Case Report

Tracheobronchomegaly–Mounier–Kuhn syndrome

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ABSTRACT

Tracheobronchomegaly is a rare disorder of uncertain etiology, characterized by marked dilatation of the trachea and major bronchi, associated with tracheal diverticulosis, bronchiectasis and recurrent respiratory tract infection. We are reporting a 60-year-old man from the Kingdom of Saudi Arabia with this condition and review the literature for such a rare entity.

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Tracheobronchomegaly is a rare disorder of uncertain etiology, characterized by marked dilatation of the trachea and major bronchi, associated with tracheal diverticulosis, bronchiectasis and recurrent respiratory tract infection. It has been first described on 1932 and since then less than 100 cases have been reported.[1] The syndrome usually appears in the fourth or fifth decade of life with symptoms similar to chronic bronchitis or bronchiectasis leading ultimately to death from recurrent infection and respiratory compromise. The diagnosis depends on the presence of large trachea and proximal bronchi by plain radiography or computed tomography scan. Tracheal diverticulosis is seen in approximately one third of the cases. Treatment is mainly supportive; however, there are few cases where insertion of dynamic tracheal stent resulted in some success.

Case Report. A 60-year-old non-smoker, retired Saudi police officer, presented to us with acute exacerbation of his chronic respiratory illness in the form of cough with purulent, foul smelling sputum of large amount, especially when bending forward. He also had a history of breathlessness, which was progressively worsened with time. He had similar episodes since the last 40-years with frequent hospital admission and emergency visits. He was labeled as a case of chronic obstructive pulmonary diseases and was managed with bronchodilators and home oxygen.

On examination, the patient was in respiratory distress, tachypneac, using accessory muscles of respiration. His oxygen saturation by pulse oximetry was 82% in room air. There was no clubbing and the cardiovascular examination was normal. Respiratory system examinations revealed diminish breath sounds and coarse inspiratory crackles over the lower third of both lung fields. The rest of the systemic examination was unremarkable. Routine laboratory investigations showed total white blood count of 5600 cells/mm^3, erythrocyte sedimentation rate of 31 mm/hour, and arterial blood gas with pH 7.39, PO_2 50 mm Hg, PCO_2 54 mm Hg and HCO_3 32 mmol/L. Chest x-ray AP and lateral (**Figure 1a & 1b**) and computerized tomography (CT) chest (**Figure 2**) with coronal and sagittal views revealed the presence of tracheobronchomegaly with tracheal transverse and sagittal diameters of 40 mm and 30 mm. The diameters of right and left main stem

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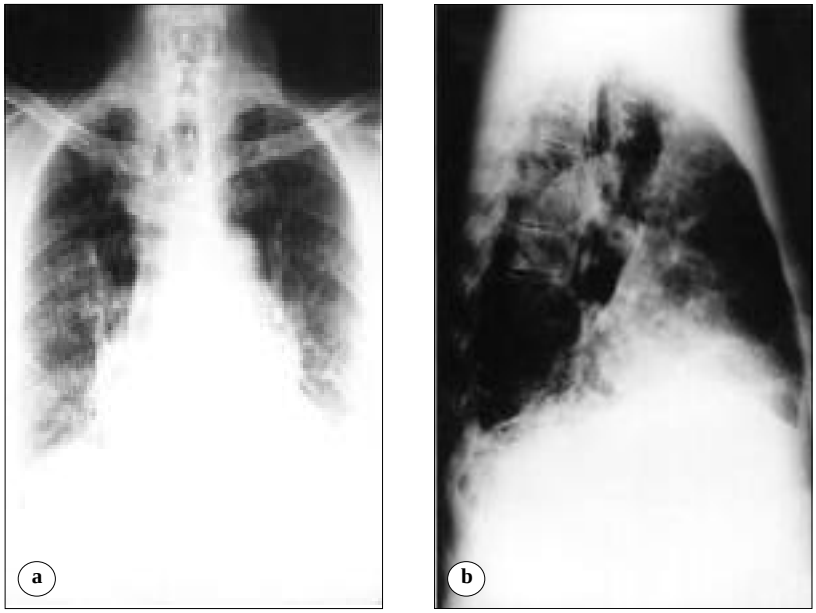

Figure 1 - Chest x-ray showing **a)** anteroposterior view of the dilated trachea and consolidation in right middle lobe and **b)** the lateral view of the dilated trachea

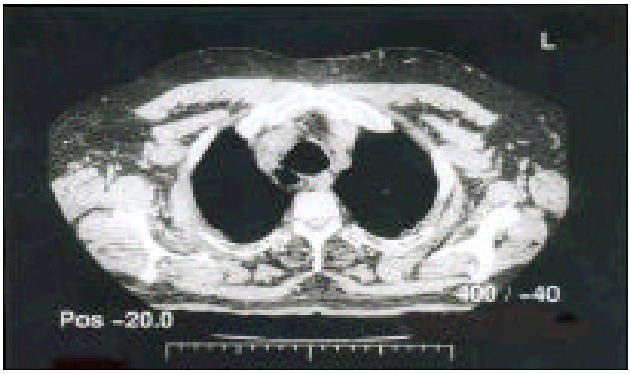

Figure 2 - Computerized tomography scan of the chest with dilated transverse diameter of the trachea and adjacent diverticula.

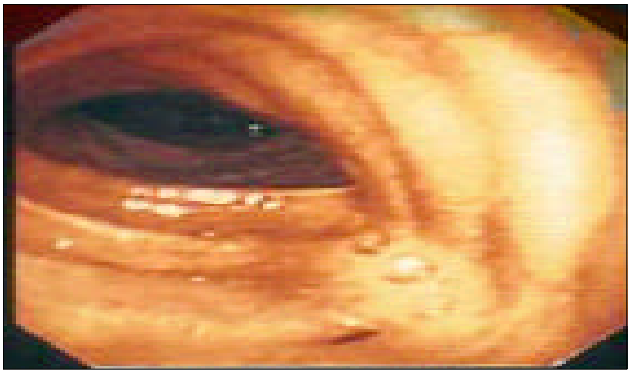

Figure 3 - Bronchoscopic picture of the trachea with tracheal dilatation, and diverticula in the posterior right tracheal wall (at 5 o'clock).

bronchi were 2.5 mm. Lower lobes showed area of bronchiectasis and consolidation. Pulmonary function test was consistent with a mixed pattern of obstructive and restrictive pulmonary diseases with forced vital capacity of 32% of predicted, forced expository volume in one second of 37% of predicted and ratio of 89%. Total lung capacity was reduced to 42% of predicted. Bronchoscopy showed a dilated trachea with tracheal diverticula in the posterior wall of the upper third of the trachea (**Figure 3**). The mucosa looked inflamed and the lower lobe was full of copious secretions. Endobronchial biopsy and bronchoalveolar lavage were compatible with chronic inflammatory changes and presence of pus cells. No pathogens were isolated from sputum or bronchial wash bacterial culture. Patient was managed with antibiotics, bronchodilator, chest physiotherapy and home oxygen with moderate improvement.

Discussion. Tracheobronchomegaly (TBM) was first recognized at autopsy and subsequently it was identified as a distinctive clinical, radiological and endoscopic entity in 1932.[1] As of 1991, only 97 cases had been reported. However, the actual number of cases may be much greater as some patients with TBM are totally asymptomatic and it is frequently overlooked in symptomatic patients based on chest x-ray alone.[2] The etiology remains uncertain.[3] A congenital connective tissue weakness, in combination with inhalation of irritants like cigarette smoking and air pollution was raised as possible factors in the development of this condition. There is also an almost invariable history of respiratory tract infection with chronic

inflammatory lung changes. However, it seems more likely that all of these factors exacerbate the condition more than causing it.[1,4,5] Johnston and Green[6] suggested an autosomal recessive pattern of inheritance in one family. A form of TBM was also seen in association with pulmonary fibrosis in adults[7] and mechanical ventilation in preterm neonates.[8] Secondary TBM was also described in association with Ehlers-Danlos syndrome, Brachmann-de Lange syndrome, Bruton-type agammaglobulinemia, Ankylosing Spondylitis, Cutis laxa and Light chain deposition disease.[9-11] Histologically, there is severe atrophy or absence of the longitudinal elastic fibers and thinning of the muscular layers of the trachea and main bronchi. This leads to dilatation of the membranous and the cartilaginous portions of the airways, mucosal herniation between the tracheal rings and subsequent tracheal diverticulosis. The airways distal to the fourth and fifth order division are usually not affected. The absence of elastin leads to collapse of the airway during expiration and inadequate cough mechanism. This alters the airway dynamics causing ineffective mucociliary clearance, retention of secretion, which predisposes the development of frequent infection, bronchiectasis and eventually fibrosis. Presentation is mainly in fourth and fifth decade of life and male predominance has been observed.[3] Clinical picture can be variable, from totally asymptomatic to advanced spectrum of infection, respiratory failure and death. Symptoms are usually similar to chronic bronchitis or bronchiectasis. Sputum is often copious, purulent and foul smelling. Spontaneous pneumothorax, hemoptysis and finger clubbing may also develop.[3,5] Tracheobronchomegaly has been classified according to the progression of the disease: type 1 - dilated trachea and major bronchi, type 2 - more obvious enlargements with eccentric configuration and early diverticula and Type 3 - diverticula extend to the main bronchi.[12] On plain chest x-ray, the increased diameter of central airways may be visible and more appreciated in the lateral view. For an adult, any diameter of trachea, right main bronchus and left main bronchus that exceeds 3 cm, 2.4 cm and 2.3 cm on a plain chest x-ray is diagnostic of TBM.[2,3] Irregular appearance of trachea airway column with protrusion between cartilages may also be found. Associated features such as central bronchiectasis, emphysematous changes and pulmonary fibrosis may be observed in advance cases.[7,13] With computed tomography scan, a transverse and sagittal diameter of the trachea exceeding 25mm and 27 mm and when the transverse diameters of the right and left main stem bronchi exceeding 21.1 and 18.4 is diagnostic of Mounier-Kuhn syndrome in men. In women, the diagnosis made when the transverse tracheal diameter of 21mm and sagittal diameter of 23mm and 20mm and 17.5mm for main stem bronchi.[14,15] Several case reports showed that the coronal and sagittal CT maybe used to evaluate the presence of diverticula which seen in one third of patients.[11,16] The experience with MRI is still limited to one case report in the literature.[17] Bronchoscopy will show dilated trachea with redundant wall with respiratory variation in central airway caliber. Pooled secretion and saccular pouch or protrusion (diverticulum) maybe found which is more common in the posterior tracheal wall.[11] As expected, pulmonary function test will reveal obstructive pattern with increased residual volume and hyperinflation with decrease diffusion capacity of carbon monoxide if lung parenchymal diseases are present. However, in advance cases, restrictive pattern secondary to fibrosis will be predominant.

Treatment is mainly supportive. Chest physiotherapy to help clear the secretions and antibiotics during infectious exacerbations is the main modalities.[2,9] There is no study to show the effectiveness of inhaled bronchodilators or corticosteroid in such condition. There is no role for surgery because of the diffuse involvement of the airways. However there was two case reports of using dynamic tracheal stent or tracheobronchial endoprosthesis with a limited success.[18,19]

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