Radiology Case Reports 20 (2025) 1268–1272

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

A rare cause of dyspnea: Mounier Kuhn syndrome ☆

Feryal El Oualladi, MD, Mohamed Labied, MD, Ismail Bouzid, MD, Chorouk Mountassir, MD, Ghizlane Lembarki, MD, Mouna Sabiri, PhD, Samira Lezar, PhD.*

Central Unit of Radiology, University Hospital Center Ibn Rochd, School of Medicine and Pharmacy of Casablanca, Hassan II University, Casablanca, Morocco

ARTICLE INFO

Article history:
Received 6 October 2024
Revised 6 November 2024
Accepted 7 November 2024

Keywords:
Mounier-Kuhn
tracheobronchomegaly
CT scan

ABSTRACT

Mounier-Kuhn syndrome, also known as tracheobronchomegaly, is a rare clinical and radiological condition characterized by tracheobronchial dilatation and recurrent respiratory infections. Patients may be asymptomatic or present with severe respiratory illnesses. A CT scan is sufficient to confirm the diagnosis. Treatment is symptomatic, and surgery is rarely indicated. We present the case of a 55-year-old patient with a 30-year history of chronic dyspnea, who presented with worsening shortness of breath, a productive cough, and episodes of bronchial superinfection. A chest CT scan, performed before and after contrast injection during both inspiratory and expiratory phases, confirmed the diagnosis of Mounier-Kuhn syndrome. This case aims to raise awareness of this rare condition and highlights a presentation where dyspnea is the primary symptom.

Teaching points

- The clinical symptoms of Mounier-Kuhn syndrome are nonspecific and primarily include recurrent respiratory infections.
- The CT scan is the reference test for diagnosing Mounier-Kuhn syndrome by measuring the diameter of the trachea and the main bronchi.
- It is necessary to consider Mounier-Kuhn syndrome for patients with recurrent respiratory infections or chronic dyspnea.

Introduction

Mounier-Kuhn syndrome, also known as tracheobronchomegaly, is a chronic airway disease. It is clinically and radiologically well-defined and characterized by dilatation of the airways. The etiology is not fully understood, but amyotrophy of smooth muscle and connective tissue remains the most likely theory. Clinical signs are nonspecific, with recurrent respiratory infections being the predominant symptom. Diagnosis is radiologic, based on computed tomography. Treatment is symptomatic.

☆ Competing Interests: There are no conflicts of interest declared by any of the authors.

* Corresponding author.
E-mail address: elouallad i.feryal@gmail.com (F. El Oualladi).

https://doi.org/10.1016/j.radcr.2024.11.015

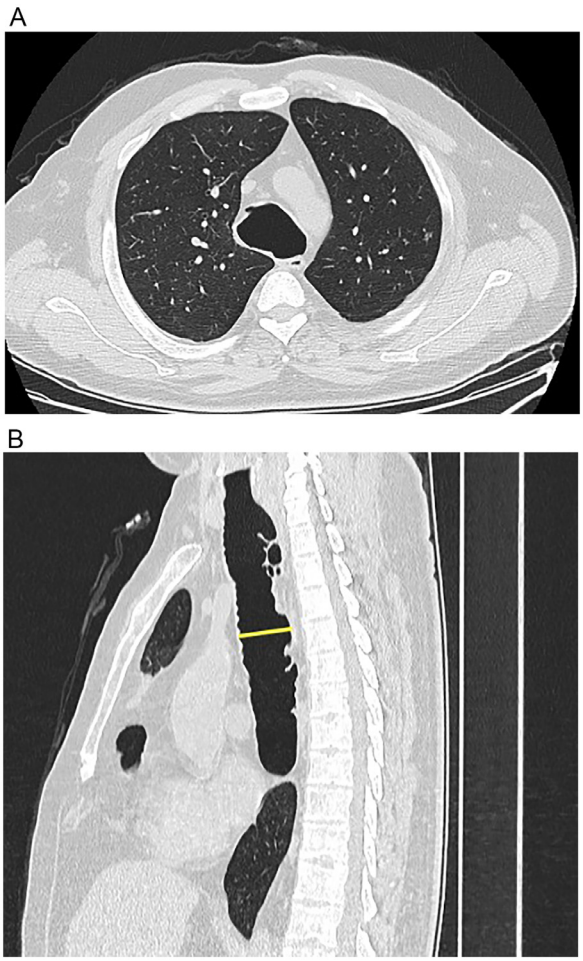

Fig. 1 – A 55-year-old male patient with Mounier-Kuhn syndrome. Chest computed tomography in parenchymal window showing dilated trachea measuring 45 mm (A) and 32 mm (B) in the transverse and sagittal planes respectively, with an irregular wall.

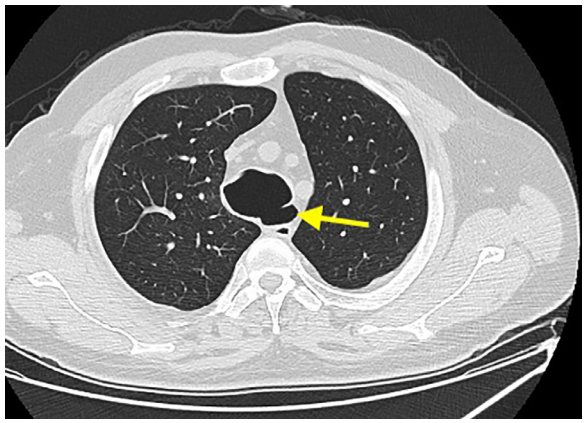

Fig. 2 – Chest computed tomography in parenchymal window showing tracheal diverticula (yellow arrow).

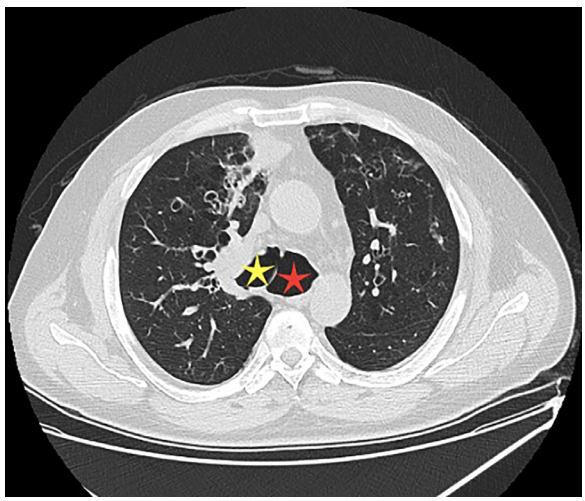

Fig. 3 – Chest computed tomography in parenchymal window showing dilated right main bronchus measured at 26 mm (yellow asterisk) and left main bronchus measured at 31 mm (red asterisk).

Case report

We report the case of a 55-year-old patient with a 30-year history of chronic stage II dyspnea and recurrent respiratory infections. A chronic smoker stopped smoking 22 years ago. The patient has no history of tuberculosis, connective tissue or autoimmune disease, and no significant personal or family medical history. He has presented over the past 4 months with worsening dyspnea, progressing to stage III, accompanied by a productive cough and frequent episodes of bronchial superinfection. Clinical examination revealed a febrile patient with an increased anteroposterior chest diameter and snoring rales on auscultation. A thoracic CT scan showed a dilated trachea measuring 45 mm in the transverse plane and 32 mm in the sagittal plane (Fig. 1), with an irregular wall and multiple tracheal diverticula (Fig. 2). The main bronchi were dilated to 26 mm on the right and 31 mm on the left (Fig. 3). Bilateral cylindrical and cystic bronchiectasis was present, with mucoid impactions (Fig. 4). The trachea's diameter on expiration was reduced to 21 mm in the anteroposterior plane (Fig. 5). Tracheal and bronchial dilatation was visible on maximum intensity projection (MIP) reconstructions (Fig. 6), and a 3D reconstruction program was used to examine the course and wall of the tracheobronchial tree (Fig. 7).

Bronchoscopy revealed a dilated, thin-walled tracheobronchial tree with diverticula and generalized collapse on expiration.

These radiologic and endoscopic findings confirmed the diagnosis of Mounier-Kuhn syndrome.

The infectious and immunological workups were negative, and the assessment for sarcoidosis was normal.

The patient showed improvement after symptomatic treatment, which included bronchodilators, mucolytics, physiotherapy, and a 7-day course of antibiotic therapy with amoxicillin and clavulanic acid.

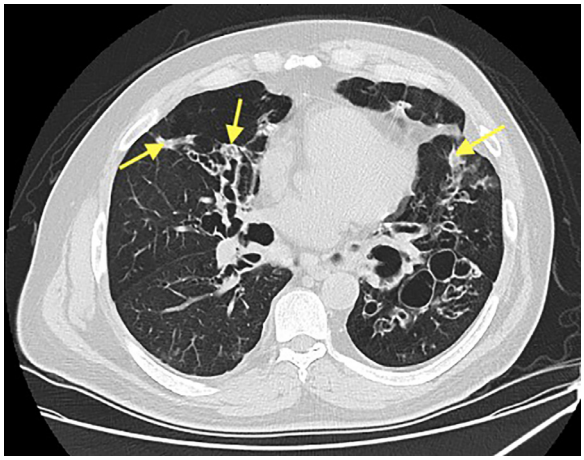

Fig. 4 – Chest computed tomography in parenchymal window showing bilateral cylindrical and cystic bronchiectasis with mucoid impaction (arrow).

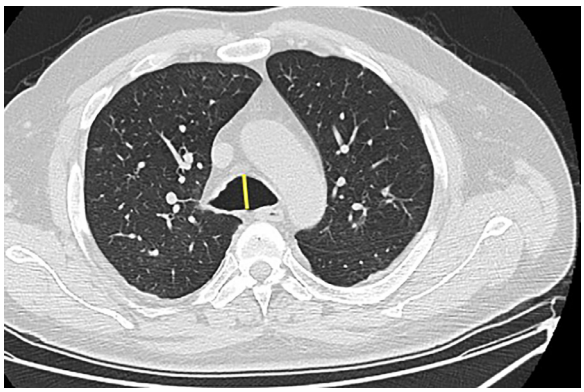

Fig. 5 – Chest CT scan in the parenchymal window with axial reconstruction showing a reduction in the anteroposterior diameter of the trachea to 21 mm during expiration.

Discussion

Mounier-Kuhn syndrome is defined as "a peculiar clinical and radiologic condition characterized by marked dilatation of the trachea and main bronchi associated with chronic respiratory infections" [1]. It is an extremely rare disease, first described by Mounier-Kuhn in 1932 [2]. The exact prevalence is unknown, but at least 300 cases have been documented in the literature [3]. The condition is more common after the third decade of life [3,4], with a notable male predominance of approximately 8:1 [3,5,6].

It is most commonly found in smokers, though cases have also been reported in nonsmokers [7]. The exact cause of this condition is unknown. Biopsy studies reveal thinning of the smooth muscle and atrophy of the elastic tissue in the trachea and mainstem bronchi [1,3].

The condition may be congenital or acquired. A congenital cause is likely, particularly when associated with other

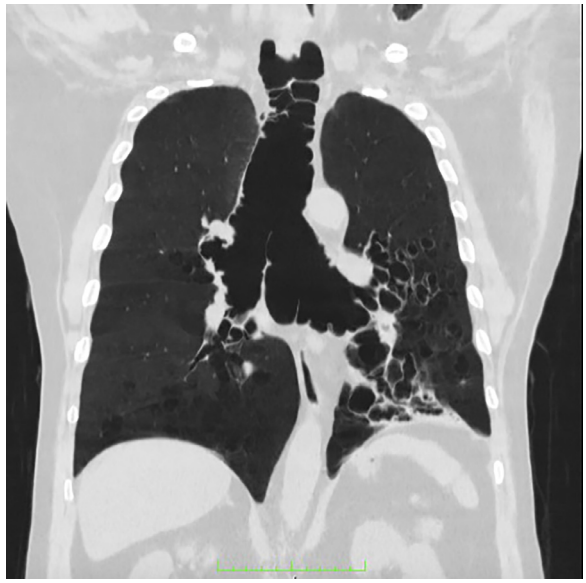

Fig. 6 – Chest CT scan with minimum intensity projection (MinIP) coronal reconstruction showing tracheobronchial dilatation and wall.

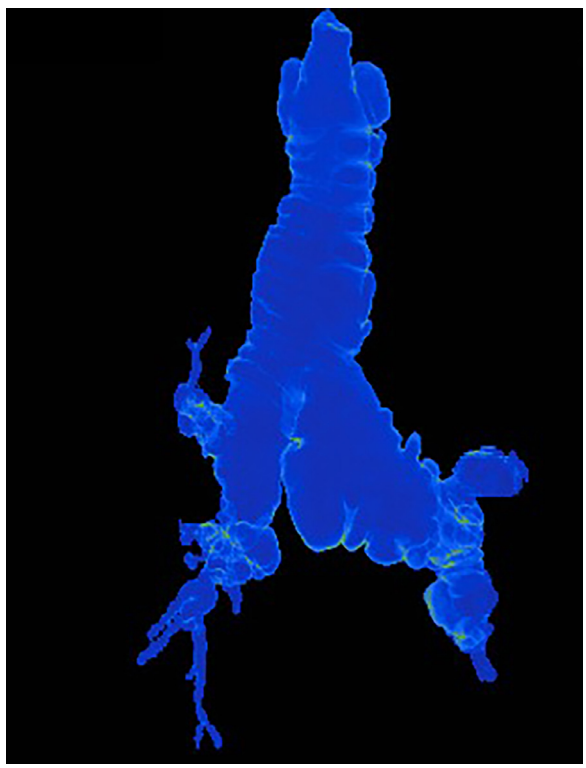

Fig. 7 – A 3D reconstruction program showing dilatation of the trachea and its diverticular wall as well as dilatation of the main bronchi.

syndromes such as Ehlers–Danlos syndrome [8], Marfan syndrome [3], Cuti Laxa in children, ataxia-telangiectasia, ankylosing spondylitis, Kenny-Caffey syndrome, and Brachmann-de Lange syndrome [3,4,6]. The sporadic nature of acquired cases is most common in adults [5,8,9].

There are 3 anatomic subtypes of the syndrome: [3,10]:

- Type 1: Diffuse symmetrical dilatation of the trachea and main bronchi.
- Type 2: Dilatation and diverticula are distinct.
- Type 3: Diverticula extends into the distal bronchi.

The clinical presentation of Mounier-Kuhn syndrome is variable [4]. Symptoms typically include a persistent productive cough, recurrent pneumonitis, dyspnea, and occasionally hemoptysis [11]. The condition progresses with increased bronchial dilatation, leading to ineffective coughing, then stagnation of bronchial secretions and suppuration of the airways.

Clinical examination may reveal bronchial rales, digital clubbing, fever, and tachycardia. Spirometry often shows variable degrees of obstruction and increased residual volume.

Bronchoscopy reveals increased trachea and mainstem bronchi diameters, with collapse noted on expiration [3].

Standard X-rays can reveal tracheal and bronchial dilatation, but CT has been the definitive diagnostic tool since its introduction in 1988 [12]. The involvement extends from the trachea to the fourth-order bronchi. Diagnosis is based on well-established measurements of the trachea and mainstem bronchi [3,6,7]:

- Transverse and sagittal diameters of the trachea are more than 25 mm and 27 mm, respectively, in men and more than 21 mm and 23 mm, respectively, in women.
- Diameters of the right and left main bronchi are beyond 18 mm and 21 mm in men and 17.4 mm and 19.8 mm in women.

The dilated airways often exhibit a deeply scalloped appearance due to the herniation of musculomembranous tissue through the cartilaginous bronchial rings, leading to the formation of diverticula. While the cervical trachea remains normal, there is a dilatation of the thoracic trachea.

The second goal of CT is to evaluate associated bronchoparenchymal and parietal lesions. Bronchiectasis is frequently observed, and parenchymal lesions of varying severity may be present, including atelectasis, pneumopathy, interstitial fibrosis, or emphysema. Sternocostal malformations have also been reported. Emphysema is often associated with pneumothorax. Pulmonary superinfections, including those caused by tuberculous and nontuberculous mycobacteria, are common.

Asymptomatic patients do not require specific treatment [4]. For patients with secretions, mucolytic therapy and respiratory physiotherapy are recommended. In cases of infection, antibiotic therapy is advised [3,6,8].

In rare cases, a Y tracheal stent or lung transplantation has been reported as a treatment option [3,6,8]. Prophylactic vaccination is also recommended.

The differential diagnosis includes Williams-Campbell syndrome, characterized by congenital cystic bronchiectasis due to insufficient cartilage in bronchi beyond the third order. In contrast to Mounier-Kuhn syndrome, the trachea and main bronchi are of the correct caliber [3]. Secondary emphysema and bronchiolitis may also be present.

When an acquired origin is suspected, a thorough review of the patient's history is essential. Conditions such as pulmonary fibrosis, diffuse interstitial pneumonia, sarcoidosis, mechanical ventilation (especially in premature infants), radiation therapy [3], and ankylosing spondylitis [13] may cause secondary tracheomegaly due to fibrotic traction during their evolution [14].

Conclusion

Mounier-Kuhn syndrome is often diagnosed late due to the nonspecific nature of its clinical symptoms. This diagnosis should be considered in case of recurrent respiratory infections or chronic dyspnea. Diagnosis is primarily radiological and is well-defined on CT scans. Treatment is primarily symptomatic.

Patient consent

Written informed consent was obtained from the patient for publication of his case.

REFERENCES

[1] Katz I, Levine M, Herman P. Tracheobronchiomegaly. The Mounier-Kuhn syndrome. Am J Roentgenol Radium Ther Nucl Med 1962;88:1084–94.
[2] Mounier-Kuhn P. Dilatation de la trachée: constatations radiographiques et bronchoscopiques. Lyon Méditerranée 1932;150:106–9.
[3] Krustins E, Kravale Z, Buls A. Mounier-Kuhn syndrome or congenital tracheobronchomegaly: a literature review. Respir Med 2013;107(12):1822–8.
[4] Celik B, Bilgin S, Yuksel C. Mounier-Kuhn syndrome: a rare cause of bronchial dilation. Tex Heart Inst J 2011;38(2):194–6.
[5] Johnston RF, Green RA. Tracheobronchiomegaly: report of five cases and demonstration of familial occurrence. Am Rev Respir Dis 1965;91(1):35–50.
[6] Menon B, Aggarwal B, Iqbal A. Mounier-Kuhn syndrome: report of 8 cases of tracheobronchomegaly with associated complications. South Med J 2008;101(1):83–7.
[7] Woodring JH, Howard RS 2nd, Rehm SR. Congenital tracheobronchomegaly (Mounier-Kuhn syndrome): a report of 10 cases and review of the literature. J Thorac Imaging 1991;6(2):1–10.
[8] Ayres JG, Pope FM, Reidy JF, Clark TJ. Abnormalities of the lungs and thoracic cage in the Ehlers-Danlos syndrome. Thorax 1985;40(4):300–5.
[9] Akgedik R, Karamanli H, Kizilirmak D, Kurt AB, Öztürk H, Yildirim BB, et al. Mounier-Kuhn syndrome (tracheobronchomegaly): an analysis of eleven cases. Clin Respir J 2018;12(3):885–9.
[10] Schwartz M, Rossoff L. Tracheobronchomegaly. Chest 1994;106(5):1589–90.
[11] Odell DD, Shah A, Gangadharan SP, Majid A, Michaud G, Herth F, et al. Airway stenting and tracheobronchoplasty improve respiratory symptoms in Mounier-Kuhn syndrome. Chest 2011;140(4):867–73.
[12] Dunne MG, Reiner B. CT features of tracheobronchomegaly. J Comput Assist Tomogr 1988;12(3):388–91.

[13] Fenlon HM, Casserley I, Sant SM, Breatnach E. Plain radiographs and thoracic high-resolution CT in patients with ankylosing spondylitis. Am J Roentgenol 1997;168: 1067–1072.

[14] Ghanei M, Peyman M, Aslani J, Zamel N. Mounier-Kuhn syndrome: a rare cause of severe bronchial dilatation with normal pulmonary function test: a case report. Respir Med 2007;101(8):1836–9.