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Case Report

Mounier Kuhn syndrome in adult: a case report

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ABSTRACT

Mounier-Kuhn syndrome or tracheobronchomegaly is a rare congenital abnormality of the trachea and main bronchi characterized by marked cystic dilatation of the tracheobronchial tree, associated with tracheal diverticulosis, bronchiectasis and recurrent lower respiratory tract infections. We report a case of 22 years adult male with history of recurrent lower respiratory tract infection.

Keywords: Mounier Kuhn syndrome, Tracheobronchomegaly

INTRODUCTION

Mounier-Kuhn syndrome is a rare clinical and radiological entity characterized by marked dilatation of the trachea and bronchi and recurrent lower respiratory tract infections. Diagnosis is usually made on CT scan. Various names are used by various authors for this condition, e.g. tracheobronchomegaly, trachiectasis, multiple tracheal diverticula, tracheobronchopathia malacia and tracheomegaly.[1]

CASE REPORT

A 22-year-old male presented to TB & chest OPD with history of chronic cough with sputum production and fever for the past 10 days.

He gave history of increased frequency of respiratory infections and increasing sputum production for last few years for which he had hospital admissions for several times and was treated with antibiotics.

He was asymptomatic in between these episodes. The patient was a non- smoker with no occupational or environmental exposure to respiratory irritants and there was no family history of a similar illness.

On physical examination the patient was well nourished, thin built with no other abnormality. Sputum and tracheal aspirate were negative for bacterial, mycobacterial, and other atypical organisms. Pulmonary function tests were entirely normal

CT scan of the chest was performed. HRCT reveals tracheobronchomegaly. The trachea was grossly dilated, with a diameter of 4.2 cm, while the right and left main bronchi had diameters of 2.2 and 2.7 cm, respectively.

Multiple diverticula and areas of scalloping were seen between the cartilaginous rings in the trachea and right and left main bronchi.

Findings suggested type 2 Mounier-Kuhn syndrome. Tubular bronchiectasis was also noted in the right middle and lower lobes. Few fibrotic opacities ware seen in bilateral lung fields at places.

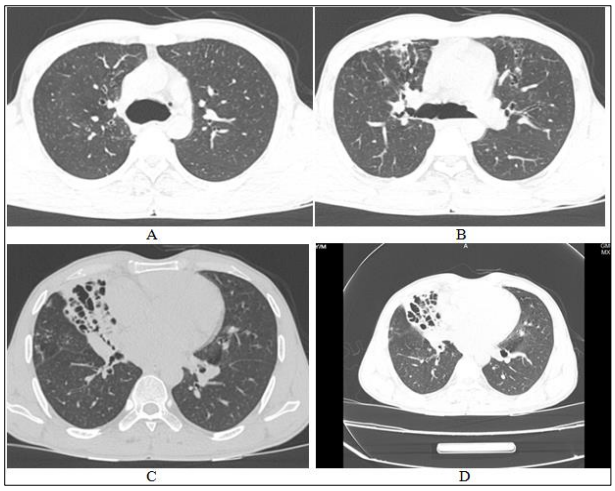

Figure 1: Axial HRCT thorax images in A & B showing dilated trachea and bronchi. C& D showing traction bronchiectasis in right middle lobe.

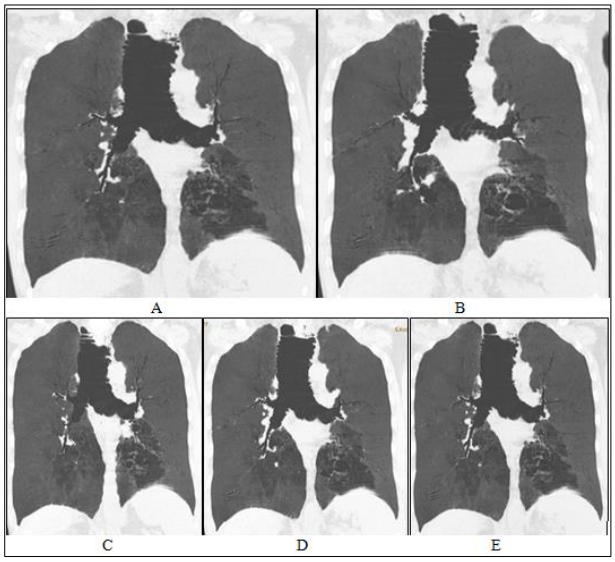

Figure 2: MIP coronal images (A-E) showing dilated trachea and bronchi. The tracheobronchial walls are corrugated in appearance suggesting diverticulosis (white arrows).

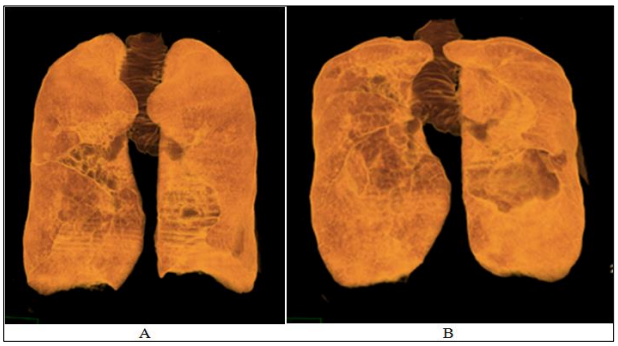

Figure 3: Volume rendered images (A & B) of the same patient showing dilated trachea and bronchi with bronchiectasis.

DISCUSSION

Mounier-Kuhn syndrome is a congenital abnormality of the trachea and main bronchi characterized by atrophy or absence of elastic fibers and thinning of muscle, which allows the trachea and main bronchi to become flaccid and markedly dilated on inspiration with narrowing or collapse on expiration or cough. The abnormal airway dynamics and pooling of secretions in broad outpouchings of redundant musculomembranous tissue between the cartilaginous rings predispose to the development of chronic pulmonary suppuration, bronchiectasis, emphysema, and pulmonary fibrosis.[2]

Mounier Kuhn syndrome was first recognized at autopsy by Czyhlarz in 1897, but it was not until 1932 that Mounier-Kuhn associated the endoscopic and radiographic appearance of the markedly enlarged airway with recurrent respiratory tract infection.[3]

The symptoms of Mounier Kuhn syndrome are nonspecific and are usually indistinguishable from those caused by chronic bronchitis or bronchiectasis. Sputum production is often copious and purulent. Associated spontaneous pneumothorax, haemoptysis, pneumonia and finger clubbing may sometimes develop.[4]

The disease predominantly occurs in males.[4] This syndrome generally presents in young adults, although the symptoms may date back to childhood. Typically, the patients have chronic cough and recurrent chest infections. Excessive sputum production with occasional hemoptysis occurs. Some patients develop mucosal herniation between the tracheal rings leading to tracheal diverticulosis. The poor clearance of secretions is associated with recurrent lower respiratory tract infections.[5]

Diagnosis of Mounier Kuhn syndrome is established by bronchoscopy and non-invasively by Computed Tomography (CT).[6] In adults, the accepted criteria for diagnosis of tracheobronchomegaly on CT are diameters of the trachea, right main bronchus and left main bronchus of >30 mm, 20 mm and 18 mm respectively.[7]

Mounier-Kuhn syndrome has 3 subtypes. In type 1, there is a slight symmetric dilation in the trachea and main bronchi. In type 2, the dilation and diverticula are distinct. In type 3, diverticular and saccular structures extend to the distal bronchi.[8]

It is a condition for which no effective therapy exists and usually it is mainly supportive. Symptomatic patients with airway collapse might require endobronchial stenting. Treatment is directed to minimizing the damage resulting from stasis and infection, and general supportive therapy, e.g., bronchodilators.[9]

In conclusion, this rare syndrome of tracheobronchomegaly should be considered in patients

with bronchiectasis associated with abnormal dilation of trachea and major bronchi on CT scans. HRCT helps in the identification of bronchiectasis and accurately determine the extent and type of the disease.

Despite long-term follow-up by physicians for the repeated episodes of chronic productive cough and lower respiratory tract infection, tracheobronchomegaly had not been diagnosed in this patient until CT was performed. Because tracheobronchomegaly can be easily overlooked on plain chest films, a CT scan should be done in patients who have chronic recurrent lower respiratory tract infection for evaluation of the underlying predisposing conditions, including tracheobronchomegaly.

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