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Case Report

Mounier-Kuhn syndrome: a rare entity of bronchiectasis

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ABSTRACT

Mounier-Kuhn syndrome (MKS) is a rare clinical and radiologic entity characterized by pathologic dilatation of the trachea and bronchi. The etiology of this disorder is uncertain and the clinical presentation is variable. The diagnosis is usually made on the basis of the characteristic CT scan findings. We report one such case in a 44-year-old man presenting with recurrent lower respiratory tract infections.

Keywords: Mounier-Kuhn Syndrome, Bronchiectasis, Recurrent pulmonary infections

INTRODUCTION

Mounier-Kuhn syndrome (MKS) is a rare clinical and radiologic entity characterized by pathologic dilatation of the trachea and bronchi.[1,2] It is a form of expiratory central airway collapse characterized by the softening of the airway wall cartilaginous structures and is usually associated with bronchiectasis and lower respiratory tract infection. We present one such rare case in middle aged man.

CASE REPORT

A forty-four years old man presented with fever, dyspnea and productive cough. He had a history of recurrent hospitalization for similar complaints since four years. He was treated symptomatically with antibiotics every time. There was no history of allergy, asthma or tuberculosis. He was neither a smoker nor alcoholic. Mild clubbing of the finger was observed. On auscultation, bilateral coarse crepitations were heard over the mammary and infrascapular areas. Heart sounds were normal with tachycardia.

His routine laboratory investigations were shown in the Table 1.

Table 1: Routine laboratory workup

Hemoglobin	16.2 gm%	ESR	40/ I hour
Total WBC count	17,800 / µL	Urea	28 mg/dl
Neutrophils	90%	Creatinine	0.9 mg/dl
Lymphocytes	08%	Random glucose	110 mg/dl
Monocytes	02%	Montoux test	Negative
Eosinophils	00%	Sputum for AFB	Negative
Platelet count	2.2 lakhs/cu.mm		

Radiograph of the chest showed enlargement of trachea and bronchi and bilateral bronchiectasis. Retrospective evaluation of an earlier chest radiographs also revealed bronchiectasis. CT scan of the chest revealed increased diameters of trachea and right and left main bronchi

(Figure 2). The lung fields were unremarkable. The antero-posterior and transverse diameters of the trachea were 3.1 cm and 3.3 cm respectively. At the level of carina the right and left main bronchi were 2.6 and 2.4 cm in diameter respectivelyThus, a diagnosis of MKS or tracheabronchomegaly with bronchiectasis was made and patient was treated symptomatically with antibiotics and chest physiotherapy.

MKS is a rare clinical and radiological entity described by Mounier and Kuhn for the first time in 1932.[1] It is known by a number of different names like trachiectasis, tracheobronchopathia malacia, tracheomegaly and multiple tracheal diverticula.[2] Most cases present in the third or later decades with recurrent respiratory tract infections as was in our case and there is male predominance.

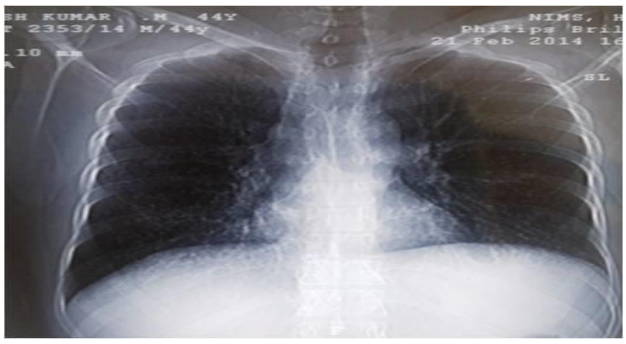

Figure 1: Radiograph of the chest showing enlargement of trachea and bronchi and bilateral bronchiectasis.

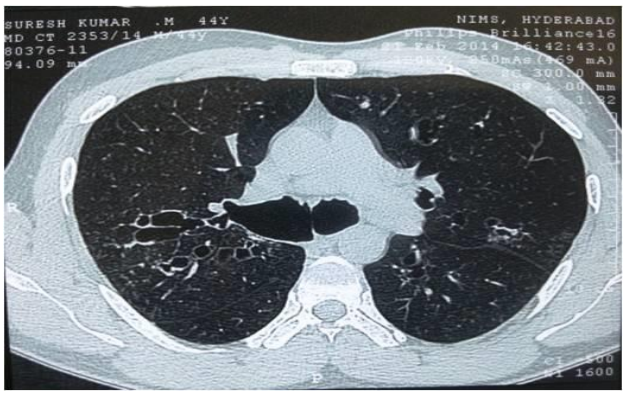

Figure 2: CT scan of the chest showing increased diameters of t right and left main bronchi with bilateral brochiectasis.

Although the etiology is uncertain, it is believed to be due to the lack of smooth muscles and elastic connective tissue in the trachea and main bronchi, leading to sacculations and formation of diverticulae between the cartilaginous rings.[3]

On CT scan, the diagnosis is made when the transverse diameter of the trachea measures greater than 3 cm and that of the right and left main bronchi exceeds 2.4 cm and 2.3 cm, respectively.[3,4] In our case also, the diagnosis was made as per the CT scan findings.

Treatment is symptomatic, and consists of respiratory physiotherapy in order to manage secretions, and antibiotics, bronchodilators, and corticosteroid treatment during exacerbations.[5]

The case is presented because of its rarity. In conclusion, MKS should be considered in patients who presented with bronchiectasis associated with abnormal dilation of trachea and major bronchi on CT scans.

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