CASE REPORT

TRACHEOBRONCHOMEGALY: A RARE CAUSE OF BILATERAL BRONCHIECTASIS

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ABSTRACT

Mounier-Kuhn syndrome, also called tracheobronchomegaly, is a very rare congenital disorder of the lung primarily characterized by an abnormal widening of the upper airways. The abnormally widened trachea and mainstem bronchi are associated with recurrent lower respiratory tract infection and copious purulent sputum production, eventually leading to bronchiectasis and other respiratory complications. Here we report a rare case of 55 year old nonsmoker male who presented with recurrent lower respiratory tract infection and bronchiectasis on chest xray, ultimately diagnosed to have bilateral bronchiectasis due to tracheobronchomegaly.

Keywords: Tracheobronchomegaly, Mounier Kuhn syndrome, Bronchiectasis

INTRODUCTION

Mounier Kuhn syndrome, or tracheobronchomegaly, is a rare clinical and radiological condition. It is characterized by distinct dilatation of the trachea and bronchi and by recurrent lower respiratory tract infections (LRTIs).[1-4] The syndrome was first described by Mounier-Kuhn in 1932, and fewer than 100 cases had been reported in the medical literature upto 1994 [4]. Recently more cases are being reported because of the wide availability of the CT scans. The cause of the condition is not clearly understood; however, in biopsy studies, congenital atrophy has been observed in the smooth muscle and elastic tissue of the trachea and main bronchi. Bronchial and tracheal diverticula can also accompany tracheobronchomegaly.[1-4] Normal tracheal inner diameter is approximately 25mm in males and 21mm in females. A diameter of more than 30mm is diagnostic of tracheobronchomegaly which is usually measured 2cm above the aortic arch.

CASE REPORT

A 55 years old male presented with complaints of recurrent cough with mucoid expectoration and breathlessness (MMRC GRADE 2) since 15-20 years which used to get aggravated on exertion and with environmental changes and was non progressive in nature.

Patient was taking inhaled formoterol fumarate and fluticasone propionate in DPI form since 3 years.There was no history of orthopnea, paroxysmal nocturnal dyspnoea, hemoptysis, tuberculosis or ATT intake in past. Patient was chronic alcoholic for 20 years but had stopped alcohol consumption for last 10 years.

On examination, patient was overweight with a BMI of 25.1 kg/m^2. Patient was afebrile and there was no pallor, icterus, cyanosis, lymphadenopathy, pedal oedema. Grade I clubbing was present. His pulse was 94/min, regular, normo-volumic & peripheral pulses were felt bilaterally. Respiratory rate was 26/min, thoracoabdominal, blood pressure was 110/74 mm Hg in right arm in supine position and oxygen saturation was 98% on room air.

On respiratory system examination upper respiratory tract showed a congested throat with deviated nasal septum to right side and inferior turbinate hypertrophy. In lower respiratory tract examination, normal vesicular breath sounds were heard in all lung fields along with bilateral extensive crepitations and occasional polyphonic rhonchi.

Chest xray PA view **(Fig 1)** was suggestive of bilateral cystic changes more in the right mid and lower zone with patchy areas of consolidation in right mid zone.

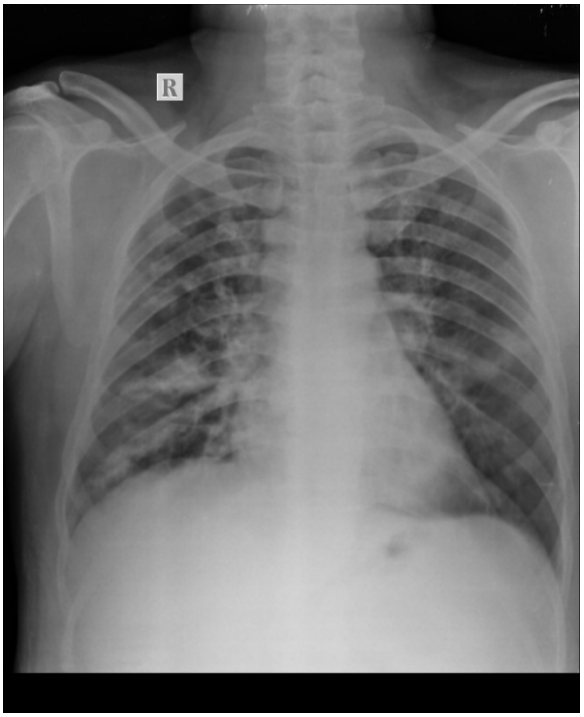

Figure 1: Chest xray showing bilateral bronchiectasis more in right mid and lower zone.

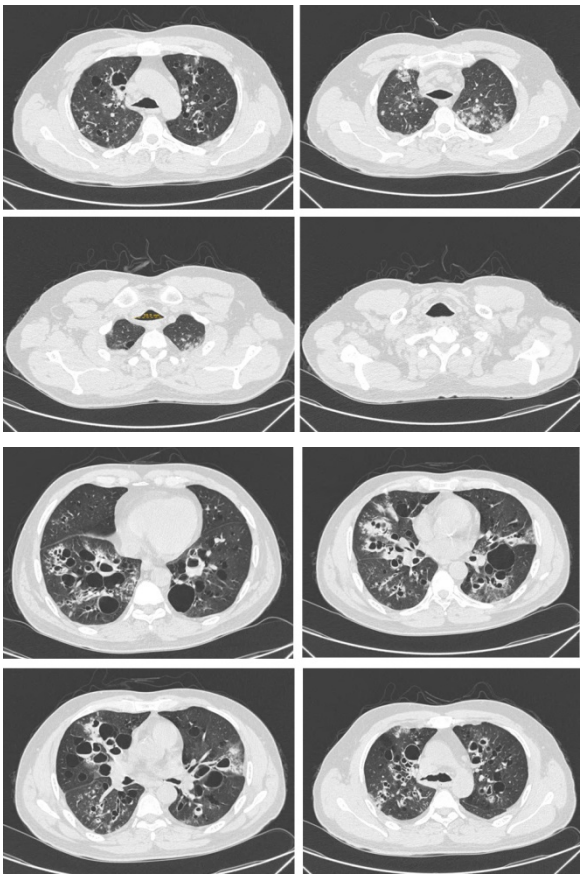

Figure 2a and 2b: HRCT image showing cystic bronchiectasis with tracheobronchomegaly.

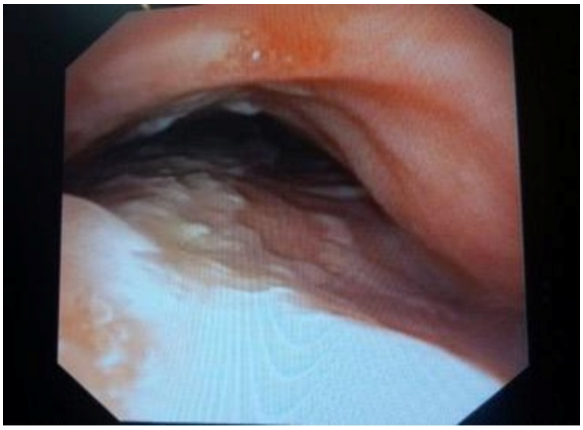

Figure 3: Bronchoscopy image showing increased tracheal diameter

His lab investigations showed Haemoglobin 11.2gm%, WBC 17100/cmm with 78% neutrophils, normal platelet counts & ESR 67mm/hr. Liver & kidney function tests were normal along with normal blood sugar & ELISA test for HIV was nonreactive. Absolute eosinophils count was 171/cmm. Arterial blood gas level was also normal with PH 7.38, HCO3 24meq/l, PAO2 92 mmhg, PACO2 42 mmhg. Pulmonary function testing revealed a forced expiratory volume in 1 sec (FEV1) of 2.03 L (58%), a forced vital capacity (FVC) of 2.39 L (55%), and FEV1/FVC of 68%.

HRCT THORAX **(Fig 2a & 2b)** showed multiple well defined cystic lesions with air fluid levels noted in few of the cysts, bronchial wall thickening, multiple ill defined opacities with surrounding ground glass opacities s/o cystic bronchiectasis with infective infiltration. The tracheal diameter measured was 36.9 mm on CT scan. Further investigations showed serum ANA negative, Mantoux test non reactive, thyroid stimulating hormone 1.20, C Reactive Protein 59.1, normal USG abdomen & pelvis and bone marrow studies.

Sputum cytology showed inflammatory smear with polymorphonuclear cells, macrophages and cell debris. No malignant cells seen. Sputum for AFB bacilli was negative Sputum culture and sensitivity showed no growth. Flexible fiberoptic bronchoscopy showed (**Fig 3**) mobile vocal cords, oedematous and hyperemic tracheal mucosa. There was increase in transverse diameter and reduction in anteroposterior diameter of trachea. The posterior membranous wall was excessively bulging into the tracheal lumen. The carina was sharp and there was no other obvious abnormality. Thick secretions present in bilateral lower lobe bronchi were aspirated into the mucous trap which showed

growth of Pseudomonas aeruginosa species on culture.

Final diagnosis of Tracheobronchomegaly causing bilateral bronchiectasis was made.

DISCUSSION

Mounier-Kuhn syndrome (also called T**racheobronchomegaly**) is a very rare congenital disorder of the lung primarily characterized by an abnormal widening of the upper airways.[10] The abnormally widened trachea and mainstem bronchi are associated with recurrent lower respiratory tract infection and copious purulent sputum production, eventually leading to bronchiectasis and other respiratory complications.

The underlying abnormality is an absence or marked atrophy of the elastic fibres and smooth muscle within the wall of the trachea and main bronchi, leading to sacculations and the formation of diverticula between the cartilaginous rings.[5,6] Although this is thought to be congenital, there is no universal agreement. Most cases present in the third or later decades with recurrent respiratory tract infections. Most cases, however, are sporadic and show no evidence of associated connective tissue disease,[8,9] as was the case in our patient also.

As a result of the flaccidity of the wall of the respiratory tree there is significant change in airway size during the different phases of respiration. During inspiration negative intra-thoracic pressure develops leading to marked enlargement of the trachea. However, as expiration commences, the reversed intra-thoracic pressure causes collapse of trachea. In addition to this dynamic change, bronchial or tracheal diverticulosis are common, as is bronchiectasis.[2] There is also an absence of the myenteric plexus of the bronchial tree.

The most sensitive imaging test is a biphasic CT with images of the trachea obtained during inspiration and expiration. Two-phase chest radiographs will also demonstrate the enlargement of the trachea on inspiration and collapse during expiration, but they are clearly less sensitive.

To consider the diagnosis, the diameter of the trachea should be greater than 30 mm: this is usually measured 20 mm above the aortic arch [1-2]. Other measurements that have been used to make the diagnosis include bronchial diameters of 20 or 24 mm (right) and 15 or 23 mm (left) have also been used. Posteriorly projecting tracheal diverticulae may also be seen.

Treatment is usually conservative with physiotherapy and postural drainage. Acute exacerbations are treated with antibiotics.

With the advent of HRCT and bronchoscopy, reporting of such cases is on increase. Singh m et al recently reported a similar case with a tracheal diameter of 45.5 mm[11]. Krustins E did a systematic analysis of 128 cases published from year 1987 to 2013. 8:1 male predominance was found in 89 identified reports (128 cases). Mean age was 53.9 years, and average tracheal diameter was 36.1 mm. No correlation between increasing age and increasing tracheal diameter was found. Bronchiectasis, tracheal diverticulosis and tracheobronchial dyskinesia were common (49.2%, 33.6% and 28.9%, respectively). Cough, dyspnea and recurrent respiratory infections (71.1%, 51.6% and 50.8%, respectively) were the most common complaints[12].

CONCLUSION

It is recommended that when no other etiologies are identified in bronchiectasis, we should carefully look for tracheal diameter in HRCT and bronchoscopy as Tracheobronchomegaly can be the cause of bilateral bronchiectasis.

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