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A case of Mounier-Kuhn syndrome in a geriatric patient

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Description

Mounier-Kuhn syndrome, or tracheobronchomegaly, is a rare clinical and radiologic condition characterized by marked tracheobronchial dilation and recurrent lower respiratory tract infections. The syndrome was first described by Mounier-Kuhn in 1932, and few cases have been reported in the medical literature [1]. The clinical presentation varies widely, from asymptomatic disease with preserved lung functions to recurrent chest infections, exertional dyspnoea to even respiratory failure in severe cases. Diagnosis is often made by using CT, through which abnormally large air passages are detected. In adults, the diagnostic criteria are diameters of the trachea >30 mm, right main bronchus >20 mm and of the left main bronchus >18 mm [2-4]. Upon pulmonary function testing, decreased bronchial flow speed, increased tidal volume, and dead spaces may be observed. Bronchoscopy can detect the pathologic processes that affect the tracheobronchial structures. Connective-tissue diseases, ataxia-telangiectasia, ankylosing spondylitis, Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome, and cutis laxa (elastolysis) are also associated with secondary tracheobronchial enlargement [2-5]. All these conditions should be considered in the differential diagnosis.

We describe the case of a 83 years old man admitted to our Hospital to undergo rehabilitation treatment after pertrochanteric proximal femoral fracture. In remote anamnesis spontaneous pneumothorax one year earlier with a diagnosis of idiopathic interstitial fibrosis, gastric ulcer 40 years earlier, conservatively treated. The patient was a no-smoker and worked as a professor. After three days, the patient developed severe respiratory insufficiency and a thorax TC scan was performed. It revealed the presence of a dilated trachea (34 mm A-P diameter, 27 mm L-L diameter) with enlarged principal bronchi (25 mm right and 23 left) in association with multiple bronchiectasis, emphysema with large bullae, interstitial fibrosis and right basal pulmonary consolidation. A clinic-radiological diagnosis of tracheobronchomegaly or Mounier-Kuhn syndrome wasmade and the patient was transferred to the emergency department because of severe respiratory distress.

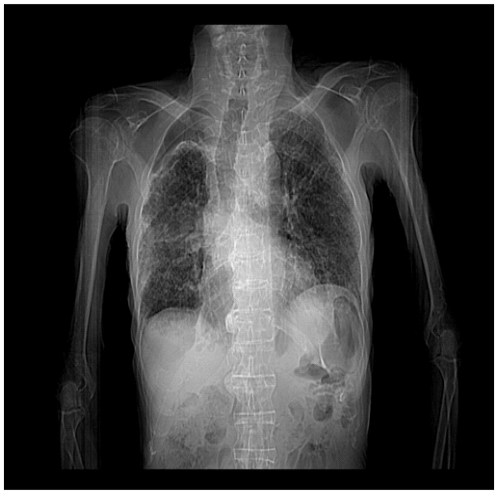

Figure 1

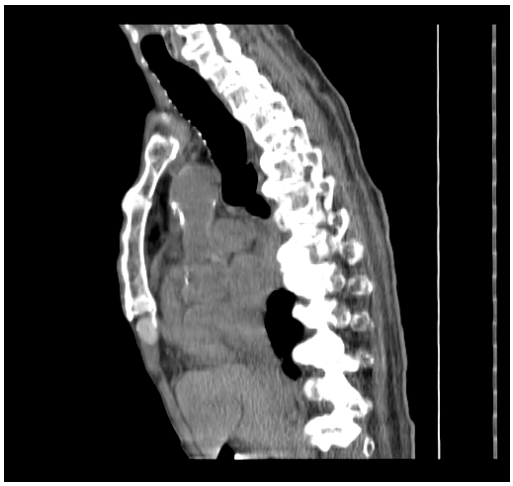

Figure 3

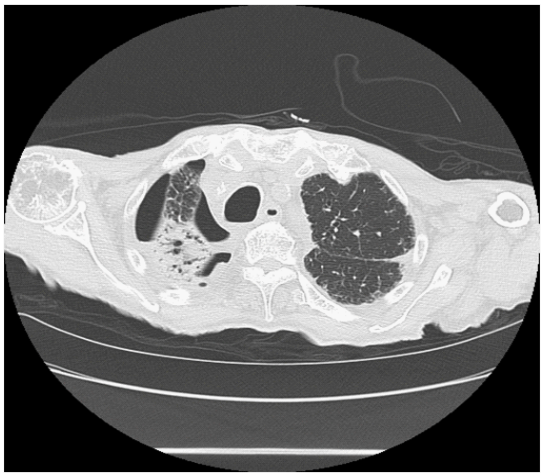

Figure 2

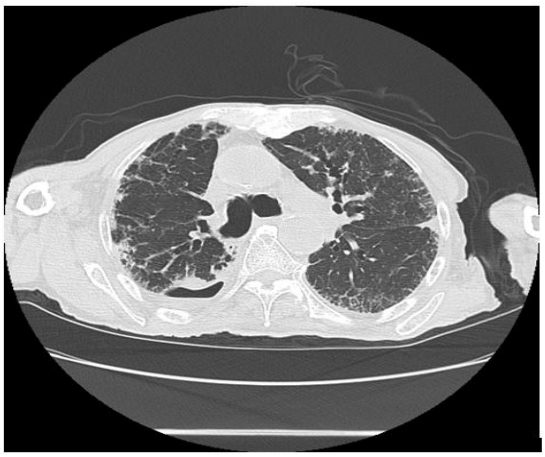

Figure 4

Figure 1-4: Large bubble of emphysema in the apical segment of the right upper lung lobe, gross pulmonary peripheral interstitial disease, multiple bronchiectasis, tracheobronchial dilatation (arrow).

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