Case Report

Mounier-Kuhn syndrome masquerading pulmonary thromboembolism in an elderly male

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ABSTRACT

Mounier-Kuhn syndrome, also referred to as tracheobronchomegaly, is a rare idiopathic clinical and radiologic disorder characterized by significant tracheobronchial dilation. It results in recurrent lower respiratory tract infections and bronchiectasis. In severe cases, patients may present with acute respiratory distress requiring hospital admission and ventilatory support. Clinical examination and chest radiography may be misleading in these patients as tracheobronchomegaly is easily overlooked on radiographs. We present an interesting report of our patient who presented with acute shortness of breath. A diagnosis of acute pulmonary thromboembolism was suggested by initial evaluation. Computed tomography pulmonary angiography was negative for thromboembolism; however, it revealed diagnosis of this rare disorder.

KEY WORDS: Computed tomography, high resolution computed tomography, Mounier-Kuhn syndrome, tracheobronchomegaly

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INTRODUCTION

Mounier-Kuhn syndrome (MKS), variously named as tracheobronchomegaly, tracheobronchiectasis, tracheocele, tracheal diverticulosis, tracheomalacia and tracheobronchopathia malacia is a rare well-defined clinical and radiologic condition.[1] The pathologic hallmark is distinct dilation of the trachea and bronchi. Associated findings include tracheal and bronchial diverticula.[2] Clinically, it presents as recurrent lower respiratory tract infections and a course marked by frequent exacerbations and remissions, though extremes of presentations are also reported with asymptomatic patients and those with severe respiratory compromise as in our patient.[3] Though the condition remains largely idiopathic, autopsy studies have implicated congenital atrophy of smooth muscle and elastic tissue of the trachea and main bronchi as one of the causative mechanisms.[4] Diagnosis is established by bronchoscopy and non-invasively by computed tomography (CT). There are established criteria for tracheal and bronchial dimensions for diagnosis of tracheobronchomegaly.[5,6] In the present report, we highlight an uncommon presentation of this syndrome and provide a brief review of the condition.

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CASE REPORT

A 70-year-old male presented with shortness of breath for 3 days. There was associated mild non-productive cough. Patient had a past history of episodes of cough with expectoration and fever on several occasion. However, the patient had never suffered such a severe respiratory embarrassment in the past. On examination, there was tachycardia (heart rate of 100 bpm), tachypnea (respiratory rate was 25/min) with SpO_2 85% and FiO_2 50%. Patient was afebrile and blood pressure was within normal range (measured on at least three occasions). On general physical examination, the patient had pallor. However, there was no cyanosis, clubbing or pedal edema. Cardiorespiratory examination was significant for bilateral crepitations and occasional bronchial breath sounds. The heart sounds were normal. Hemogram revealed mild anemia (hemoglobin

11.5 g%). However, the total and differential leukocyte counts as well as platelet counts were normal. Routine biochemical tests including renal and liver function tests were normal. Chest radiograph (CXR) revealed patchy areas of bronchiectasis in both lungs. In addition, there were fibrotic opacities, more on the right side [Figure 1]. Echocardiography revealed dilatation of the right atrium (RA) and ventricle (RV). Associated findings included decreased contractility of RV and moderate tricuspid regurgitation. A diagnosis of acute pulmonary thromboembolism (PTE) was suggested. Doppler ultrasound evaluation of bilateral lower limbs was negative for deep venous thrombosis. Well's pre-test probability score was 4.5 (pulmonary embolism more likely than alternate diagnosis and heart rate more than 100). CT pulmonary angiography (CTPA) was subsequently performed to establish the diagnosis. CTPA excluded the diagnosis of acute PTE; however, high resolution CT reconstructions revealed massive enlargement of trachea (maximum diameter of 33 mm) and right main bronchus (maximum diameter of 25 mm) with evidence of tracheal and bronchial diverticula. There was extensive bronchiectasis involving the right upper lobe with peripheral fibrotic opacities suggestive of parenchymal scarring [Figures 2 and 3]. The main pulmonary artery was dilated. Further, the echocardiography findings of dilatation of RA and RV were confirmed. The pulmonary artery hypertension (PAH) was explained to be due to extensive parenchymal disease. Retrospective evaluation of a CXR done 1 year back revealed tracheabronchomegaly. The lung fields were unremarkable. Thus, a diagnosis of tracheabronchomegaly with bronchiectasis and PAH was made and patient was put on oxygen therapy and chest physiotherapy.

DISCUSSION

MKS is a rare condition characterized by tracheobronchial dilation. It is presumed to be due to atrophy of the musculo-elastic tissues in the trachea and bronchial walls with involvement occurring at different levels, from the trachea down to the 4th order branches. Primarily a disease of middle aged males, it has varied clinical presentation ranging from asymptomatic disease with well-preserved lung function to severe respiratory failure and death. However, majority of the patients have a disease course punctuated by several exacerbations and remissions due to recurrent infections and bronchiectasis.[3] The exact etiology is unclear, though tracheobronchomegaly may be due to familial susceptibility. Although most reported cases are sporadic, autosomal recessive inheritance has been postulated by some authors.[7] Regardless of the cause; tracheobronchomegaly leads to ineffective cough and impairment of mucociliary function. This leads to accumulation of secretions and recurrent lower respiratory tract infection. Three subtypes have been described.[4] Type 1 MKS is characterised by slight symmetric dilation in the trachea and main bronchi. Dilation and diverticula is a distinct feature of Type 2 disease. Type 3 MKS represents the severest sub-type with diverticulae and saccular structures extending to the distal bronchi.

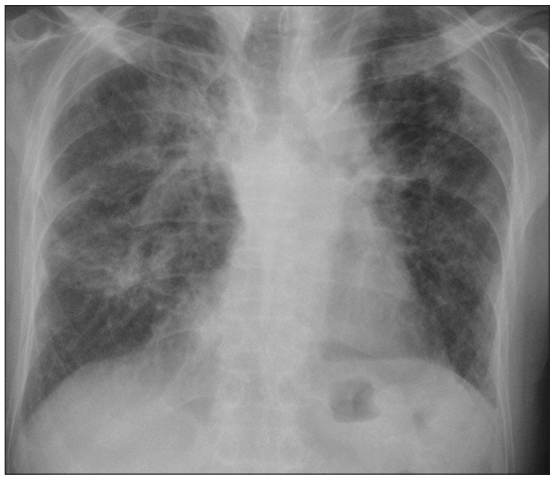

Figure 1: Frontal chest radiograph shows patchy areas of bronchiectasis in both lungs with fibrotic opacities, predominantly affecting the right lung. Also, note the markedly dilated trachea and right main bronchus

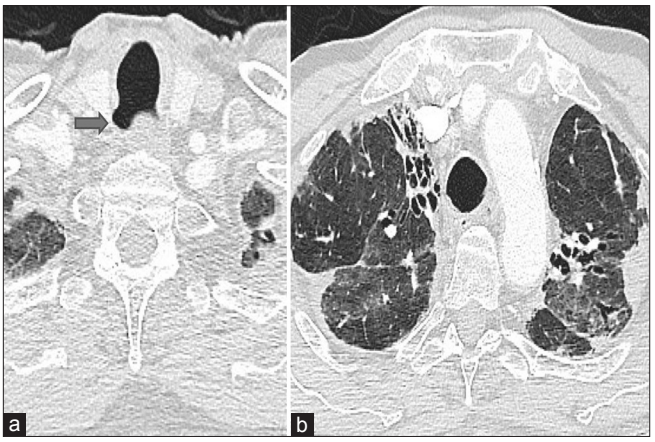

Figure 2: Axial high resolution computed tomography reconstructions image at the level of the trachea (a) demonstrates tracheomegaly with sacculation (arrow) from the posterior aspect. Another axial image at a lower level, (b) demonstrates bilateral upper lobe bronchiectasis, mosaic attenuation and peripheral scarring

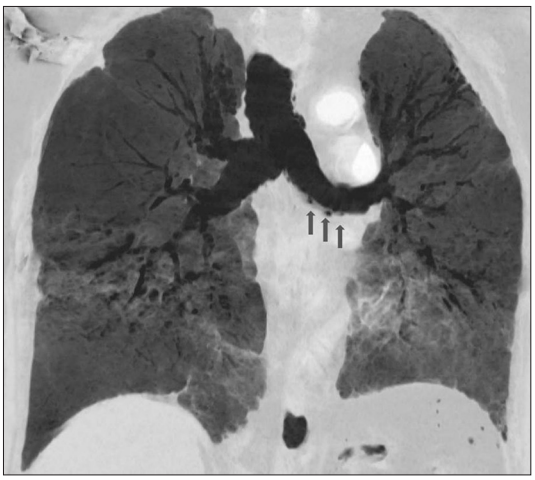

Figure 3: Thick coronal minimum intensity projection (minIP) demonstrates tracheobronchomegaly with tracheal and bronchial diverticulosis and bronchiectasis

Imaging features are diagnostic in MKS. CT allows direct measurement of the tracheobronchial tree at multiple levels as well as demonstrates secondary features including diverticuli, sacculation, bronchiectasis and parenchymal scarring. Tracheal diverticula develop most commonly in the posterior membranous trachea. In adults, the accepted criteria for diagnosis of tracheobronchomegaly on CT are diameters of the trachea, right main bronchus and left main bronchus of >30 mm, 20 mm and 18 mm respectively.[5] Though CXR can also demonstrate tracheobronchomegaly, this finding is rarely appreciated prospectively. Bronchography is excellent in demonstrating the extent of disease, but is obsolete now. The criteria on CXR and bronchography include tracheal, right bronchial and left bronchial diameters of 30 mm, 24 mm and 23 mm respectively.[6]

Rigid or flexible bronchoscopy demonstrates the pathologic dilatation of the trachea and main bronchi during inspiration and constriction during expiration and coughing. It is useful in confirming the diagnosis when CT findings are equivocal; however, it is invasive. Ancillary investigations to support the diagnosis include pulmonary function testing (PFTs). PFTs demonstrate a reduction in bronchial flow speed, increased tidal volume and dead spaces. Differential diagnoses include connective-tissue disorders, ankylosing spondylitis, Ehlers-Danlos syndrome, Marfan syndrome, cutis laxa (elastolysis) etc., conditions associated with secondary tracheobronchial enlargement.[8] These patients carry the stigmata of underlying diseases and have a relatively young age at diagnosis.

Treatment is guided by disease severity. Basic aim in all patients is to prevent further damage to the airway tract. This is achieved by cessation of smoking, minimizing exposure to the industrial and occupational irritants and pollutants. Another important approach in these patients is optimization of concomitant cardiopulmonary conditions such as asthma, chronic obstructive pulmonary disease and chronic bronchitis. Asymptomatic or patients with mild symptoms require no specific treatment. In symptomatic patients, chest physiotherapy allows clearance of secretions. Antibiotics are employed during exacerbations to control infections. Tracheal stenting is helpful in severe cases; surgery has virtually no role, considering the diffuse nature of the disease.[9]

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