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Radiology

Incidental Finding of Mounier-Kuhn Syndrome

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Abstract **Case Report**

Tracheobronchomegaly (TBM) or Mounier - Kuhn syndrome is a rare disorder of uncertain etiology characterized by marked dilatation of the trachea and bronchi and recurrent lower respiratory tract infection. We report the case of a 56-year-old male presenting with a chronic cough.

Keywords: Tracheobronchomegaly (TBM), Mounier-Kuhn syndrome, uncertain etiology, unproductive cough.

INTRODUCTION

Mounier-Kuhn syndrome, also known as tracheobronchopathia malacia, tracheomegaly, and multiple tracheal diverticula, is a rare clinical and radiological finding, defined as a defect in the development of connective tissue and muscle in the trachea and bronchi, resulting in irreversible tracheobronchomegaly, and recurrent lower respiratory tract infection.

Symptoms are unspecific and diverse ranging from asymptomatic to severe respiratory failure. Diagnosis is based on a CT scan. Management of Mounier-Kuhn syndrome focuses on symptomatic relief and prevention of complications.

CASE REPORT

We report the case of a 58-year-old man, consulting for a 2-year history of persistent and unproductive cough, further interrogation found recurrent pulmonary infection with no smoking history. Clinical exam and chest X- ray were reported to be normal.

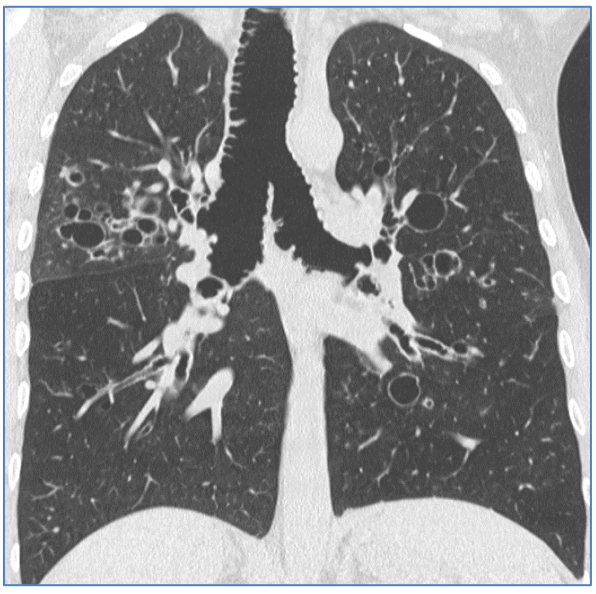

Figure 1: Coronal Chest CT scan objectifying diverticular tracheobronchomegaly.

A non-contrast chest CT was performed revealing multiple diverticula and areas of scalloping between the cartilaginous rings in the trachea and main bronchi, the trachea was grossly dilated with a transverse diameter of 35 mm (Figure 1), while the right and left main bronchi had a diameter of 24 mm and 22 mm respectively (Figure 2), associated with bilateral Cystic bronchiectasis (Figure 4), confirming the diagnosis of Mounier-Kuhn syndrome.

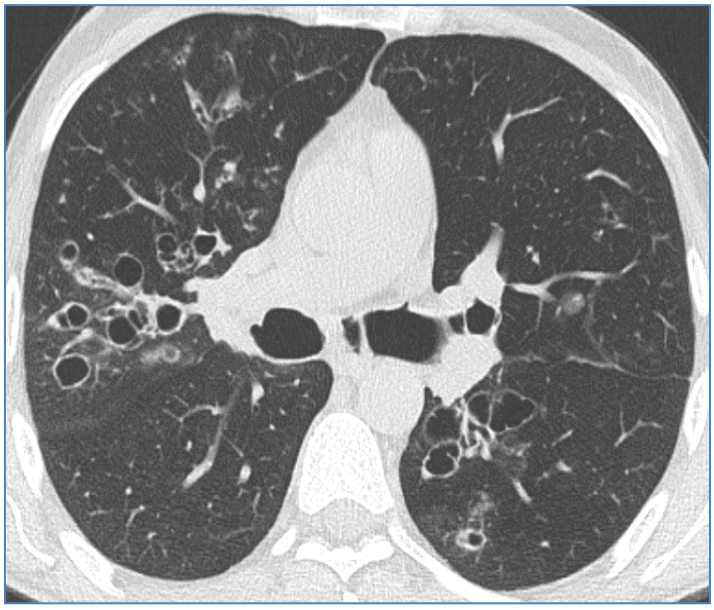

Figure 2: axial chest Ct scan noting bilateral cystic bronchiectasis

DISCUSSION

Mounier-Kuhn is a rare syndrome first described endoscopically and radiologically in 1932, by Pierre-Louis Mounier-Kuhn. Its actual incidence may be underappreciated due to the lack of specific symptoms [1]. Tracheobronchomegaly results from the atrophy of smooth muscles and elastic connective tissue in the trachea and main bronchi, leading to sacculations and diverticulum formation between the cartilaginous rings, with no certain etiology [2].

It is reported sporadically amongst male adults, which argues in favor of the acquired origin of the pathology. Several contributing factors have been framed: barotrauma during infancy, exposition to irritants, notably tobacco, and pollution [4]. However, some studies speculate this syndrome to be a recessive inheritance trait, due to some cases being reported among siblings and cousins and its association with Ehlers-Danlos syndrome and cutis laxa in children [3]. Our case proved no family history of TBM, congenital or associated syndrome, and no personal or professional exposure to tobacco or particulate pollution.

Diagnosis is based on measurements on CT scan, it is defined by an increase in the transverse and sagittal diameter of the trachea beyond 25 and 27 mm, and/or an increase in the diameter of the right and left main bronchus beyond 18 and 21 mm. The same definition applies to women with respective measurements of 21, 23, 17.4, and 19.8 mm. An increase in the cross-sectional area of the trachea beyond 371 mm^2 for men and 299 mm^2 for women also defines the disease. For our patient, the transverse diameter of the trachea was 35 mm, the diameter of the left main bronchus at 22 mm, and of the right main bronchus at 24 mm [5].

The Dilated airways often present a scalloped appearance that is secondary to herniation of musculo-membranous tissue through the bronchial cartilaginous rings. Large diverticular or sacculiform formations have also been reported in the posterior-basal territory of the trachea and the stem bronchi. For our patient, the dilation was harmonious on the chest CT scan; on the other hand, endoscopic bronchial exploration was not performed. Chronic and recurrent infection usually results in central bronchiectasis, which may be cylindrical, cystic, or varicose.

Mounier-Kuhn syndrome could be associated with nasal polyposis and polymalformative genetic syndrome including bilateral ptosis, epicanthus, micrognathia, and excess upper lip skin. None of these signs was reported in our patient [2]. The most common respiratory complications are bronchiectasis, tracheobronchomalacia, and emphysema, while the most common non–non-respiratory comorbidity is Gastroesophageal reflux disease (GERD) [6]. Our patient presented with cystic bronchiectasis.

CONCLUSION

Mounier-Kuhn syndrome is a rare condition of which congenital or acquired origin is still debated. Diagnosis and impact assessment is necessary and based

on a CT chest exam. Treatment is supportive aiming to treat and prevent bronchopulmonary infections and to free the airways by physiotherapy.

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