Mounier-Kuhn Syndrome: Case of Cough, Recurrent Chest Infections with Bronchiectasis

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ABSTRACT

Mounier Kuhn syndrome (MKS) is a rare airway disease characterized by dilated trachea and the main bronchi. Our case report is to report a 57-year-old male, ex-smoker with chronic cough and recurrent chest infections. CT thorax showed dilated trachea, main bronchi with central bronchiectasis and emphysema and forced spirometry reveals obstructive airway disease. The treatment is supportive, clinical presentation is nonspecific and the diagnosis is based on radiological findings.

INTRODUCTION

Mounier Kuhn syndrome (MKS) is a rare airway disease defined with dilated trachea and the main bronchi with ambiguous etiology and variable clinical presentation. Although there are no specific criteria for clinical diagnosis of MKS, it is based on radiological imaging findings, lung physiological studies and bronchoscopy[1,2].

Here we report a case with diagnosis of MKS to increase the knowledge of this rare disease and toemphasis the need for early diagnosis to provide the appropriate managemnt.

THE CASE

A 57-year-old gentleman who was evaluated due to recurrent moderate to severe chest infections requiring multiple courses of antibiotics and steroids with a necessitated need for medical admissions for more than four times per year for acute management. His respiratory symptoms started more than ten years which are progressive in severity. He has chronic productive cough, shortness of breath even at rest, persistent wheeze worse with exertion and episodes of intermittent hemoptysis with no other respiratory red flags. He does not report ankle swelling, orthopnea, or paroxysmal nocturnal dyspnea. He denied symptoms of connective tissue disorders and never treated for tuberculosis.

He is known for atrial fibrillation, congestive heart failure with EF 40%, obesity hypoventilation syndrome on nocturnal BIPAP and increased BMI. He is an ex- smoker with 40 pack year history and works as a lorry driver. He was conscripted and worked in the military for three years and worked in chemical factories and he is unknown about his chemical exposure at that time. He has no tuberculosis contact or history of use of drugs with pulmonary toxicity and no known allergy or pet exposures.

There was no evidence of finger clubbing, connective tissue signs, pulmonary hypertension signs. He was euvolemic. On auscultation, there was reduced air entry with expiratory rhonchi bilaterally. Heart sounds were normal.

Blood investigations are shown in table 1. A posterior-anterior chest radiograph showed evidence of trachemogaly (Figure 1). MKS is diagnosed based on radiological findings and it is defined by an increase in the transverse and sagittal diameter of the trachea beyond 25 mm and 27 mm, respectively, and or an increase in the diameter of the right and left main bronchi beyond 18 and 21 mm. Our patient was diagnosed with MKS based on radiological findings; computed tomography (CT) chest showed dilated trachea 3.7 cm in diameter above the level of the aortic arch, right main bronchus measures 3.5 cm in diameter and left main bronchus measures 3.9 cm in diameter, dilated lobar and segmental bronchi, central proximal bronchiectasis, and emphysema with multiple bullae and blebs. (Figure 2, 3). Spirometry revealed severe obstructive airway disease with negative bronchodilator response, normal gas transfer and normal lung volumes.

Table 1: Summarized blood investigations

Laboratory investigation	Result	Normal reference range
Full blood count & renal function	Within normal	
Albumin	42	(40-49)
Complement C 4	0.5	(0.14-0.54 g/L)
Complement C 3	1.6	(0.75-1.65 g/L)
Immunoglobulin G	11.4	(7-16 g/L)
Immunoglobulin A	3.17	(0.74 g/L)
Immunoglobulin M	0.57	(0.4- 2.3 g/L)
Alpha 1 antitrypsin	1.34	(0.9- 2 g/L)
Connective tissue screen	Negative	
ENA screen	Negative	
ANCA screening	Negative	
RF	Negative	
RAST for Grass	Positive	(Associated with seasonal allergic rhinitis- Methods fluroenzyme immunoassay)
RAST Aspergillus	0.14 (negative)	(0.00- 0.34 KuA/L Fluroenzyme immunoassays)
RAST Feathers	Negative	(Fluroenzyme immunoassays)
RAST trees	Negative	(Fluroenzyme immunoassays)
Budgerigar IgG	5.6	(0-7.9 mgA/l)
Pigeon Ig G	10.8	(0- 37.9 mgA/l)
Micropolyspora Feani Ig G	2.6	(0-9.9 mgA/l)
Ig G Anti Aspergillus	158↑	(0-39.9 mgA/L)
Total I g E	108 ↑	0-80 Ku/ L

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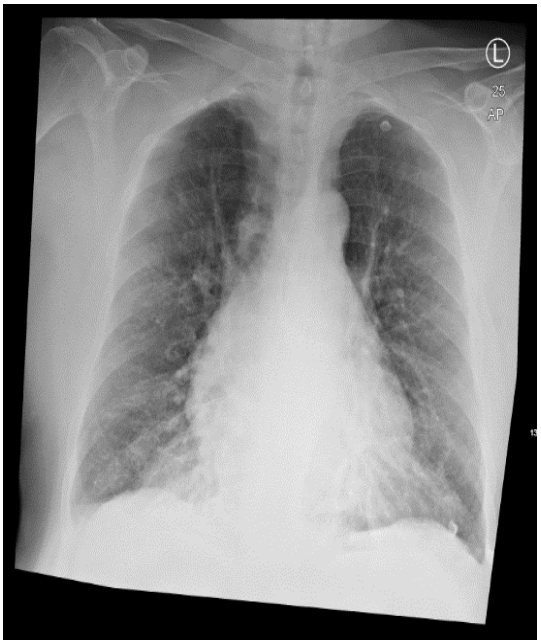

Figure 1: Dilated trachea

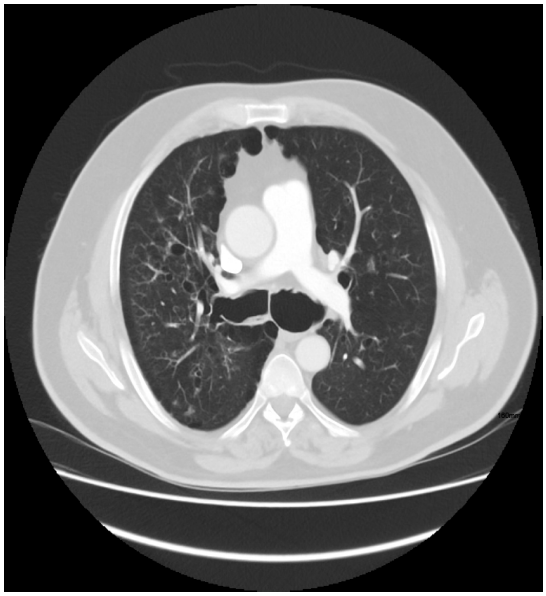

Figure 2: CT thorax Dilated right and left main bronchi

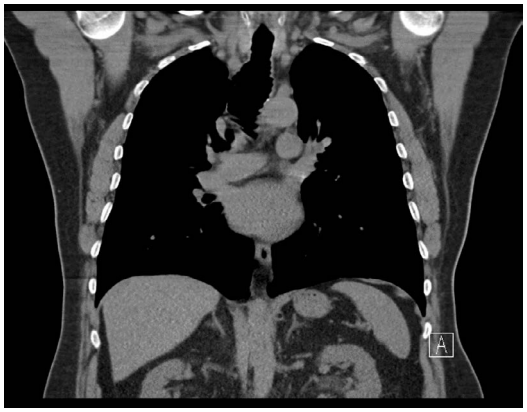

Figure 3: CT thorax: Dilated trachea

The clinical history of recurrent chest infections with radiological findings in CT thorax is consistent with tracheobronchomegaly (Mounier Kuhn syndrome, MKS). MKS management is supportive, and this includes treating and preventing infections. He was managed with inhaled corticosteroid/long-acting beta agonist, inhaled tiotropium, azithromycin 250 mg on Monday, Wednesday, and Friday, mucolytics and airway clearance device. He was referred for chest physiotherapy and pulmonary rehabilitation and advised to be up to date with pneumococcal, flu and COVID 19 vaccination. He was also referred for dietician to help to reduce his weight as his BMI 34 kg/ m^2. He was on BIPAP for obesity hypoventilation syndrome and Bi – level positive pressure and this would help to clear secretions from airways, and he was advised to continue follow up on non-invasive ventilation clinic. He has regular followed up in respiratory clinic.

DISCUSSION

MKS is characterized by dilation of the trachea and main bronchi during inspiration and narrowing and collapsibility of the airways during expiration and this is due loss or atrophy of the elastic fibers and smooth muscles tissues of the trachea and main bronchi[5]. The consequence of this change in the dynamic airway physiology is accumulation of secretions and outpouching of redundant musculomembranous tissue between the cartilaginous rings predisposing to the development of chronic pulmonary suppuration, diverticula formation, bronchiectasis, emphysema, and pulmonary fibrosis[4].

The diagnosis of MKS in our patient was reached through multidisciplinary meeting and was based on radiological imaging findings which showed dilated trachea and main bronchi with emphysema and bronchiectasis as complications of the disease.

The disease tends to be more in males than females with 8:1 ratio. The etiology of MKS is unknown but thought to be inherited with no identified gene yet and smoking and pollutants could be predisposing factors due to chronic inflammation[2,3]. Our patient was an ex-smoker with a negative family history of MKS. He was diagnosed at the age of 42 after a ten-year history of respiratory symptoms and this is attributed to unspecified clinical presentation.

MKS is associated with connective tissue diseases such as rheumatoid arthritis and polyarteritis nodosa, Marfan's syndrome and Ehlers-Danlos syndrome[4]. He has no clinical features suggestive of connective tissue diseases and connective tissue screening, ENA and ANCA screening were negative. Genetic tests were not done to exclude inherited disorders.

Treatment of MS is supportive consisting of chest physiotherapy for airway clearance techniques and to treat chest infections. Cessation of smoking, vaccination, minimizing exposure to industrial and occupational irritants and pollutants is likely to be advantageous[5]. In those with tracheomalacia as complication of MKS, tracheal stenting, surgical tracheoplasty, laser treatment and long-term positive airway pressure have all been tried. Double lung transplant has been performed in end stage disease[4].

CONCLUSION

MKS is a rare disease and probably underdiagnosed as the clinical presentations are non-specific and variable. The diagnosis is based on radiological imaging and no specific criteria for diagnosis yet and the treatment is mainly supportive.

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