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Pulmonology

A Case of Mounier Kuhn Syndrome with Bronchial Asthma

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Abstract **Case Report**

Tracheobronchomegaly (Mounier–Kuhn syndrome) is a rare disease characterized by dilatation of the trachea and major bronchi because of atrophy or absence of elastic fibers and smooth muscle cells. The clinical signs are diverse and not very specific. Imaging provides a positive diagnosis. We present a case of 40 year old male who presented with intermittent episodes of cough with productive sputum, wheezing and breathing difficulty since last 35 years and he 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; Bronchial asthma; muscle cells.

INTRODUCTION

Mounier Kuhn syndrome (MKS) or Tracheobronchomegaly, first described in 1932, is defined as a characteristic clinico-radiological condition consisting of marked dilatation of the trachea and main bronchi, and frequently associated with reccurent lower respiratory tract infection [1]. MKS is considered to be extremely rare nonetheless, some patients may be totally asymptomatic and are not diagnosed, and those with symptoms frequently are overlooked if chest radiographs alone are used for diagnosis.The clinical signs are diverse and not very specific. Imaging provides a positive diagnosis [2].

CASE REPORT

We report the case of a 40 years old non smoker male patient, married with two children, who was exposed to silica dust and had a family history of atopy. He was referred to the allergology consultation of our Department, for the management of his asthma, revealed by a symptomatology dating back to the age of 34 years old, made of a predominantly nocturnal wheezing dyspnea, chest tightness, respiratory discomfort and a chronic cough, with seasonal exacerbations. His asthma was classified as severe, persistent with frequent exacerbations despite a combination of high dose ICS, LABA, anticholinergic and antileukotriene.

The physical examination revealed a saturation at 94% and bilateral diffuse sibilant and ronchi. The pulmonary function test showed a severe airway obstruction (FEV1 : 19%) with reversibility of 23%.

The chest X-ray showed a significant increase in the diameter of the trachea with bilateral diffuse cystic lesions.

A chest CT showed a dilated trachea and the two main bronchus, measuring respectively 30mm, 27 and 24mm of maximal diameters, with undulated wall and individualization of partial diverticula more marked posteriorly. Bilateral and diffuse cylindrical, monoliform and cystic bonchectasias involving lobar, segmental and sub-segmental bronchi.

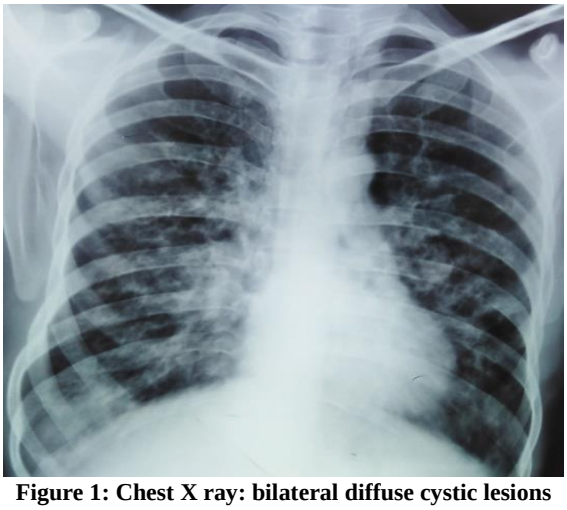

Figure 1: Chest X ray: bilateral diffuse cystic lesions

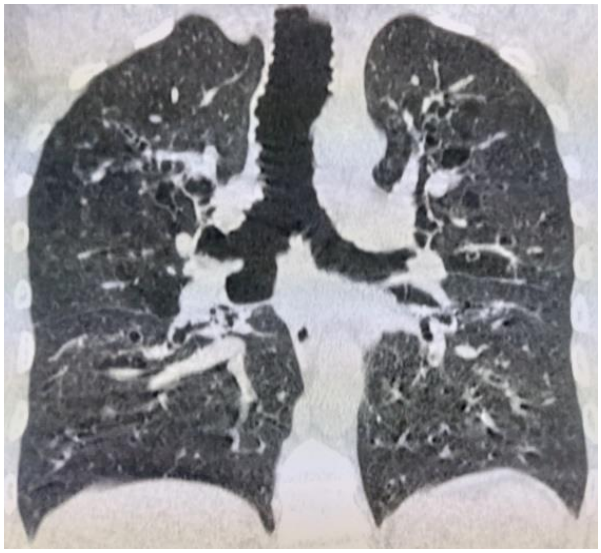

Figure 2: CT image of Coronal section of lung showing dilated trachea and Right and left main bronchus

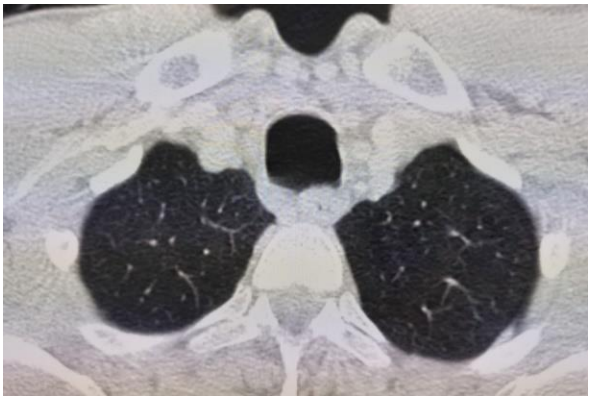

Figure 3: CT image of transverse section of lung showing dilated trachea

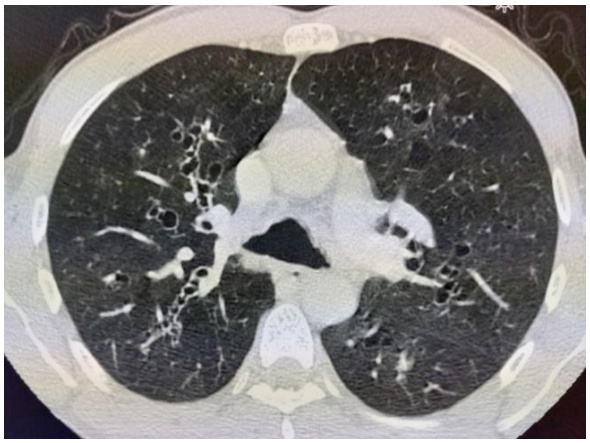

Figure 4: CT Image showing bilateral bronchiectasis

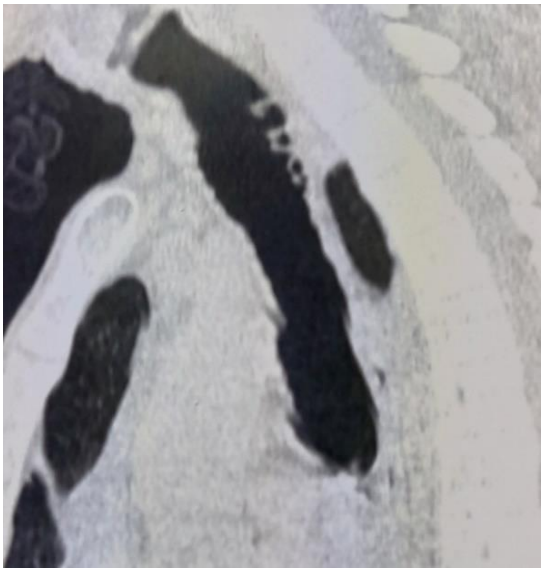

Figure 5: CT Image showing multiple small diverticula along the posterior tracheal and wall

DISCUSSION

MKS is a rare disorder of unknown physiopathology while autopsy studies suggested a congenital atrophy of the elastic and smooth muscle tissues of the trachea and main bronchi, resulting in dilatation [3]. Some authors require the elimination of any condition that may cause acquired MKS before retaining the diagnosis, including pulmonary fibrosis, mechanical ventilation, especially in newborn premature infants, and radiation therapy [1].

It is a rare pathology with an estimated prevalence between 0.4 and 1.6 % of patients with respiratory symptoms [4, 5], for most patients the diagnosis is not made until after the third decade of life [1]. As is the case of our patient.

Most patients appear to be smokers, even if nonsmokers have been reported [6]. Our patient has never smoked but was exposed to wood smoke.

Spirometry reveals varying degrees of obstruction and increased residual capacity [7], in somes cases lung function was normal [1]. Our patient had a severe airway obctruction with reversibility of 23%.

MKS can be seen on standard chest radiographs [6] where the size of the trachea may exceed the width of the spine, but it is common to not to be seen at all until a CT scan is performed [8]. CT is the golden standard for confirming the diagnosis. The chest X-ray of our patient showed a significant increase in the diameter of the trachea with bilateral diffuse cystic lesions.

On bronchoscopy, the increased tracheal diameter and expiratory collapse due to tracheomalacia are visible and the redundant tracheal wall may even obstruct the view [9].

Differentes suggestions had been made to classify the cases of MKS:
Based on anatomical appearance [1]
Type I: shows diffuse symmetrical enlargement of both trachea and bronchi.
Type II: the enlargement is more eccentric with pronounced diverticula and an abrupt change to normal bronchial size.
Type III: where the diverticula may extend to more distal bronchi.

Our patient is type I.
Based on clinical presentations [10]:
Type 1A : Infants who have underwent fetal endoscopic tracheal occlusion as a therapy for prenatally diagnosed severe congenital diaphragmatic hernia.
Type 1B : infants who had prolonged intubation.
Type 2A : Patients who develop MKS after multiple pulmonary infections
Type 2B : Patients who develop MKS after being diagnosed with pulmonary fibrosis
Type 3 : Patients with evidence of extra-pulmonary elastolysis
Type 4 : Patients with no clear predisposing factors

Our patient is type 4

The variability of the disease is best represented by two cases of long-term follow-up. One patient suffered from recurrent febrile respiratory infections but maintained normal lung function, while in the other patient fever was infrequent but lung function steadily decreased reaching severe obstruction [1].

Bullous emphysema is common in MKS and can cause pneumothorax [11]. Bronchiectasis is a common co-existence, affecting 45 % non-Type 1 TBM patients [10]. As is the case of our patient.

The treatment of MKS is mainly symptomatic, which includes vaccinations, treatment of infections, as well as elimination of secretions with mucolytic treatment and physical therapy [17].

For patients with tracheobronchomalacia, noninvasive continuous positive airway pressure ventilation [12], airway stenting, surgical tracheoplasty [13, 14], and laser treatment [18] have all been tried. A double lung transplant was performed in the terminal phase [19].

Our patient was put on step five of GINA treatment recommendations, physiotherapy, vaccination, and follow-up consultations were programmed.

CONCLUSION

This case highlights the importance of underlying comorbidities in patients with asthma who respond poorly to treatment. It also emphasizes the importance of early diagnosis of MKS to preserve lung function.

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