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TRACHEOBRONCHOPATHIA OSTEOCHONDROPLASTICA

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ABSTRACT

Tracheobronchopathia osteochondroplastica (TO) is a rare and usually benign disorder affecting the trachea and occasionally the bronchi.

A case of TO was diagnosed in a 61 year old male farmer presenting with pulmonary infections and prolonged productive cough. A bronchoscopy revealed multiple nodular excrescences aong the anterolateral wall of the trachea and main bronchi. CT scans showed moderate narrowing and distortion of the trachea. Biopsies revealed squamous metaplasia, and calcification and ossification in the epithelial surface. The literature on the subject is reviewed here. The severity of TO ranges from no symptoms to severe dyspnea, hemoptysis or pneumonitis. The etiology and pathogenesis is unknown. Treatment is seldom necessary. Awareness of the condition as a differential diagnosis to neoplasms is important to avoid unnecessary surgery or chemotherapy.

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INTRODUCTION

Tracheobronchopathia osteochondroplastica (TO) is a rare disorder of the laryngotracheobronchial tree. The condition is benign, characterized by submucosal nodules containing combinations of cartilaginous, osseous, and hematopoietic tissue and calcified cellular protein matrix, protruding into the bronchial lumen.

The nodules generally have a diameter of 1-3 mm and are located mainly in the lower third or half of the trachea and in the upper part of the main bronchi. In some cases, TO may also affect the larynx and subglottic trachea. In pronounced cases, the bronchoscopic picture is characteristically described as a beaded, spiculated, rock garden, or cobblestoned appearance. The overlying mucosa is often the site of squamous metaplasia and typically the posterior wall is spared.[1]

The cause of this disorder is still unknown. The first cases were described by Rokitansky (1855), and since then, approximately 300 cases have been reported, 82 from Finland. [2-5]

Many cases are asymptomatic, and before the advent of bronchoscopy it was mainly diagnosed incidentally postmortem. The characteristic symptoms of TO are cough, hemoptysis, dyspnea especially on exercise, recurrent respiratory infections and asthma-like symptoms. Only rarely can TO be suspected from a conventional chest X-

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ray. Tomography and CT-scan can be suggestive.[6]

CASE REPORT

A 61-year-old male farmer presented with fever, chills and a 15 year history of recurrent productive cough. Intermittently he had experienced chest pain provoked by deep breathing. He had suffered from recurrent respiratory infections for many years. He had never smoked. His past history included epigastic pain due to duodenal ulcer that had been treated with H_2 receptor antagonists. Otherwise the patient was well and did not require any permanent medication. History revealed no allergies and there were no respiratory complaints within the family.

On physical exam he was thin and cachectic (weight 57 kg, height 1.76 m), pulse rate 69 beats/min, blood pressure 100/60 mmHg, temperature 37.8°C, and respiratory rate 15 breaths/min, with decrease of breath sounds on the right side and fine inspiratory crackles in the lower left lung field. The erythrocyte sedimentation rate was 70 mm/h, but routine blood and serum biochemistry studies were within normal limits.

The chest roentgenogram showed a reticulofibrotic infiltration in both basal lung fields (Fig.1). A thoracic computerized tomographic (CT) scan showed moderate narrowing and distortion of the trachea, with changes in it's caliber (Fig. 2). Room air blood gas analysis gave the following results: PaO_2 = 87.6 mmHg, $PaCO_2$ = 36.8 mmHg, pH=7.42. Lung function tests revealed normal values:
FVC = 3.80 liters (100.8%), FEV_1 = 3.35 liters (111.8%), FEV_1/FVC= 84%. However, on forced flow-volume curves, inspiratory flows were always limited to values lower than the normal expiratory flows: Peak inspiratory flow = 1.98 L/s (25.2%), peak expiratory flow=7.03 L/s (89%) (Fig. 3).

On bronchoscopy the larynx and vocal cords were normal. From the subglottic trachea downwards, the mucous membrane was lumpy and stiff. Only the posterior membranous portion of the trachea was intact. The hard whitish nodule was seen from 2 cm beneath the subglottis of the trachea to the right and left main bronchi and the proximal lobar bronchi. Bronchial lavage and a considerable number of biopsies were taken from the carina, trachea and both sides of the bronchial tree. Bronchial lavage revealed no tumor cells, and no growth of any pathogens or mycobacteria. Biopsies were performed with difficulty because of the consistency of the calcific nodules and revealed a moderate mucosal squamous metaplasia, calcification and ossification in the submucosal area (Figs. 4,5).

Annual clinical and endoscopic follow up for 3 years showed no evidence of functional impairment, and confirmed that the endotracheal and bronchial lesions were not progressive.

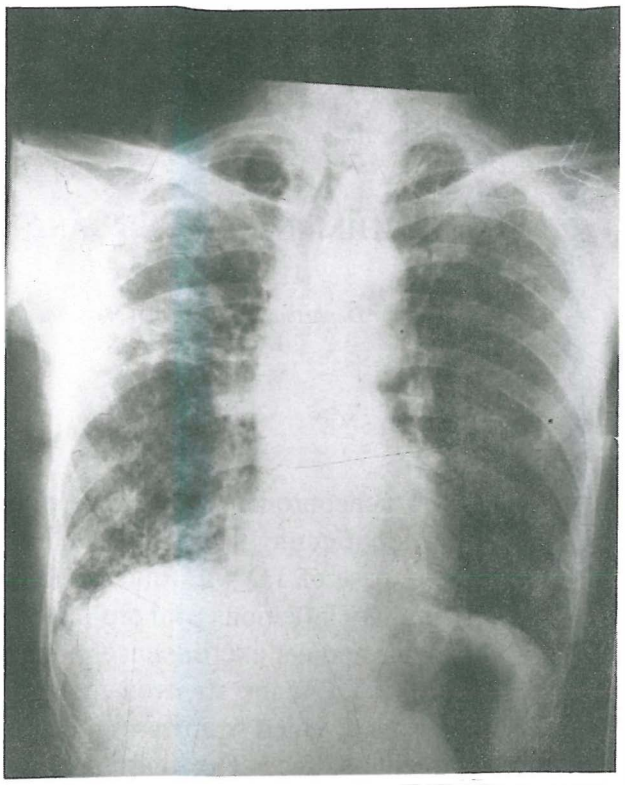

Fig. 1. Chest x-ray film showing reticulofibrotic changes of both lung fields.

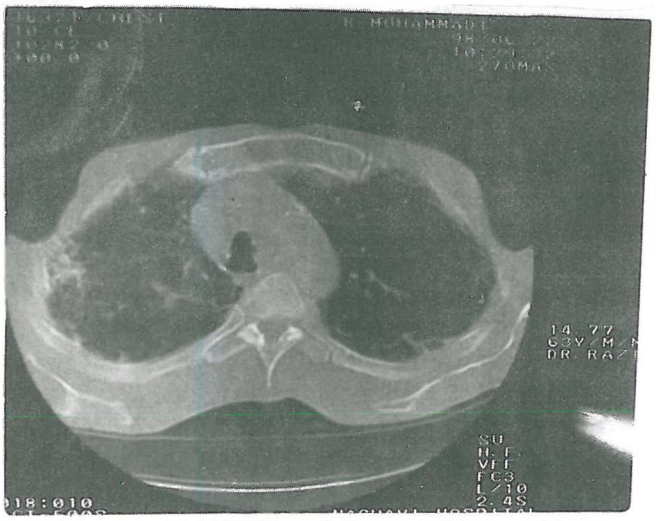

Fig. 2. CT scans show a tracheal lumen distorted and reduced in transverse diameter.

DISCUSSION

TO is an uncommon disease first named by Aschoff in 1910.[7] TO is a benign and rare disease, and only 371 cases have been reported.[8] The incidence of TO is approximately

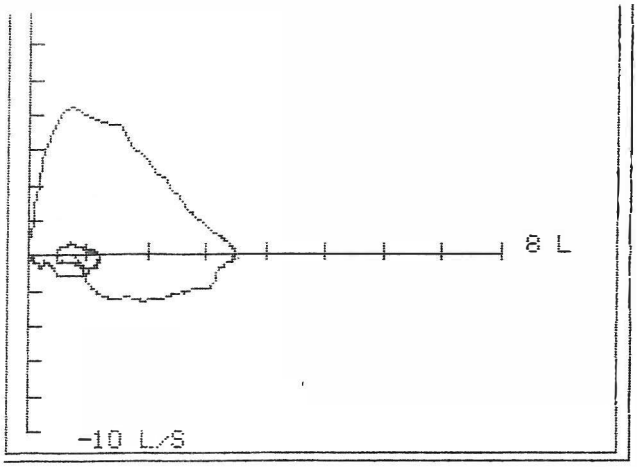

Fig. 3. Inspiratory and expiratory flow-volume curve with limited flows during inspiratory forced maneuver. A variable extrathoracic obstruction is suggested by the low inspiratory forced flows despite normal expiratory flows.

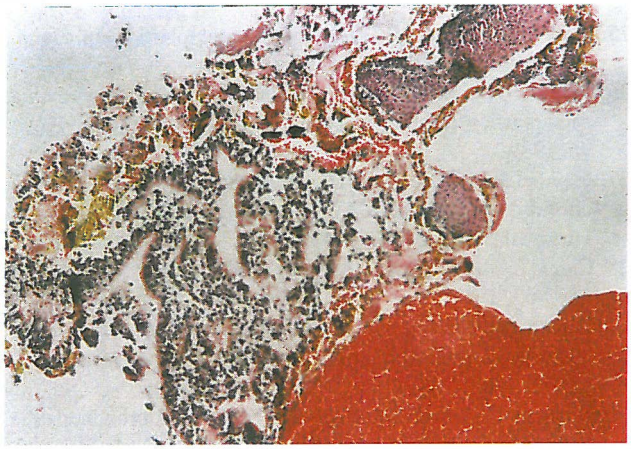

Fig. 4. Squamous metaplasia in the epithelial surface (H&E, ×40).

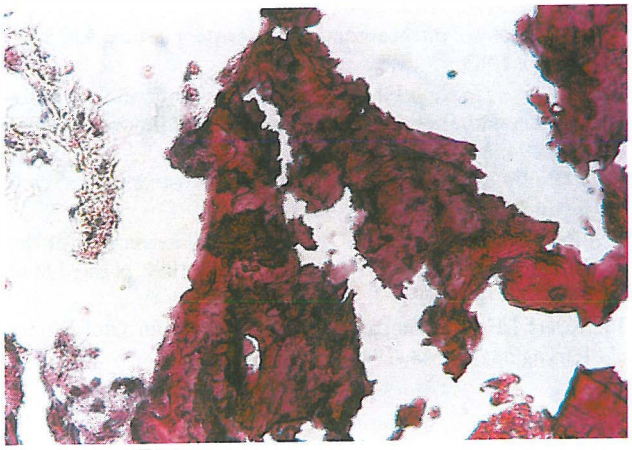

Fig. 5. Calcification and ossification in the submucosal area (H&E, ×40).

3/1000 at autopsy,[9,10] while data from bronchoscopy surveys are more varying, ranging from 1/125 to 1/5000. [11,12] Most cases of TO reported in the literature, however, were diagnosed only at autopsy.[13] There is no sexual predominance. TO is usually discovered in adults of more than 50 years of age, but has already been described in children.[3] No clear link was established between smoking and TO in a review of the literature up to 1985.[14]

The etiology of TO remains obscure. Association between TO and upper respiratory tract infections such as atropic/ozena have been reported.[3,15,16] Concomitant findings of amyloid have led to the hypothesis that TO may be a late stage of primary amyloidosis of the lung.[17-19] Moreover, a few cases have been associated with lung cancer,[20-22] and a few with thyroid tumor or thymoma.[23,24] Some predisposing genetic factors and inheritance have been suggested to play a role in the development of TO.[25,26]

Local metabolic or inflammatory factors, infections, and chemical irritation have also been suggested as etiologic factors.[25,27] TO is often asymptomatic, but can cause dry cough, dyspnea, inspiratory stridor, dry throat, dysphonia, bronchial obstruction, hemoptysis, recurrent pneumonia, and occasionally difficult tracheal intubation before surgery. The severity of symptoms in TO depends on the extent of tracheal/bronchial obstruction leading to dyspnea, and complicating respiratory tract infections. Several cases of infectious lung disease with a fatal outcome have been reported as a complication to TO. [28]

Retrospective examination of chest roentgenograms can occasionally show a fine scalloped calcified border on the internal walls of the trachea on the lateral aspect,[29,30] but this wasn't obvious in this case.

The computed tomographic findings in TO are also pathognomonic.[31] CT of the trachea and major bronchi reveals calcific nodules projecting into the lumen, thus destroying their normal configuration. These calcific deposits are absent in the posterior wall. [29,32]

Subclinical variable extrathoracic obstruction can be shown on inspiratroy flow-volume curves (Fig. 3). Lung function test in TO has no characteristic pattern and will be normal results in many cases.[5,12,15] Van Nierop found normal spirometry results in all of his four reported cases and normal volume- flow curves in two of them.[12] Flow-volume loop is often sensitive and can be helpful in following the course of TO. [30]

TO has repeatedly expressed itself as a middle lobe syndrome,[20,33] and clinically TO is a differential diagnosis for bronchial/tracheal tumor and chronic obstructive pulmonary disease. The differential diagnosis of the bronchoscopic picture of multiple nodules includes TO, amyloidosis, endobronchial sarcoidosis, calcifying lesions of tuberculosis, papillomatosis and tracheobronchial calcinosis. Treatment is symptomatic, including antibiotics for infectious complications, and in some cases, stenosis

can be relieved by bronchoscopic removal of the obstructing lesions. Successful segmental tracheal resection has been reported,[34] and recently temporary surgical insertion of a stent (T-Y tube) has been performed with success in the case of saber-sheath tracheal stenosis.[35] One case of successful relief of severe recurrent hemoptysis has been reported 9 months following radioactive therapy of 750 R.[36]

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