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Tracheopathia Osteoplastica*

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Tracheopathia osteoplastica (TPO), or tracheobronchopathia osteochondroplastica, is a rare benign disease characterized by multiple cartilaginous or bony projections into the tracheobronchial lumen. The cause of this condition is unknown. However, among the possible causes, a genetic factor has been suggested, and the familial occurrence of TPO has been reported (1, 2). More than 200 patients with this unusual condition have been reported in the medical literature. In the majority of reported cases, TPO is diagnosed only at autopsy, having been unsuspected during life (3, 4). This case report describes a patient with a long medical history of chronic obstructive pulmonary disease (COPD), whose TPO was diagnosed by bronchoscopy.

A 54-year-old male was admitted to the hospital for shortness of breath. His past medical history consisted of COPD, several episodes of pneumonia, hemoptysis and hypertension. He was neither a smoker nor an alcoholic. On physical examination, the patient was afebrile, with a heart rate of 92 beats/min, a respiratory rate of 20/min, and a blood pressure of 160/100 mmHg. Auscultation of the chest indicated a mild reduction in breathing sounds bilaterally, and a prolonged expiratory phase. Findings from the rest of his examination were unremarkable. Laboratory test results were within normal limits except for a erythrocyte sedimentation rate of 44 mm/h, and a hemoglobin level of 11.5 g/dL. ECG was normal. Pulmonary function tests revealed moderate obstruction, which was associated with mild hypoxemia. Diffusing

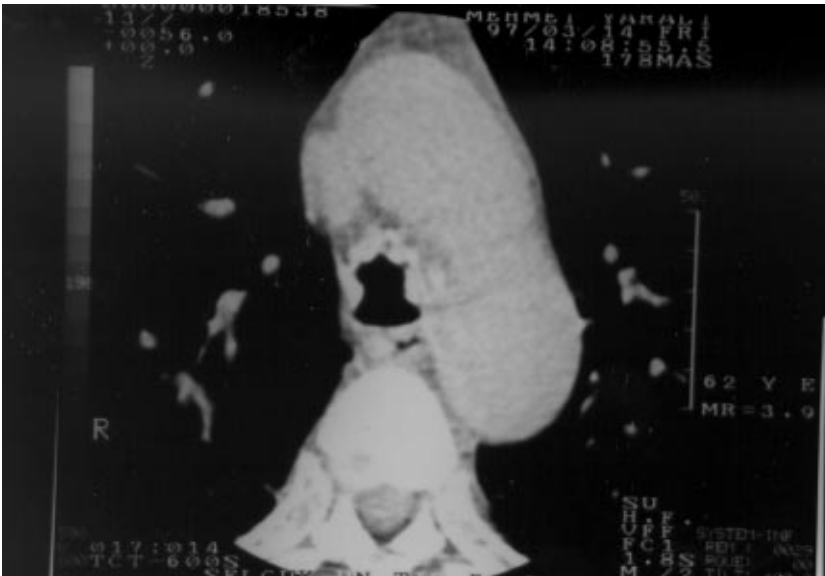


Figure 1. CT scan shows the mucosal irregularity with calcification in anterior and lateral walls of the trachea. The posterior membranous wall is intact.

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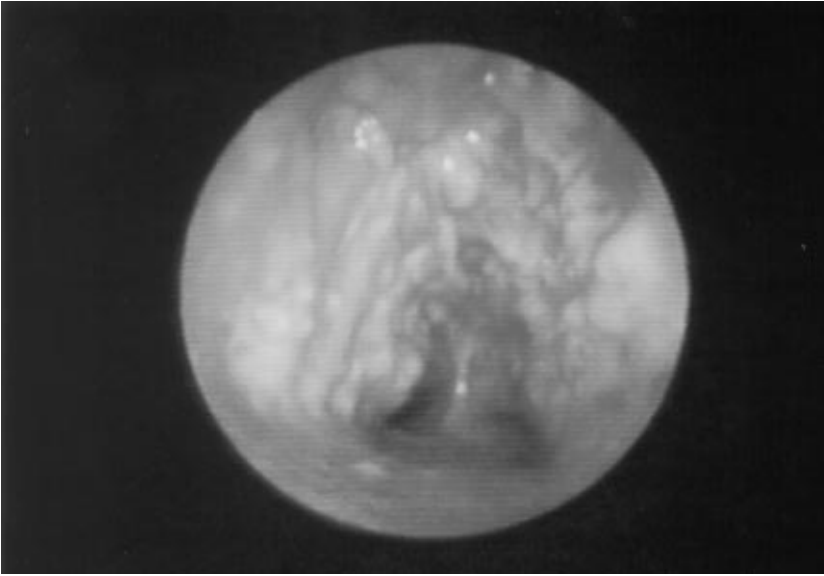


Figure 2. Bronchoscopic view of the multiple nodules obstructing the tracheal lumen. Overlying mucosa appears normal.

capacity (DLCO) and the DLCO/VA ratio were also moderately reduced. The flow-volume loop demonstrated a mild reduction of inspiratory phase. A chest radiograph disclosed multiple pleural calcifications. A CT scan of the chest demonstrated irregularity of the tracheal lumen, thickening and multiple, broad-based protrusions with calcifications in the tracheal wall. The posterior membranous wall was intact (Figure 1).

In addition, bilateral pleural thickening, pleural plaques, and emphysematous changes in the parenchyma were noted. No hilar or mediastinal adenopathy was seen. The patient subsequently underwent a bronchoscopy, which showed whitish, elevated, hard nodules situated in the lower two thirds of the anterior and lateral walls of the trachea. The posterior membranous wall was intact. The lesions extended to the first portion of the right and left major bronchi. The tracheal mucosa overlying the lesions was thin, but had a normal appearance (Figure 2). A bronchoscopic diagnosis of tracheopathia osteoplastica (TPO) was made. Microscopic examination of biopsy specimens from nodular lesions demonstrated normal mucosa with focal calcifications in the submucosa; no malignant cells were seen. These pathologic findings confirmed the bronchoscopic diagnosis of TPO. Since familial occurrence has been reported in the literature, the patient's three sons, ranging in age from 25 to 30 years, subsequently underwent bronchoscopy, but there were no abnormal findings. Cytogenetic analysis of the patient showed the short arm of the Y chromosome to be longer than normal.

The etiology and pathogenesis of TPO are unclear. Several authors have cited the association of this

condition with amyloidosis and ozena (1, 4, 5). The first report of familial occurrence of TPO was published in 1989 (6). The disease is more prevalent in men, and most patients are over 50 years of age. However, it also has been described in children. The majority of patients are asymptomatic for many years. Occasionally, gradually increasing dyspnea, hoarseness, stridor, wheezing, and hemoptysis and recurrent pulmonary infections are present. TPO is often initially misdiagnosed as asthma or bronchitis (1, 4, 6, 7). The correct diagnosis is made most easily during bronchoscopy. The frequency of TPO has been reported as 1 in 2000-5000 bronchoscopies (3, 6, 8). Many authors have stated that once the gross appearance of TPO has been observed through the bronchoscope, the diagnosis is certain. In various reports the bronchoscopic findings have been described as having the appearance of cobblestone or a stalactite cave (1, 4, 6). This condition occurs most commonly in the lower three-fourths of the trachea and often extends to involve the first portion of the major bronchi (3). A rare involvement of the larynx has also been described (9). The bronchoscope passing over the nodules produces an unforgettable grating sound (1). Pathologically, the nodules are usually confined to those portions of the tracheal and bronchial walls that normally contain cartilage (anterior and lateral walls). The posterior membranous portion of the trachea is usually intact. Histologically, the nodules demonstrate normal mucosa with focal calcification and heterotopic bone formation (1, 3, 6). Tomographic examination is more accurate than chest radiography in detecting calcification and ossification (10). Abnormalities of pulmonary function tests are dependent on the degree of involvement of the

airway lumen (6). It has been suggested that inspiratory and expiratory flow volume loops may provide useful information for the diagnosis and follow-up of this condition (11). The present case had a slight decrease in inspiratory flows. Treatment of TPO is symptomatic. It involves regular tracheobronchial toilet, and treating infection episodes with the appropriate antibiotics (1). Satisfactory results were obtained by Nd:YAG laser therapy (12). Death has been reported as a result of obstruction in patients with TPO (6).

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