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Case Report: Mounier-Kuhn Syndrome

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Abstract: Mounier-Kuhn syndrome also known as Tracheobronchomegaly is a rare disorder of the respiratory airways characterized by marked enlargement of the trachea and main bronchi, bronchiectasis, and recurrent respiratory tract infections. The cause of this disorder is uncertain and the clinical presentation is diverse. It's usually diagnosed on the basis of the characteristic CT scan findings. We are reporting a case in a 29-year-old man presenting with productive cough with copious greenish sputum of no bad odor for the last month.
Keywords: Mounier-Kuhn syndrome, tracheo bronchomegaly, bronchiectasis.

INTRODUCTION

The Mounier-Kuhn syndrome is a rare clinical and radiological entity described by Mounier and Kuhn for the first time in 1932 [1].The syndrome is characterized by marked tracheobronchial dilatation. Mostly presents in the third or later decades with recurrent respiratory tract infections. The cause is uncertain, but it is believed to be due to the lack of smooth muscle and elastic connective tissue in the trachea and main bronchi [2].

Lung diseases which cause severe fibrosis of the upper lobes may also exert tracheal traction and result in tracheal enlargement. Secondary tracheobronchial enlargement may occur in certain other conditions such as Marfan syndrome, Ehlers-Danlos syndrome, Kenny-Caffey syndrome, ataxia-telangiectasia, connective tissue diseases, Brachmann-de Lange syndrome, Bruton-type agammaglobulinemia, ankylosing spondylitis, cutis laxa, and light chain deposition disease [3] .Most cases, however, are sporadic and show no evidence of associated connective tissue disease [4].

On CT scan, the diagnosis is made when the transverse diameter of the trachea measures greater than 3 cm and that of the right and left main bronchi exceeds 2.4 cm and 2.3 cm, respectively. Apart from the tracheobronchial enlargement, diverticula are also seen between the cartilaginous rings [5].

CASE REPORT

A 29-year-old man presented to Shaab Cardiothoracic Center in Khartoum, Sudan with productive cough with copious greenish sputum of no bad odor for the last month. He has history of recurrent chest infections since late childhood. Presenting with productive cough, low grade fever, malaise, poor appetite and one kilogram weight loss in a month. He has mild exertional dyspnoea and some times audible wheezes, No chest pain and no palpitations. There were no clinical, radiological or laboratory evidences for any secondary cause of tracheobronchial enlargement.

He was quite normal in between these attacks. The patient wasn't smoker and there was no family history of a similar condition. His Chest X-Ray revealed tracheobronchial enlargement and bilateral bronchiectasis and tracheal diverticulosis (Fig-1A and B).

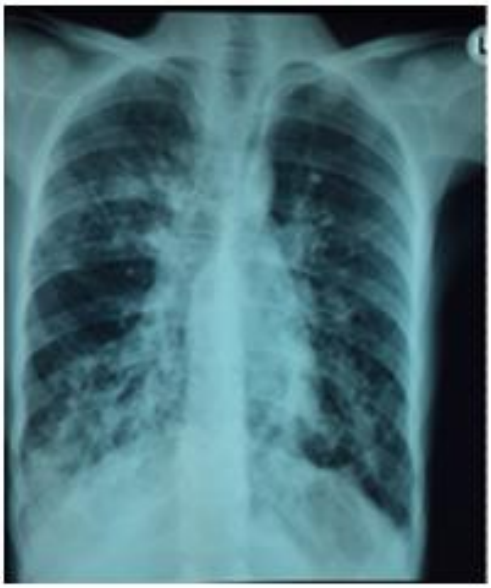

Fig-1 A

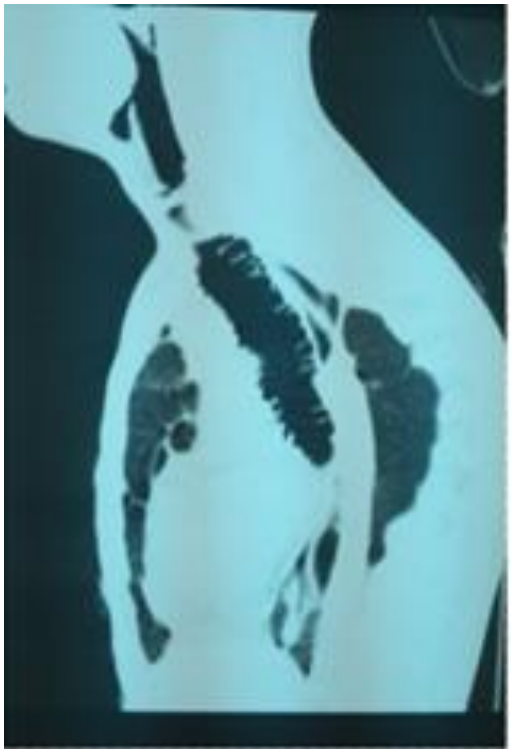

Fig-1 B

CT scan of the chest (Figure 2) showed tracheo bronchomegaly and cystic bronchiectasis was seen in the lung parenchyma bilaterally. The trachea was markedly dilated, with a diameter of 41.5 mm (Figure 2), while the right and left main bronchi had diameters of 39.6 and 26.4 mm respectively (Figure 3).

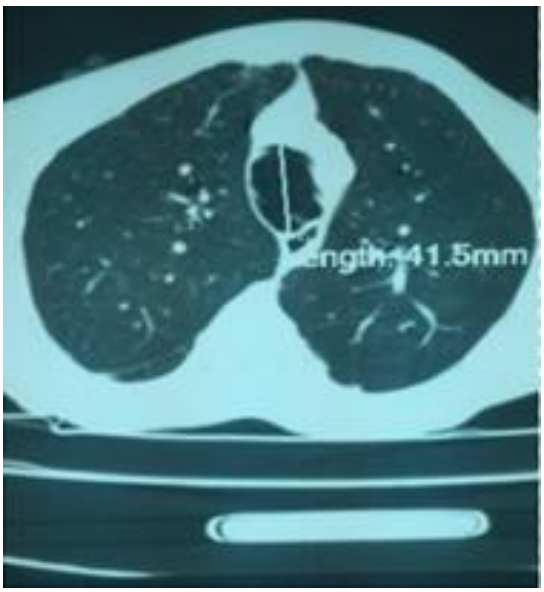

Fig-2

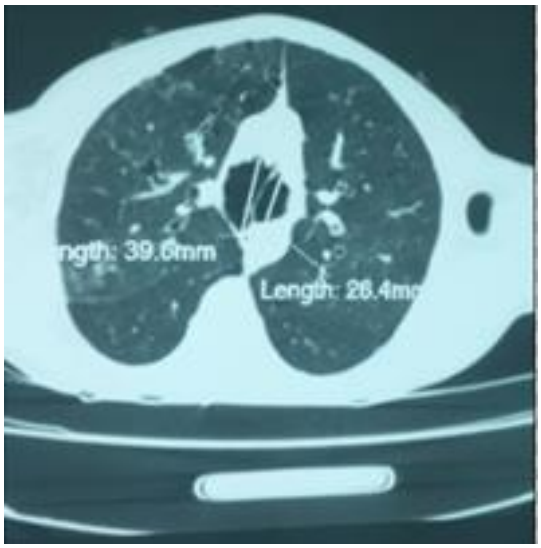

Fig-3

Fiberoptic bronchoscope revealed a dilated trachea and main bronchi and collapse of trachea with coughing (Figure 4).Histopathology of bronchial biopsy showed thin basement membrane with muscle fibrosis (Figure 5).

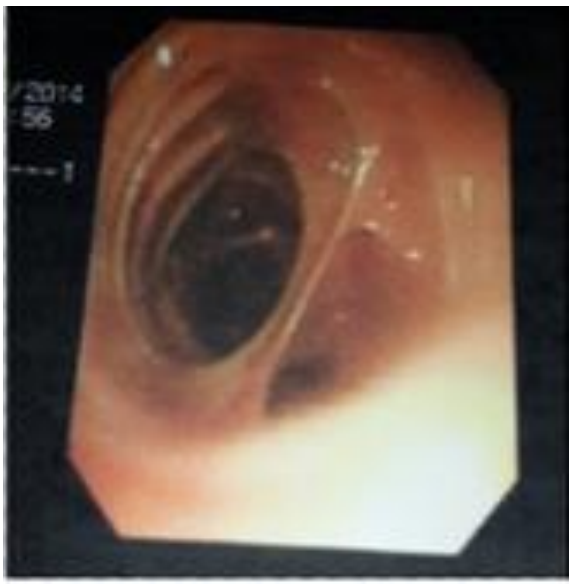

Fig-4

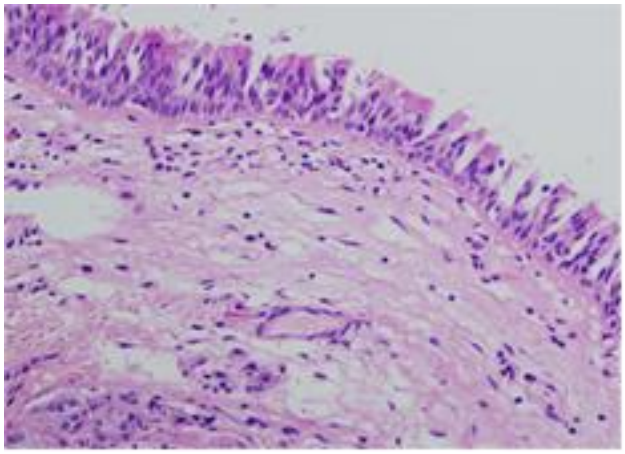

Fig-5

DISCUSSION

A 29-year-old man presented with one month productive cough, low grade fever, malaise, poor appetite, weight loss, mild exertional dyspnoea and audible wheezes, no chest pain or palpitations. He has history of recurrent chest infections since late childhood. There were no evidences for any secondary cause of tracheobronchial enlargement. He was quite normal in between these attacks. The patient wasn't smoker and no family history of a similar condition. Chest X-Ray and CT revealed tracheobronchial enlargement, bilateral bronchiectasis and tracheal diverticulosis. In CT the trachea and main bronchi were markedly dilated. Fiberoptic bronchoscope showed a dilated trachea and main bronchi. Histopathology showed thin basement membrane with muscle fibrosis. Diagnosis can be made by measuring the tracheal diameter, using only data from chest X-rays, in which the trachea can seen in profile and thus the diameter determined. However, CT of the chest, however, makes this measurement more accurate. Treatment of Mounier-Kuhn syndrome comprises physiotherapy to assist in clearing secretions and appropriate antibiotics during infective flare-ups. Severe cases have been reported that involved the use of stenting of the airways. Surgical resection is of little benefit because the disease process is rarely confined to one anatomic area [6]. Inhaled bronchodilators and corticosteroids also are ineffective for the treatment of this syndrome [7].

CONCLUSION

As tracheo bronchomegaly can be overlooked on plain films, patients who have chronic respiratory infections should have a CT scan done in to rule out underlying predisposing conditions such as this.

REFERENCES

1. Mounier-Kuhn P; Dilatation de la trachea: Constatations radio-graphiques et bronchoscopiques. Lyon Medical, 1932; 150:106–109.
2. Aaby GV, Blake HA; Tracheo bronchomegaly. Ann Thorac Surg, 1966; 2:64–70.
3. Karalezli A, Yurdakul M, Misirli F; Mounier-Kuhn syndrome: a case report (tracheobronchomegaly) [in Turkish]. Turkiye Klinikleri J Med Sci, 1999; 19(1):37–39.
4. Menon B, Aggarwal B, Iqbal A; Mounier-Kuhn syndrome: report of 8 cases of tracheo bronchomegaly with associated complications. South Med J, 2008; 101(1):83–87.
5. Dunne MG, Reiner B; CT features of tracheo bronchomegaly. J Comput Assist Tomogr, 1988; 12(3):388–391.
6. Mostafa G, Mohammadreza P, Jafar AZ; Noe. Mounier Kuhn syndrome: a rare cause of severe bronchial dilatation with normal pulmonary function test: a case report. Respiratory Medicine, 2007; 1836–1839.
7. Ghanei M, Peyman M, Aslani J, Zamel N; Mounier-Kuhn syndrome: a rare cause of severe bronchial dilatation with normal pulmonary function test: a case report. Respir Med, 2007; 101(8):1836–1839.