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IMAGES IN OTORHINOLARYNGOLOGY

Mounier–Kuhn syndrome☆

Síndrome de Mounier-Kuhn

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Mounier–Kuhn syndrome or tracheobronchomegaly is a rare clinical and radiological entity characterized by marked dilatation of the trachea and bronchi and recurrent lower respiratory tract infections.

Here we report a case of Mounier–Kuhn syndrome presenting with frequent respiratory infection.

A 56-year-old man came with productive cough and progressive breathlessness since 2 months. He had history of recurrent lower respiratory infections in the past. He had smoked 4 cigarettes per day and stopped smoking 3 months back. Patient was afebrile. He was tachypneic and tachycardic. Examination of Chest revealed the presence of bilateral rales, more on basal regions. The chest radiograph showed ill-defined shadows bilaterally on lower zones. A computed tomography (CT) scan of thorax was performed. The CT scannogram showed enlargement of the trachea and multiple cystic lesions of varying size throughout lung fields (Fig. 1). Axial CT images demonstrated marked dilatation of the intrathoracic trachea, bilaterally dilated bronchi with multiple diverticula seen between the cartilaginous rings in the trachea and right and left main bronchi and bilateral multiple cystic lesions throughout the lung fields (Figs. 2 and 3). These CT findings are characteristic of Mounier–Kuhn syndrome or tracheobronchomegaly.

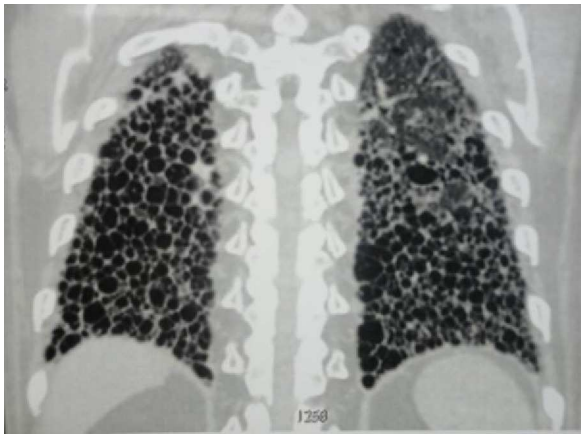

Figure 1 CT scannogram shows enlargement of the trachnea and bilateral cystic bronchiectasis.

Mounier–Kuhn syndrome is characterized by a significant dilation of the trachea and main bronchi due to atrophy or absence of the elastic fibres or smooth muscle that leads to laxity to the walls of the airway and the formation of diverticula and bronchiectasis. The syndrome was

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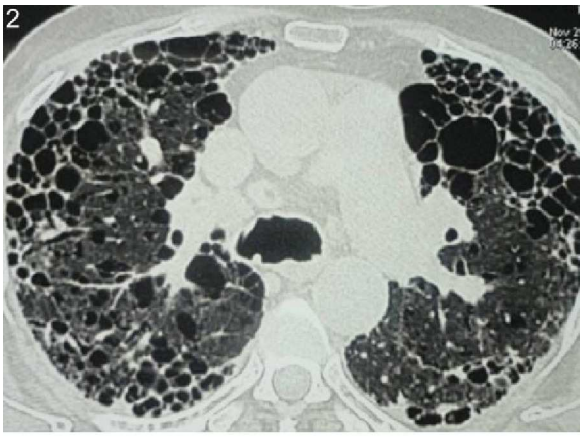

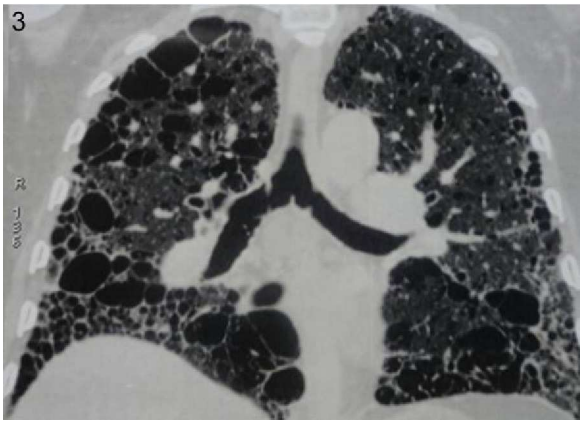

Figures 2 and 3 Axial CT show marked dilatation of the trachea, left and right bronchi with multiple diverticula (arrow heads), and bilateral cystic bronchiectasis.

first described by Mounier–Kuhn in 1932. It is predominantly seen in males between the ages of 25 and 50 years. The clinical presentation varies from recurrent respiratory infections, pneumonia, to severe respiratory failure and death. Ineffective cough secondary to pathologic dilation in the tracheobronchial tree and the impaired mucociliary activity lead to recurrent lower respiratory tract infections. Secondary causes of tracheobronchial enlargement are Marfan syndrome, Ehlers–Danlos syndrome, ataxia-telangiectasia, connective tissue diseases, ankylosing spondylitis, cutis laxa, and light chain deposition diseases. The diagnosis can usually be made by Computed tomography of the chest when the transverse diameter of the trachea measures greater than 30 mm and that of the right and left main bronchi exceeds 20 mm and 18 mm, respectively. Physiotherapy and appropriate administration of antibiotics during infectious exacerbations is the mainstay of treatment.