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CASE REPORT

Mounier-Kuhn syndrome: A rare cause of severe bronchial dilatation with normal pulmonary function test: A case report

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Summary

Tracheobronchomegaly (TBM) (Mounier-Kuhn syndrome) is dilatation of the trachea and major bronchi because of atrophy or absence of elastic fibers and smooth muscle cells. We present a case of TBM with normal pulmonary function test (PFT).

The patient was a 37-year-old man with increasing productive cough and without fever, wheezes, chest pain, weight loss or any respiratory disease. Chest helical computed tomography (CT) scan showed tracheomegaly with transversal diameters of the trachea of 44 mm. CT scan showed collapse of the trachea. Few large diverticular out-pouching and openings in the trachea was seen in bronchoscopy. PFT results were normal.

PFT in large airway disorders may be normal while abnormalities may indicate underlying small airway disorder. An underlying small airway disorders is responsible for abnormal reports in PFT of these patients. We may need to re-evaluate the role of PFT within follow-up of patients with large airway disorder.

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Introduction

Tracheobronchomegaly (TBM), also called Mounier-Kuhn syndrome, consists in dilatation of the trachea and major

bronchi because of atrophy or absence of their elastic fibers and smooth muscle cells.¹ Firstly was described in 1932.² Pathologically, there is loss of cartilage and muscle within the trachea and bronchi, resulting in dilatation. Because of the loss of inherent tracheal wall support, diverticula develop, commonly in posterior part.³ The etiology of disease was not exactly determined but previous authors suggested congenital nature of disease

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sometimes associated with Ehlers-Danlos syndrome in adults and cutis laxa in children.⁴ There are fewer reports that suggest an acquired origin of the disease, noting that almost 50% of patients show no symptoms until the third decade of life.⁵ Clinical suspicion of this syndrome is based on the construction of an exact case history together with appropriate evaluation of the chest CT which is able to diagnose and measure the abnormal amplitude of the airways.^{6,7}

The symptoms of TBM are nonspecific, with sputum production secondary to bronchiectasis and lower respiratory tract infection⁸ and on a plain chest radiograph, the increased caliber of the central airways may be visible.

In previous studies, pulmonary function tests (PFTs) reveal an obstructive pattern and increased residual volume.⁹ In addition recurrent episodes of pneumonia lead to a progressive loss of pulmonary function.¹⁰ But there is no definite description of PFT due to Mounier-Kuhn syndrome while there are some various reports in type and pattern of this test.

Here we present a case of TBM with normal PFT. To our knowledge and based on literature review we did not find any report that noted such a combination.

Case report

A 37-year-old man has been referred to our setting with increasing productive cough. Although the severity of

symptoms recovered after hospitalization but he never experienced complete recovery so far. During the last 15 years, he had experienced an increased expectoration of mucoid sputum that became purulent during infectious exacerbations, sometimes with bloody streaked sputum. He complained of increase of his productive cough from last year. He denied fever, wheezes, chest pain and weight loss. No other respiratory illness was present. His parents and siblings were normal. He never smoked and he worked as a stucco worker after partial recovery of his disease. Vital signs were normal. Finger clubbing was not present. Clinical respiratory examination disclosed normal breath sounds over both lungs but mild inspiratory crackles at the lower third of both lung fields. The results of blood analysis were within normal limits. A chest helical CT scan (Fig. 1) were performed, showing tracheomegaly with transversal diameters of the trachea of 44 mm. Computed tomography (CT) performed at end expiration also showed significant collapse of the trachea. A bronchoscopy was performed and disclosed few large diverticular out-pouching and openings in the posterior and lateral wall of trachea; some of the openings could be easily penetrated by the tip of bronchoscope (Fig. 2). Sputum and tracheal aspirate results were negative for mycobacteria. Forced expiratory volume in 1s in (FEV₁%), forced vital capacity (FVC) were equal to 91% and 87%. Other PFT values (Vital Capacity and FEV₁/FVC) were normal too.

PFT and flow volume loop was within normal range (Table 1).

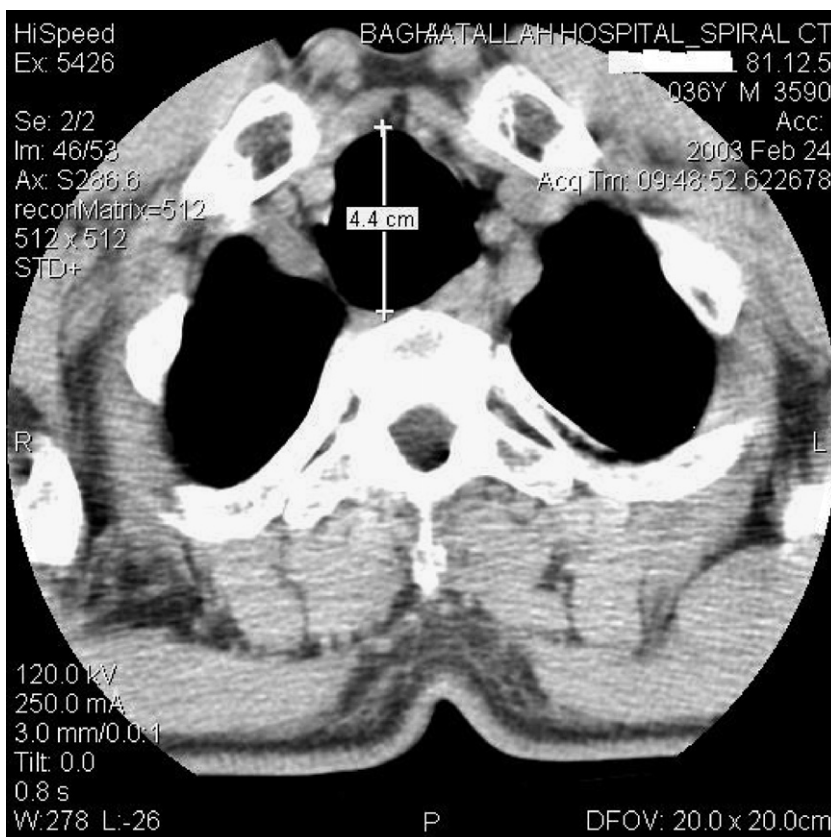


Figure 1 Tracheomegaly in tracheal CT scan of the patient.

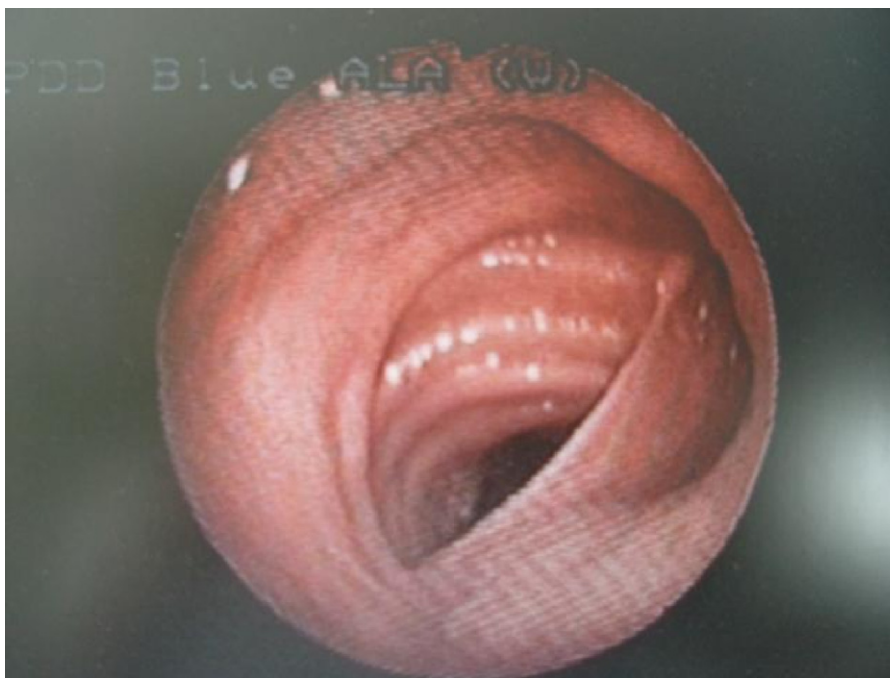


Figure 2 Bronchoscopic view of trachea in the patient with tracheomegaly.

Discussion

To our knowledge this is the first case reporting Mounier-Kuhn syndrome while normal PFT.

TBM is a rare disorder of uncertain aetiology while autopsy studies suggested a congenital defect or atrophy of the elastic and smooth muscle tissues of the trachea and main bronchi, resulting in dilatation.¹¹ Because of the weakened trachea, diverticula develop, commonly posteriorly, with secretion retention. Alterations lead to the collapse of the respiratory tract during forced exhalation, making expectoration by coughing of little use. The airways distal to the fourth-order or fifth-order division are usually of normal diameter.¹² The inciting factors of Mounier-Kuhn syndrome are not known. Acquired forms have been described as a complication of pulmonary fibrosis in adults and of mechanical ventilation in preterm neonates. No evidence of an impact of cigarette smoking has been shown. Nevertheless, the majority of cases are sporadic without an association.¹² The symptoms of TBM are not specific. Generally the syndrome appears in men during their third or fourth decades of life, with sputum secondary to bronchiectasis and lower respiratory tract infections.¹³ Subjects with this disorder are also predisposed to phlogistic bronchopulmonary pathologies such as bronchitis, emphysema, and pulmonary fibrosis.¹⁴ The diagnosis is difficult to establish with regular chest X-ray particularly in the lateral position except one may demonstrate, an increased diameter of the trachea, the right and the left main bronchus that exceed 3.0, 2.4, and 2.3 cm, respectively. Chest CT-scan permits one to evaluate the full extent of the disease, and the presence of associated tracheal diverticulosis, or the presence of areas of bronchiectasis. Management and treatment of patients affected by Mounier-

Table 1 Results of pulmonary function test

	Predict	Best	% Pred
FVC (l)	4.75	4.13	87
FEV ₁ (l)	3.72	3.39	91
FEV ₁ /FVC			%104
FEF 25–75% (l/Min)	240	206	86

Kuhn syndrome are difficult.¹⁵ Physiotherapy is useful to clean secretions; antibiotic therapy controls infectious exacerbations due to herniation of mucosa into saccular diverticulae. The centralized and diffuse nature of the disease prevents an extensive surgical approach, which is limited to complications, such as pneumothorax or tracheal stenosis.^{16,17}

PFTs in excessive dynamic airway collapse might show diminished expiratory flow, typical notching on the flow-volume (FV) loop, dynamic airway compression (calculated as slow vital capacity minus FVC), a biphasic FV loop or flow oscillations. These findings are neither sensitive nor specific. Overall, few studies rigorously report results of PFTs in these patients.

In evaluation of patients with tracheobronchomalacia it has been shown that abnormality in HRCT and PFT highly probable due to underlying small airway or parenchymal disorder. Tracheobronchomalacia as a large airway disease lonely could not able to impair PFT.¹⁸

In the case reported here, the patient was affected by tracheobronchomegaly while interestingly all pulmonary function indices are normal.

In this case as a large airway affected patient, despite severity of disorder, PFT result was within normal range. Probably it is logical to consider concurrent and underlying small airway disorders are responsible for abnormal reports in pulmonary function of these patients. All together PFT in large airway may be normal while abnormalities in this test have to trigger to consider underlying small airway disorder. Also findings of this study indicate that we may need to re-evaluate the role of pulmonary function screening within follow-up of patients with large airway disorder.

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