Irregularity of the Trachea and Atelectasis in a Middle-aged Man*

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A 62-year-old man, a professional painter, was referred to the Division of Pulmonary Medicine because of an abnormal chest roentgenogram (Fig 1). Except for mild hoarseness of several months duration he was asymptomatic, and specifically denied having cough, hemoptysis, or dyspnea on exertion. He had always enjoyed good health and had stopped smoking eight years previously after accumulating 20 pack-years.

Physical examination revealed the lungs to be clear, with vesicular breath sounds and no localized changes in fremitus or percussion. There was no adenopathy. Spirometry, plethysmographic lung volumes, gas diffusion, flow volume loops, and airway mechanics were normal, except for a slightly diminished specific conductance.

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Diagnosis: Tracheobronchopathia osteochondroplastica

Figure 1 shows a band of atelectasis in the right mid-lung field and irregularity of the tracheal air column. The tomogram (Fig 2) reveals numerous polypoid excrescences along the walls of the proximal airways including the trachea, both mainstem bronchi, and the bronchus intermedius.

The roentgenographic differential of multiple rounded defects confined to the major airways (with or without obstruction) includes tracheal papillomatosis, fibroepithelial polyposis, amyloidosis, neoplasm metastatic to bronchial mucosa, and tracheobronchopathia. Papillomas and fibromas generally are more discrete, well-rounded, and less sessile than the nodules of tracheobronchopathia. Neoplasm may cause marked mucosal irregularity, although it tends to be less diffuse and symmetric by comparison. Calcification, clinically and pathologically characteristic in this condition, is not always apparent roentgenographically. The nodular tracheobronchial form of amyloidosis may be impossible to distinguish clearly from tracheobronchopathia. Endobronchial amyloid deposits often are diffuse and may be calcified as well.

Tracheobronchopathia is a benign tumorous condition characterized by mucosal metaplasia and submucous ossification, often with hematopoietic lamellar bone. The process probably originates in the internal perichondrium and is believed to represent ecchondrosis of the tracheal cartilagenous rings. The disease is often asymptomatic, but may be associated with hoarseness, persistent wheezing, chronic cough, and recurrent pneumonia. As many as 25 percent of patients may experience hemoptysis. The hoarseness in our patient was probably due to an area of leukoplakia observed on the right vocal cord. At endoscopy he had the characteristic findings of a rigid trachea (Fig 3), with numerous hard nodules projecting into the lumen. These diminished in number and size more peripherally, few being seen at the segmental level. There was no endobronchial obstruction, and his atelectasis cleared spontaneously. Bronchial biopsy findings revealed subepithelial fragments of mature bone and cartilage.

Tracheobronchopathia is compatible with a normal life, the clinical course most closely resembling bronchiectasis. Pulmonary function data rarely have been reported, but expiratory flow limitation, as well as extrathoracic airway obstruction as determined by flow volume loop, has been described. The etiology of tracheobronchopathia is unknown. An association with amyloidosis has been proposed, but is unproved. Special stains for amyloid were negative in our case, and we were unable to make this diagnosis.

REFERENCES

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