Tracheobronchopathia osteochondroplastica: an unusual cause of chronic cough

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Learning point for clinicians

• Tracheobronchopathia osteochondroplastica (TO) is a rare disease characterized by submucosal osteocartilaginous nodules of the trachea and major bronchi.
• TO should be considered in patients with chronic cough and tracheal irregularities in imaging.
• Computed tomography and endoscopic findings are usually highly suggestive of TO.
• Atypical forms can be confused with tracheal amyloidosis, sarcoidosis or neoplasms.

Introduction

Chronic cough is a common reason for consultation and management presents a challenge for the clinician. Various rare causes of chronic cough should be considered when the classical etiologies have been eliminated. These rare causes of chronic cough include benign lesions of the trachea such as Tracheobronchopathia osteochondroplastica (TO).1 TO is a rare disease of unknown origin characterized by multiple submucosal nodules osteocartilaginous of the trachea and major airways. We report a case of TO incidentally discovered at imaging in old woman suffering of chronic cough.

Clinical case

A 46-year-old, no smoker woman, with a 5 years medical history of episodic dry cough, consult for a chronic abdominal pain with weight loss during 5 months. The physical examination was unremarkable, except hepatomegaly. There was no lymphadenopathy or splenomegaly. Routine laboratory findings revealed low hemoglobin level with elevated serum transaminases and alkaline phosphatase levels. Serology for toxoplasmosis, brucellosis, hepatitis and leishmaniasis were negative. The tuberculin test was strongly positive. Repeated sputum and urine acid-fast bacilli smears and cultures were negative. Liver ultrasound showed heterogeneous texture with few lymph nodes at porta hepatitis. Chest X-ray was normal. Computed tomography of the chest and abdomen confirmed multiple small hypodense no enhancing lesions in the liver and showed narrowing of the trachea and tracheal wall thickening with calcifications (Figure 1A and B, see Video). Histopathological examination of the percutaneous biopsy of the liver, revealed epithelioid cell granulomas with multinecled Langhan’s with caseating necrosis suggestive of granulomatous hepatic inflammation of tuberculous etiology. Bronchoscopy revealed a mild narrowing of the trachea with white mucosal granularity and nodules (Figure 1C and D). Nodules and mucosal elevations were less noted in the distal trachea. Histopathological examination of tracheal nodule biopsy showed normal tracheal mucosa with new cartilage and ossification in the submucous position. A diagnosis of hepatic tuberculosis associated to TO was made. Antituberculous therapy, including isoniazid, rifampicin, ethambutol and pyrazinamide, was initiated. Within a few weeks, progressive improvement of the general condition of the patient was observed but not chronic cough.

Discussion

TO is a rare benign condition characterized by growths of osseous and cartilaginous nodules arising in the submucosa of the trachea and major airways.2 This condition named ‘tracheopathia osteoplastica’ by Aschoff in 1910 was first described by Wilks in 1857 as ‘Ossific deposits on the larynx, trachea and bronchi’ in a 38-year-old man who died of pulmonary
Frequently, it is incidentally discovered at imaging or on intubation, but in some cases chronic cough, hemoptysis and obstructive symptoms such as wheezing or dyspnea can be suggestive. In our case, despite presence of chronic cough, the disease was incidentally discovered at imaging during exploration of a chronic abdominal pain.

Besides the case described by Wilks only one case of association with active tuberculosis has been reported in literature. Although the relative rarity of this disorder of unknown cause, the association with tuberculosis seems to be a coincidence. The theories regarding growths of osseous and cartilaginous nodules include metaplasia of submucosal tissue, cartilaginous exostosis of the tracheal rings and chronic airway inflammation. The imaging and endoscopic appearance are highly suggestive, but in atypical cases diagnosis can be confusing with other diseases such as primary tracheobronchial amyloidosis, polychondritis, sarcoidosis, neoplasms or papillomas. However, in tracheobronchial amyloidosis, the posterior wall of the trachea can be reached. In confusing cases, histopathologic examination after Congo red stain can be helpful. There is no effective treatment for TO.

In symptomatic patients, improvement can be achieved after administration of inhaled bronchodilator, humidification of the airways and treatment of infections of the respiratory system. For patients with severe airway narrowing and airflow obstruction, interventional bronchoscopy, radiotherapy and surgical resection therapy can be proposed.

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References