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CASE REPORT

A Case of Mounier-Kuhn Syndrome

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ABSTRACT

Mounier-Kuhn syndrome is an uncommon respiratory syndrome characterised by dilatation of tracheobronchial pathway. We report the case of a 25-year-old man who had history of recurrent chest infection in form of cough and fever since childhood. Computed tomography of chest revealed tracheal dilation and hence diagnosis of this rare syndrome was made.

INTRODUCTION

Mounier-Kuhn syndrome, or tracheobronchomegaly, is a rare disorder characterized by recurrent lower respiratory tract infections due marked dilatation of tracheobronchial tree causing. The diagnosis can be done with the use of computed tomography (CT) of thorax. Its clinical features varies from being asymptomatic to severe respiratory distress. However therapy remains conservative.

CASE

Twenty five year old man presented in pre-anaesthetic clinic planned for operation for uncomplicated inguinal hernia with complaints of recurrent lower respiratory tract infections since childhood, with each episodes consisting of fever and cough. He used to take medication in the form of antibiotics but after being asymptomatic for few days he again used to have same complaints. He was a non smoker and family history was not contributory. The X-ray of the chest showed enlargement of the trachea and bronchi and bilateral bronchiectasis.

Contrast enhanced CT (CECT) of the thorax was done which revealed dilatation of the entire trachea and both main bronchi. The internal diameters of trachea, right main bronchus and left main bronchus were measured to be 37 mm, 26 mm and 35 mm respectively. There were multiple posterior diverticula arising from inter-cartilaginous regions. The lung window also revealed scattered areas of cylindrical bronchiectasis, mucus plugging with centrilobular nodules

in both lungs. (Fig. 1A, B,C). Patient did not gave consent for bronchoscopy and did not turned up for pulmonary function test.

The diagnosis of mounier Kuhn syndrome was hence made.

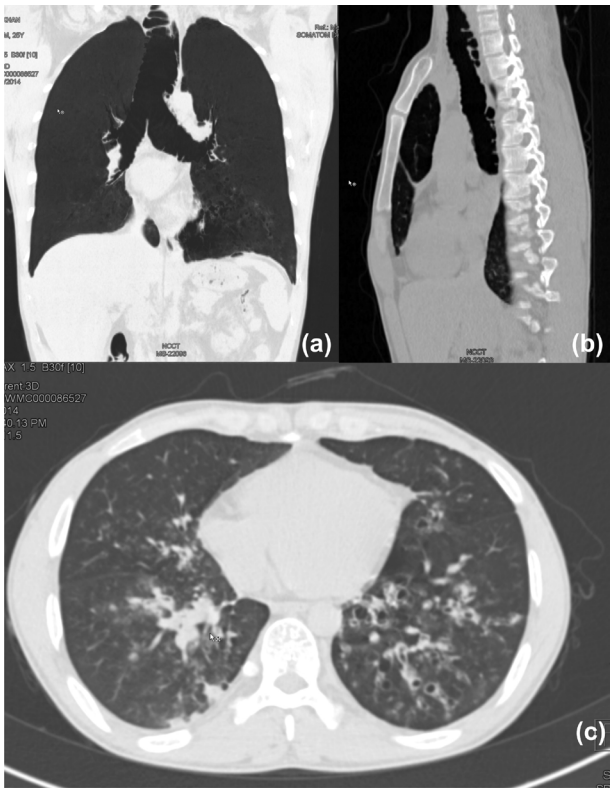

Fig. 1 (a,b,c): CECT thorax revealed dilatation of the entire trachea and both main bronchi with the internal diameters of trachea, right main bronchus and left main bronchus to be 37 mm, 26 mm and 35 mm respectively. There were multiple posterior diverticula arising from inter-cartilaginous regions. The lung window also revealed scattered areas of cylindrical bronchiectasis, mucus plugging with centrilobular nodules in both lungs

DISCUSSION

Mounier-Kuhn syndrome is characterized by dilatation of tracheobronchial pathway at different levels, from the trachea to fourth bronchial branch as a consequence of atrophy of the muscular and elastic tissues in the trachea and main bronchial wall.[1,2] It is more common in third or fourth decade of life with male preponderance. The clinical presentation varies widely, from being asymptomatic to severe respiratory distress and death. Pathologic dilation in the tracheobronchial tree with defect in mucociliary activity leads to mucous plugging in lower respiratory tract causing recurrent symptoms. In severe cases it could lead to recurrent pneumonia consequently leading to fibrosis of tracheobronchial tree.

Various conditions are associated as a cause of secondary cause of tracheobronchial dilatation, such as Connective-tissue diseases, ataxia-telangiectasia, ankylosing spondylitis, Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome and cutis laxa (elastolysis).[2,3]

Computed tomography (CT) of thorax is diagnostic which shows abnormally dilated air passages. For diagnosis in adults, the criteria includes diameters of the trachea, >30 mm; of the right main bronchus, 20 mm; and of the left main bronchus, 18 mm.[2,4,5]

Treatment in symptomatic includes chest physiotherapy and avoiding exposure to pollutants with antibiotics during acute attacks.[3] Surgery is seldom used as the disease involve whole of tracheobronchial pathway.[2,3]

Mounier-Kuhn syndrome is a rare disease but should be kept as a differential diagnosis in patients coming with recurrent lower respiratory tract infections.

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