Case Report

Tracheopathia Osteochondroplastica

Kashif Hussain, MD, *Department of Pulmonary-Critical Care, University of Wisconsin Hospital, Madison, Wisconsin*
Steven Gilbert, MD, *Department of Pulmonary Medicine, Marshfield Clinic, Marshfield, Wisconsin*

ABSTRACT

The case of a male, 61 years of age, presenting with occasional hemoptysis and shortness of breath (duration of 1 year) is reported. Congestive heart failure was presumed and supported by chest x-ray and echocardiography. The patient improved with diuretic and angiotensin converting enzyme (ACE) inhibitor therapy, but continued to experience cough and occasional hemoptysis. Bronchoscopy revealed numerous firm nodular projections within the trachea with distribution along the cartilaginous rings. Tracheopathia osteochondroplastica (TPO) was diagnosed. TPO is an uncommon, benign, but slowly progressive disease of unknown etiology. It is characterized by endoluminal projection of cartilaginous and bony nodules arising in the submucosa of the trachea. Involvement may extend to lobar or segmental bronchi. TPO should be considered in cases where cough, dyspnea, persistent pulmonary infection, hoarseness, or recurrent hemoptysis remain after appropriate treatment of other presumptive underlying causes.

INTRODUCTION

Chronic worsening cough and exertional dyspnea can be hallmarks of asthma, chronic bronchitis, congestive heart failure, or a neoplasm. When such a cause is positively identified and treated, yet the cough and dyspnea persist, other causes might be at play. The case of a male, 61 years of age, who presented with symptoms of congestive heart failure that was subsequently confirmed by chest x-ray and echocardiography is reported. Appropriate therapy improved the clinical symptoms of heart failure; however, a cough with intermittent hemoptysis persisted. Bronchoscopy revealed the uncommon entity of tracheopathia osteochondroplastica (TPO).

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Kashif Hussain, MD
Department of Pulmonary/Critical Care
University of Wisconsin Hospital
600 Highland Avenue
Madison, WI 53791
Telephone: 608-833-5885
Fax: 775-244-1330
Email: skhussain@hotmail.com

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CASE HISTORY

A male Caucasian, 61 years of age, presented complaining of worsening shortness of breath, exertional dyspnea, persistent cough and occasional hemoptysis over the previous year. Previous treatment with prednisone and inhaled bronchodilator had no notable response. Seven months previously, the patient had been treated for pneumonia with subsequent improvement following antibiotic therapy.

A lifelong nonsmoker, the patient reported tightness in the chest, occasional wheezing, and a cough occasionally productive of clear sputum with blood intermixed. Concomitantly, the patient related orthopnea and paroxysmal nocturnal dyspnea. Echocardiography showed severely reduced left ventricular function. A chest x-ray revealed cardiomegaly with distension of the pulmonary vasculature. The clinical and radiographic pictures were consistent with congestive heart failure as the predominant underlying process.

The patient showed subsequent clinical improvement following treatment with a diuretic, an angiotensin converting enzyme (ACE) inhibitor and digoxin. However, coughing and intermittent hemoptysis continued, prompting pulmonary consultation.

Bronchoscopy revealed numerous firm nodular projections within the trachea with distribution along the cartilaginous rings and sparing of the membranous trachea (figure 1). There were a few smaller nodules within the mainstem bronchi. The remainder of the bronchial tree was sequentially inspected to the subsegmental level with all orifices patent and no other endobronchial abnormalities seen. A cytological specimen obtained at the time of bronchoscopy revealed no tumor cells. The appearance and pattern of distribution of the nodules was consistent with TPO. No airway obstruction was evident. The patient was advised to continue on treatment for congestive heart failure.

DISCUSSION

In retrospect, this case presented several signs consistent with symptomatic TPO: chronic persistent cough unresponsive to cough suppressants, antihistamines and decongestants or bronchodilators; exertional dyspnea; recurrent hemoptysis; a history of pulmonary infection; and detection

at an age over 50 years.¹⁻⁴ In this instance, the symptoms were concomitant with and masked by the symptoms associated with chronic congestive heart failure.

The true incidence of TPO is not known with certainty. Most cases appear to be asymptomatic and nonneoplastic. It may only be detected incidentally upon intubation or when tomographic, magnetic resonance, or bronchoscopic imaging is performed for other unrelated conditions.^{2,5-9} In one report, TPO was an incidental finding at autopsy in 1 in 400 cases.¹⁰ In other reports, TPO was an incidental finding upon bronchoscopic examination in 4 of 550 and 9 of 2,180 examinations.^{3,11} This places the incidence at 2 to 7 cases in 1,000.

Even though the progression of the disease is slow and considered benign, by the time it becomes symptomatic, pulmonary function can become compromised. Most commonly, symptomatic TPO is accompanied by cough, hoarseness, hemoptysis, exertion dyspnea and wheezing.¹² The cough is thought to result from turbulent air flow in the trachea, increased airway sensitivity, and impaired ciliary clearance caused by nodules that may become so numerous as to appear as a "rock garden."^{13,14} Tracheal stenosis may be detectable by spirometry in advanced cases.³ Decreased respiratory exchange and pulmonary clearance can lead to life threatening persistent pneumonia or other airway infections, and/or acute respiratory failure.¹⁵

In a recent French retrospective multicenter study of 41 patients, endoscopic follow-up over a mean interval of 7 ± 10 years revealed progression of lesions in 45% of patients, which was minimal in 28% of cases, but significant in 17%.¹² TPO is thus usually a benign disorder, but not a static one. Disease progression resulting in severe tracheal stenosis has been reported,^{3,16} but is rare. Only one case has been reported of rapidly progressing TPO where tracheal stenosis advanced from 8 mm to 3 mm in 6 weeks.¹⁷

Differential diagnosis should include endobronchial neoplasm, calcification of deep parts of tracheobronchial amyloidosis, endobronchial sarcoidosis, calcificating lesions of tuberculosis, papillomatosis, tracheobronchial calcinosis, and changes in the trachea and bronchi due to ageing that may exhibit calcification of the tracheal cartilage, but in which there is no thickening of the wall of the trachea.¹⁸⁻²¹ In TPO, the posterior membrane is typically spared.

Figure 1. Bronchoscopic appearance exhibiting numerous firm nodular projections within the trachea with distribution along the cartilaginous rings sparing the dorsal pars membranacea.

Definitive diagnosis may be made by bronchoscopy, computed tomography or magnetic resonance imaging. Biopsy histopathology is desirable, but not necessary for diagnosis. Obtaining a specimen with a flexible bronchoscope may be difficult due to the bony nature of the nodules.^{14,22-24}

Histopathology shows that the nodules are calcifications, chondrifications or ossifications of the upper layer of the mucous membrane.⁷ The calcifications and ossifications may have foci of bone marrow with active areas of hemopoiesis.²⁵ Ossifications are lamellar-type bone covered by normal mucosa or squamous metaplasia that may connect by bone, cartilage or connective tissue, to the perichondrium of the tracheal rings.^{9,14}

The etiology of the nodules remains unknown. There is no apparent association with a history of smoking. An infective association with atrophic rhinitis and ozena accompanied by atypical microorganisms, such as *Mycobacterium avium* or *M. tuberculosis* and *Klebsiella ozaenae* has sometimes been reported.^{12,26-29} Others report an occupational exposure to silica accompanied by pulmonary silicosis.^{30,31} Some authors have suggested that TPO is related to end stage primary tracheobronchial amyloidosis,^{32,33} but this has not been proven. Familial occurrence of TPO has been described.³⁴ Whatever the trigger, there is histopathologic evidence that bone morphogenetic protein-2 acting synergistically with transforming growth factor- β 1, plays an important role in nodule formation in the tracheal submucosa.³⁵

The first descriptions of TPO have been attributed to works by Rokitsky in 1855, Luschka in 1856 and Wilks in 1857.³⁶⁻³⁸ Yet, the only therapy currently available is symptomatic treatment with centrally-acting antitussive agents.³⁹ It also seems reasonable to consider prophylactic or periodic antibiotics if the course is complicated by recurrent lower respiratory infection. Others have attempted surgical or bronchoscopic removal, laser ablation (CO₂ or holmium: yttrium-aluminum-garnet [Ho:YAG]), cryotherapy, and external beam irradiation in cases causing significant airway compromise.^{4,5,25,40-42}

While the incidence rates of TPO remain uncertain, its occurrence is probably more common than currently recognized. Awareness of this entity may help explain cases of persistent cough, exertional dyspnea, recurrent episodes of hemoptysis, and persistent pulmonary infections, especially when other presumptive causes of these symptoms have been treated or ruled out.

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