RARE ASSOCIATION OF MOUNIER-KUHN SYNDROME WITH PANLOBULAR EMPHYSEMA

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HOW TO CITE THIS ARTICLE: Suhas HS, Utpat K. Rare association of Mounier-Kuhn syndrome with panlobular emphysema. J. Evid. Based Med. Healthc. 2017; 4(52), 3212-3214. DOI: 10.18410/jebmh/2017/637

PRESENTATION OF CASE

Mounier-Kuhn Syndrome (MKS) is an unwonted disorder of obscure aetiology signalised by constellation of marked tracheal dilatation, tracheal diverticulum and associated bronchiectasis. Its pathogenesis is postulated to be enrooted in aberrant connective tissue proliferation. Chronic cough with sputum production, recurrent lower respiratory tract infections and spontaneous pneumothoraces trademark the clinical picture. Bronchiectasis is the customary pattern of lung involvement. However, pulmonary fibrosis and emphysema could also could be encountered infrequently. We hereby report a divergent presentation of this syndrome with exhibition of a combination of bronchiectasis and emphysema.

Keywords- Mounier-Kuhn Syndrome, Emphysema, Tracheobronchomegaly.

A 30-year-old man non-addict was symptomatic with complaints of cough with expectoration copious in amount with postural variation and dyspnoea on exertion since childhood with history of infective exacerbations 2 to 3 per year. There was past history of empirical antituberculous therapy intake for 3 months. There were no history of similar complaints in family. There was no history suggestive of malabsorption or history of consanguineous marriage in parents. On general examination, grade 2 clubbing was seen. There were no other features suggestive of connective tissue disorder. On respiratory system examination, there was presence of bilateral coarse crackles. Examination of other systems was within normal limits.

Chest x-ray was suggestive of bilateral cystic opacities with hyperlucent area in left lower zone (Figure 1). On HRCT (high resolution computerised tomography of thorax), thorax examination revealed the presence of bilateral cystic bronchiectasis with air fluid levels within and tracheobronchomegaly with diameter of trachea, right main bronchus and left main bronchus being 32.3 mm, 21.9 mm and 18.1 mm, respectively. There was presence of diffuse centrilobular emphysematous changes in both lungs along with presence of panlobular emphysema seen in left lower lobe. Expiratory scans revealed bilateral patchy air trapping (Figure 2a, 2b, 2c). Bronchoscopy was suggestive of tracheal diverticulum with increased collapsibility of the airway (Figure 3). Spirometry was suggestive of obstructive abnormality with poor bronchodilator reversibility. Alpha-1 antitrypsin was 240 mg/dL (100-300 mg/dL). Patient was managed with adequate chest physiotherapy for postural drainage of secretions and initiated on inhalers for airway involvement.

Financial or Other, Competing Interest: None.
Submission 14-06-2017, Peer Review 22-06-2017,
Acceptance 28-06-2017, Published 29-06-2017.
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DOI: 10.18410/jebmh/2017/637

DIFFERENTIAL DIAGNOSIS

Ehlers-Danlos syndrome, Marfan's syndrome and alpha-1 antitrypsin deficiency.

CLINICAL DIAGNOSIS

Bilateral bronchiectasis secondary to Mounier-Kuhn syndrome with panlobular emphysema.

PATHOLOGICAL DISCUSSION

Tracheobronchomegaly (TBM) commonly known Mounier-Kuhn Syndrome (MKS) is a condition where there is profound tracheobronchial dilatation.[1,2] It is caused due to atrophy of the muscular and elastic tissues in the trachea and main bronchial wall. Clinical manifestations can vary from being asymptomatic to often a very severe presentation in the form of respiratory failure. It is often associated with recurrent episodes of respiratory tract infection, which results in destruction of underlying cartilage leading to tracheal diverticulosis and bronchiectasis. It is more commonly seen in men and is usually diagnosed in the 3rd or 4th decades of life. Imaging features are diagnostic in MKS. Computerised tomography of thorax helps in direct measurement of the tracheobronchial tree at multiple levels and also to demonstrate secondary features including diverticulum, sacculation, bronchiectasis and parenchymal scarring. In adults, the accepted criteria for diagnosis of tracheobronchomegaly on CT are diameters of the trachea, right main bronchus and left main bronchus of >30 mm, 20 mm and 18 mm, respectively.[3] Hallmark features of MKS include a combination of upper airway manifestations in the form of tracheobronchomegaly and tracheal diverticulum and lower respiratory tract and lung parenchymal manifestations in the form of bronchiectasis and rarely lung fibrosis and emphysema. There may be a permutation and combination of these components in individual patients. Our patient was, however, unique due to the fact that all these manifestations were present synchronously in our patient.

Association of tracheobronchomegaly with emphysema has been reported only sporadically in literature.[4] There are meagre differentials, which can be considered in these

scenarios, namely connective tissue disorders like Ehlers-Danlos syndrome, Marfan's syndrome and alpha-1 antitrypsin deficiency. Once all these causes of emphysema are excluded, then it can be said to be secondary to MKS. It has been postulated that due to repeated infective exacerbations in this entity leads to inflammatory activity that is responsible for causing destruction of underlying cartilage and the attachments of alveoli leading to development of emphysema. Therapy involves respiratory physiotherapy for clearing secretions and antibiotic use during infectious exacerbations. Tracheal stenting has been helpful in severe cases, however, surgery is rarely performed, because of the diffuse nature of the disease.[5,6] Thus, our case was a noteworthy example of a rarer manifestation of an already rare syndrome. Clinicians need to maintain a high index of suspicion for this syndrome whilst dealing with a case of emphysema occurring in a young patient without any other predisposing causes.

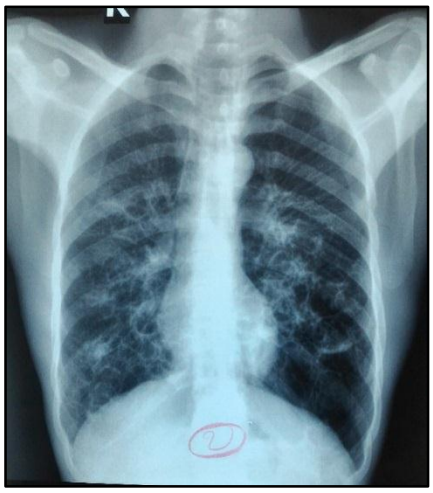

Figure 1. Chest x-ray Showing the Presence of Bilateral Cystic Opacities with Hyperluscent Area in Left Lower Zone

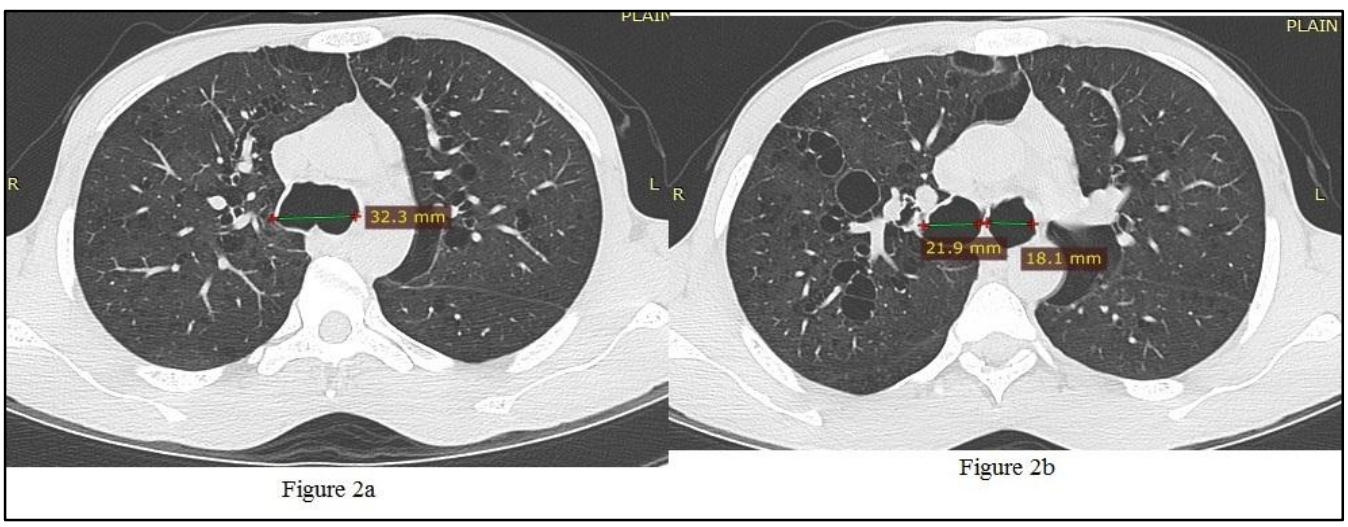

Figure 2a, 2b. HRCT Showing the Presence of Bilateral Cystic Bronchiectasis with Air Fluid Levels within and Tracheobronchomegaly with Presence of Diffuse Centrlobular and Panlobular Emphysema.

Thick MIP Images

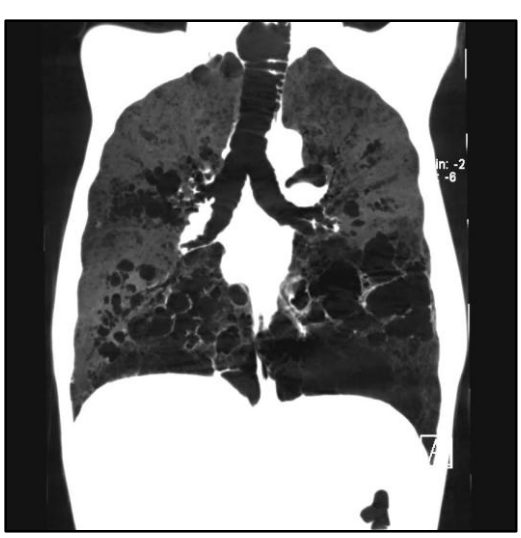

Figure 2c. Thick Mip Images Showing Better Delineation of Tracheobronchomegaly

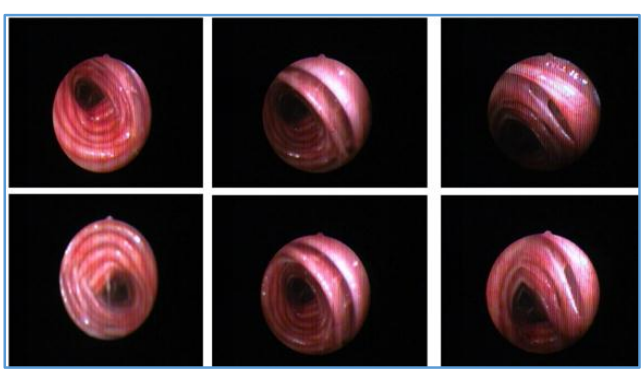

Figure 3. Bronchoscopy Showing Tracheal Diverticuli with Increased Collapsibility of the Airway

Final Diagnosis

Mounier-Kuhn syndrome with panlobular emphysema.

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