

Tracheobronchomegaly

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Clinical summary

A 48 year old man occasional smoker was symptomatic since 4 years with cough with copious amount of expectoration and exertional dyspnoea and recurrent infective exacerbations. There was past history of tuberculosis 15 years back treated with empirical antituberculosis therapy for a period of 1 year. There was no significant family history. Physical examination revealed presence of post exercise desaturation, grade III clubbing and coarse crackles in bilateral lung fields. Chest Radiograph (CXR) showed the presence of bilateral cystic opacities. High Resolution Computerised Tomography (HRCT) of the chest (Figure) (1a) (1b) revealed bilateral cystic bronchiectasis and tracheobronchomegaly with tracheal, right and left main bronchus dimensions being 30 mm, 27.2 mm, 22 mm respectively. Spirometry showed Forced Vital Capacity (FVC) of 1.55 L (of predicted), Forced Expiratory Volume in 1 second (FEV1) of 1.02 L (32% of predicted), and FEV1/FVC of 66%. A diagnosis of tracheobronchomegaly-Mounier Kuhn Syndrome (MKS) was made.



Figure1: 1(a) and 1(b): Axial sections of CT thorax showing transverse diameter of trachea and right and left main bronchus.

Discussion

Tracheobronchomegaly (TBM) commonly known Mounier Kuhn Syndrome (MKS) is a condition characterized by significant tracheobronchial dilatation [1]. 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 respiratory failure. It is often associated with recurrent episodes of respiratory tract infection, tracheal diverticulosis and bronchiectasis. It is more common in men and is usually diagnosed in the 3rd or 4th decades of life. Imaging features are diagnostic in MKS. Computerised tomography of thorax aids in direct measurement of the tracheobronchial tree at multiple levels and also to demonstrate secondary features including diverticuli, sacculations, 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 >30mm, 20 mm and 18 mm respectively. A classification system has been suggested which classifies TBM as: Type 1A consisting of infants who develop TBM after having undergone fetoscopic tracheal occlusion, and Type 1B patients are infants and children who develop TBM after prolonged intubation. Type 2 individuals develop TBM following recurrent

pulmonary infections (2A) or pulmonary fibrosis (2B). Type 3 represents TBM with evidence of extra-pulmonary elastolysis, and Type 4 denotes the development of TBM with no clear predisposing factors [2]. Therapy involves respiratory physiotherapy for clearing secretions and antibiotic use during infectious exacerbations. Tracheal stenting has been helpful in severe cases however surgery is seldom performed because of the diffuse nature of the disease [3-5].

References

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