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Case Report

Mounier-Kuhn Syndrome – a rare cause of bronchiectasis in a Sri Lankan patient

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Introduction

Mounier-Kuhn syndrome (MKS) was first described by Mounier-Kuhn in 1932 [1]. It is characterized by the presence of a dilated trachea and proximal airways resulting in recurrent respiratory tract infections and associated complication. A patient with this syndrome has not been reported in Sri Lanka. We present an elderly male patient with chronic shortness of breath due this rare syndrome, complicated by bronchiectasis.

Case report

A 67-year-old male patient was evaluated for chronic shortness of breath, sputum production and wheezing for more than 20 years. He was a retired businessman with a past history of 15 pack years of smoking. He had no past history of tuberculosis. He denied any other environmental or occupational exposures or a family history of chronic lung disease. Physical examination showed generalized wasting, grade 2 clubbing, bilateral basal coarse crepitations and wheezing.

His basic blood investigations were within normal limits. His pulmonary functions revealed FVC – 2.88L (76%), FEV1- 1.96L (72%) and FEV1/FVC – 68%. Chest radiograph showed bilateral bronchiectatic changes involving the bases. A high-resolution CT of the chest was ordered for further evaluation. It showed gross dilatation of the trachea (37mm) with dilated main bronchi (Right – 18mm, Left 20mm). Additionally, there were features of bilateral bronchiectasis (Figure 01).

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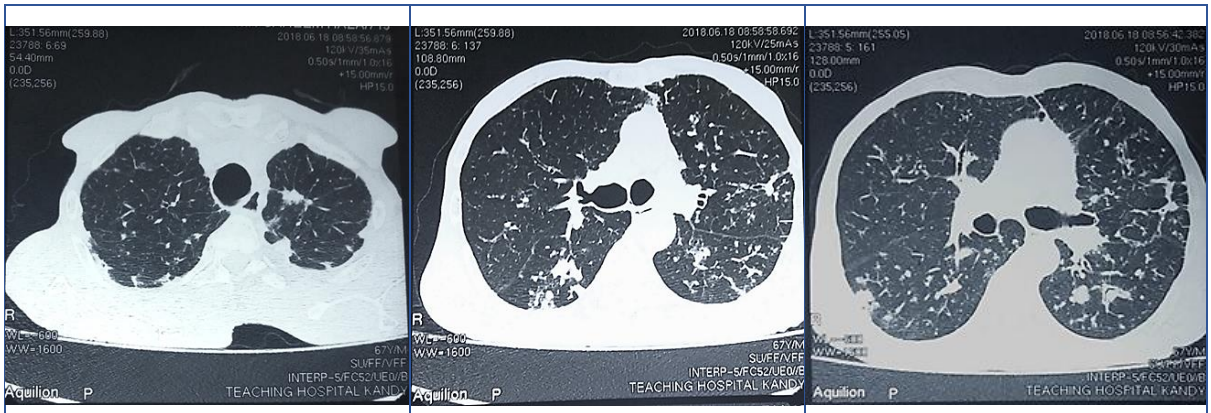

A – Dilated trachea | B – Dilated right main bronchus | C – Dilated left main bronchus

Figure 01

A diagnosis of MKS with chronic obstructive airway disease and secondary bronchiectasis was made. He was treated with inhaled bronchodilators for optimization of chronic obstructive airway disease and referred for chest physiotherapy. He was offered pneumococcal and influenza vaccinations. During the next six months of follow up he had one infective exacerbation, but otherwise remained stable.

Discussion

MKS is also known as tracheobronchomegaly. Main pathology in the syndrome is tracheobronchial dilatation secondary to atrophy of the muscular and elastic tissue of the large airways including trachea and main bronchi [2]. The aetiology is not well understood. In some adult patients it is associated with Ehlers Danlos syndrome and in children with cutis laxa [3]. It is mostly seen in African and American countries and commoner among men. Usual presentation of the disease is frequent lower respiratory tract infections or features of bronchiectasis. Patients can present at any age but commonly they present around the 3rd and 4th decades of life. Cases of childhood MKS have been reported. Hubbard *et al.* have reported the youngest case of MKS in a 15 month child [4].

There are three subtypes of MKS. Type 1 has only dilatation of the airways. Type 2 and 3 have diverticula in addition to the dilatation. Formation of diverticula is a result of the lack of tracheal wall support and they more commonly form in the posterior parts of airways [5]. In type 2 MKS the diverticula are shorter and in type 3 they extend up to distal bronchi [6].

The diameter of the normal trachea is between 15-22mm [7]. Accepted CT criteria to diagnose MKS include a tracheal diameter more than 30mm, right main bronchus diameter more than 20mm and left main bronchus diameter of 18mm [8]. In our patient the maximum tracheal diameter was 37mm and the left main bronchus diameter was 20mm consistent with the diagnosis of MKS. However, the diameter of the right main bronchus was 18mm which was slightly lower. There were no visible diverticula in the CT which favoured the diagnosis of type 1 MKS. Bronchoscopy would have been more informative but was not performed as it is invasive and offered no additional benefit to

the patient. Our patient also had CT features of bilateral bronchiectasis, which is a known complication of undetected long standing MKS [3,8].

In most patients with MKS, pulmonary function tests reveal an obstructive pattern which was seen in our patient as well. Ghanei *et al*. have described a patient with MKS with normal pulmonary function tests [3]. Therefore, the obstructive pattern may not be specific for the MKS and could be a manifestation of underlying COPD or bronchiectasis.

There is no specific treatment for MKS. Chest physiotherapy, control of infections and optimization of other comorbidities including asthma and COPD can be helpful to prevent further damage to the airways. One of the major problems associated with the disease is the inability to expectorate due to collapse of the airways. In such severe cases tracheal stenting and tracheobronchoplasty may be helpful [9]. Tracheobronchoplasty is a novel treatment option where the redundant posterior wall of the trachea is strengthened with a mesh via a thoracotomy. Odell DD *et al*. have described a series of patients with MKS subjected to stenting and tracheobronchoplasty [10]. They recommend initial stenting of patients for a trial period of several weeks followed by the offer of surgical intervention to those who show significant improvement. In their series of patients, 9 patients who were subjected to tracheobronchoplasty had significant clinical improvement with acceptable rates of complications. Our patient was managed with chest physiotherapy and inhalers for COPD. So far, he remains stable and is able to perform his normal day to day activities with minimal symptoms. Therefore, we did not consider further surgical intervention.

Knowledge of MKS is important to both physicians and radiologists as the diagnosis is easily overlooked. Early detection of the disease, optimization of management and surgical intervention in selected patients can result in reduced morbidity and mortality.

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