



## A rare case of Mounier-Kuhn syndrome with bronchial asthma

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### ABSTRACT

Tracheobronchomegaly (Mounier-Kuhn syndrome) is dilatation of the trachea and major bronchi because of atrophy or absence of elastic fibers and smooth muscle cells. Patients presenting to primary care physician with lower respiratory tract symptoms like dyspnoea and wheezing are often diagnosed with COPD or asthma and started on treatment. There are uncommon conditions like Mounier-Kuhn syndrome (MKS) that can present with similar symptoms and it is important for the primary care physician to keep them in mind when evaluating the patients. It is important to identify MKS, as early recognition can lead to better management and prevention of complications. Very few cases of MKS with Bronchial Asthma have been reported so far. We present a case of 65 year old male who presented with intermittent episodes of cough with productive sputum, wheezing and breathing difficulty since last 35 years and was treated for Bronchial Asthma since last 25 years. He was later diagnosed with Mounier-Kuhn syndrome with Bronchial Asthma by clinical history supported by Radiological evidences.

**Keywords:** Mounier-Kuhn syndrome (MKS); Elastic fibers ;Tracheobronchomegaly ; Bronchial Asthma

### INTRODUCTION

Tracheobronchomegaly is a well-defined clinical and radiologic entity characterized by marked dilatation of the trachea and the central bronchi and frequently associated with recurrent lower respiratory tract infection.(1,2)The condition was first recognized at autopsy by Czyhlarz(1) 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.(2)Tracheobronchomegaly is believed to be extremely rare. To date, only few hundreds of cases have been reported in the literature. However, some patients with tracheobronchomegaly may be totally asymptomatic and are not detected, whereas those with symptoms frequently are overlooked if chest radiographs alone are used for diagnosis.

We report a 65 year old male who presented with intermittent symptoms of cough with expectoration, shortness of breath, wheezing since last 35 years.

### CASE REPORT:

A 65 year old male came to outpatient department of Department of Respiratory Medicine in SriAurobindo Medical College, Indore with complaints of fever since last 5 days and intermittent episodes of shortness of breath, cough with expectoration and wheezing since last 35 years. Shortness of breath has progressed from MMRC Grade 2 to MMRC Grade 3 in last 4 days. He was diagnosed with Bronchial Asthma with secondary infection on basis of typical history of atopy, seasonal exacerbations, positive family history and pulmonary function test which was done 8 years back which showed reversible airway obstruction(FEV1=51% , FEV1/FVC =58.84 with reversibility of 16%.

After it, he was started on regular medications with MDI and bronchodilators but was partially relieved. But since last 2 months his symptoms worsened. He never smoked and worked as a Farmer. The patient had a history of recurrent lower respiratory tract infections since early childhood.

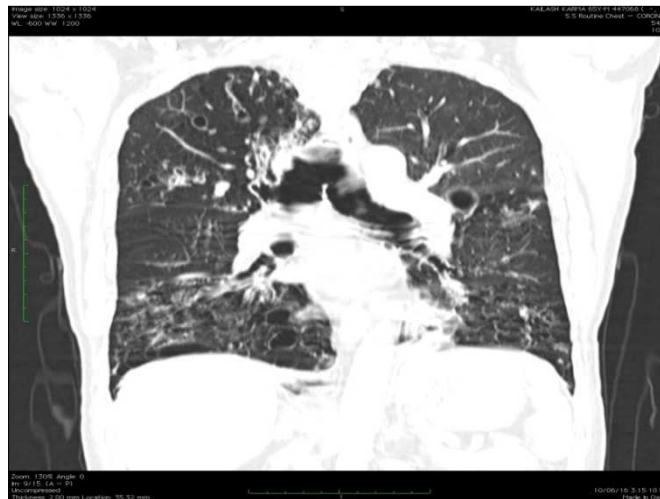
Physical examination revealed fever (99.8<sup>0</sup> F), Tachypnoea (24/min) ,Bilateral diffuse rhonchi and bilateral crepts (Right >Left).

The chest X-ray revealed a significant increase in the diameter of the trachea together with cystic lesions in left mid zone at hilar level(Fig.1)



**Fig.1.Chest X ray (PA view)**

CT Chest (Fig.2) shows dilated trachea (35cm) with dilated proximal bronchi(25 cm)and left main bronchus (24 cm) with mild proximal cylindrical bronchiectasis with wall thickening. Also there is widespread cystic bronchiectasis in both lungs sparing left upper lobe. Many of the cystic areas showed luminal air fluid levels. There were patchy areas of peribronchial nodularity in tree in bud pattern in both lungs around the areas of bronchiectasis.



**Fig.2(a)CT image of Coronal section of lung showing dilated trachea and Right and left main bronchus**



**Fig.2(b)CT Image showing bilateral cystic bronchiectasis**

His arterial blood gas study revealed mild hypoxemia with a Pao<sub>2</sub> of 70 mm Hg on room air. 2D Echo had very poor echo window and mild PAH (PASP=34mm Hg). His sputum culture sensitivity showed growth of *Pseudomonas aeruginosa* sensitive to Aminoglycosides and Penicillin group of drugs. Pulmonary Function Test revealed (FEV<sub>1</sub>=40.1 ; FEV<sub>1</sub>/FVC=50.30) with reversibility of 2% with interpretation of severe large and small airway obstruction with poor reversibility. Serum IgE levels were normal and skin prick test for *Aspergillus* was negative. Finally a diagnosis of Bronchial Asthma with Mounier-Kuhn syndrome with secondary infection was made on Clinical and Radiological evidences. He was treated with Chest physiotherapy ,antibiotics and supportive treatment to which he responded well .

### DISCUSSION

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.[1]TBM has been described by a variety of names, including Mounier–Kuhn syndrome, tracheal diverticulosis, tracheobronchiectasis, tracheocele, tracheomalacia, and tracheobronchopathiamalacia[1,2,3].The common denominator is the dilatation of trachea and central bronchi with an abrupt transition to a normal caliber of the peripheral airways.[4]

For an adult, an increased diameter of the trachea, the right and the left main bronchus that exceeds 3.0, 2.4, and 2.3 cm, respectively, on a standard chest radiograph or bronchogram is diagnostic for tracheobronchomegaly because these are the upper limits of the means plus three standard deviations.[5]

It is more common in men and is typically diagnosed in the 3rd or 4th decades of life. The clinical presentation varies widely, from minimal disease in which lung function is preserved to severe respiratory failure and death. Involvement occurs at different levels, from the trachea down to the 4th bronchial branch[4,6]. Although its cause is not fully known, tracheobronchomegaly is reportedly linked to familial susceptibility and is perhaps inherited through an autosomal recessive mechanism. Cases are often sporadic.[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.[6] The main problems associated with this disease are ineffective cough consequent to pathologic dilation in the tracheobronchial tree and the impairment of mucociliary activity. These cause difficulty in expectorating secretions and lead to recurrent LRTIs.[7] The symptoms of Mounier-Kuhn syndrome are nonspecific. In the absence of infection, the disease can develop asymptotically. Bronchiectasis and LRTIs are clinically prominent, and recurrent pneumonia and fibrosis can develop.[4,6]

Connective-tissue diseases, Ataxia-telangiectasia, Ankylosing spondylitis, Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome, and cutis laxa [elastolysis] are also associated with secondary tracheobronchial enlargement.[8,9] All of these conditions should be considered in the differential diagnosis.

The described case report offers a rare insight into the natural course of the progression of an undiagnosed Mounier-Kuhn Syndrome, with an evaluation of the changes in imaging and pulmonary functions over time. Advanced cases of MKS have a very poor prognosis and are usually noted on autopsy. These patients tend to have severe recalcitrant mucus plugs, tracheal diverticuli and fibrotic changes which eventually lead to respiratory failure and their demise. Tracheal stenting has proved beneficial in patients with multiple tracheal diverticuli.[10,11]. Since MKS involves diffuse areas of the airway, surgery on selective parts of the bronchial tree is very difficult. However a few rare cases of lung transplantation have been reported in the literature with no proven benefit in mortality.[12,13]

This case supports early diagnosis in patients with Mounier-Kuhn Syndrome, so that a decline in respiratory function resulting from possible complications of Mounier-Kuhn Syndrome may be prevented.

### CONCLUSION

Known cases of asthma are always overlooked for the associated diseases . This case report enlightens the necessity to look for an alternative diagnosis or associated disease in all those asthma cases who respond poorly to treatment so that associated disease could be identified at an early stage and its progression could be prevented. However more work has to be done in terms of treatment and follow up.

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