Tracheobronchopathia Osteochondroplastica Presenting as Right Middle Lobe Collapse*
Diagnosis by Bronchoscopy and Computerized Tomography

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Tracheobronchopathia osteochondroplastica (TO) is a rarely described disorder which historically has not been frequently recognized antemortem. Studies by computerized tomography (CT) and bronchoscopy now permit a definitive antemortem diagnosis and can obviate a more invasive diagnostic evaluation. We describe two cases of TO presenting as right middle lobe collapse, discuss the clinical and pathologic features, and outline an approach to its evaluation using CT and bronchoscopic study. (Chest 1988; 94:842-44)

Tracheobronchopathia osteochondroplastica (TO) is a rare disorder characterized by numerous osseous and cartilaginous submucosal nodules projecting into the lumen of the trachea and bronchi. In the past a diagnosis of TO was most frequently established at autopsy, and any symptoms were secondary to airway obstruction and its sequelae. However, with the advent of new pulmonary diagnostic techniques including bronchoscopic examination and CT, TO can be recognized antemortem. However, the CT appearance of TO is not always diagnostic, and other causes must be considered. We report the radiographic and bronchoscopic findings in two recent cases of TO presenting as right middle lobe (RML) collapse and review the clinical and pathologic features of this rare disorder.

Case Reports

Case 1

A 64-year-old woman with stage 1B, grade 3 poorly differentiated adenocarcinoma of the endometrium underwent a transabdominal hysterectomy and bilateral salpingo-oophorectomy. A routine preoperative chest roentgenogram revealed right middle lobe atelectasis that did not resolve postoperatively. The patient had a 15 pack-year history of cigarette use. She denied having any acute chest symptoms, recurrent respiratory tract infections, dyspnea on exertion, or hemoptysis. She had a history of tuberculous peritonitis at age 18 years treated by one year in a sanitarium. She occasionally noted a productive cough, which she attributed to smoking. Physical examination was unremarkable. There was no clubbing or lymphadenopathy. Results of laboratory studies were normal. Since the atelectasis failed to respond to conservative therapy, she underwent a fiberoptic bronchoscopic evaluation, at which 3 to 4-mm nodules were seen scattered throughout the airways to the level of the segmental bronchi. The orifice of the RML was more than 90 percent obstructed by a spicule-like lesion extending from the medial wall of the RML bronchus. The overlying mucosa appeared normal, without evidence of inflammation. The biopsy specimen of this site revealed a glistening, pale white surface with minimal bleeding. Multiple biopsies were performed, resulting in more than 50 percent patency of the RML bronchus. Microscopic examination of the biopsy material revealed fragments of normal cartilage with normal overlying mucosa (Fig 1). A chest roentgenogram obtained two months later revealed persistent RML atelectasis. Cultures for Mycobacterium tuberculosis were negative.

Case 2

A 79-year-old man was admitted for treatment of recurrent urinary tract infections and evaluation of 11.25-kg (25-lb) weight loss. A chest roentgenogram taken on admission showed RML collapse, enlarged main pulmonary arteries, and evidence of hyperinflation. Previous chest roentgenograms taken at another institution several years earlier did not demonstrate the RML collapse. The patient had a 50 pack-year history of smoking cigarettes and had dyspnea on climbing one flight of stairs. He denied chronic cough, sputum production, hemoptysis, or chest pain. No history of tuberculosis

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Figure 1. Biopsy specimen of RML lesion from case 1 (original magnification × 63).
were elicted. On examination he was a cachectic man. Breath sounds were symmetrically decreased, and rales were present over the right anterior lung field. He had no palpable lymphadenopathy or clubbing. A chest CT scan without contrast (Fig 2) confirmed RML collapse and demonstrated multiple nodular densities in the bronchial walls, most prominent in the left main stem bronchus, with no involvement of the trachea. A fiberoptic bronchoscopic study was performed to evaluate further the RML collapse and the nodular densities. Numerous nodular lesions were seen, beginning at the midtrachea and extending into the subsegmental bronchi bilaterally. The posterior tracheal wall was spared. The lesions ranged from 1 to 5 mm in diameter and occurred most frequently over cartilaginous rings. The lesions were firm, and the overlying mucosa appeared normal. The lesions narrowed the diameter of the bronchi, but involvement was greatest at the middle right and upper lobes. There was 90% narrowing of the right upper lobe orifice. The RML orifice was slit-like and would not admit the bronchoscope or a cytologic brush. Biopsy of the lesions produced minimal bleeding, and pathologic examination revealed normal bronchial mucosa with no osseous or cartilaginous elements.

DISCUSSION

TO was originally described by Wilks° in 1857. A review of the literature by Martin1 in 1974 brought the number of reported cases to 245; however, the number of cases of TO reported in the United States is small.

Right middle lobe collapse is a common radiographic finding. The differential diagnosis includes malignant and benign tumors and infection (tuberculosis, bronchiectasis, bacterial pneumonia, and fungal infections). When the duration of the collapse is unknown, evaluation with either CT scan or bronchoscopic study is indicated. In our two cases this led to the unexpected diagnosis of TO and prompted a review of the literature on this rare disorder.

The submucosal nodules of TO contain elements of elastic cartilage present in different stages of development, which may be associated with calcified cartilage or osseous tissue. These nodules most commonly involve the lower two thirds of the trachea, rarely extending proximally to involve the larynx.

Extension distally into the major bronchi is common. Lobar bronchi are occasionally involved, and rarely the process extends into segmental bronchi. The nodules can be numerous and diffusely located or only few. They are 1 to 3 mm in diameter, but can be plaque-like or, as in our first case, pedunculated. They are firm and frequently difficult to biopsy through the fiberoptic bronchoscope, which may explain the absence of osseous or cartilaginous elements on histologic examination in our second case.

The origin of the lesion of TO remains unclear. Dalgaard3 and Martin1 reviewed the theories regarding the etiology of TO, and both favored metaplasia of submucosal connective tissue. Founier and Pieterse4 reported a case where histologic examination of TO nodules revealed the lesions originating from a tracheal ring, suggesting eccchondrosis and exostosis of tracheal cartilage. Harma and Suurkari5 have suggested chronic irritation and infection as a potential etiology, since 23 of 30 cases they reviewed had a history of atrophic rhinitis or pharyngitis. Other proposed etiologies of TO include metabolic abnormalities, degenerative processes related to aging, and amyloidosis.

Most patients with TO are asymptomatic throughout their lives. If symptoms occur, they are usually the result of airway obstruction, most commonly causing cough, sputum, dyspnea, and recurrent respiratory tract infections. Hemoptysis secondary to an acute infection, bronchiectasis, or ulceration of a nodule also occurs. Physical examination results are usually normal. The nodules and calcifications of TO are not appreciated on routine chest roentgenography. Radiographic abnormalities, if present, consist of atelectasis (ranging from small areas of involvement to lobar collapse) and infiltrates.

In 1981 Lundgren and Stjernberg6 reported pulmonary function data in eight cases of TO. Six patients were nonsmokers, and two were exsmokers. No history of asthma was reported. All eight patients demonstrated airflow obstruction on pulmonary function testing, with one having combined restrictive and obstructive ventilatory deficits. Three patients had decreased inspiratory flows, possibly due to narrowing of the trachea and large airways. Decreased inspiratory airflow with normal expiratory flow rates has also been reported.7

The differential diagnosis of multiple nodular lesions of the trachea and bronchi is limited to TO, papillomatosis, amyloidosis, and endobronchial sarcoidosis. Traditionally, TO was diagnosed incidentally at autopsy, but with the advent of flexible fiberoptic bronchoscopy and CT scans, the diagnosis is increasingly established antemortem. Three previous reports have illustrated the utility of CT in establishing the diagnosis of TO.8-10 In these cases chest CT revealed multiple, nodular, submucosal irregularities, some of which were calcifi-
fied, with sparing of the posterior tracheal wall. In our second case, the diagnosis was originally suggested by chest CT which showed multiple nodular densities of the main bronchial walls, most prominently on the left (Fig 1). In contrast to the marked tracheal abnormalities seen at bronchoscopy, the trachea appeared normal on CT. The presence of calcifications in the nodules could not be conclusively demonstrated.

On bronchoscopic examination the lesions of diffuse TO are numerous, appearing as discrete nodules 1 to 3 mm in diameter, which may coalesce into plaque-like lesions. They may also appear as spicules protruding into the airway. Reduction of airway caliber or complete obstruction can result from the lesions. The mucosa overlying the nodules usually appears normal unless a nodule has ulcerated. Milder forms of TO may be overlooked at bronchoscopic evaluation, since there may only be a few localized nodules or spicules.

While there is no specific treatment for TO, the lesions do not appear to have a malignant potential. Most patients experience little morbidity from this disease and the disease appears to progress quite slowly. Van Nierop et al.11 reported a case where bronchoscopy and pulmonary function testing were repeated eight years after the initial evaluation, and no progression could be documented. However, recurrent respiratory tract infections due to local obstruction may be a complication, potentially leading to bronchiectasis. Hemoptysis is usually self-limited. Endobronchial resection has been suggested as a potential form of therapy when significant obstruction of an airway exists.11 In our first case endoscopic removal of the obstruction using forceps biopsy did not result in reexpansion of the RML, possibly reflecting the chronicity of the atelectasis. In addition to endobronchial resection of obstructing lesions, laser therapy may be another possible option for improving patency of airways with local involvement from TO.

Tracheobronchopathia osteochondroplastica should be considered in the differential diagnosis as an unusual cause of hemoptysis, persistent atelectasis, and recurrent segmental or lobar infection. The bronchoscopic appearance of TO is highly suggestive and confirmed by the appropriate pathologic findings. While the CT can be highly suggestive, it is not always diagnostic. In contrast to previous reports of the CT appearance of TO, we were unable to identify calcification of the nodules. Additionally, even in the presence of calcified lesions, posterior tracheal wall involvement necessitates consideration of other diagnostic possibilities, especially tracheobronchial amyloidosis. Since calcifications can be present in amyloidosis pathologically, it is conceivable that the CT appearance of amyloidosis and TO may be similar. However, involvement of the posterior tracheal wall is more common in amyloidosis and should only occur in very advanced cases of TO.12

Therefore, in evaluating the CT appearance of suspected TO, the presence of multiple, calcified nodules that spare the posterior tracheal wall can be considered pathognomonic of TO. In cases with posterior tracheal wall involvement, the possibility of tracheobronchial amyloidosis should be considered regardless of whether calcifications are demonstrated by CT or not. If calcifications are absent, papillomatosis and endobronchial sarcoidosis are diagnostic possibilities as well. In these instances bronchoscopy should be employed to distinguish among TO, tracheobronchial amyloidosis, papillomatosis, and endobronchial sarcoidosis.

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