

Case Report

Clinical and Radiological Manifestations of Congenital Tracheobronchomegaly (Mounier - Kuhn Syndrome)

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Abstract

Background

Mounier-Kuhn Syndrome is a rare congenital abnormality characterized by pathological dilation of the trachea and major bronchi. Symptoms can range from minimal with preserved lung function to severe respiratory failure. Early recognition is important as it can lead to better management and prevention of complications. The diagnosis is usually made on the basis of the characteristic CT scan findings.

Case report

We report the case of a 57-year old man with chronic obstructive pulmonary disease and recurrent lower respiratory tract infections. Chest CT showed tracheobronchomegaly with multiple diverticula of the trachea and main bronchi – findings typical of Mounier-Kuhn Syndrome.

Conclusion

The diagnosis of Mounier-Kuhn syndrome should be considered in the differential diagnosis of patients with recurrent lower respiratory tract infections.

Keywords: Mounier-Kuhn Syndrome; Tracheobronchomegal; Computed Tomography

Introduction

Mounier-Kuhn Syndrome is a rare congenital abnormality characterized by pathological dilation of the trachea and major bronchi, often accompanied with tracheal diverticulosis, bronchiectasis, bullae and recurrent pulmonary infections. It was first described by a french physician Pierre Mouni-

er Kuhn in 1932 [1]. The incidence was estimated to be in the range of 0.5 to 1.5% with male preponderance [2]. The cause of the condition is not clearly understood; however, in biopsy studies, the absence or marked atrophy of the elastic fibers and smooth muscle of the trachea and main bronchi has been observed. Symptoms can range from minimal with preserved lung function to severe respiratory failure. Since

clinical picture is non-specific, diagnosis is made radiologically. Chest radiography may establish the diagnosis if tracheal dilatation is evident; however, CT is usually required for accurate airway measurement and to assess complications. Early recognition is important as it can lead to better management and prevention of complications.

Case report

A 57-year old man underwent chest CT imaging for the first time with complaints of recurrent lower respiratory tract infections over the last ten years, presenting as episodes of productive cough with fever. He had recently been diagnosed as chronic obstructive pulmonary disease.



Figure 1. Coronal and axial chest CT showing tracheobronchomegaly.

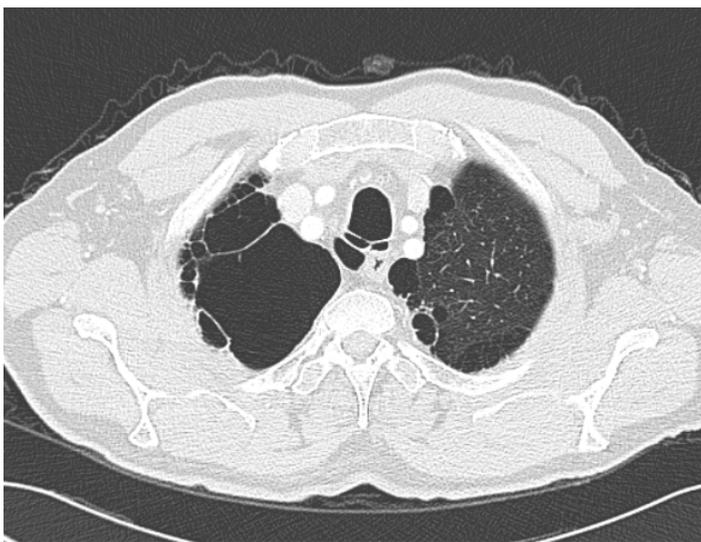


Figure 2. Coronal and axial chest CT showing multiple diverticula.

The patient was ex-smoker with no occupational exposure to respiratory irritants. There was no family history of a lung illness. Pulmonary function test were normal. Previous chest radiographs were stated to be normal, without any pulmonary infiltrates.

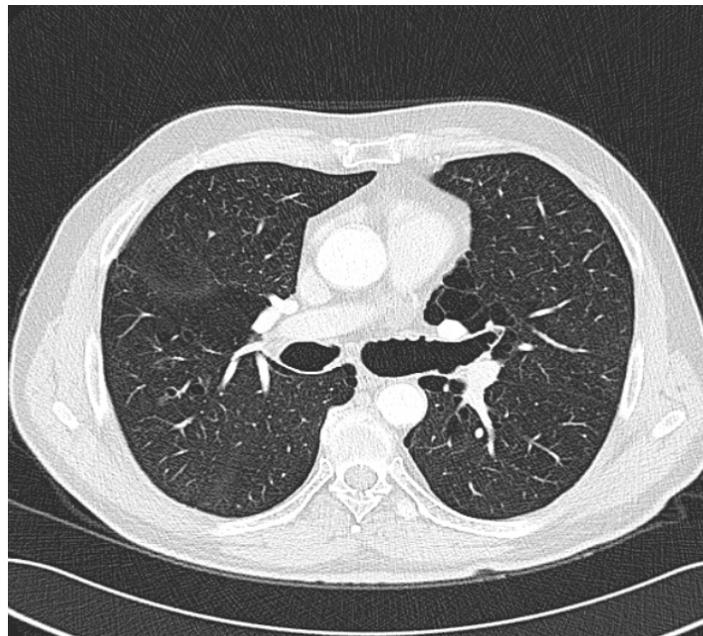


Figure 3. Coronal and axial chest CT showing bullous emphysema.

Chest CT scan showed evident tracheobronchomegaly. The trachea was dilated, with a diameter of 3.4 cm, while the right and left main bronchi had diameters of 2.1 and 2.3 cm, respectively. Multiple diverticula and areas of scalloping were seen between the cartilaginous rings in the trachea and in right and left main bronchi. The largest posterior tracheal diverticula was measuring 2x1 cm. Additional bullous emphysema and several cystic bronchiectasis were seen in the lung parenchyma bilaterally (Figures 1-3).

Discussion

Mounier-Kuhn syndrome is a rare congenital abnormality of unknown cause that is characterized by marked dilatation of the trachea and main bronchi, often accompanied with tracheal diverticulosis, emphysematous bullae, bronchiectasis, and recurrent pulmonary infections. The underlying abnormality is the absence or marked atrophy of the elastic fibers and smooth muscle within the wall of trachea and main bronchi. These airways are thus flaccid and markedly dilated on inspiration with narrowing or collapse on expiration or cough. The abnormal airway dynamics predisposes to mucosal herniation between tracheal rings, leading to tracheal diverticulosis and promoting frequent bacterial colonization. There is also increase in dead space and tidal volume. The airways distal to the fourth-order division are usually normal in diameter.

The condition is more common in men and is typically diagnosed in the 3rd or 4th decades of life [3]. Although the majority of cases appear to be sporadic, a familial form has been described [2]. Secondary tracheobronchomegaly was also described in association with Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, cutis laxa in children and with connective tissue diseases [4].

A broad spectrum of clinical abnormalities has been documented in Mounier-Kuhn syndrome ranging from minimal disease with good preservation of pulmonary function to progressive disease in the form of bronchiectasis, emphysema and pulmonary fibrosis leading to respiratory failure and death [3,5]. The typical clinical findings are non-specific such as dyspnea, cough, recurrent bronchitis or pneumonia. As the clinical symptoms are similar to chronic bronchitis many patients carry the clinical diagnosis of chronic obstructive pulmonary disease as seen in our case [6,7]. Spontaneous pneumothorax may also develop [8,9].

The most sensitive imaging test is a biphasic chest CT with images of the trachea obtained during inspiration and expiration. However, in most cases a chest CT during inspiration is sufficient for accurate airway measurement and to assess complications. Plain chest radiography may also demonstrate the enlargement of the trachea which is usually best seen in the lateral projection. For an adult, any diameter of the trachea, right main bronchus, and left main bronchus that exceeds 3.0 cm, 2.0 cm, and 1.8 cm, respectively, is diagnostic of Mounier-Kuhn syndrome [10-12].

Asymptomatic patients require no specific therapy. Smoking cessation and avoiding air pollutants is beneficial. The management of symptomatic patients is limited to physiotherapy to assist in clearing secretions and appropriate antibiotic therapy during infectious exacerbations [10]. There is no study to show the effectiveness of inhaled bronchodilators or corticosteroids. Few cases reported lung transplantation in Mounier-Kuhn syndrome, but there was no proven benefit in regards to morbidity and mortality [10]. Tracheal stenting can be useful in advanced cases to prevent expiratory collapse and there are reports of tracheobronchial endoprosthesis being used with some success [13,14]. The patients with Mounier-Kuhn syndrome require specific airway management during general anesthesia. There may come to severe peritubal air leakage due to the abnormally dilated trachea or to tracheal injury after intubation due to the weakened airways [15,16]. In one report a tracheal stenosis developed in a patient with Mounier-Kuhn syndrome being intubated for 15 days [17].

Conclusion

In conclusion, the diagnosis of Mounier-Kuhn syndrome should be considered in the differential diagnosis of patients

with recurrent lower respiratory tract infections. Since clinical picture is similar to chronic bronchitis many patients carry the clinical diagnosis of chronic obstructive pulmonary disease. The diagnosis is made radiologically with CT as the gold standard, and treatment is mainly supportive.

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