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Case report

Mounier-Kuhn syndrome: A variable course disease

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ABSTRACT

Mounier-Kuhn syndrome or tracheobronchomegaly is a rare disease characterized by marked dilation of the trachea and proximal bronchi with recurrent lower tract respiratory infections. Computed tomography and bronchoscopy are the key tools to accomplish the diagnosis. This is a condition with a clinical polymorphism, symptoms vary from minor with preserved respiratory function, to very severe with life threatening exacerbations leading to respiratory failure and premature death. The treatment is mainly symptomatic, stenting or surgery are reserved to extreme cases.Herein, we report two cases of the same condition with different clinical signs and diverse outcome.

1. Introduction

Mounier-Kuhn syndrome (MKS) or Tracheobronchomegaly is a rare disease, to date, less than 400 cases have been reported [1]. MKS is characterized by a marked dilation of the trachea and stem bronchi, sometimes associated with the presence of tracheal diverticula and recurrent lower respiratory tract infections. Tracheal dilations have been described since 1897 [2] but the first clinical description was made by Mounier-Kuhn in 1932 [3]. In MKS, there is a clinical and prognostic polymorphism; symptoms vary from minor to major with severe infectious exacerbations and respiratory failure leading to premature death.

We present the cases of two of our patients diagnosed with MKS. Each one of them presented a different evolution, illustrating at best the heterogeneity of MKS's manifestations.

2. Case 1

A 46-year-old female patient with a history of chronic cough and sputum emission since the age of fourteen was admitted for right chest pain with purulent sputum. She had no history of tobacco smoking or noxious fume exposure. No past history of tuberculosis was found. On physical examination, she had bilateral crackles at the lower field of both lungs. The chest x-ray showed dilation of the tracheal clarity and bilateral bronchiectatic lesions in the bases (Fig. 1). The routine blood analysis and blood gas analysis were within normal rates. The pulmonary functions tests were within normal limits (FVC = 2.45L (85%), FEV1 = 2.22L (90%) and FEV1/FVC = 91%). For further investigation, a thoracic Computed Tomography was ordered. It disclosed dilation in the trachea and in the main bronchi along with cystic bronchiectasis in both lung bases. Bronchoscopy confirmed the presence of an irregular dilation of the bronchial wall of the trachea and stem bronchi with exaggerated collapsibility upon cough or expiration along with abundant secretions. There was no growth on culture tests of the aspirated bronchial secretions. The diagnosis of tracheabronchomegaly was made. The patient was treated with antibiotics (amoxicillin-clavulanic acid) and chest physiotherapy. She was advised Pneumococcal and Influenza vaccination. The course was marked by the occurrence of moderate infectious episodes that did not requiring hospitalization. After twelve years of follow up, a control CT scan revealed multiple tracheal diverticulain addition to the dilation of the trachea (42 mm) and the main bronchi (Right = 22 mm, Left = 24 mm) and the pre-existent bronchiectasis (Fig. 2. The pulmonary function showed a mild obstructive ventilatory deficit was found FVC = 1.89L (73%), FEV1 = 1.34L (62%) and FEV1/FVC = 71%. Otherwise, the patient was clinically stable.

3. Case 2

A 37-year-old non-smoking patient with a history of type two

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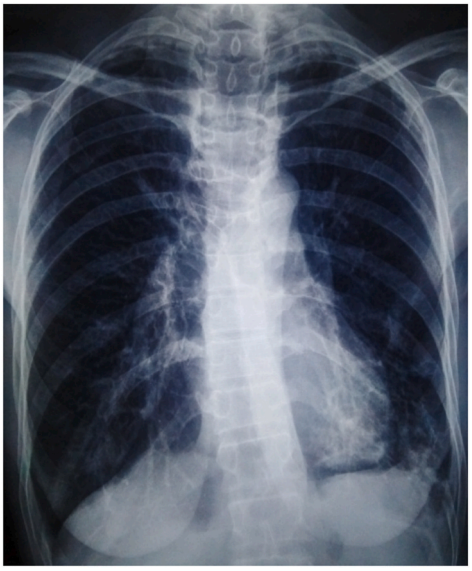

Fig. 1. Chest x-ray showing dilation of the tracheal shadow and bilateral cystic lesions in the bases.

diabetes and asthma was hospitalized in the Pulmonology Department for fever, dyspnea and purulent sputum. The patient's parents were blood related. There was not any particular occupational exposure. On physical examination, the patient presented fever at 38.8 °C with accelerated respiration and desaturation at 88% in ambient air. The chest radiograph showed bilateral interstitial opacities, bilateral pleural effusion and dilated tracheal shadow (Fig. 3). The respiratory functional assessment revealed a severe restrictive pattern FVC 1.7 L (32%), FEV1 1.3L (30%), FEV1/FVC 88%, TLC 53%, DLCO 44%. Chest computed

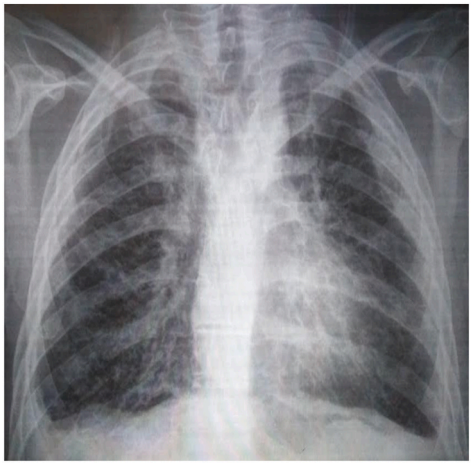

Fig. 3. Bilateral interstitial opacities, bilateral pleural effusion and dilation of the tracheal clarity on chest radiograph.

tomography (CT) showed tracheobronchial dilation associated with multiple diverticula. CT confirmed also the presence of diffuse interstitial lesions consisting with fibrosis (Fig. 4). Bronchoscopy demonstrated the presence of multiple diverticula in the posterior wall of the trachea (Fig. 5). The patient received 21 days of imipenem then discharged after a clinical biological and radiological improvement. The evolution was marked by the recurrence of bronchial infections and the appearance of chronic respiratory failure requiring long-term oxygen therapy. After three years of follow up, the patient had an acute respiratory failure with septic shock leading to his death despite adequate reanimation.

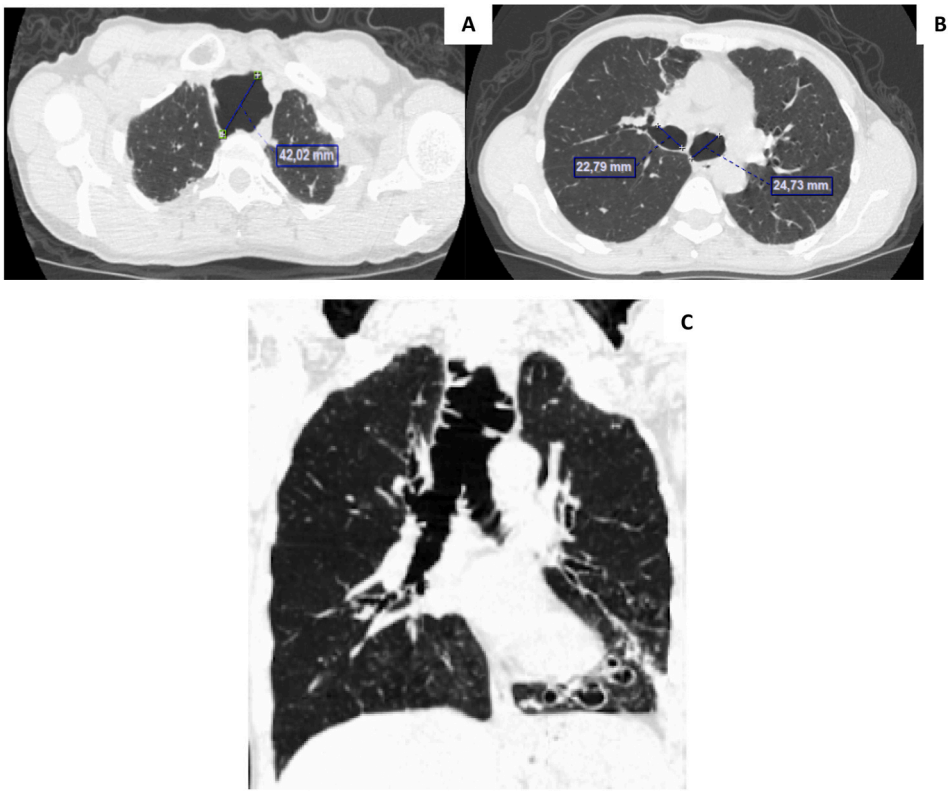

Fig. 2. Thoracic Computed Tomography: dilation in the trachea (42 mm) with multiple tracheal diverticula (A, C) and dilatation in the main bronchi (B) along with cystic bronchiectasis (C).

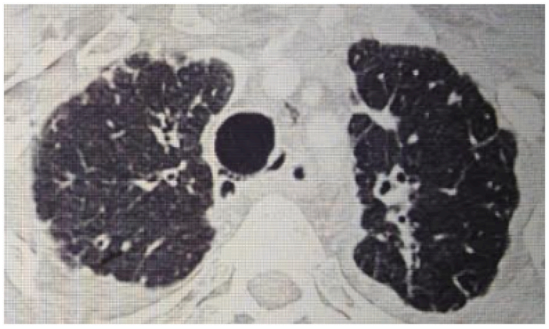

Fig. 4. Chest computed tomography showing tracheal dilatation associated with multiple diverticula.

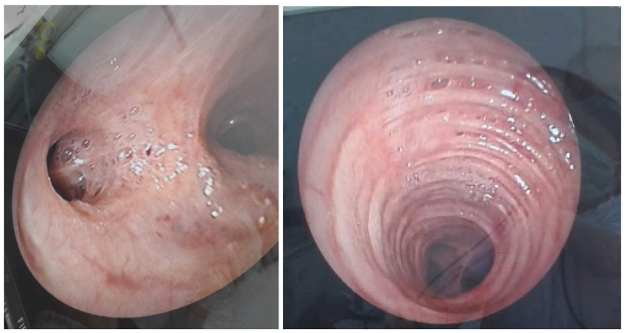

Fig. 5. Multiple diverticula in the posterior wall of the trachea and main right bronchus on bronchoscopy.

4. Discussion

Tracheobronchomegaly is a rare condition characterized by marked dilation of the trachea and recurrent respiratory infections.

Although the cause of MKS is not well known, the few biopsy and necropsy studies of the tracheal wall under showed a thinning of the muscularis mucosae, with loss of elastic fibers [4].

Some authors claim that cases are sporadic [5] while others link MKS to a familial susceptibility likely through an autosomal recessive mechanism [4]. As for our two patients, we did not find any similar cases in the family history.

MKS is more frequent in male patients aged between the third and the sixth grade [4,6and7].

It is frequently associated with multiple tracheal diverticula, which can be congenital or acquired. Often we only find congenital diverticula and their pathogenesis remains undetermined [8]. Acquired diverticula can appear at any level of the organ but are more frequent at the posterolateral wall on the junction between the intra-thoracic and extra-thoracic portion of the trachea, like the case of our two patients, which may result from an anatomical defect at this level [8], or from the increase of intraluminal pressure due to chronic cough [9].

In MKS, the clinical features are non-specific and the follow-up courses are diverse. Some patients are asymptomatic with normal respiratory function, while others present with chronic cough and recurrent lower tract respiratory infections and they progress rapidly to a chronic respiratory failure [8,9].

Computed tomography and bronchoscopy confirm the diagnosis of MKS. On CT scan, the diagnosis of MKS is made when the transverse diameter of trachea exceed 30 mm and that of right and left main bronchi measure more than 24 mm and 23 mm, respectively [10].

When the diagnosis remains uncertain, three-dimensional reconstruction CT and virtual bronchoscopy can help identify the connection between the diverticulum and the tracheal lumen and define the extent of the lesion, especially in the presence of numerous tracheal overflows [11].

Bronchoscopy remains a strong diagnostic tool that can detect the dilation in the trachea and main bronchi during inspiration, and their constriction and even collapse during expiration and coughing [4].

There is no standardized classification of MKS. Initially, Schwartz classified tracheobronchomegaly into three radiological subtypes. In type 1, there is a slight symmetrical dilation in the trachea and main bronchi. In type 2, the dilation and the diverticula are distinct. In type 3, the diverticular and saccular structures extend to the distal bronchi [12]. Payandeh suggested a Clinical Classification Scheme for MKS, identifying six subtypes [13]. Type 1A consisted of infants who developed MKS after having undergone fetoscopic tracheal occlusion, and Type 1B patients include infants and children who developed MKS after prolonged intubation. Type 2 individuals develop MKS following recurrent pulmonary infections (2A) or pulmonary fibrosis (2B). Type 3 includes patients of MKS with evidence of extra-pulmonary elastolysis. Persons with Type 4 MKS have no clear predisposing factors.

Pulmonary function testing may be normal or may show an obstructive ventilatory defect with elevated residual volume [4].

The factors affecting disease progression in patients have not been fully studied; therefore the prognosis of patients with MKS remains largely unknown [1]. The most frequent pulmonary complications of MKS are bronchiectasis, bullous emphysema, recurrent pneumonia, and aspergillosis [1]. Infection with atypical organisms, including tuberculous and non-tuberculous mycobacteria, may complicate some cases [7].

The main differential diagnosis are laryngocele, pharyngocele, Zencker's diverticulum, apical pulmonary hernia and bullous emphysema [8]. In fact, laryngocele is an air-containing evagination of laryngeal mucous membrane [14]. Likewise, pharyngocele and Zencker's diverticulum are both a protrusion of mucous membrane from the pharynx [15]. In all three syndromes, there is no tracheal diverticula. All the same, lung hernia is a protrusion of the lung outside the thoracic wall [16], and bullous emphysema is characterized by damaged alveoli that distend to form exceptionally large air spaces [17]. In these two pathologies, the trachea and bronchi are intact. Williams-Campbell syndrome is also a differential diagnosis for MKS, characterized by congenital bronchiectasis cysts resulting from a deficiency of cartilage in the fourth and sixth order bronchi. However, in this syndrome the trachea and the main bronchi are of normal caliber [10].

Asymptomatic patients do not require any specific treatment. Tobacco cessation is highly beneficial. Minimizing exposure to industrial and occupational irritants and pollutants are necessary. In symptomatic patients, therapy is essentially supportive including massage and postural drainage physiotherapy and antibiotic use during infectious exacerbations [12]. The pneumococcal and influenza vaccines are recommended regardless of age and symptomatology [6]. In extreme severe cases, stenting or more rarely surgery may be considered. Few instances of successful laser tracheoplasty to prevent the collapse of the trachea have been reported [6].

5. Conclusion

The diagnosis of MKS is easily overlooked and requires collaboration between pulmonologists and radiologists. The early detection of the disease and the optimization of the management may result in a reduced morbidity and mortality.

Declaration of competing interest

The Authors declare that there are no competing interests.

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Abbreviations

CT: computed tomography
MKS: Mounier-Khun syndrome
FVC: Forced Vital Capacity
FEV1: Forced Expiratory Volume in one second
TLC: Total Lung Capacity
DLCO: diffusing capacity of the lung for carbon monoxide