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Case report

Mounier-Kuhn syndrome (Tracheobronchomegaly): Radiological diagnosis ☆

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ABSTRACT

Mounier Kuhn Syndrome or tracheobronchomegaly is a rare disease, characterized by dilatation of the trachea and the main bronchi. Our study concerns a case of 67-years old male patient, smoker, with a chronic cough. Chest scan was requested; it shows an enlargement of the tracheal clarity and the two main bronchi. Meanwhile, the Functional respiratory exploration was normal. The treatment includes mucolytics and pulmonary physiotherapy. Mounier-Kuhn syndrome is rare and the clinical signs are not specific, the positive diagnosis is purely radiological. The treatment is about to free the airways to prevent infection.

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Introduction

The Mounier-Kuhn syndrome is a rare disease, defined as a dilatation of the trachea and main bronchi and/or proximal bronchi. The etiopathogenic studies concluded for a defect in the development of connective tissue and smooth muscle in the trachea and bronchi, resulting in tracheobronchomegaly.

The clinical symptoms are diverse and not specific, going from asymptomatic to severe respiratory failure. The medical imaging techniques make it possible to make a positive diagnosis and to establish the pulmonary impact. However, there

is no curative treatment, it is an irreversible disease, and only supportive treatments are proposed.

Case report

Our Patient is a non-smoker 67-year-old man, with a BMI of 25, without any other toxic habits, but with a medical history of: Diabetic type II for 11 years on Antidiabetics pills, Dysthyroidism for 06 years on treatment; and without any history of recurrent bronchopulmonary infections in childhood.

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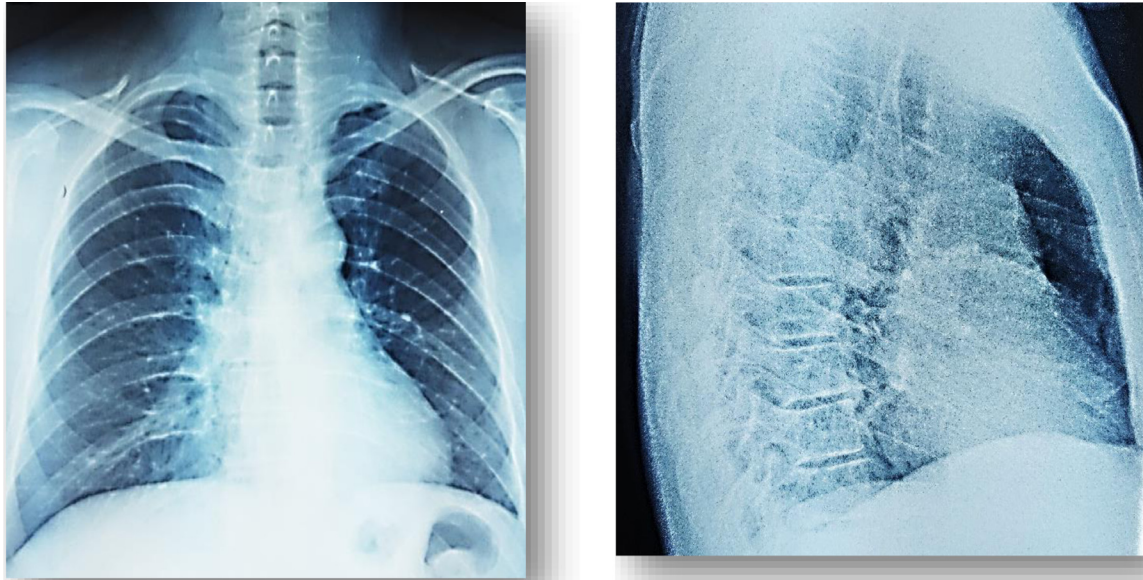


Fig. 1 – (A) Frontal chest x-ray: enlargement of the tracheal clarity and of the 2 main bronchi, thin-walled hyperclitics at the left base, reduction in the volume of the left thoracic hemfield compared to the right. (B) Lateral chest x-ray: enlargement of the tracheal clarity.

The patient shows up with a chronic cough for 1 year, most often wet and productive, with abundant sputum and Green-yellow phlegm, without hemoptysis, without dyspnea, without chest pain, and without fever. All the symptoms were progressing without any decrease of general condition. The patient was motivated for several consultations; he was treated as a bronchopulmonary infection with antibiotics several times, with an improvement of his sputum appearance but with a persistence of the productive trait of the cough. He was also treated with antihistamines and inhaled corticosteroids without any real improvement. Because of the chronic persistent cough, which is interfering with the patient's daily life activities, a chest X-ray and a chest CT scan were requested and the patient was referred to our department for additional care.

The patient, when he came to the consultation, has just an isolated productive cough with abundant white sputum without any sign of infection and without any other respiratory or extra-respiratory symptoms.

The Clinical exam found an eupneic patient with a respiratory rate of 18 bpm, a normal heart rate of 86 bpm, with a 97M of oxygen saturation. Pleuropulmonary exam noted some diffuse ronchi sounds on both of lung fields. Cervical auscultation revealed a slight sound of rasping. The rest of the physical exam was normal. In addition, there is no ptosis or excess of the skin in the upper lip.

The frontal chest X-ray shows an enlargement of the tracheal clarity and the 2 main bronchi with thin-walled clarity at the base of the left lung (Fig. 1 A). The lateral chest X-ray also shows an enlargement of the tracheal clarity.

The chest scan, performed before and after injection of contrast product, with slices of 0.5 mm thick (Figs. 2, 3, and 4), then by coronal reconstruction (Fig. 5) shows a harmonious enlargement of the cervico-thoracic trachea and the 2 main bronchi. With an anteroposterior diameter of the trachea at

24.4 mm and transverse at 34.7 mm, with the presence of a focus of cylindrical bronchiectasis at the lower left lobe.

The Functional respiratory exploration was normal, The Six-minute Walk Test, and the spirometry test were also normal.

Our patient was put on BDLA treatment for his cough, physiotherapy was prescribed to free his bronchial tubes from secretions, a vaccination was planned, and follow-up consultations were scheduled.

Discussion

Mounier-Kuhn syndrome was first described, endoscopically and radiologically, by Pierre-Louis Mounier-Kuhn in 1932, associated with recurrent pulmonary infections [1], then described anatomically as tracheobronchomegaly by Katz et al. in 1962 [2].

Studies have concluded that this pathology is due to the atrophy of the elastic fibers [3] of the trachea and bronchi, leading to thinning of the smooth muscle and ultimately to dilation [4,5].

About the etiology of the tracheobronchomegaly, it is still debatable. The debate persists between the congenital and the acquired cause. It is believed that Mounier-Kuhn could be an autosomal recessive inheritance trait, because of cases that have been detected in siblings and cousins, but no genes have been yet identified [4,5], and because of the association with Ehlers Danlos syndrome and cutis laxa in children. However, tracheobronchomegaly appears more frequently in adults with a sporadic character, which argues in favor of the acquired origin of the pathology [6]. Several contributing factors have been put forward, such as barotrauma during intensive neonatal ventilation with oxygen therapy [7,8], or expo-

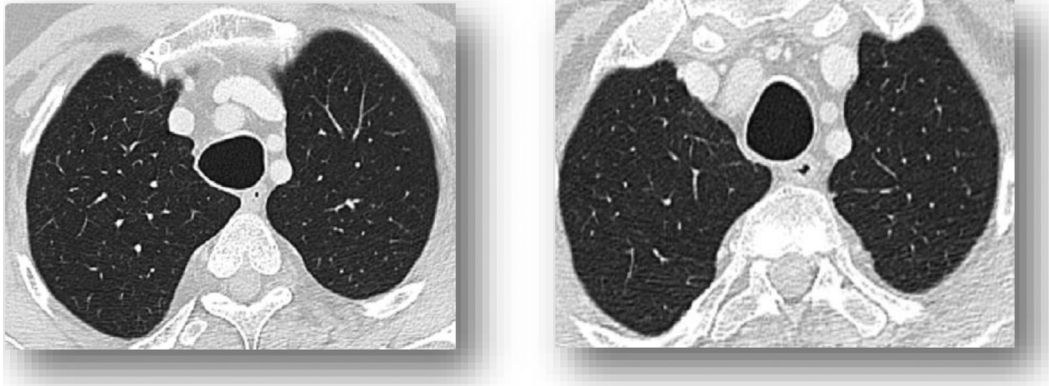


Fig. 2 – Chest CT scan: axial slices through the parenchymal window passing through the trachea. The transverse diameter of the trachea is 34.7 mm; the anteroposterior diameter is 24.4 mm.

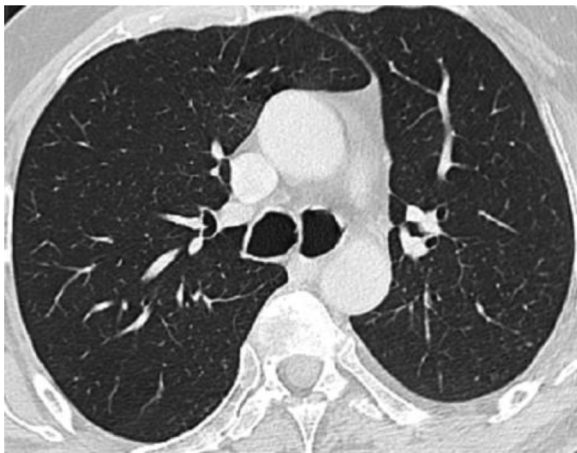


Fig. 3 – Chest CT scan: axial view through the parenchymal window passing through the tracheal bifurcation.

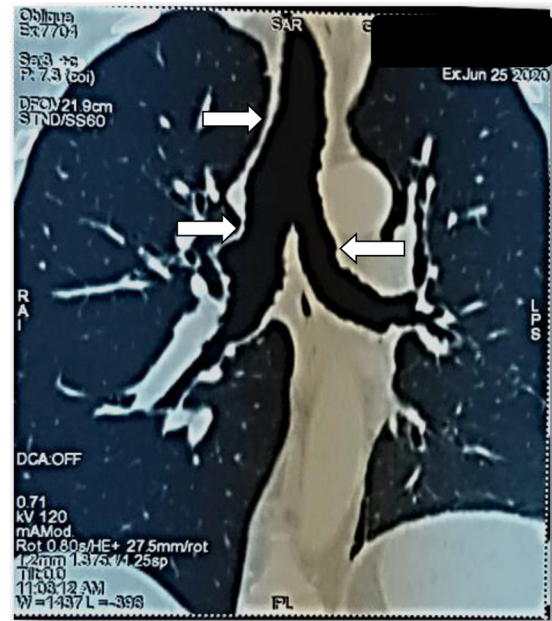


Fig. 5 – Chest CT scan: coronal reconstruction according to a plane passing through the trachea and the stem bronchi objectifying the tracheobronchomegaly.



Fig. 4 – Chest CT scan: axial view through the parenchymal window passing through the 2 main bronchi, the left main bronchus at 15 mm and the right main bronchus at 17 mm.

sition to certain irritants of the bronchial membrane, notably tobacco, and pollution.

For our patient, there is no similar case in the family, no congenital syndrome or associated syndrome, no notion of smoking and no exposition to a particulate pollution.

In a study about 128 cases over a period of 30 years, the mean age of the diagnosis was 54 years with few symptoms in childhood [9]. No correlation has been proven between the age of diagnosis and the degree of tracheal dilation [10]. This pathology is 8 times more common in men [9]. Note that for our patient, who is male, aged 67, the diagnosis has been made in adulthood. Bronchopulmonary infections did not start to be frequent until 1 year ago.

The diagnosis of Mounier-Kuhn syndrome is based on well-coded measurements of the trachea and the main bronchi. These measurements can be made by a chest x-ray but are much more precise on CT [7,11].

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 in women with respective measurements of 21, 23, 17.4, and 19.8 mm [12,13]. An increase in the cross-sectional area of the trachea beyond 371 mm² for men and 299 mm² for women also defines the disease [14]. For our patient, the transverse diameter of the trachea was 34.7 mm, the anteroposterior diameter at 24, the diameter of the left main bronchus at 15 mm, and of the right main bronchus at 17 mm.

The Dilated airways have often a scalloped appearance that is secondary to herniation of musculo-membranous tissue through the bronchial cartilaginous rings. Large diverticular or sacculiform formations may also be observed, especially in the posterior and postero-basal territory of the trachea and the stem bronchi [15]. For our patient, the dilation was harmonious on the chest CT scan; on the other hand, endoscopic bronchial exploration was not performed.

Mounier-Kuhn syndrome has 3 subtypes. In type I, there is a slight symmetrical dilation of the trachea and/or the main bronchi. In type II, the dilation and diverticula are distinct. In type III, the diverticular and sacculiform structures extend to the distal bronchi [16]. Our patient then presents type I.

In Mounier-Kuhn Syndrome, the symptomatology is not very specific. Can be poor or simply absent, or it manifests mainly by recurrent bronchopulmonary infections [17]. The cough is typically productive, sometimes associated with hemoptysis. Over the course of the process, the cough becomes ineffective with the increased dilation, which leads to stagnation of bronchial secretions and superinfection of the airways [6]. This is the case with our patient, who has a chronic productive cough, with abundant sputum, sometimes white sometimes green-yellow, and with a notion of recurrent bronchopulmonary infections but recent.

Mounier-Kuhn syndrome may also include sinonasal polyposis and polymalformative genetic syndrome including bilateral ptosis, epicanthus, micrognathism and excess skin of the upper lip [18]. None of these signs was reported in our patient.

A wide range of pulmonary and non-pulmonary clinical consequences associated with Mounier-Kuhn syndrome have been described [19]. The most common respiratory complications are bronchiectasis, tracheobronchomalacia, and emphysema, while the most common non-respiratory comorbidity was Gastroesophageal reflux disease (GERD) [20]. Only GERD was reported by our patient.

Spirometry may be normal or most frequently shows obstructive ventilatory disorder with increased capacity of pulmonary Function (CPT) and Residual volume (RV) as an abnormality [21]. For our patient, the spirometry was normal.

The treatment of Mounier-Kuhn syndrome is mainly supportive, which includes prevention with vaccinations, treatment of infections with antibiotics, as well as elimination of secretions with mucolytics and pulmonary physiotherapy; This is the treatment we recommended for our patient.

For patients with tracheobronchomalacia, long-term continuous positive airway pressure [19], airway stenting [10,20], surgical tracheoplasty [22], and laser treatment [17] have all been tried. A double lung transplant was performed in the terminal phase [21].

Conclusion

Mounier-Kuhn syndrome or tracheobronchomegaly is a very rare condition whose congenital or acquired origin is still debated, but most likely due to atrophy of the elastic fibers. The clinical signs are not very specific and the positive diagnosis is purely radiological. The impact assessment is necessary and based on an analysis of the lung parenchyma by a CT chest exam and on a functional respiratory analysis by spirometry. Treatment aims to treat bronchopulmonary infections or better prevent them and to free the airways by physiotherapy.

Patient consent

The patient confirmed the consent for publication of our case report.

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