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(CASE REPORT)

Hypoxemic pneumonia revealing Mounier-Kuhn syndrome: A case report

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Abstract

Mounier-Kuhn syndrome, or tracheobronchomegaly (TBM), is named after P. Mounier-Kuhn, who first described the condition in 1937 [1]. It is a rare disease with an origin that is still debated, whether congenital or acquired. The condition is characterized by a marked dilation of the trachea and proximal bronchi, resulting from thinning of the smooth muscle layer and atrophy or absence of elastic fibers. TBM is significantly more common in men [2]. The average age at diagnosis ranges from 30 to 60 years [3]. Although the exact cause of TBM remains to be determined, smoking and environmental pollutants are considered exacerbating factors that lead to airway inflammation [3].

Chronic cough with sputum production, accompanied by recurrent lower respiratory tract infections, characterizes the clinical presentation [4]. The clinical signs are not very specific. The radiological diagnosis is often straightforward on chest X-ray and CT scan. It is based on the measurement of the diameter or surface area of the trachea and main bronchi. Imaging is also essential for assessing the impact on the lung parenchyma. There is no curative treatment available [5]. We present the case of a 48-year-old man who was admitted to our pulmonology department for the management of acute pneumonia. During clinical evaluation and supplementary tests, tracheal and main bronchial dilation were noted, leading to the fortuitous discovery of Mounier-Kuhn syndrome.

Keywords: Mounier-Kuhn Syndrome; Tracheobronchomegaly; Pneumonia; Bronchial Dilation

1. Introduction

Mounier-Kuhn syndrome, or tracheobronchomegaly, is defined as significant dilation of the trachea and main bronchi. It is a rare condition secondary to a developmental defect of the connective tissue and smooth muscle of the trachea and bronchi, leading to tracheobronchomegaly. The clinical signs are varied and non-specific. The diagnosis relies on imaging, particularly chest X-ray and computed tomography (CT).

2. Case Presentation

We report the clinical case of a 48-year-old male patient, from a non-consanguineous marriage, with no toxic habits, who presented to the emergency department with respiratory symptoms including productive cough, fever, and progressive dyspnea. Oxygenation was impaired, with a SpO2 of 83% on room air. On auscultation, bilateral diffuse crackles were noted. Physical examination also revealed morphological abnormalities, such as loose, wrinkled skin, short and wide fingers and toes, as well as limb length inequality (Figure 2), along with short stature and a barrel-shaped chest.

The chest X-ray shows marked enlargement of the tracheal air column and both main bronchi, with a well-defined alveolar opacity occupying the upper two-thirds of the right hemithorax, along with diffuse bilateral alveolar images, possibly indicative of a right lobar pneumonia (Figure 1a). Tuberculosis screening was negative, and arterial blood gas

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analysis revealed type II respiratory failure with hypoxia (PaO_2: 55.5 mmHg, PaCo2: 57,1). The chest CT scan revealed (Figure 3):

- Tracheobronchomegaly with an undulating wall and the presence of posterior wall diverticula.
- Bilateral cystic and cylindrical bronchiectasis with mucous impaction.
- Pulmonary consolidation visible in the right Fowler position.

The clinical and radiological investigation led to the diagnosis of a coincidental finding of tracheobronchomegaly syndrome with associated malformations. The diagnostic criteria are based on the combination of physical malformations, respiratory symptoms, and characteristic radiological abnormalities.

The infectious workup at admission was positive, with a white blood cell count of 27,200/mm^3 and a C-reactive protein (CRP) level of 428 mg/L, showing improvement under antibiotic therapy. Respiratory PCR testing was positive for Acinetobacter baumannii, Haemophilus influenzae, Serratia, Streptococcus pneumoniae, rhinovirus, and enterovirus. Hepatic and renal function tests were unremarkable, and glucose and hemoglobin A1c (HbA1c) levels were normal.

The progression of the pneumonia was favorable with intravenous antibiotics (amoxicillin-clavulanic acid) for 7 days, along with respiratory physiotherapy, oxygen therapy, and non-invasive ventilation. There was significant clinical, biological, and radiological improvement. Follow-up chest X-ray (Figure 1b) showed early radiological clearing and a reduction in opacity.

In terms of prevention, it is essential to regularly monitor respiratory functions, vaccinate against respiratory infections (pneumococcus, influenza), initiate treatment promptly in case of infection, and promote respiratory hygiene. Early and appropriate management of infections is crucial to prevent serious complications.

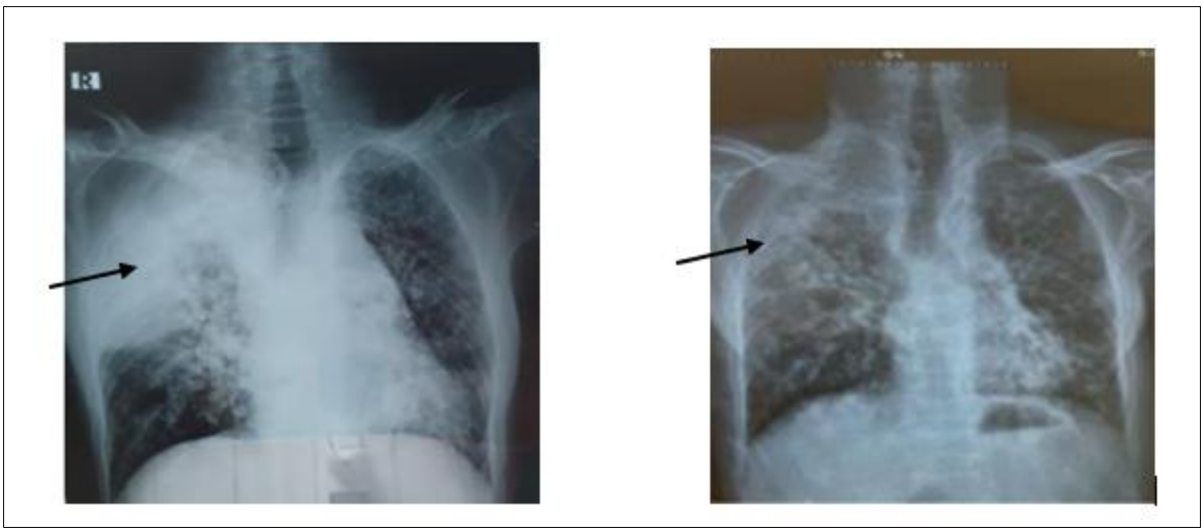

Figure 1a **Figure 1b**

Figure 1 and b The chest X-rays taken during the hospitalization show significant progression of the pulmonary involvement. Figure 1a shows a right lobar pneumonia, while Figure 1b shows a regression of the opacity, becoming less dense or less extensive

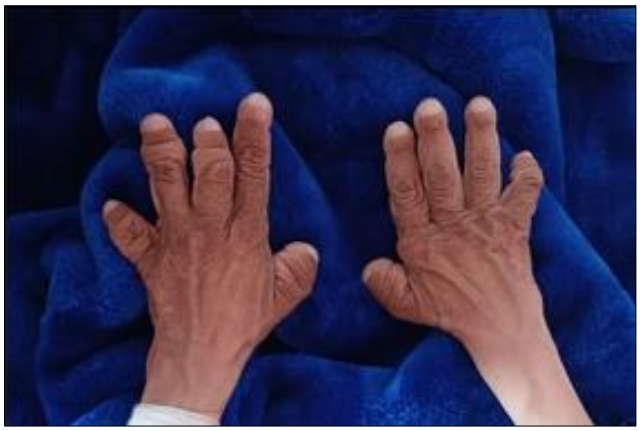

Figure 2 The following image shows morphological abnormalities of the patient's hand, including loose and wrinkled skin, short and wide fingers, as well as finger length inequality

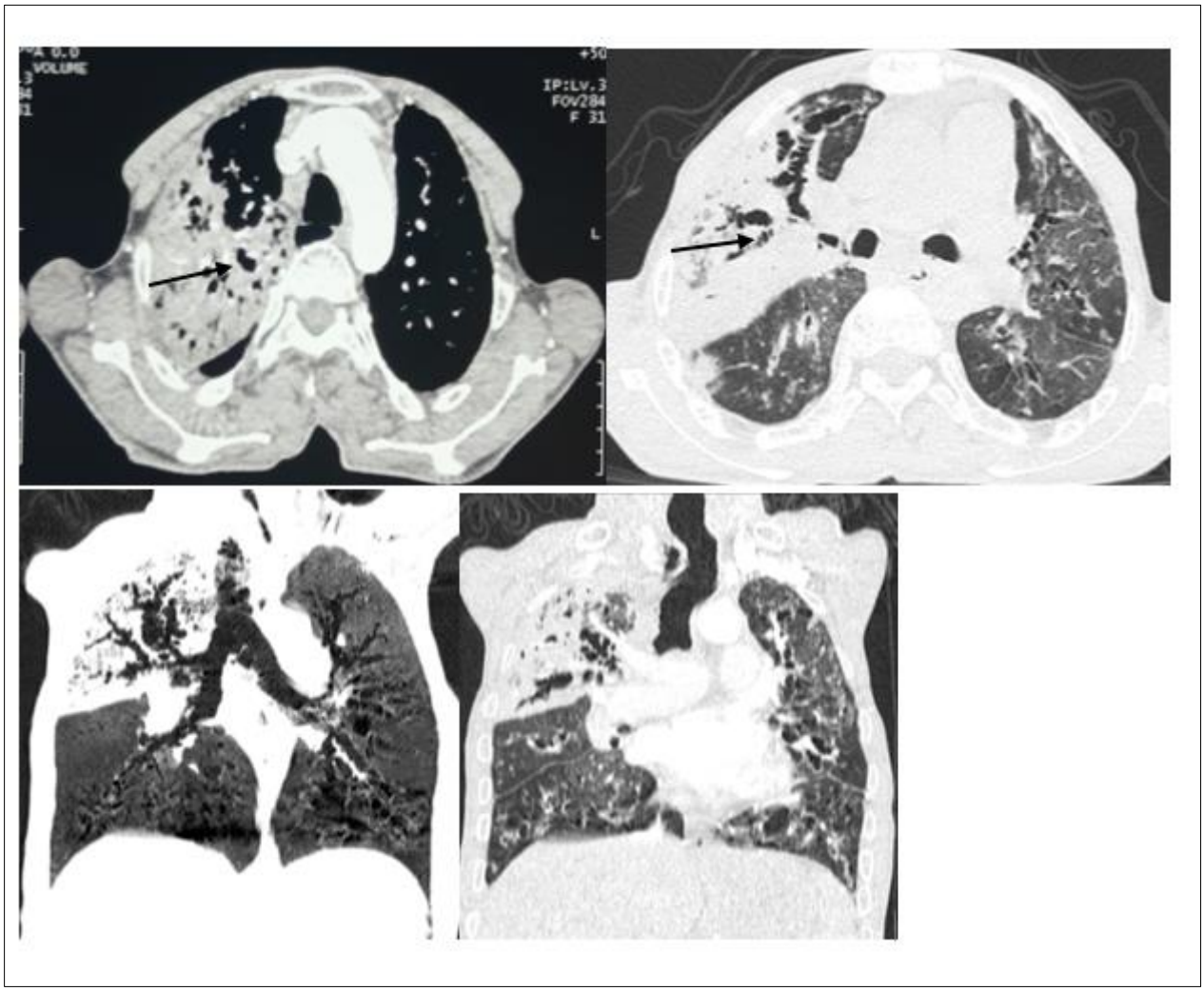

Figure 3 The chest computed tomography (CT) scan performed during hospitalization precisely characterized tracheobronchomegaly with a focus of pulmonary consolidation visible in the right Fowler position, along with bilateral cystic and cylindrical bronchiectasis (black arrow)

3. Discussion

Mounier-Kuhn Syndrome, also known as tracheobronchomegaly (TBM), is a rare but well-defined condition both clinically and radiologically. This syndrome is primarily characterized by marked dilation of the trachea and main bronchi. This abnormal dilation is often associated with other respiratory abnormalities such as bronchiectasis, tracheal diverticula, and recurrent broncho-pulmonary infections. These manifestations contribute to the clinical severity of the disease, particularly due to the stasis of secretions and the increased risk of infections in the airways. While tracheal dilation is the hallmark feature of Mounier-Kuhn syndrome, it can also present with additional abnormalities. Indeed, nasosinusal polyposis is frequently observed in patients with this syndrome. Other clinical signs may also be present as part of a polymalformative syndrome, including bilateral ptosis, epicanthus, micrognathia, and excess skin of the upper lip. These signs, though atypical, can guide the clinician toward a more accurate diagnosis of Mounier-Kuhn syndrome, especially in cases associated with syndromic malformations. Mounier-Kuhn syndrome was first described in 1932 by Mounier-Kuhn, who made the first endoscopic and radiological description of this condition. It was not until 1962 that Katz et al. introduced the term "tracheobronchomegaly" to refer to this pathology, thus emphasizing the involvement of the trachea and bronchi in the pathological process [6].

It is a rare condition, with fewer than 100 cases reported in the literature to date [7]. It primarily affects young adults (in their 3rd to 4th decade) with a clear male predominance [8, 9]. Rare cases have been reported in children and elderly individuals.

The etiopathogenesis of TBM is uncertain. The congenital cause, with an autosomal recessive transmission pattern, is likely given its association with Ehlers-Danlos syndrome in adults and cutis laxa in children. It can also be associated with Kenny-Caffey syndrome, Brachmann-de Lange syndrome, as well as rheumatological disorders such as ankylosing spondylitis and rheumatoid arthritis [3]. However, TBM frequently presents in adults in a sporadic form, which supports the theory of an acquired origin for the condition [10]. Several predisposing factors have been suggested, such as barotrauma during neonatal intensive ventilation with oxygen therapy [11, 12], or exposure to certain bronchial irritants, primarily tobacco and pollution.

This dilation is due to an abnormality in the elastic and muscular tissues of the tracheobronchial wall [13]. Clinically, patients present with persistent cough and recurrent pneumonias. The cough is typically productive and sometimes associated with hemoptysis. The disease generally progresses to chronic bronchitis. Endoscopy has limited value due to the extent of the dilation, and diagnosis relies primarily on radiological investigations. Tracheobronchography, at a minimum, is rarely performed [14].

The diagnosis of TBM is made through imaging examination. The diagnosis is confirmed by measuring the trachea, which can be done with a chest X-ray, but high-resolution computed tomography (HRCT) is the gold standard. Chest X-rays show dilation of the trachea and bronchi, with an irregularly undulating or scalloped appearance of the air columns, along with associated complications such as diverticula, bronchiectasis, and infections [15]. Measurement of the trachea and main bronchi can be performed using CT. In adults with TBM, the diagnostic criteria on CT include an increase in the tracheal diameter beyond 30 mm and/or an increase in the diameters of the right and left main bronchi beyond 18 mm and 21 mm, respectively [16]. Tracheobronchomegaly has been classified into three subtypes. In type 1, there is diffuse and symmetrical dilation of the trachea and main bronchi, relatively subtle in appearance. Type 2 shows more evident dilation with bizarre, eccentric configurations, and diverticula may also be present. In type 3, the diverticula or sacculations extend into the distal bronchi [17].

The secondary objective of CT is to assess associated broncho-parenchymal and wall lesions [18]: bronchiectasis is frequently associated, and more or less severe parenchymal lesions may be intertwined (atelectasis, pneumonia, interstitial fibrosis, or emphysema). Sternocostal malformations have also been reported. Mounier-Kuhn syndrome should be differentiated from Williams-Campbell syndrome, which features congenital cystic bronchiectasis resulting from a deficiency of cartilage in the fourth and sixth order bronchi. However, in this syndrome, the trachea and main bronchi are of normal caliber [6]. The condition can be tolerated for a long time, but progression to chronic respiratory failure is inevitable. Treatment is based on respiratory physiotherapy and antibiotic therapy in case of infection. Treatment with continuous positive airway pressure (CPAP) or the insertion of an endobronchial prosthesis is sometimes indicated in severe cases [19, 20].

4. Conclusion

Mounier-Kuhn syndrome remains a rare and complex condition, often underdiagnosed, but early management is crucial for improving the long-term prognosis of patients. A better understanding of the pathophysiology of this disease could help develop new therapeutic strategies, and larger epidemiological studies could contribute to better identifying risk factors and associated comorbidities.

Advances in diagnostic techniques, particularly through radiology and bronchoscopy, have improved the ability to identify this condition and initiate earlier treatments. The role of clinicians in recognizing this pathology is therefore crucial to optimize patient management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

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