of recurrent bronchial infections due to bronchiectasis, who was diagnosed with Mounier-Kuhn syndrome as an adult during her first admission to hospital.

The 63-year-old woman was admitted to the pneumology department with fever, cough with purulent expectoration, and dyspnea that had begun 4 days earlier. She had a history of recurrent lower respiratory infections since childhood.

The physical examination revealed that the patient was in good general health, with tachypnea at rest and normal auscultation. Laboratory tests revealed 13.7×10^3 leukocytes/μL with no left shift, and C-reactive protein at 5.78 mg/dL. Arterial blood gases while breathing normal air revealed a pH of 7.45, PCO_2 of 33.6 mm Hg, PO_2 of 56 mm Hg, and HCO_3 of 23.1 mm/L.

A chest x-ray showed possible atelectasis of the right lower lobe and an increase in the diameter of the trachea of approximately 4 cm, with pseudodiverticula that suggested tracheobronchomegaly (Figure A). It was confirmed using a computerized axial tomography (CAT) scan of the chest, which showed a trachea of 4.2 cm in diameter (Figure B) with a thin wall, main right bronchial diameter of 2.5 cm and left bronchial diameter of 2.3 cm (Figure C). It also showed diverticula between the cartilaginous rings and cylindrical bronchiectasis in the middle lobe and the lingula. The expiration tests revealed a tracheobronchial collapse. During the bronchoscopy, in addition to the anatomical abnormalities, we found suppuration in the main bronchi with thick mucus plugs and complete distortion of the bronchial anatomy due to ectasis and prominent scarred cartilage. No obvious growth of any pathogens was observed in the samples obtained. Antigen tests for pneumococcus and legionella in the urine were negative.

The patient made good progress with empirical antibiotic treatment administered for 7 days, and the atelectasis remitted after fiberoptic bronchoscopy and respiratory physical therapy. The patient was discharged and followed up in an outpatient setting.

Mounier-Kuhn syndrome is characterized by a significant dilation of the trachea and main bronchi due to atrophy or absence of the elastic fibres or smooth muscle that leads to laxity to the walls of the airway and the formation of diverticula and bronchiectasis. At the same time, decreased mucociliary clearance and difficulty in coughing favors recurrent infections.[3]

The prevalence of Mounier-Kuhn syndrome is relatively low, affecting between 1% and 4.5% of the population,[4] and occurs predominantly in males between the ages of 25 and 50 years. The etiology is poorly understood, but several hypotheses have been postulated: a congenital origin, since cases have been described in children, and an acquired origin, as in our case, given the age of the patient, although the time of evolution of the disease was unknown. This syndrome has been known to be associated with Ehlers-Danlos in adults, and cutis laxa in children, as well as other connective tissue diseases.[3]

The clinical profile of this syndrome varies, with recurrent infections being the most frequent signs,[4] although cases do exist in the literature including dyspnea on exertion, some asymptomatic cases described in autopsies, and others that are diagnosed incidentally.

The radiological diagnosis using a chest CAT scan depends on the presence of a transverse tracheal diameter greater than 3 cm along with a right bronchial diameter of 2.4 cm and left diameter of 2.3 cm.[5] In our case, the transverse diameter of the trachea measured 4.2 cm, the right main bronchus measured 2.5 cm, and the left 2.3 cm.

Until the development of the CAT scans, bronchoscopy was the gold standard for making the diagnosis, as it was able to evaluate tracheal dynamics, but with CAT scans, in addition to being able to

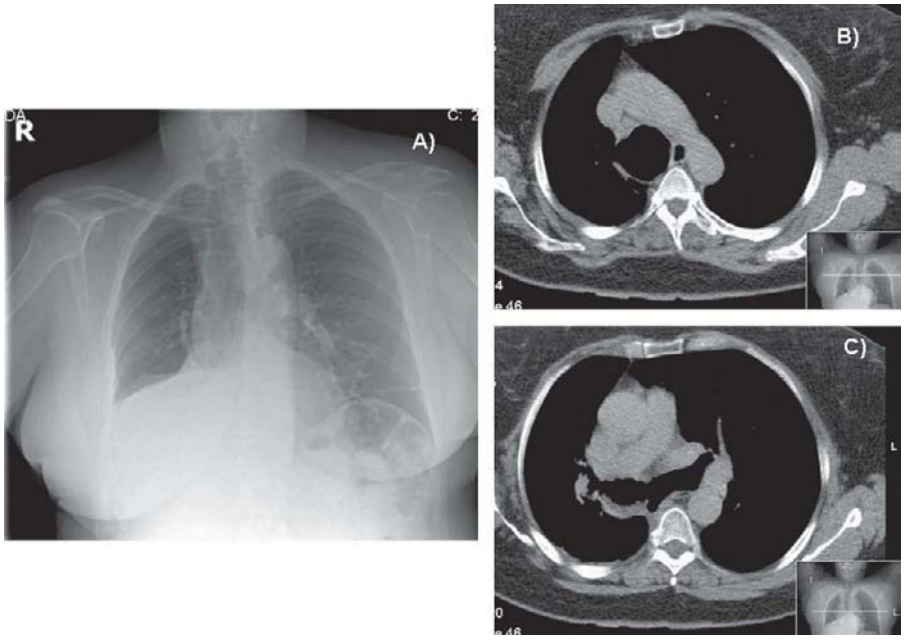

Figure. A) Chest X-ray showing atelectasis in the right lower lobe and enlargement of the tracheal air column. B) Computerised axial tomography showing a large increase in the diameter of the trachea, with thinning of the walls and pseudodiverticula. C) Computerised axial tomography showing a major enlargement of the main bronchi.

evaluate expiratory tracheal dynamics, we can ascertain the extent of the disease and evaluate the presence of tracheal diverticula and areas of associated bronchiectasis, as in our case.

Treatment is symptomatic, and consists of respiratory physical therapy in order to manage secretions, and antibiotic, bronchodilator, and corticosteroid treatment during exacerbations.[6]

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