International Journal of Medicine and Medical Sciences Vol. 4(10), pp. 211-213, December 2012
Available online at http://www.academicjournals.org/IJMMS
DOI: 10.5897/IJMMS12.111
ISSN 2006-9723 ©2012 Academic Journals

Case Report

Tracheobronchopatia osteochondroplastica: An underdiagnosed central airway disease

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Accepted 6 November, 2012

In the population of patients who do not smoke, do not take an angiotensin-converting-enzyme inhibitor, and have a normal chest radiograph, upper airway cough syndrome, asthma, gastroesophageal reflux disease and chronic bronchitis are responsible for approximately 99% of cases of chronic cough. The diagnosis is often based upon the patient's response to empiric therapy; nevertheless, there still remains 1% with an undiagnosed etiology. We report a case of a 76 year old female who had presented with persistent cough for roughly four decades requiring hospitalizations on multiple occasions. Despite receiving empiric therapy and the standard of care management for bronchial asthma, her symptoms were progressively getting worse. Physical examination and laboratory workup were unremarkable. Pulmonary function test was abnormal; the flow volume loop demonstrated a flattening of the inspiratory phase, which resulted into why an extrathoracic obstruction was suggested to require further workup. A chest computed tomography revealed a proliferation of bone and cartilage in the anterior and lateral walls of the trachea sparing the posterior wall. Multiple submucosal sessile cartilaginous nodules were detected by flexible bronchospy. Tissue sampling resulted in a mucosal squamous metaplasia, all findings consistent with tracheobronchopatia osteochondroplastica. The patient was treated with laser therapy with physiological and clinical response.

Key words: Tracheobronchopatia osteochondroplastica, extrathoracic obstruction, osteocartilaginous nodules, central airway obstruction, chronic cough.

INTRODUCTION

Tracheobronchopathia osteochondroplastica is an idiopathic rare disease of the trachea and major bronchi characterized by multiple submucosal osteocartilaginous nodules that protrude into the lumen sparing the posterior wall. The first case was described in 1857 as "Ossific deposits on the larynx, trachea and bronchi" found incidentally in a patient at an autopsy (Abu-Hijleh et al., 2008; Leske et al., 2001). Cases reported in 1863 were termed as "ecchondrosis and exostosis" (Leske et al., 2001). It was not until 1910 when the entity was named "tracheobronchopathia osteochondroplastica" due to its clinical presentation (Leske et al., 2001).

TBO is an unknown entity seen with a frequency of 0.4% at bronchoscopy (Chroneou et al., 2008). There is no gender predominance and the disease typically manifest in patients in their mid-50s. The clinical presentation is variable, ranging from incidental diagnosis in asymptomatic patients to devastating disease, causing central airway obstruction (Tukiainen et al., 1998). Due to recent advances in modern technology, the disease has been well recognized early in its course and prompt treatment implemented.

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Abbreviations: **TBO,** Tracheobronchopatia osteochondroplastica; **PFT,** pulmonary function test; **FEV1,** forced expiratory volume in one second; **FVC,** forced vital capacity.

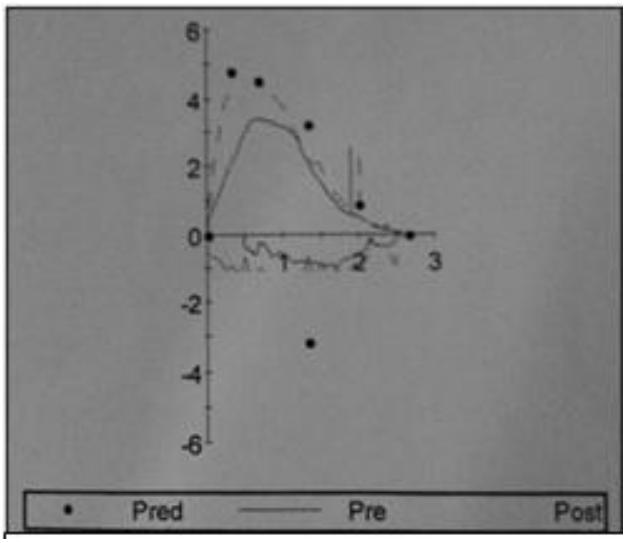

Figure 1. Flow volume loop demonstrating flattening of the inspiratory phase.

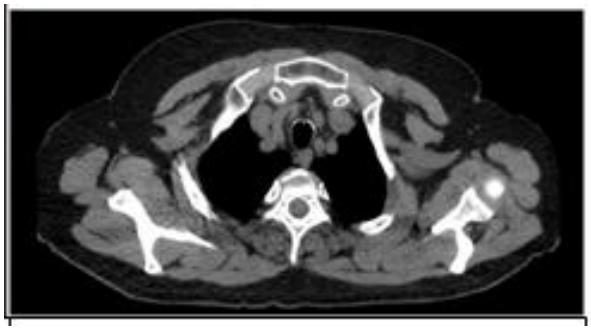

Figure 2. Chest tomography with poliferation of bone and cartilage in the anterior trachea

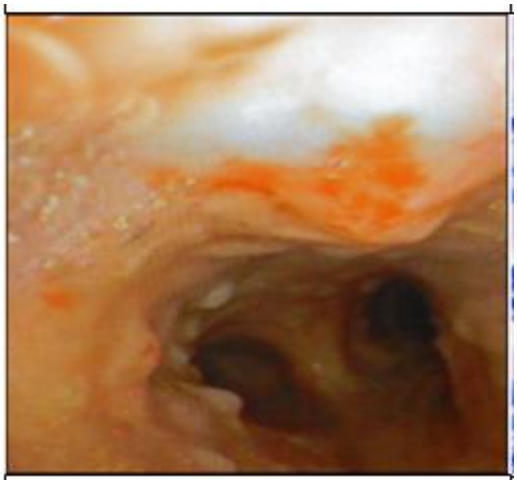

Figure 3. Bronchoscopy displaying multiple submucosal sessile cartilaginous nodules.

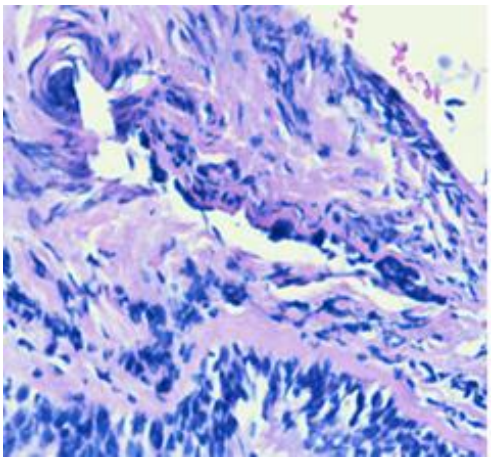

Figure 4. Giesma stain exhibiting mucosal squamous metaplasia.

CASE REPORT

We present a case of a 76 year-old female, non-smoker, with a past medical history of arterial hypertension, hypothyroidism, dyslipidemia and bladder cancer with tumor resection at the age of 35. Neither chemotherapy nor radiotherapy was necessary for the above condition at the time of diagnosis and no occupational or environmental exposures to fumes or dust was pertinent in the history. The patient sought medical attention after complaining of persistent nonproductive cough for the last forty years requiring hospitalizations on several occasions. Within this period, she always had been treated with the usual management for bronchial asthma since her clinical manifestations were misdiagnosed with this obstructive and chronic disease.

Despite receiving the standard of care management for bronchial asthma, her symptoms had been worsening over the past few years. At age 73, as part of the initial evaluation, the physical examination and laboratory workup were unremarkable including complete blood count, complete metabolic panel and thyroid function. No abnormal findings on chest auscultation such as inspiratory/expiratory rhonchi, wheezing, stridor, nor crackles were detected. PFT was significant for FEV_1 1.93 L (94% predicted value) and FEV_1/FVC 76% (101% predicted value), nevertheless the flow volume loop was remarkable for flattening of the inspiratory phase (Figure 1), reason why an extrathoracic obstruction was suggested leading to further studies. These included a chest computed tomography (Figure 2) which revealed a proliferation of bone and cartilage in the anterior and lateral walls of the trachea just to the level of the carina and proximal half of bilateral main bronchi sparing the posterior wall suggestive of TBO. In order to have a definite diagnosis of the condition, a bronchoscopy (Figure 3) was performed which was remarkable for multiple submucosal sessile cartilaginous nodules. The biopsy (Figure 4) demonstrated mucosal squamous metaplasia, commonly seen in TBO. The patient was treated with laser therapy with physiological and clinical response. Further, PFT demonstrated an improvement of the previously seen obstructive impairment and flattening of the inspiratory phase.

DISCUSSION

The etiology remains uncertain, though several theories

about the pathogenesis have been linked to bone morphogenetic protein-2 (BMP-2) and transforming growth factor β1 (TGF-β1) (Prince et al., 2002). Due to the indistinct symptomatology which chronic cough appears to be the most common clinical presentation, the disease can go unnoticed and misdiagnosed. Nevertheless, with advances in techniques including the development of bronchoscopy and airway images, the detection has been increasing although still the overall incidence remains low. Treatment is reserve for symptomatic patients. It is palliative and it includes local laser, mechanical debulking using a rigid bronchoscope, and endobronchial stent placement. The outcomes are variable. An untreated condition can lead to a devastating outcome due to worsening of the obstructive disease for which clinicians should consider as a differential diagnosis. Hence, primary care physicians should be aware that obstructive patterns in the PFTs not only are secondary to conditions such as Chronic obstructive pulmonary disease (COPD) or bronchial asthma.

ACKNOWLEDGMENTS

A special note of thanks to Dr. Samuel Suárez Báez, MD, FCCP at the Ashford Medical Center who made this paper possible and for sharing his enthusiasm and resources. Thank you for your commitment to education. I would like to express my gratitude to the Center for Technological Support in Academia (CATA, by its Spanish acronym) Medical Science Campus-UPR for facilitating photography service.

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