Mounier-Kuhn syndrome: radiological findings and clinical presentation

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Abstract

Mounier-Kuhn syndrome is a rare disease characterized by recurrent respiratory infections. The present report describes a case of this disease with analysis of chest radiography and high resolution computed tomography showing increased caliber of the trachea, main bronchi and central bronchiectasis. Such changes, in association with clinical data, suggest the diagnosis.

Keywords: Tracheobronchomegaly; Mounier-Kuhn; High resolution computed tomography; Radiology.

CASE REPORT

A male, 33-year-old painter, active smoker (two cigarettes packs a day), with a previous history of treatment for pulmonary tuberculosis in 1979, presented fever, nocturnal sudoresis, productive cough, chest pain, dyspnea and chest wheezing for two months, was referred to the institution for pneumological evaluation. At clinical examination, the patient showed nail clubbing (Hippocratic fingers), lung auscultation with bilateral crepitus in the lower thirds. Spirometry demonstrated severe obstructive ventilatory disorder: FVC = 3.40 (75%), FEV₁ = 1.49 (40%), TI = 39.1%.

Chest radiography demonstrated increase in the lung volume and transparency, with signs of reticular interstitial infiltrate predominating in the central regions. The trachea presented increased caliber, with a maximum transverse diameter of 40.0 mm (Figure 1). High resolution CT demonstrated details of the parenchymal alterations particularly characterized by predominantly cystic bronchiectasis clustered in the central regions, diffuse perfusional alterations in the pulmonary parenchyma (mosaic pattern of attenuation), besides increased caliber of the trachea and of the main bronchi which also presented jagged contours corresponding to tracheobronchial diverticulosis (Figure 2).

Over the years, the patient presented a progressive worsening of the dyspnea with chronic suppuration (sputum analysis demonstrated Pseudomonas aeruginosa colonization) and several hospital admissions because of infective exacerbation. In 2009, the patient was admitted to the hospital, progressing to fatal respiratory failure.
DISCUSSION

The diagnosis of tracheobronchomegaly may be achieved when the diameters of the trachea and of the right and left main bronchi are > 3.0, 2.4 and 2.3 cm, respectively\(^1\). Frequently, TBM is associated with tracheal diverticulosis and bronchiectasis. The tracheal diverticulum may be originated from increased compliance of the tracheal wall and development of a redundant membranous tissue\(^4\). The disease can be diagnosed by means of plain chest radiography in association with clinical findings, but the investigation with computed tomography allows a more accurate evaluation of abnormalities in the affected airways as well as of the disease extent\(^5\). The diminished clearing of secretions observed in this syndrome causes its accumulation, leading to repetitive respiratory infections which results in the development of the mentioned anatomic alterations and consequent functional impairment, rang-
ing from minimal functional damage to respiratory failure leading to death\(^{(1,3)}\). Respiratory function testing demonstrate an obstructive pattern, increased residual volume and decreased CO\(_2\) diffusion in the presence of parenchymal disease; in advanced cases, a predominance of a restrictive pattern secondary to fibrosis is observed\(^{(1)}\). The treatment is generally supportive, by means of respiratory physiotherapy, appropriate antibiotic therapy for recurrent infections and tobacco use cessation. Success has been achieved in few cases with tracheal stenting to prevent expiratory collapse\(^{(6)}\).

**CONCLUSION**

Suspicion of tracheobronchomegaly should be raised in patients with chronic pulmonary suppuration, repetitive respiratory infections and ineffective cough and whose chest radiography demonstrates increased tracheal caliber. High resolution computed tomography (HRCT) plays a relevant role in the accurate evaluation of abnormalities present in the lung parenchyma.

**REFERENCES**