

CASE REPORTS

Mounier-Kuhn syndrome in an older patient

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Abstract

Respiratory problems in older adults are frequently labelled as being due to chronic obstructive airways disease (COAD). However where the presentation may be suggestive of another pathology due to the clinical history or type of pathogen isolated from the sputum, then consideration should be given to appropriate imaging. We describe a case of an older lady labelled for many years as COAD but who was a lifelong non-smoker and had *Pseudomonas* in her sputum. A CT of thorax for suspected bronchiectasis revealed tracheobronchomegaly (Mounier-Kuhn syndrome). This is the oldest case of Mounier-Kuhn syndrome at presentation and highlights the importance of appropriate history taking and investigation of older people.

Keywords: *Mounier-Kuhn syndrome, oldest case, tracheobronchomegaly, elderly*

A 79-year-old lady presented with productive cough and dyspnoea. She reported a 6-month history of recurrent respiratory tract infections. She had recently been diagnosed with chronic obstructive airways disease. She had no previous respiratory history but reported a productive cough on most days over the past 20 years. She was a lifelong non-smoker with no occupational or environmental exposure to respiratory irritants.

She had been independent in her activities of daily life up until 6 months previously. On examination, she was cachectic but afebrile, and not clubbed. She had bibasal crepitations more marked on the right. Her routine bloods were normal with O₂ saturation at 92% on room air. No abnormality was noted in the chest x-ray. Her sputum grew a pan-sensitive *Pseudomonas* and methicillin-resistant *Staphylococcus aureus* (MRSA).

A CT scan of the thorax was performed because of the suspicion of bronchiectasis. It demonstrated marked dilatation of the intrathoracic trachea with sagittal diameter of at least 40 mm. There was extensive diverticulosis of the trachea. Both bronchi were dilated, with the left main bronchus having a maximum diameter of 23 mm, and the right, 20 mm. There was minimal bronchiectasis in the right upper lobe. The airway dimensions exceed the diagnostic criteria for tracheobronchomegaly [Mounier-Kuhn (MK) syndrome].

She was commenced on intravenous antibiotics and nebulised colomycin and referred for chest physiotherapy. Recovery was good and she was discharged home 2 weeks later.

Discussion

Tracheobronchomegaly is a rare disorder characterised by pathological dilation of the trachea and major bronchi. Its clinical, radiological and endoscopic features were first described in 1932 by Mounier-Kuhn [1]. Around 100 case reports can be found in the literature. As the clinical symptoms are similar to chronic bronchitis and bronchiectasis the true incidence of the disease may be higher. Many patients carry the clinical diagnosis of COAD.

The aetiology remains uncertain. Early autopsy studies suggested a congenital defect of the smooth muscle tissue of the trachea and main bronchi [2]. Although there is a known association with Ehlers-Danlos in adults and cutis laxa in children [3] the majority of patients do not have either of these diseases suggesting a further, as yet unknown, factor in the aetiology. Histology shows atrophy of cartilage and smooth muscle. The resultant laxity of connective tissue leads to tracheal diverticulosis [4]. In addition dilated airways lead to an inefficient cough mechanism with impairment of mucociliary clearance.

Resultant inflammation combined with enlarged weakened airways leads to a cycle of recurrent pneumonia and

Table 1. Diagnostic criteria for tracheobronchomegaly with relative airway diameters in millimetres [5]

	Trachea (transverse/ sagittal) (mm)	Left main bronchus (mm)	Right main bronchus (mm)
Male	25/27	18	21
Female	21/23	17.4	19.8

fibrosis. The primary disease process spares fourth and fifth order bronchial divisions, bronchiectasis is, therefore, not a core feature but results from repeated infections.

Given the non-specific clinical pattern, diagnosis is made radiologically. Chest x-ray may establish the diagnosis if tracheal dilatation is evident; however, CT is usually required for accurate airway measurement and to assess for complications, e.g. tracheal diverticulosis, scarring and bronchiectasis. The original guidelines for diagnosis of MK are based on CXR (see Table 1), however, with the advent of CT, diagnosis is now accepted on CT showing left main bronchus >20 mm, right >15.

Symptoms are non-specific with chronic sputum production and recurrent respiratory tract infections. Presentation is usually in the third to fourth decade with a male predominance [6]. While there are reported cases of patients aged 18 months to 79 years old, it would appear that most cases diagnosed in later life had a long misdiagnosis of COAD [7, 8]. The authors believe that this is the oldest initial presentation and diagnosis of tracheobronchomegaly.

Treatment is mainly supportive with no specific therapy for asymptomatic patients.

Physiotherapy and postural drainage assist in clearing secretions, and appropriate administration of antibiotics during infectious exacerbations is the mainstay of treatment [9]. There is no study to show the effectiveness of either inhaled bronchodilators or corticosteroids.

Tracheal stenting can be useful in advanced cases and there are reports of tracheobronchial endoprosthesis being used with some success [8, 10].

- The clinical symptoms are similar to chronic bronchitis and bronchiectasis. The diagnosis is made radiologically with CT as the gold standard, and treatment is mainly supportive

Conflicts of interest

None

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Key points

- Tracheobronchomegaly is a rare disorder although possibly under-diagnosed.