Case Report

A 54-Year-Old Man with Tracheomegaly, Tracheal Diverticulas and Bronchiectasis — Mounier-Kuhn Syndrome

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Abstract

Mounier-Kuhn syndrome is a rare idiopathic clinical, radiological and bronchoscopic disorder characterised by abnormal dilatation of the tracheo-bronchial tree. The usual presentation is with recurrent lower respiratory tract infections. Herein, we report the case of an adult male who was diagnosed to have Mounier-Kuhn syndrome based on radiographic finding of a tracheal diameter of 45.5 mm on computed tomography and dynamic complete collapse of the tracheo-bronchial tree on forced expiration, observed during ronchoscopy. **[Indian J Chest Dis Allied Sci 2015;57:113-115]**

Key words: Mounier-Kuhn syndrome, Tracheomegaly, Bronchoscopy, Dynamic collapse.

Introduction

Mounier-Kuhn syndrome, also called as tracheo-bronchomegaly, is a rare condition characterised by marked dilatation of the trachea and proximal bronchi. The condition was first described in 1932 by Mounier-Kuhn.[1] A recent literature review revealed nearly 200 published cases of the syndrome.[2] On histopathological examination, the condition is characterised by atrophy of elastic fibres. Also thinning of muscularis layer has been demonstrated.[3]

Herein, we report the case of an adult male who was diagnosed to have Mounier-Kuhn syndrome based on characteristic clinical presentation, radiographic findings and classical findings on bronchoscopy.

Case Report

A 54-year-old male presented with a 30-year history of repeated attacks of colds, cough with sputum and episodic breathlessness with wheezing. He used to have episodic increase in purulence and quantity of sputum (about ≥200 mL/day), 2-3 times a year, especially during the winter season. His breathlessness had become persistent for the past 10 years. He worked as an auto-rickshaw driver, and had smoked 20 *bidis*/day for 20 years before quitting 10 years ago. He had been diagnosed earlier to have bronchial asthma with bronchiectasis for which he was being treated.

On examination, clubbing was present. Auscultation revealed bilateral inspiratory crepitations and polyphonic rhonchi with prolonged expiration. His arterial oxygen saturation on room air by pulse oximetry was 95%; blood counts were within normal limits. Pulmonary function testing revealed a sharp rise to peak expiratory flow rate (PEFR) with a post-bronchodilator PEFR of 4.63 L/s (71%) and a rapid decline in the flow. Oscillations were also seen in the descending limb of flow volume loop with a forced expiratory volume in the first second (FEV_1) of 1.00 L (48%), a forced vital capacity (FVC) of 2.16 (85%) and the ratio of FEV_1/FVC of 46% (Figure 1).

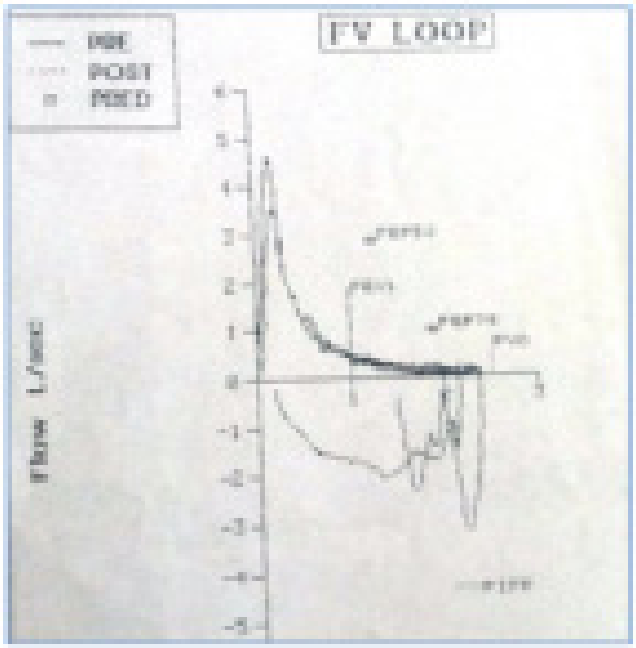

Figure 1. Spirometry showing a rapid fall from peak with oscillations in descending limb of expiratory curve.

[Received: July 21, 2014; accepted: August 21, 2014]

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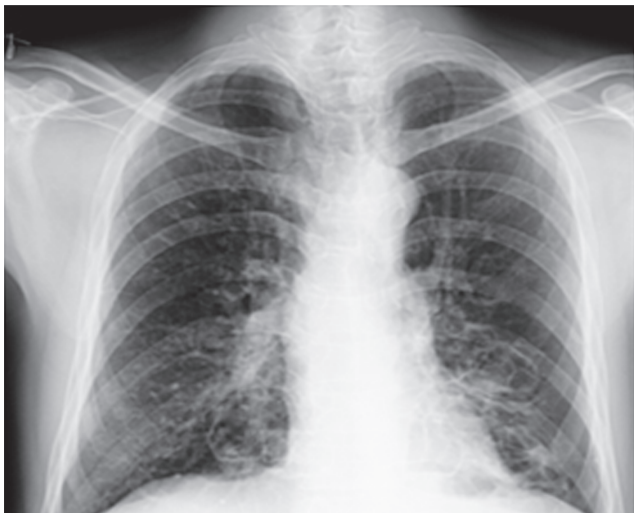

Figure 2. Chest radiograph (postero-anterior view) showing tracheomegaly with bilateral bronchiectasis.

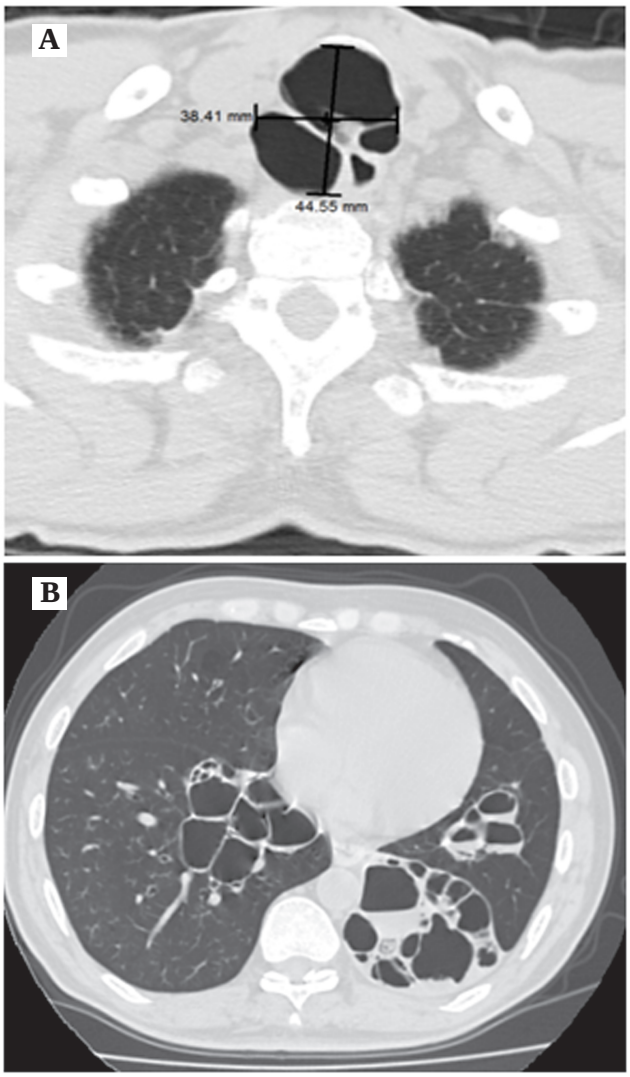

Figure 3. High resolution computed tomography (HRCT) of chest showing (A) coronal diameter of 38.4 mm and saggital diameter of 44.5 mm of the trachea with multiple tracheal diverticulas, and (B) bilateral bronchiectasis.

Chest radiograph revealed enlargement of the trachea and bilateral bronchiectasis (Figure 2). Computed tomography (CT) of the chest showed the coronal and sagittal diametres of the trachea to be 38.4 mm and 45.5 mm, respectively, multiple tracheal diverticula and bilateral bronchiectasis (Figure 3A and B).

Fibreoptic bronchoscopy revealed dilatation of the trachea with multiple diverticulae with blind ends in its posterior wall (Figure 4A). A nodule was noted in the right lateral tracheal wall, biopsy of which revealed only submucosal gland hyperplasia (Figure 4B). The proximal tracheo-bronchial tree as well as the major bronchi showed almost complete collapse on forced expiration (Figures 4C, 4D, 4E, and 4F).

On the basis of characterstic radiological and bronchoscopic findings, the patient was diagnosed to have Mounier-Kuhn syndrome and was discharged on treatment with oral antibiotics.

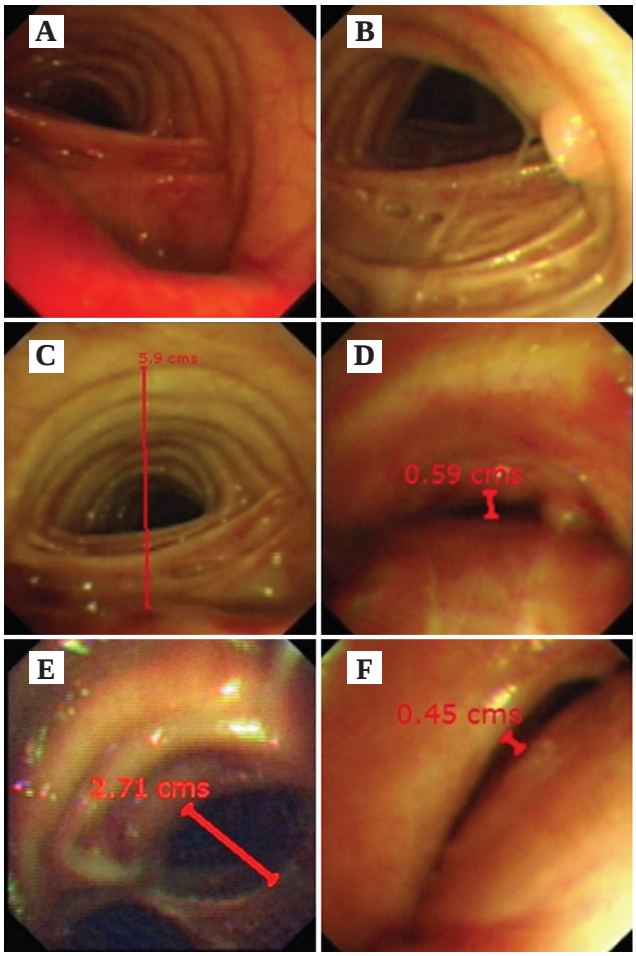

Figure 4. Fibreoptic bronchoscopy showing (A) multiple tracheal diverticula, (B) a tracheal nodule, (C & D) dynamic collapse of trachea and (E & F) dynamic collapse of main bronchi.

Discussion

Mounier-Kuhn syndrome is a rare disorder characterised by pathological dilation of the tracheo-bronchial tree due to atrophy of the elastic and muscular tissue.[3] The aetiology of the syndrome is unknown. In some cases there is a known association with Ehlers-Danlos in adults and *cutis laxa* in children suggesting a possible correlation with connective tissue disorder while majority of the cases are sporadic.[4] In our case, the patient denied any history suggestive of a connective tissue disorder. The disease is predominantly seen in men in the third and fourth decades of life.

The clinical presentation of the syndrome is non-specific with most of the symptoms related to recurrent infections, secondary to diverticula and bronchiectasis. The enlarged weakened airways with poor cough reflex leads to recurrent episodes of pneumonia further leading to bronchiectasis and fibrosis.[5] The presentation in our case was also secondary to repeated episodes of purulent sputum.

Pulmonary function testing may be useful in the evaluation of patient but these are not useful diagnostically. The various patterns described are low peak flow with a rapid decrease in flow related to dynamic central airway collapse, notching of the expiratory curve, oscillations and a reduction in maximum voluntary ventilation.[6] The present case also showed a reduction in peak expiratory flow with rapid decline and oscillations in expiratory curve.

Radiographically, the diagnostic criteria for tracheo-bronchomegaly in adults, defined as upper limit of the mean plus three standard deviations of the normal values, include tracheal diameter exceeding 30 mm, or the left and the right mainstem bronchi exceeding 23 mm and 24 mm in diameter, respectively.[7] In the present case, the coronal and sagittal diameters of the trachea were 38.4 mm and 44.5 mm, respectively, thus fulfilling the diagnostic criteria for tracheomegaly.

Bronchoscopic demonstration of dynamic tracheal or bronchial collapse is considered the "gold standard" for diagnosing tracheomalacia. Most studies have used a cut-off greater than 50% reduction in cross-sectional area of the trachea during expiration, although some have suggested a cut-off greater than 70% narrowing on forced expiration, as a diagnostic criteria.[6] Our case showed almost complete collapse of the treachea on forced expiration.

Tracheomalacia has been classified into 3 types: type 1 (relatively subtle, symmetric, diffuse enlargement of the tracheobronchial tree); type 2 (more obvious enlargement with diverticula and bizarre eccentric configurations); and type 3 (diverticula and sacculi in the trachea extending to the bronchial tree.[8] The present case fits into type 3 due to the presence of multiple diverticuli and sacculi in the tracheo-bronchial tree.[8]

Treatment aims at smoking cessation, effective treatment of infections, treatment of associated co-morbidities and airway stents in those with severe symptoms.[2]

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