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Manuscript Type: Images of Interest

Title: Mounier-Kuhn Syndrome

Authors:

Irfan Ismail Ayub MD, DM, MRCP(UK)
Professor,
Department of Pulmonary Medicine,
Sri Ramachandra Medical College and Research Institute, Chennai, India

Krishnamoorthy Vengadakrishnan MD
Professor,
Department of Internal Medicine,
Sri Ramachandra Medical College and Research Institute, Chennai, India

Corresponding author:

Irfan Ismail Ayub
Department of Pulmonary Medicine,
First Floor Link, G Block,
Sri Ramachandra Medical College and Research Institute, SRIHER,
No.1 Ramachandra Nagar, Porur, Chennai 600 116, Tamilnadu, India
Phone: +919884282400
Email: iia@rediffmail.com

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Author contributions: Both IIA and KV conceived the article and both IIA and KV contributed equally to writing and revising the manuscript.
Mounier-Kuhn Syndrome

Sixty-two-year-old male presented with increasing cough, purulent sputum and breathlessness for three days. Over the last thirty years, he has been having recurring cough with sputum, often interrupted by exacerbations, requiring multiple outpatient and emergency visits, and on several occasions, admissions for antibiotics, oxygen and bronchodilator therapy. Clinical examination revealed tachypnoea with peripheral saturation of 91%, tachycardia, pallor, digital clubbing, with bilateral crackles and wheeze on chest auscultation. Blood investigations and arterial blood gas analysis revealed anaemia and hypoxemia, respectively. Sputum microbiology, sweat chloride and serum immunoglobulin profile were non-contributory. Chest radiograph showed bilateral lower zone cystic changes (yellow circles) with airspace opacities (white arrows). Computed tomography scan of chest revealed tracheomegaly (tracheal diameter 31.29 millimetre) with multiple tracheal and bronchial wall diverticula (red arrows), upper lobe emphysema (E), bilateral bronchiectasis (yellow arrows), and right lung consolidation (P). Flexible bronchoscopy confirmed multiple tracheal and bronchial wall diverticulosis (white arrows), with visible overlying atrophied longitudinal smooth muscle (black arrows), consistent with Mounier-Kuhn syndrome. Bronchoalveolar lavage from middle lobe grew Klebsiella pneumoniae. He was discharged following clinical
and radiological improvement after appropriate antibiotic, oxygen and bronchodilator therapy. He continues to have reasonable symptom control with regular home nebulized bronchodilator therapy and chest physiotherapy, and has completed vaccination for pneumococcus and influenza.

First described in 1932, Mounier-Kuhn syndrome is a rare clinical entity characterized by tracheobronchomegaly, which in turn leads to ineffective airway mucociliary clearance, recurrent lower respiratory tract infections and bronchiectasis. Around 200 cases have been reported in medical literature, with less than twenty from the Indian subcontinent. Pathological examination of the airways in these patients have demonstrated atrophy of the elastic and smooth muscle fibres. Subsequent to this, there is loss of the architectural framework of the central airways, leading to tracheobronchomegaly, and outpouchings of the walls and development of blind diverticula. The diagnosis is often made on computed tomography scan of the chest, demonstrating a tracheal diameter above 30 millimetre (mm) (measured 2 centimetre above the aortic arch) and a right and left bronchial diameter of more than 20 and 18 mm respectively. Flexible bronchoscopy, when performed, may aid in the diagnosis, by demonstrating airway secretions and central airway diverticulosis. Clinical management of these patients
involves treatment of infective exacerbations with antibiotics and supportive therapies, and long-term application of regular airway clearance techniques.[1,2]

References:


Figure caption
Figure 1 - Chest radiograph postero-anterior view shows bilateral lower zone cystic changes (yellow circles) with air space opacities (white arrows).

Figure 2 - (A) and (B) Axial high resolution computed tomography (HRCT) scan of chest (A) at the level of mid trachea 20 millimetres (mm) above arch of aorta showing tracheomegaly with a measured transverse diameter of 31.29 mm, with bilateral paraseptal emphysema (E), and (B) at the level of carina showing bilateral bronchial dilatation, multiple bronchial diverticula (red arrows), bilateral upper
lobe paraseptal emphysema (E) and bronchiectasis (yellow arrows), and right upper lobe consolidation (P). (C) Flexible bronchoscopy showing lower trachea and carina, with diverticula over posterior wall of trachea, and medial wall of both main bronchus (white arrows), along with atrophy of the longitudinal smooth muscle seen over the posterior tracheal wall (black arrows).