Clinics in Surgery

Case Report
Published: 11 Jul, 2016

Tracheobronchopathia Osteochondroplastica in a Patient with Leukoplakia

*Dhillon KV and Akst ML**

Department of Otolaryngology-Division of Laryngology, Johns Hopkins University, USA

Abstract

Objectives: We present a case of Tracheobronchopathia osteochondroplastica (TBOC) found incidentally in a patient being managed for recurrent vocal fold leukoplakia and dysplasia. Tracheobronchopathia osteochondroplastica is a rare disease of the airway.

Methods: Single case report

Results: We review the clinical manifestations, diagnosis, pathophysiology and treatment for our patient with this rare disease.

Conclusion: Tracheobronchopathia Osteochondroplastica is a rare benign disease of the airway which usually presents incidentally with no evidence of symptom. It is important to consider this disease on the differential of patients with classic airway findings.

Introduction

Tracheobronchopathia osteochondroplastica (TBOC) is a rare benign disease of the tracheobronchial tree that is characterized by multiple submucosal osseous and cartilaginous nodules along the entire length of the trachea. Histopathology has demonstrated these lesions as calcium phosphate deposits that form islands underneath the mucosa of the tracheal lumen. The overall incidence of this disease varies as reported in the literature—on average 1:125 to 1:6000 cases during bronchoscopy [1]. The presentation is often asymptomatic or may instead include non-specific respiratory complaints, with cough and dyspnea the most common. The etiology of TBOC is unclear, but it is considered secondary to chronic airway inflammation. The diagnosis is made on bronchoscopic findings and at times, CT imaging of the chest, with little role in histopathology to confirm findings other than to exclude other pathologies. Treatment is symptomatic, ranging from cough suppression to excision and dilation depending on severity of the airway compromise [2,3].

In this report, we present a case of tracheobronchopathia osteochondroplastica found incidentally in a patient being managed for recurrent vocal fold leukoplakia and dysplasia. We review the clinical manifestations, diagnosis, pathophysiology and treatment for our patient with this rare disease.

Case Presentation

A 77 year-old male presented with dysphonia and history of prior outside surgery for leukoplakia, with outside pathologic diagnosis of dysplasia. At time of presentation to our clinic, transoral stroboscopy revealed scar of the right vocal fold consistent with his prior surgery and persistent/recurrent leukoplakia. The patient had no complaints of dyspnea, cough or inspiratory stridor. With persistent disease and desire to re-establish pathologic diagnosis, the patient was taken to the operating room for suspension microlaryngoscopy and microflap excision of diseased vocal fold epithelium. During microlaryngoscopy, views of the anterior subglottis demonstrated a small nodular submucosal lesion along the anterior tracheal wall just beneath the cricoid cartilage. This anterior tracheal lesion was biopsied during the procedure. While the vocal fold leukoplakia was found on histopathology to be dysplasia without evidence of invasion, the final pathology of the proximal tracheal lesion was chronic inflammation.

Serial follow-up for the patient's vocal fold dysplasia was recommended, and approximately three years after surgery, recurrence of the vocal fold leukoplakia was noted. The patient elected in-office KTP laser treatment of his lesion, and with benefit of laryngeal anesthesia, tracheobronchoscopy was done with a flexible scope at the time of the procedure; of note, all prior exams in the office had been done with a rigid scope for magnified, brilliantly illuminated visualization of his vocal

OPEN ACCESS

Correspondence:
Lee M Akst, Department of Otolaryngology-Division of Laryngology, Johns Hopkins University, 601 N Caroline Street 6th floor, Baltimore, MD 2123, USA, Tel: 410 955 1654; E-mail: Lakst1@jhmi.edu

Received Date: *19 Jun 2016*
Accepted Date: *06 Jul 2016*
Published Date: *11 Jul 2016*

Citation:
Dhillon KV, Akst ML. Tracheobronchopathia Osteochondroplastica in a Patient with Leukoplakia. Clin Surg. 2016; 1: 1071.

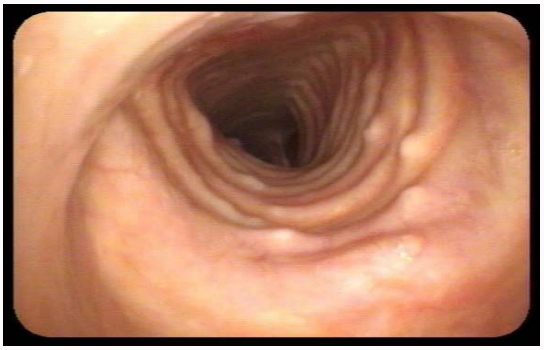

Figure 1: Characteristic for TBOC.

folds, and this was the first time a flexible scope had been used for this patient in this practice. Findings of tracheobronchoscopy (Figure 1) were characteristic for TBOC. The patient never endorsed any airway symptoms, cough, hemoptysis, pneumonia etc, and therefore no further treatment was indicated. To the present day, the patient continues to be monitored for his vocal cord leukoplakia with no evidence of disease and no evidence of any airway complaints.

Discussion

The existing knowledge about the rare entity of TBOC is largely from case reports and case series, with the first report of TBOC based on autopsy findings. The condition was described by Wilks in 1857 as ossific deposits in the anterior portion of the trachea [4]. In 1896, Von Schroetter became the first to use a laryngeal mirror to document these lesions on a living patient [5]. Shortly thereafter, Killian described these lesions using bronchoscopy in 1899 [2]. In 1910, Ascoff was credited for coining the name tracheobronchopathia osteochondroplastica [4].

The largest case study described is by Leske et al with 41 patients [6]. Most other case series are smaller, between one and three patients, all with variable presentations.

Overall the disease entity is rare, but its relevance is important in the laryngology field with respect to presentation and evaluation on bronchoscopy.

The best approximation of incidence of TBOC is cited as 11 per 10,000 cases [7]. There is no difference in distribution between males and females, and family history does not appear to play role. The average age of diagnosis is between the fourth and seventh decade [2]. It is a benign disease, with most cases incidentally found on bronchoscopy or autopsy. Patients are normally asymptomatic on diagnosis, and it is thought that those who become symptomatic may have acquired a superimposed infection or progressive airway stenosis secondary to the lesions. In turn, they present with non-specific complaints, the most common ones cited being cough and dyspnea with exertion [7].

The etiology of TBOC is unknown. Theories have been formulated regarding the pathophysiology of the disease. These have included chondrosis and exostosis of the tracheal cartilaginous rings or metaplasia of the submucosal stroma [7]. Associations with other diseases such as atrophic rhinitis and neoplastic processes have been reported, but there is no science to consistently demonstrate this relationship [2,7].

The histopathology of these lesions demonstrates calcium and phosphate deposits within small islands under the tracheal mucosa. More recently, factors such as bone morphogenetic protein 2 and transforming growth factor beta 1 have been implicated in potentially playing a role in formation of new bone and cartilage [5]. These calcium and bone deposits are in direct contact with the natural C-shaped anatomical structure of the tracheal rings - and therefore TBOC deposits are limited to the anterior and lateral walls of the trachea. The posterior wall of the trachea is membranous and is therefore spared [7]. This is an important trait in narrowing the differential diagnosis of tracheal lesions, and posterior wall involvement of the trachea increases suspicion for other disease pathologies including amyloidosis, sarcoidosis, polychondritis and malignancies such as lung cancer [7,8]. The role of biopsy is to confirm diagnosis and evaluate for associated conditions. Bronchoscopy is considered the gold standard for the diagnosis of TBOC.

Visualization of the bony or cartilaginous nodules on the tracheal walls is diagnostic of TBOC, more commonly found in the distal 2/3 of the trachea [8]. Some case studies have discussed involvement of the proximal trachea including subglottis and larynx [8], in which laryngoscopy may be sufficient to establish a diagnosis.

Though laryngoscopy should be pathognomonic for the condition, its rarity means that many bronchoscopists are unfamiliar with its appearance, and biopsy is often performed [9].

The role of imaging in diagnosis is uncertain. Jabbardarjani et al. [3] demonstrated the low yield of chest X-ray in detection of tracheal lesions. Computed tomography (CT) of the chest plays a role in demonstrating the sessile submucosal nodules along the anterior and lateral tracheal cartilaginous rings, when present. Such findings however may not be universally present [3]. Other ancillary tests such as pulmonary function tests and laboratory tests such as CRP and ESR are non-specific [1].

There is no specific treatment for TBOC with most treatment reserved for patients presenting with severe airway obstruction presenting with debilitating symptoms. A recent case series demonstrated all symptomatic patients (n=10) underwent removal of nodules with the use of Nd:YAG laser [3]. The other options include surgical resection, mechanical debulking with the use of rigid bronchoscope, stent placement, and medial management including inhaled steroids, antibiotics and expectorants [1-3,8]. Medical therapies are considered palliative or for symptomatic treatment only. There is a relatively good prognosis for these patients, with slow progression of the disease. Leske et al. [6] reported that over 55% of the patients do not show any disease progressive following diagnosis. Treatment modalities should be implemented on a case-by-case basis pending the severity of the disease.

Our case presentation is an incidental finding of TBOC on bronchoscopy in a patient that was otherwise asymptomatic from an airway standpoint. Our goals at this time are continued surveillance of these lesions, as there is evidence for progression of disease as documented in other case reports. Given the rarity of this disease, it is important to consider those who have been undiagnosed. Tracheobronchopathia osteochondroplastica is a disease that should remain on the differential when there are diffuse lesions along the tracheal wall. Treatment is done on a case-by-case basis, but continued surveillance of findings and symptoms is the mainstay in management for the majority of these patients.

References

1. Simmons C, Vinh D, Donovan DT, Ongkasuwan J. Tracheobronchopathia Osteochondroplastica Laryngoscope. 2015.
2. Agarwal A, Klair J, Joiner A, Meena NK. Nodular trachea: tracheobronchopathia osteochondroplastica. BMJ Case Rep. 2015; 2015.
3. Jabbardarjani HR, Radpey B, Kharabian S, Masjedi MR. Tracheobronchopathia osteochondroplastica: presentation of ten cases and review of the literature. Lung. 2008; 186: 293–297.
4. Wilks S. Ossific deposits on the larynx, trachea and bronchi. Trans Pathol Soc Lond. 1857; 8: 88.
5. Tajima K, Yamakawa M, Katagiri T, Sasaki H. Immunohistochemical detection of bone morphogenetic protein-2 and transforming growth factor beta-1 in tracheopathia osteochondroplastica. Virchows Archiv. 1997; 431: 359-363.
6. Leske V, Lazor R, Coetmeur D, Crestani B, Chatté G, Cordier JF, et al. Grouped'Etudeset de Recherchesur les Maladies "Orphelines" Pulmonaires (GERM"O"P). Tracheobronchopathia osteochondroplastica: a study of 41 patients. Medicine. 2001; 80: 378–390.
7. Nielsen SW, Stevens JR, Dion GR, Howard NS. Dyspnea, Dysphonia, and Cough: Varied Presentations of Tracheobronchopathia Osteochondroplastica. Ann Otol Rhinol Laryngol. 2015; 124: 829-833.
8. Ulasli1 SS, Kupeli K. Tracheobronchopathia osteochondroplastica: a review of the literature. Clin Respir J. 2014.
9. Abu-Hijleh M, Lee D, Braman SS. Tracheobronchopathia osteochondroplastica: a rare large airway disorder. Lung. 2008; 186: 353-359.