Size matters: an unusual case report

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Mounier-Kuhn syndrome is a rare clinical entity characterized by dilation of the tracheobronchial tree leading to recurrent lower respiratory tract infections. The clinical spectrum ranges from asymptomatic patients to those with severe respiratory failure. The disease is diagnosed by the use of computerized tomography scans, bronchoscopy, and pulmonary function tests of the affected patients. There is no specific medical or surgical therapy for the entity, and care is largely supportive, targeted towards prevention of infections in such patients. We describe the case of a 27-year-old male who presented to us with complaints of paroxysmal episodes of cough with expectoration on and off since childhood. Computerized tomography scans revealed dilation of the trachea (diameter 33.7 mm) with bronchiectatic changes noted in the right lower lung lobe. Virtual bronchoscopy revealed tracheal scalloping and diverticulae which further substantiated the diagnosis of Mounier-Kuhn syndrome. Pulmonary function test result was normal.

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Introduction

Mounier-Kuhn syndrome is a rare clinical condition characterized by dilation of the trachea and bronchi which manifests as recurrent lower respiratory tract infections (LRTIs). It was first described in 1932 by Mounier-Kuhn [1], and since then, there are less than 100 documented cases in medical literature [2]. Although the underlying etiology is not well understood, pathogenesis involves the atrophy of smooth muscle and elastic fibres lining the wall of the trachea and bronchi. This leads to tracheobronchomegaly and may even cause tracheal and bronchial diverticulae. The diverticulae hamper effective mucociliary clearance leading to retention of airway secretions that act as a nidus of infection in such patients. As the entity is less commonly identified, these patients are treated as recurrent LRTIs and face significant morbidity and mortality risks.

Case history

A 27-year-old male presented with complaints of paroxysmal episodes of cough with minimal expectoration since childhood. The expectoration was colourless and odourless with no hemoptysis. Medical records revealed multiple hospital admissions for LRTI treated with intravenous antibiotics. He was not a smoker, and there was no history of asthma, atopy or tuberculosis in the patient or any of his contacts.

On examination, his vitals were stable (pulse rate 84/min, blood pressure 110/72 mmHg, respiratory rate 20/min, saturation: 94% on room air, and afebrile). General physical examination was significant for grade 1 clubbing. Examination of the respiratory system revealed bilateral rhonchi and coarse crepitations over bilateral mammary and interscapular region on auscultation. Remainder of the clinical examination was within normal limits.

Investigations

A chest radiography revealed gross enlargement of the trachea. Computerized tomography scan (Fig. 1) revealed tracheal diameter as 33.7 mm at the level of the arch of aorta and 35.2 mm at the carina, with an internal diameter of 21.8 mm (right main bronchus) and 31.1 mm (left main bronchus), respectively, along with bronchiectatic segments in the right lower lobe. Virtual bronchoscopy revealed dilated trachea and bilateral bronchi with diverticulae protruding along the posterior tracheal wall (Fig. 2). Tracheal scalloping was also noted.

Pulmonary function test result was normal.

Laboratory evaluation showed normal levels of serum calcium and 24-h urinary calcium levels. Serum angiotensin-converting enzyme levels were also normal, ruling out sarcoidosis. Serum antinuclear antibiodies, rheumatoid factor (RF), and

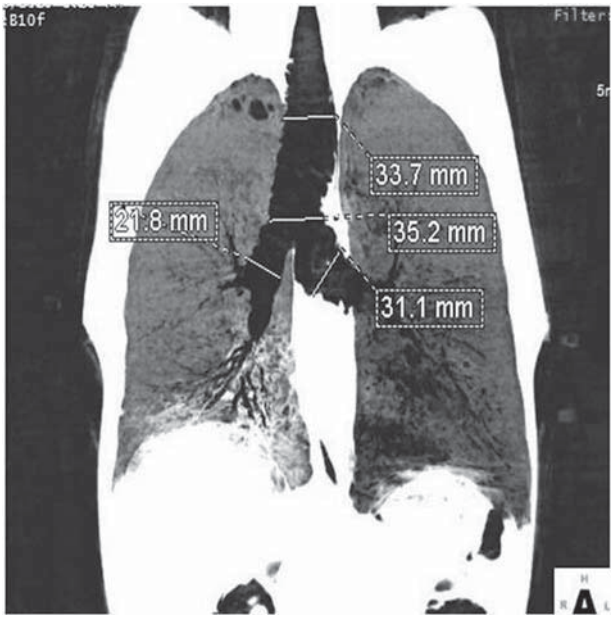
antineutrophil cytoplasmic antibodies were also negative making an autoimmune disease unlikely. Total Immunoglobulin E titres and titres of Immunoglobulin G against *Aspergillus fumigatus* were negative. Thus, with the more commonly encountered causes of secondary airway dilation ruled out, we focussed on congenital causes of airway dilation. This approach was also supported by the fact that the patient had been symptomatic since childhood and only the respiratory system was predominantly affected.

Discussion

Mounier-Kuhn syndrome is characterized by tracheobronchial dilation usually manifesting as recurrent LRTIs as the patient is unable to clear his airway secretions. Bronchoscopy shows enlarged trachea and main bronchi during inspiration, and constriction or collapse during expiration and coughing [2]. Biopsy of the trachea or bronchi may show thinning of the muscularis mucosa and atrophy of longitudinal and elastic fibres [3]. Diverticulae arise from protrusion of redundant nonmuscular segments in the transverse band of muscles between the tracheal cartilages.

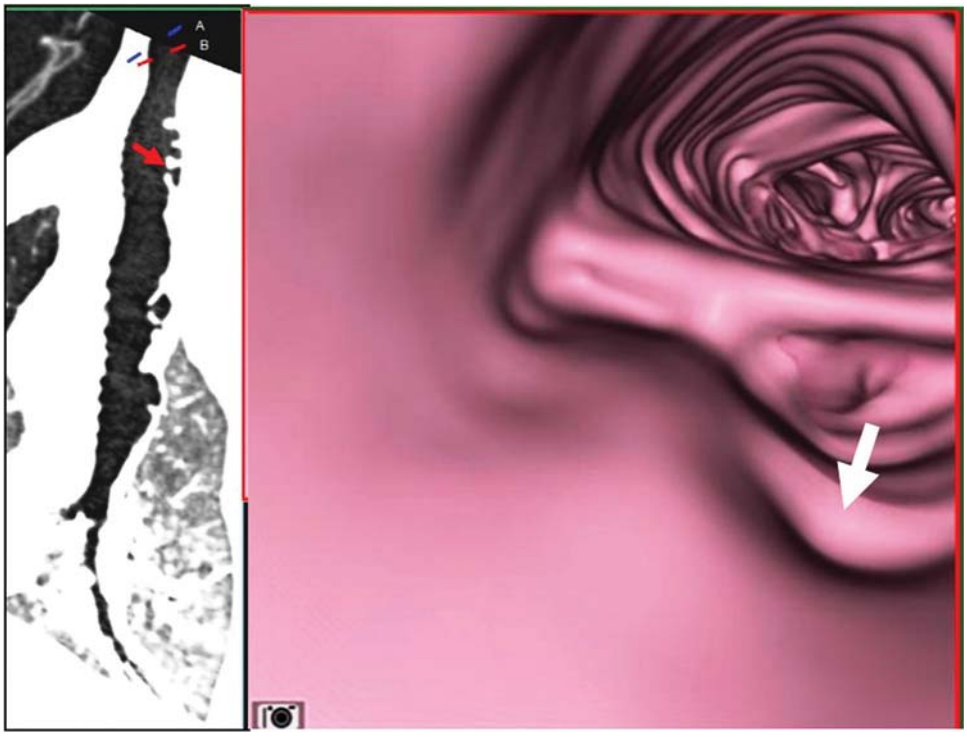
The disease manifests more often in the third and fourth decade of life and affects men more than women. The presentation varies from an incidental

Figure 1



Computerized tomography scan of chest showing the dilated airways with corresponding measurements (cited in text).

Figure 2



Virtual bronchoscopy of patient with tracheal scalloping (marked with red arrow) with corresponding tracheal diverticulae noted on the posterior tracheal wall (marked with white arrow).

finding in an asymptomatic patient with preserved lung function to severe respiratory failure.

The diagnosis is primarily based on clinical suspicion aided by radiological findings and bronchoscopy. The chest radiography reveals gross enlargement of the trachea, whereas computerized tomography may show dilated airways with diverticulae. The upper limit of the airway diameter (usually considered at 2 cm above the top of the aortic arch) is a transverse and sagittal tracheal diameter exceeding 25 and 27 mm, respectively, or the left and right mainstem bronchi exceeding 18 or 21 mm in diameter, respectively, in men. The respective figures for women are 21, 23, 17.4, and 19.8 mm, respectively [4] (Fig. 1). Additionally, features such as bronchiectasis or pulmonary fibrosis as a result of recurrent infection may be seen.

There are three known variants of Mounier-Kuhn disease [5]:

(1) Type 1: symmetrical enlargement of trachea and central bronchi.
(2) Type 2: eccentric dilatation of trachea with prominent diverticulae.
(3) Type 3: it is the rarest type with diverticulae extending to distal bronchi.

Differentials include Marfan syndrome, ataxia telangiectasia, Ehlers-Danlos syndrome, allergic bronchopulmonary aspergillosis and sarcoidosis, which cause secondary bronchial enlargement [6].

Asymptomatic patients require no specific treatment. Therapy is supportive, largely limited to respiratory physiotherapy for clearing secretions and to antibiotic use during infectious exacerbations [6]. Tracheal stenting, laser ablation and even lung transplantation have been attempted, without much benefit [6].

The etiology of Mounier-Kuhn syndrome remains unclear. It is usually attributed to congenital absence of smooth muscle or atrophy of elastic fibres and smooth muscle of airways, but alternative etiologies such as disappearance of connective tissue network of airway wall and enhanced matrix metalloproteinase activity owing to chronic inflammation have been proposed [7]. However, it has not been possible to verify the multiple hypothesized etiologies owing to the small number of cases, as well as the scarcity of documented biopsies.

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Conflicts of interest
There are no conflicts of interest.

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