

Case Report: Mounier–Kuhn Syndrome

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Abstract

Tracheobronchomegaly (TBM) or Mounier - Kuhn syndrome is a rare disorder of uncertain aetiology characterized by marked dilatation of trachea and bronchi and recurrent lower respiratory tract infection. We report a case in a 20 year old male presenting with recurrent lower respiratory tract infection.

Keywords: Mounier–kuhn syndrome; Tracheobronchomegaly (TBM)

Introduction

Mounier–Kuhn syndrome or Tracheobronchomegaly is a rare clinical and radiological entity characterized by marked dilatation of the trachea and bronchi and recurrent lower respiratory tract infection [1-3].

It is known by a number of different names like trachiectasis, tracheobronchopathia malacia, tracheomegaly and multiple tracheal diverticula.

Case Report

A 20 year old male patient was admitted with complaints of recurrent lower respiratory tract infection since 8 years of age, presenting as episodes of productive cough with fever. He was asymptomatic in between these intervals. Since last 4 years, the frequency of lower respiratory tract infections have increased with increased productive cough and gradual onset of breathlessness. He had never smoked. There was no significant occupational history. There was no family history of similar illness or significant occupational history.

Patient had undergone chest x-ray and other routine blood investigations previously, but previous medical records were not available and this disease was not suspected till now. Physical examination revealed increased body temperature with decreased breath sound in both lungs with inspiratory crepitations, more pronounced in mid and lower lung field. Finger clubbing was present.

Laboratory investigations revealed mild leukocytosis (12.8×10^3 leukocytes/ μ L) with ESR (20 mm/hr) and C- reactive protein (18.9) in borderline range. Sputum was negative for acid fast bacilli. Pulmonary function testing revealed a forced expiratory volume in 1 sec (FEV1) of 1.98 L (50%), a forced vital capacity (FVC) of 2.7 L (59%), and FEV1/FVC of 0.73.

Chest radiograph revealed enlargement of trachea and bronchi and bilateral bronchiectasis.

CT scan of chest was performed. The scanogram (Figure 1) revealed tracheobronchomegaly. The trachea was grossly dilated with transverse diameter of 3.3 cm and sagittal dimension of 3.95 cm (Figure 2), while right and left main bronchi had diameter of 2.83 cm and 3.60 cm respectively (Figure 3).

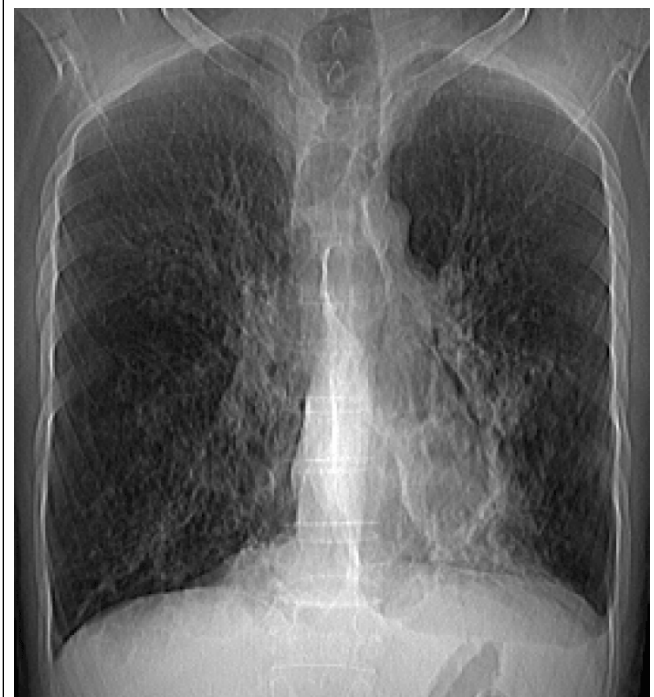


Figure 1: Scanogram of chest showing dilated trachea and main bronchi

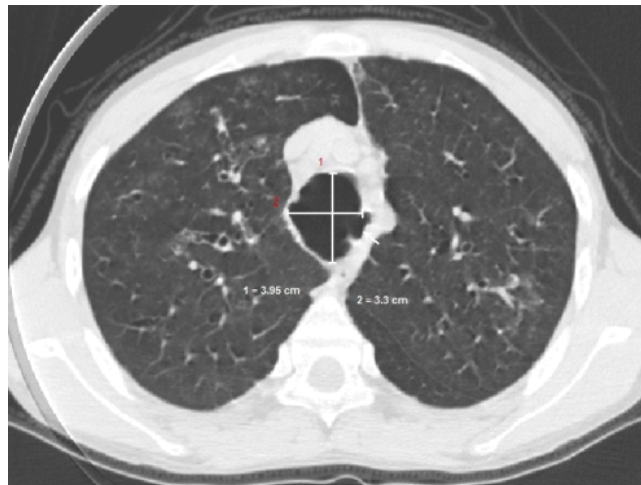


Figure 2: High resolution computed tomography of chest shows abnormal dilatation of trachea in both transverse and antero-posterior diameter. Diverticulae are also seen (marked by arrow)

Multiple diverticulae and areas of scalloping were seen between the cartilaginous rings in trachea and main bronchi (Figure 2 and 3). Cystic bronchiectasis was seen in bilateral lung parenchyma (Figure 4).



Figure 3: High resolution computed tomography of chest shows dilatation of both main bronchi associated with sacculations

Fibreoptic bronchoscopy revealed dilated trachea and widening of bronchial tree bilaterally with flaccid walls and diverticular outpouchings in tracheal and main bronchi region. Airway flaccidity was seen up to second generation bronchi with partial collapse during expiration.



Figure 4: High resolution computed tomography of chest in lung window shows extensive bilateral cylindrical and varicose bronchiectasis

Discussion

Congenital Tracheobronchomegaly or the Mounier–Kuhn syndrome is a rare clinical and radiological entity. In 1932, Mounier – Kuhn correlated its endoscopic and radiographic appearance for the first time [4].

Incidence of TBM is rare, however the actual number of cases may be much greater as some patients with TBM are totally asymptomatic, whereas in those with symptoms, TBM is frequently overlooked when only chest radiographs are made [5].

Patients usually present with recurrent respiratory tract infections and there is male predominance. Although the aetiology is uncertain, it is believed to be due to the lack of smooth muscles and elastic connective tissue in the trachea and main bronchi, leading to sacculations and formation of diverticulae between the cartilaginous rings [6,7].

Disorders such as Sarcoidosis, usual interstitial pneumonia and cystic fibrosis, which causes severe fibrosis of the upper lobes, may also exert sufficient tracheal traction to result in tracheal enlargement. Certain other condition such as Marfan syndrome, Ehler – danlos syndrome, Kenny – coffey syndrome, ataxia telangiectasia, connective tissue diseases, Brachmann – de lange syndrome, Bruton type agammaglobulinaemia, ankylosing spondylitis, cutis laxa and light chain deposition diseases are also associated with secondary tracheobronchial enlargement [8]. However most cases are sporadic and idiopathic and show no evidence of associated connective tissue disease [8], as was the case in our patient.

On CT scan, the diagnosis is made when the transverse diameter of trachea measures greater than 3.0 cm and that of right and left main bronchi exceeds 2.4 cm and 2.3 cm respectively [9]. The diameter in present case were 3.3, 2.8 and 3.6 cm respectively. Apart from Tracheobronchomegaly, tracheal diverticulosis, secondary to protrusion of redundant musculo membranous tissue between the cartilaginous rings, may result in an irregular, corrugated or scalloped appearance of trachea and occasionally the main bronchi. Chronic

infection usually results in central bronchiectasis, which may be cylindrical, cystic or varicose [9].

The experience with the use of magnetic resonance imaging (MRI) in TBM is limited [10]. There is no specific therapy for asymptomatic patients with supportive treatment in symptomatic patients. Physiotherapy, postural drainage and antibiotics for clearing of secretions and suppressing infectious exacerbations are the mainstay of treatment. There is no definite role of either inhaled bronchodilators or corticosteroids.

Surgery is rarely done due to diffuse nature of disease with tracheal stenting being useful in advanced cases with reports of tracheobronchial endoprosthesis being used with some success [11-13].

As Tracheobronchomegaly are often overlooked on plain chest X-Ray, patients with chronic and recurrent lower respiratory tract infection should have a CT scan done to rule out underlying predisposing factor like tracheobronchomegaly as is in this case.

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