

Case Report

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CT scan Diagnosis of Mounier-Kuhn Syndrome in A Patient with Recurrent Respiratory Infections: About A Rare Case Report

Benfaddoul O*, El Khalifa M, Bellasri S, Slioui B, Roukhsi R, Hammoune N, Mouhsine A and Atmane E

Department of Radiology, Avicenne military hospital, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco

ABSTRACT

Tracheobronchomegaly or Mounier-Kuhn syndrome is a very rare condition characterized by dilatation of the trachea and main bronchi, bronchiectasis and recurrent respiratory tract infections. The causality of this syndrome is unclear and the clinical representation is variable. The diagnosis is mostly established on the basis of characteristic CT scan findings. We report the case of a 65-year-old female with a history of recurrent lower respiratory tract infections, that was not initially diagnosed by repeated chest radiographs, but was later identified by CT scan.

*Corresponding author

Benfaddoul O, Department of Radiology, Avicenne military hospital, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakech, Morocco.
Tel: +212 667248367; E-mail: benfaddoulwalid@gmail.com

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Introduction

Mounier-Kuhn syndrome, or tracheobronchomegaly, is a uncommon clinical and radiological condition, characterized by marked enlargement of the trachea and bronchi and recurrent lower respiratory tract infections, this syndrome was first defined by Mounier-Kuhn in 1932, but less than 100 cases have been published in the literature. The underlying cause of the condition is not yet clearly understood; however, congenital atrophy of the smooth muscle and elastic tissue of the trachea and main bronchi has been identified by biopsy studies [1,2]. The clinical presentation is quite variable and it may occur in several different age groups and imaging plays a major role in the diagnosis of such rare but easily identified disease [3,4].

Case Report

The patient was a 65-year-old woman with a medical history of recurrent low respiratory tract infections. She was admitted to the emergency room for a productive cough, wheezing, dyspnea, hypoxemia accompanied by fever and weight loss. Physical examination revealed pulmonary cachexia, bilateral coarse crackles, and wheezing, Laboratory investigations were performed and showed a neutrophilic leukocytosis and elevated c-reactive protein, a thoracic CT scan was performed and showed a rippled and dilated appearance of the trachea with a diameter of 32 mm and multiple tracheal diverticula between the cartilage rings, (Figure 1 and 2), dilation of the main stem bronchi reaching the distal bronchi, measuring respectively 22 mm and 23 mm (Figure 2 and 3), associated with emphysematous changes in both lung apices, consolidation in the upper lobes and the lingula, honeycombing, traction bronchiectasis and interstitial thickening predominantly in the upper lobes, middle lobe and the lingula.



Figure 1: Computed tomography scan of chest (axial) showing dilated trachea (white arrow)



Figure 2: Computed tomography scan of chest (coronal) showing a rippled and dilated trachea with multiple tracheal diverticula between the cartilage rings and dilation of the main stem bronchi reaching the distal bronchi



Figure 3: Computed tomography scan of chest (coronal) showing dilated main stem bronchi (red arrow) and distal bronchiectasis (white arrows) (Type 3 changes).

Discussion

Mounier-Kuhn syndrome is characterized by a distinctive dilatation of the trachea and bronchi, it is more frequent in males and is generally diagnosed in the 3rd or 4th decade of life [5]. It is due to atrophy of the muscular and elastic tissues of the trachea and main bronchial wall; atrophy or absent elastic fibers and thinning of the muscle, allows the trachea and main bronchi to become loose and significantly dilated on inspiration, with shrinkage or collapse on expiration or coughing. Abnormal airway dynamics and accumulation of secretions in large openings of the redundant musculo-membranous tissue between the cartilage rings predispose to the development of chronic pulmonary suppuration, emphysema, bronchiectasis, and pulmonary fibrosis [6]. Involvement can occur at different levels, from the trachea to the 4th bronchial branch, despite the fact that its etiology is not fully understood, tracheobronchomegaly is thought to be associated with a familial susceptibility and possibly inherited through an autosomal recessive mechanism [7]. Mounier-Kuhn syndrome includes 3 subtypes; in type 1, there is a slight symmetrical enlargement of the trachea and the main bronchi, type 2 has a more evident dilation with diverticula that are clearly distinct and in type 3, the diverticular and saccular structures reach the distal bronchi [8].

A large range of clinical manifestations has been documented in association with Mounier-Kuhn syndrome, the clinical picture is wide and nonspecific and a large range of clinical manifestations has been documented in association with Mounier-Kuhn syndrome; varying from minimal disease with good preservation of lung function to progressive disease resulting in respiratory failure and death [9]. The principal issues involved in this condition are an ineffective cough secondary to the pathological dilation of the tracheobronchial tree and impaired mucociliary activity, these factors together lead to difficulty in expectorating secretions and result in recurrent lower respiratory infections. In the absence of infection, it may develop asymptotically.

The diagnosis is often established using computed tomography, which identifies the abnormally enlarged respiratory tract. In adults, the radiological criteria are: in men, 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 stem

bronchus beyond 18 and 21 mm, respectively. The same definition applies in women with measurements of 21, 23, 17.4 and 19.8 mm respectively.

An increase in tracheal cross-sectional area beyond 371 mm² in men and 299 mm² in women also defines the condition. The enlarged airways frequently have a deeply festooned appearance that is secondary to herniation of Musculo-membranous tissue through the cartilaginous bronchial rings, large diverticular or sacculiform formations may also be seen especially in the posterior and posterobasal territory of the trachea and stem bronchi [10].

The second purpose of the CT scan is to assess the associated broncho-parenchymal and parietal lesions; bronchiectasis is frequently associated and more or less severe parenchymal lesions may be intermixed (atelectasis, pneumopathy, interstitial fibrosis or emphysema). Sternocostal malformations have also been reported [11].

Mounier-Kuhn syndrome can be debated in view of Williams-Campbell syndrome, which involves congenital cystic bronchiectasis resulting from cartilage deficiency in the fourth and sixth order bronchi. However, in this syndrome the trachea and main bronchi are of a normal caliber.

In asymptomatic patients, no specific treatment is required. Smoking cessation is very beneficial, as well as reducing exposure to industrial and professional irritants and pollutants. In the presence of symptoms, the treatment is limited to respiratory physiotherapy to flush secretions and the use of antibiotics in the case of infectious exacerbations. While tracheal stenting has been beneficial in severe cases, surgery is rarely considered given the diffuse nature of the disease. Lung transplantation has not demonstrated any benefits regarding the risk of major morbidity and death [12].

The condition can be tolerated for a long time but the evolution towards chronic respiratory failure is inevitable.

Conclusion

Mounier-Kuhn syndrome or tracheobronchomegaly is a very rare condition of which the congenital or acquired origin is still debated. The clinical manifestations are not very specific and the radiological diagnosis is often made very easily on CT scan. Computed tomography plays a key role in the diagnosis and the evaluation of the repercussions through careful analysis of central respiratory tract and pulmonary parenchyma.

Disclosure of interest:

The authors declare that they have no conflicts of interest concerning this article.

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