

## CASE REPORT

### MOUNIER KUHN SYNDROME: A RARE CAUSE OF BRONCHIECTASIS

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#### HOW TO CITE THIS ARTICLE:

Sachin Khanduri, Deepak Agrawal, Bhumika Singh, GarimaVarshney, Fahimul Huda. "Mounier Kuhn Syndrome: A Rare Cause of Bronchiectasis". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 03 January 08; Page: 485-488, DOI:10.14260/jemds/2015/71

**ABSTRACT:** Mounier Kuhn syndrome, also known as tracheobronchomegaly is a rare idiopathic 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. The diagnosis is easily missed as tracheobronchomegaly is overlooked on plain chest x-ray. Here we present an interesting report of our patient who presented with recurrent cough and shortness of breath. A diagnosis of bronchiectasis was first thought of. Computed tomography scan revealed a final diagnosis of this rare disorder. Management of the disease is only supportive and depends on the severity of the condition.

**KEYWORDS:** Mounier Kuhn, Tracheobronchomegaly, Bronchiectasis, Computed tomography.

**INTRODUCTION:** Mounier Kuhn syndrome or tracheobronchiomegaly also known as tracheocele, tracheobronchiectasis and tracheomalacia is a very rare entity presenting as recurrent lower respiratory tract infection.<sup>(1)</sup> Due to this nonspecific presentation it is a possible differential for various diseases. The focal findings of the condition being dilatation of trachea along with mainstem bronchi, down till the 4<sup>th</sup> bronchi level. Bronchiectasis is the usual diagnosis in these cases. It is diagnosed on x-ray, bronchoscopy but decisively on computed tomography scan.

**CASE REPORT:** A 28 year old male came to our outpatient department with cough, fever, dyspnoea and chest pain since one month. There had been episodic exacerbations of these complaints since three years. There were generalized constitutional symptoms of fatigue, malaise and weight loss. The patient had no family history of chronic obstructive pulmonary diseases. He had always been a non-smoker and worked as manual labourer.

On examination, there was tachycardia, tachypnoea and reduced oxygen saturation (SpO<sub>2</sub> 84%). Physical examination revealed pallor. Antero-posterior diameter of his chest had increased. Auscultation disclosed inspiratory crepitations in both lower lung fields. There was no cyanosis or clubbing. His blood work showed mild anemia (Hb- 11.1 gm %) and leucocytosis (TLC-12,500 mm<sup>3</sup>), RBC-5.9 million cells/mm<sup>3</sup>. MCV, MCH and MCHC were within normal limits. Renal and liver function tests were also within normal range.

On radiological examination, chest x-ray showed prominent trachea with emphysematous lung fields. The hilum and pleural spaces were normal. Areas of honeycomb shadowing in bilateral lower zones were also noted. Patchy areas of fibrotic opacities were noted on the left lower zones. Computed tomography scans of the chest (Plain) revealed dilated trachea measuring approximately 3 cm with dilated mainstem bronchi (Figure 1). Multiple sacculations were noted in the proximal part of thoracic part of trachea involving posterolateral walls (Figure 2& 3). Bilateral lung fields were hyperinflated with retrosternal and prevertebral herniations of lung fields with areas of air trapping in both lung fields with centiacinar emphysematous changes with few paraseptal changes. Patchy

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areas of fibronodular changes are noted in left lower lobe with tractional and tubular bronchiectasis in bilateral lung fields (Figure 4).

Spirometry showed no significant ambivalent findings. His sputum was negative for AFB. On the basis of above findings, patient was diagnosed as Mounier Kuhn syndrome with associated emphysema with bronchiectasis. He discharged from the hospital after 7 days of oral levofloxacin therapy.

**DISCUSSION:** P. Mounier Kuhn was the first to introduce this syndrome in 1932, later broadly understood as tracheobronchiomegaly. The focal findings of the condition being dilatation of trachea along with mainstream bronchi, down till the 4<sup>th</sup> bronchi level. Men in their 3<sup>rd</sup> - 4<sup>th</sup> decade of life, presenting with recurrent lower respiratory tract infection is the usual presentation. However, this disease has a wide range of presentation from mild cough to chronic bronchitis, bronchiectasis to respiratory failure followed by the eventual demise.<sup>(2,3)</sup> Although of unknown etiology, associated with underlying conditions such as Ehlers-Danlos syndrome, cutis laxa, Marfan's, Kenny-Caffey syndrome is considered. The majority of cases are sporadic, however autosomal recessive pattern is sometimes speculated. The pathology of this condition is atrophy of the musculo-elastic tissue of the trachea and bronchi leading to their dilatation and causes ineffective mucociliary action and impaired cough reflex. Furthermore, during inspiration due to negative intrathoracic pressure, there is marked dilatation of trachea which collapses during expiration.<sup>(2,3)</sup> This subsequently leads to accumulation of secretions and frequent infections i.e. bronchiectasis, recurrent pneumonia and emphysema like the picture. Various subtypes of this disease are type 1 which is symmetrical dilatation of trachea, while type 2 is dilatation plus diverticula formation and type 3 is diverticula with sacculations extending till distal bronchi, as in our case. Chest x-ray could diagnose the condition, as positive findings being enlarged trachea and bronchi with bronchiectatic changes, but it has poor predictive value. Computed tomography scan is the diagnostic method of choice with criteria on measurements being trachea: >30 mm, right main bronchus: >20mm and left main bronchus: >18mm. Sacculations in the trachea and bronchi with hyperinflated lung fields alongside the fibrobronchiectasis are the main associated findings. Fiberoptic bronchoscopy will help in appreciating the deranged airway dynamics in inspiration/expiration.<sup>(4)</sup>

General guidelines for treatment of disease is cessation of smoking, as low as possible exposure with inhaled irritant pollutants and managing simultaneously existing cardiopulmonary diseases like chronic bronchitis, chronic obstructive pulmonary disease. <sup>(5,6,7)</sup> Physiotherapy increases clearance of secretions. Tracheal stenting is the need of the hour in severe cases. Surgery is rendered irrelevant as the disease is widespread.

**CONCLUSION:** A 28 year old non-smoker male came with cough, fever, dyspnoea and chest pain since one month with chest x-ray showed prominent trachea with emphysematous lung fields. On further evaluation computed tomography scan of the chest (plain) revealed dilated trachea and mainstem bronchi with multiple sacculations in the proximal part of thoracic part of trachea involving posterolateral walls with tractional and tubular bronchiectasis in bilateral lung fields which confirms it as a case of Mounier-Kuhn syndrome which is a rare cause of bronchiectasis and should be considered in differentials of lower respiratory tract infections.

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**Figure 1:** Topogram of chest CT scan shows the prominent trachea with honeycombing in lower ones (Arrows).

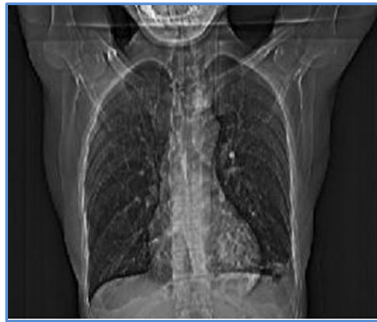


Figure 1

**Figure 2:** Plain chest CT scans mediastinal window (2A) and lung window (2B), axial image shows dilated trachea (black arrow) with multiple sacculations (black broad arrow) in the proximal part of thoracic part of trachea involving posterolateral walls.

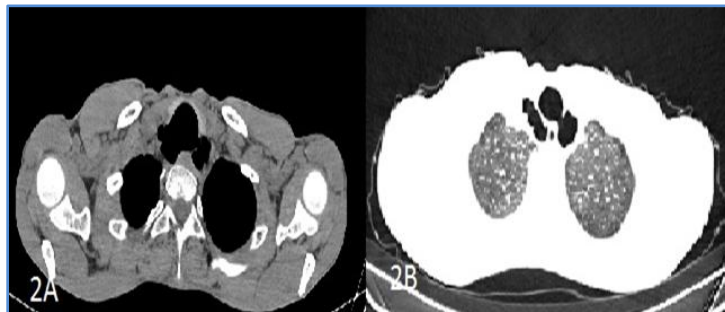


Figure 2

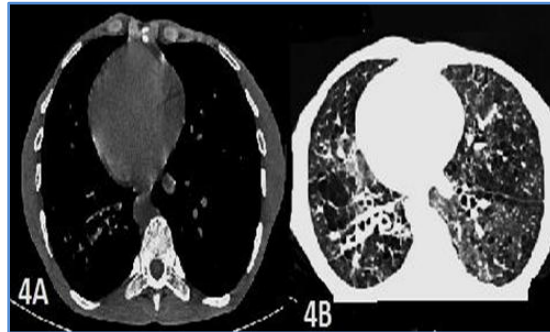
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**Figure 3:** Plain chest CT scan mediastinal window, axial image shows dilated both mainstem bronchi (white arrow).



**Figure 3**

**Figure 4:** Plain chest CT scans mediastinal window (4A) and lung window (4B), axial image shows tractional and tubular bronchiectasis (black arrow) in bilateral lower lobes with hyperinflated lung fields with areas of air trapping.



**Figure 4**

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Date of Submission: 23/12/2014.  
Date of Peer Review: 24/12/2014.  
Date of Acceptance: 31/12/2014.  
Date of Publishing: 07/01/2015.