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Tracheobronchomegaly (Mounier-Kuhn Syndrome): A Rare Cause of Recurrent Pulmonary Infections – Imaging Case Review

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Abstract | **Case Report**

Mounier-Kuhn syndrome, or tracheobronchomegaly, is a rare congenital disorder caused by structural weakness of the tracheobronchial wall, leading to abnormal dilatation of the trachea and main bronchi. It predisposes patients to impaired mucociliary clearance, recurrent infections, and progressive pulmonary dysfunction. We present the case of a 40-year-old man with chronic productive cough and repeated episodes of lower respiratory tract infections resistant to standard medical therapy. Chest CT revealed marked dilatation of the trachea (>30 mm) and main bronchi (20 mm right, 18 mm left), associated with diffuse bronchiectasis, bronchial wall thickening, mucus plugging, and centrilobular micronodules. These findings were consistent with Mounier-Kuhn syndrome. Recognition of this entity is essential, as its clinical presentation is nonspecific, and diagnosis relies primarily on CT-based morphometric criteria. Management is supportive, focusing on infection prevention, prompt antibiotic therapy, and pulmonary physiotherapy.

Keywords: Mounier-Kuhn syndrome, Tracheobronchomegaly, Bronchiectasis, Recurrent respiratory infections, CT imaging, Airway dilatation.

INTRODUCTION

Tracheobronchomegaly, also known as Mounier-Kuhn syndrome, is a rare congenital disorder characterized by significant dilatation of the trachea and main bronchi due structural weakness of the connective tissue of the tracheobronchial wall often leading to recurrent respiratory tract infections, bronchiectasis, and progressive pulmonary dysfunction. The symptoms remain nonspecific and computed tomography is the main diagnosis method,

CASE PRESENTATION

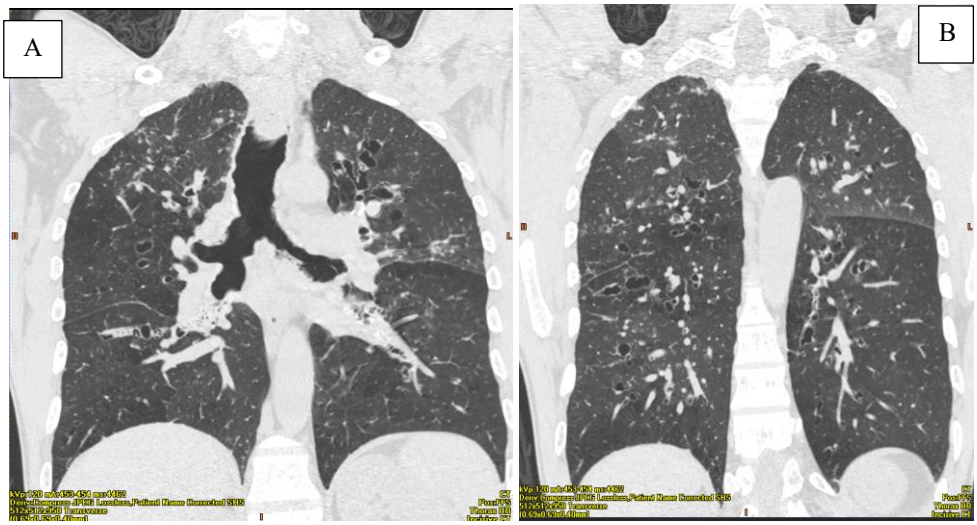
We report the case of a 40-year-old male patient, with a long-standing history of recurrent episodes of bronchitis and lower respiratory tract infections, characterized by productive cough, exertional dyspnea, and repeated courses of antibiotic therapy. Despite adequate medical management, his respiratory symptoms persisted, warranting further diagnostic evaluation.

Imaging Findings:

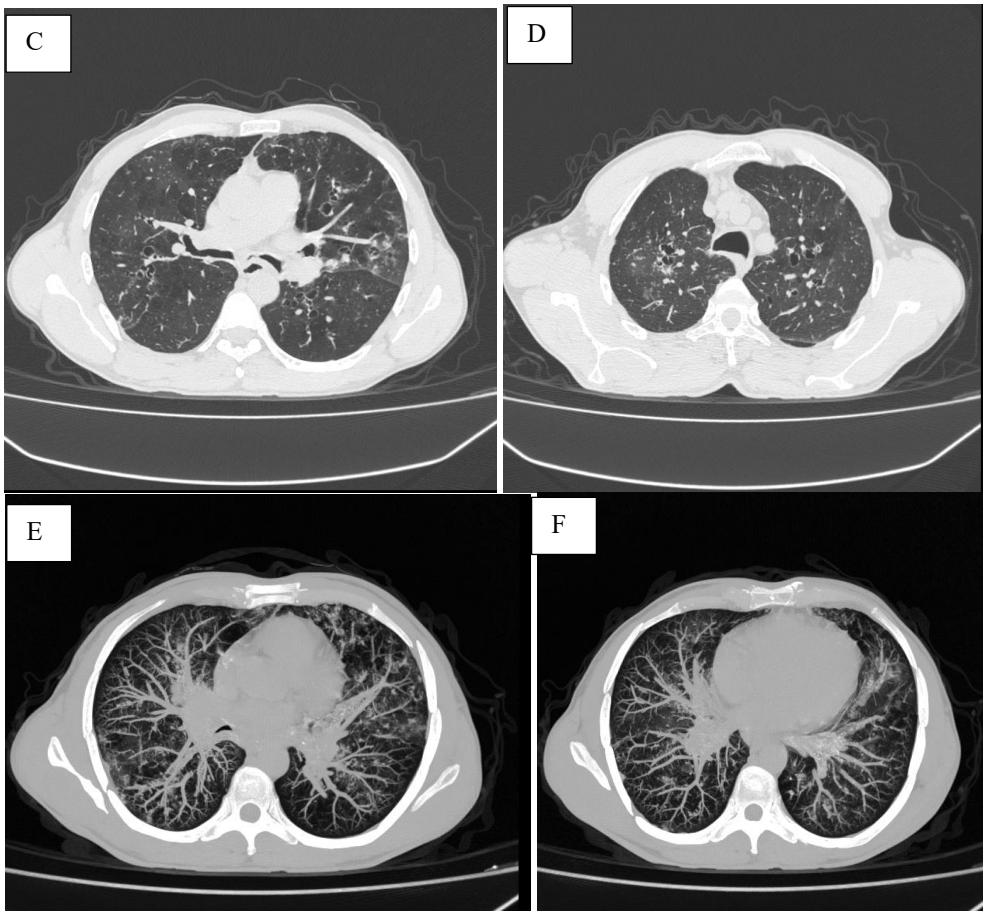
Chest CT demonstrated a markedly enlarged trachea, with a transverse diameter exceeding 30 mm, accompanied by dilatation of the main bronchi, measuring more than 20 mm on the right and 18 mm on the left. These airway changes were associated with diffuse bronchiectasis and bronchial wall thickening.

In addition, mucus plugging was observed within the dilated bronchi. No evidence of endobronchial obstruction or extrinsic compression was identified. (figures A-F)

In conjunction with the patient's clinical history of recurrent lower respiratory tract infections and chronic bronchitic symptoms, the constellation of imaging findings was considered highly suggestive of Mounier-Kuhn syndrome (tracheobronchomegaly).



A-B: Coronal images in the lung parenchymal window demonstrating marked tracheal and and main bronchi dilatation (A) associated bronchiectasis (B)



C–D. **Axial CT images in the lung parenchymal window demonstrate bronchiectasis associated with bronchial wall thickening.**

E–F. **Axial maximum intensity projection (MIP) reconstructions reveal multiple centrilobular micronodules along the segmental and subsegmental bronchi.**

DISCUSSION

Mounier-Kuhn syndrome is a disorder of the tracheobronchial wall, characterized by atrophy or deficiency of elastic fibers and smooth muscle, resulting in abnormal airway compliance and marked dilatation. Although the exact etiopathogenesis remains unclear, the

structural weakening of the airway wall is believed to underlie the tracheobronchomegaly observed in affected patients. [1]

It remains a rare disorder, with fewer than 100 cases documented in the literature to date. The first endoscopic and radiological description of this condition was provided by Mounier-Kuhn in 1932. Subsequently, in 1962, Katz and colleagues introduced the term 'tracheobronchomegaly.' The disease typically affects young adults, most often during the third or fourth decade of life, with a clear male predominance. [2]

The clinical presentation of Mounier-Kuhn syndrome is often nonspecific and may range from mild or even absent symptoms to recurrent lower respiratory tract infections. Patients typically exhibit a chronic productive cough, which may occasionally be accompanied by hemoptysis. [3]

As airway dilatation progresses, cough efficiency diminishes, resulting in impaired clearance of bronchial secretions and recurrent pulmonary infections [6]. This pattern was observed in our patient, who presented with a chronic productive cough, along with a history of recurrent pulmonary infections, particularly in recent episodes

The diagnosis of tracheobronchomegaly (TBM) is primarily based on standardized measurements of the trachea and main bronchi.

While these measurements can be obtained on chest radiographs, computed tomography (CT) provides greater accuracy and detail [4]

In men, TBM is defined by a transverse and sagittal tracheal diameter exceeding 25 mm and 27 mm, respectively, and/or a right and left main bronchus diameter greater than 18 mm and 21 mm. In women, the corresponding thresholds are 21 mm and 23 mm for the trachea [5], and 17.4 mm and 19.8 mm for the main bronchi [6].

Dilated airways often demonstrate a markedly scalloped or irregular appearance due to herniation of the musculo-membranous tissue through the cartilaginous bronchial rings. Large diverticular or saccular formations may also be observed, predominantly along the posterior and postero-basal regions of the trachea and main bronchi [7].

In addition to assessing airway dilatation, CT allows detailed evaluation of associated bronchial and parenchymal abnormalities. Bronchiectasis is commonly present, often accompanied by bronchial wall thickening, reflecting chronic airway inflammation. Secondary changes related to recurrent infections, such as mucus plugging, areas of consolidation, and atelectasis, may also be observed. Parenchymal alterations can range from mild post-infectious scarring to interstitial fibrosis or emphysematous changes. [8]

Differential diagnoses include acquired tracheomegaly (post-tracheostomy, prolonged ventilation, or connective tissue disorders such as Ehlers-Danlos and Marfan syndromes).

Treatment is mainly supportive:

Management of Mounier-Kuhn syndrome is primarily supportive, focusing on prevention of infections through appropriate vaccinations, prompt treatment of acute infections with antibiotics, and facilitation of airway clearance using mucolytic agents and pulmonary physiotherapy [9]

CONCLUSION

Mounier-Kuhn syndrome is a rare but important cause of recurrent lower respiratory tract infections. Awareness of its imaging features, particularly marked dilatation of the trachea and main bronchi on CT, is essential for timely diagnosis. Radiologists play a key role in recognizing this condition and guiding clinical management.

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