

Mounier Kuhn Syndrome: A Rare Variety of Bronchial Dilatation

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Abstract

Background: Mounier Kuhn's syndrome is a rare condition. It is characterised by an increase in the diameter of the trachea and main bronchi. Imaging plays a fundamental role in confirming the diagnosis. This syndrome is little known and has not benefit of enough studies allowing a better comprehension of its pathophysiology and a performing therapeutic approach.

Case presentation: In this paper, we report the case of a 53-year-old man suffering from recurrent respiratory infections. Imaging has made the diagnosis of Mounier Kuhn syndrome by showing an increase in the diameter of the trachea and main bronchi. The treatment was based on physiotherapy and appropriate antibiotic therapy in case of infectious outbreak.

Conclusions: This long-neglected "orphan disease" should receive more research attention in order to provide better understanding of its aetiology, pathogenesis and multimodality treatment

Keywords: Mounier Kuhn syndrome, Tracheal dilatation, bronchial dilatation, congenital and rare disease, radiological diagnosis

BACKGROUND:

Mounier Kuhn's syndrome is a rare and congenital disease. It is characterized by an increase in the size of the trachea and main bronchi caused by a congenital disorder of the connective tissue and smooth muscles of these structures, the distal bronchial tree is usually normal. Its diagnosis is accomplished with the use of Computed tomography (CT). It can be associated with other congenital conditions, which makes the understanding of its physiopathology more complicated. Its clinical features vary from being asymptomatic to severe respiratory distress.

However, therapy remains conservative [1].

CASE REPORT:

Mr AB, a 53 years old non-smoking patient and father of 7 children, with a history of recurrent respiratory infections. He was admitted for a cough and purulent sputum evolving in a context of deterioration of the general state. Clinical examination showed bilateral ptosis, polypnea at 22 breathes per minute, sonorous bilateral rales in pleuro-pulmonary auscultation and a fever at 38.5 °C. Chest X-ray showed tracheal dilatation (figure 1).

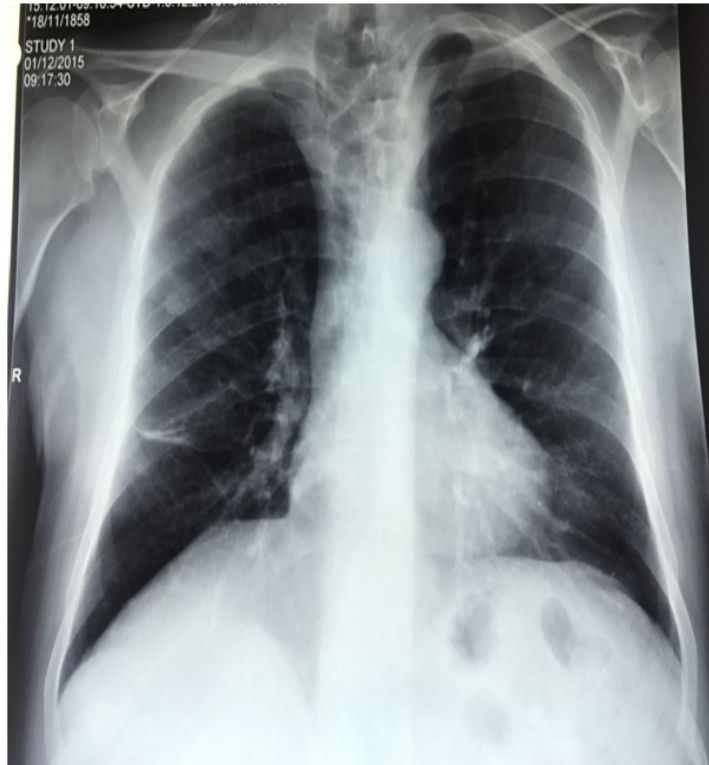


Fig 1: *Chest X-ray showing tracheal dilatation*

Blood tests found a CRP at 75 mg/l, white blood cells at 12,500/ mm³ predominantly of neutrophils at full blood count. Thoracic computed tomography was conducted (figures 2, 3, and 4) and revealed tracheal

dilatation with a diameter of 35 mm (whereas the normal diameter should not exceed 25 mm in humans), and dilation of main stem bronchi with a diameter of 25 mm on the right and 26 mm on the left.

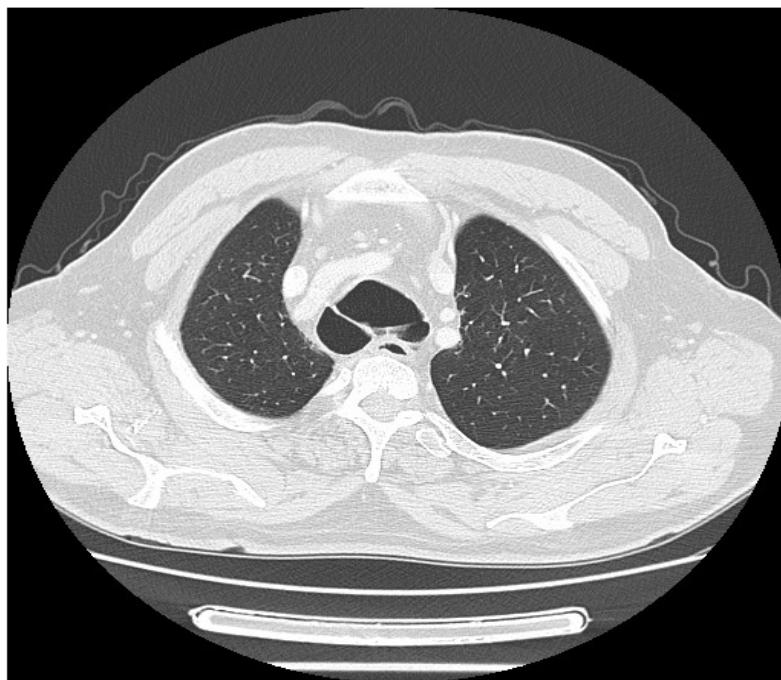


Fig 2. *CT scan showing tracheal dilatation.*



Fig 3. CT scan showing an increase in the diameter of both main bronchi.



Fig 4. CT image reconstruction confirming Mounier Kuhn's syndrome

Bronchial fibroscopy demonstrated an inflammatory aspect of the mucosa with tracheomalacia and tracheal diverticulum (figure 5). Spirometry was suggestive of obstructive abnormality with poor bronchodilator reversibility, Alpha-1 antitrypsin was 255 mg/dL (100-300 mg/dL), nasal endoscopy did not reveal any polyposis and the

spermogram was not performed (patient, father of 7 children). The diagnosis of a Mounier Kuhn syndrome has been made. The evolution under antibiotic treatment (of the episode of infection) and adequate chest physiotherapy for postural drainage of secretions was marked by clinical and biological improvement.

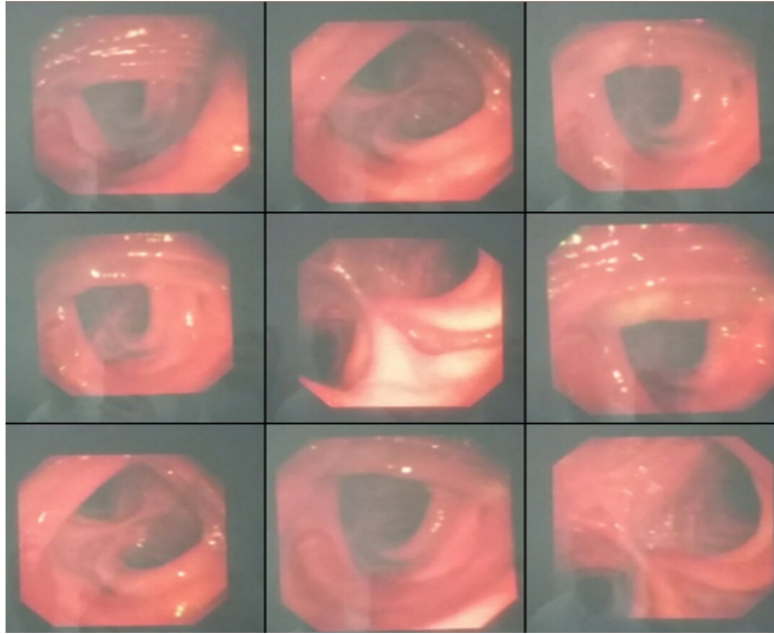


Fig 5. *endoscopic appearance of tracheomalacia and tracheal diverticulum*

DISCUSSION

Mounier Kuhn syndrome is a rare congenital disorder. It is characterised by a developmental defect in the connective tissue and smooth muscle resulting in increased tracheal diameter and enlarged main bronchi. There is also an absence of the myenteric plexus of the bronchial tree. Diagnosis usually takes place in adulthood. No correlation between increasing age and increasing tracheal diameter was found [2].

The syndrome was first described by Mounier Kuhn in 1932 and the term tracheobronchomegaly was introduced by Katz and al. in 1962. The mechanism is not known. It is a rare disease that can come in congenital form: familial forms were described with possible recessive inheritance, or in acquired form: Pulmonary fibrosis in adults and after mechanical ventilation in premature babies (Barotrauma). Some patients develop mucosal herniation between the tracheal rings leading to tracheal diverticulosis. The poor clearance of secretions is associated with recurrent lower respiratory tract infections [3]. There is also increase in deadspace and tidal volume [4].

This syndrome predominates in humans in the third and fourth decade [5]. Our presentation is that of a 53-year-old patient. The symptomatology is non-specific, poor or absent and is manifested basically by recurrent bronchopulmonary infections [6] and it is the same clinical symptomatology reported in our observation.

The clinical examination can be strictly normal, as it can show nasosinus polyposis and a polymalformative syndrome with bilateral ptosis, epicanthus, micrognathia and excess skin of the upper lip. Our patient presents only bilateral ptosis. Asthenospermia or azoospermia has been reported in some patients with Mounier-Kuhn syndrome.

Chest X-ray can help diagnosis by showing an increase in the size of the trachea beyond 3 cm especially in profile incidence [7].

Thoracic computed tomography confirms the diagnosis, it finds an increase in the tracheal transverse diameter upper than 3 cm in adults, and an increase in the right and left main bronchi greater than 2.4 cm and 2.3 cm respectively. It also serves to reveal cylindrical or cystic proximal bronchiectasis, extending to the fourth-order bronchi. The distal bronchial tree is usually normal.

Magnetic resonance imaging (MRI) does not have any supplementary advantage in diagnosis [7].

Bronchoscopy may show dilated proximal airways with purulent secretions, tracheal diverticulae and tracheomalacia. Recent data would suggest that bronchoscopy with confocal microscopy may aid diagnosis by permitting real-time analysis of the bronchial mucosal microstructure, demonstrating a deficiency of elastin fibres within the bronchial wall. Bronchoscopy can be difficult to perform

because of important airway obstruction due to tracheomalacia [7].

Spirometry can show different degrees of obstruction or increased residual function [7].

Mounier-Kuhn syndrome has 3 subtypes. In type 1, there is a slight symmetric dilation in the trachea and main bronchi. In type 2, the dilation and diverticula are distinct. In type 3, diverticular and saccular structures extend to the distal bronchi [8]. Our patient is classed in type 2.

Literature reviews have discussed the possibility of associating Mounier Kuhn's syndrome with other congenital pathologies such as Kenny-Caffey syndrome, Marfan's syndrome, hypogammaglobulinemia, Ataxia-telangiectasia, Ankylosing spondylitis, Brachmann-de Lange syndrome and cutis laxa [8, 9]. This association makes the understanding of the mechanism of the occurrence of Mounier Kuhn syndrome more complicated. Most frequent pulmonary complications are bullous emphysema, aspergillosis and pneumothorax [7, 10]. Our patient has a Mounier Kuhn syndrome without any other association or complication.

The main differential diagnosis in our observation is Williams-Campbell syndrome which is a rare condition characterized by the absence of cartilaginous rings in the sub segmental bronchi, resulting in dilation of the cystic bilateral bronchi which collapse on expiration and induce a hyperinflation. It usually affects the bronchi beyond third division (fourth to sixth order) [11]. The caliber of the trachea and main bronchi is strictly normal: this concept is fundamental, it characterizes the Williams Campbell syndrome and allows to exclude this diagnosis in our observation.

Treatment is based on physiotherapy and appropriate antibiotic therapy in case of outbreak infection (which is often the result of atypical organisms, tuberculous or non-tuberculous mycobacteria). Prevention of recurrent infections will prevent progression to irreversible pulmonary fibrosis [12]. Central impairment excludes any surgical treatment, and the placement of tracheobronchial prosthesis was reported in advanced cases. Lung transplantation has been performed in at least two cases of Mounier-Kuhn syndrome, one of who unfortunately died in the post-operative period.

CONCLUSION

Mounier-Kuhn syndrome or tracheobronchomegaly is a very rare condition whose congenital or acquired origin is still controversial. The clinical signs are not very specific and; it should be kept as a differential diagnosis in patients coming with recurrent lower respiratory tract infections. The radiological diagnosis is easy, based on a careful analysis of the central airways and pulmonary parenchyma by CT examination. However more work has to be done in terms of treatment and follow up of this orphan disease.

ABBREVIATIONS

CT: computed tomography

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

AUTHORS' CONTRIBUTIONS

Fatima Zahra Mrabet drafted this manuscript under Hicham Souhi's supervision. Jihane Achrane, Hanane Ouazzani, Ismail Rhorfi and Ahmed Abid have made substantial contributions to acquisition of data

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Citation: Fatima Zahra Mrabet, Jihane Achrane, Hicham Souhi, et al. Mounier Kuhn Syndrome: A Rare Variety of Bronchial Dilatation. *Archives of Pulmonology and Respiratory Medicine*. 2019; 2(1): 11-16.

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