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Tracheobronchopathia osteochondroplastica in the setting of COVID-19

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Abstract

Tracheobronchopathia osteoplastica (TO) is a rare, benign disease of unknown etiology, primarily affecting the major tracheobronchial tree, characterized by irregular nodular calcifications of the cartilaginous component of the inner wall of the tracheobronchial tree while sparing the posterior wall, leading to progressive narrowing of the airway. We report the case of a 60-year-old male otherwise healthy nonsmoker, who complained of chronic breathing discomfort and recurrent chest infections and was found to have TO according to radiographic, microlaryngoscopic, and biopsy findings. He experienced a flare up with worsening of disease progression after years of being in stable condition, after his infection with SARS-CoV-2.

Keywords

Tracheobronchopathia osteoplastica, larynx, stridor, COVID-19

Introduction

Tracheobronchopathia osteoplastica is an uncommon non-neoplastic disease that affects the trachea and major bronchi. The etiology remains unknown, with no genetic predisposition documented. The condition is more common in males, with diagnosis usually being made between ages 40 and 60, with no association to smoking. It is characterized by the development of cartilaginous or bony submucosal nodules, mainly involving the airway cartilages and extending into the tracheobronchial lumen. The membranous posterior wall of the trachea typically is spared due to its lack of cartilage support.[1]

In most cases, the disease is asymptomatic, and it is often discovered incidentally either on autopsy, or with intubation, bronchoscopy, or radiographic imaging. When symptomatic, it most commonly presents as persistent or recurrent cough, exertional dyspnea, recurrent lower respiratory tract infections, and occasionally hemoptysis.[1]

Due to the resemblance in presentation to diseases such as bronchial asthma, the diagnosis of TO is often missed or delayed, with patients being managed as cases of asthma with no improvement in their condition. Therefore, it is important to increase awareness of this condition to improve patient outcomes and guide healthcare workers in their diagnostic approach.

Case report

A 60-year-old otherwise healthy male non-smoker, presented to our tertiary center complaining of chronic shortness of breath on exertion and cough of many years, with a history of recurrent hospital admissions due to bronchopneumonia.

His condition began with mild symptoms, gradually worsening with time, and leading to his admission to the internal medicine department multiple times for bronchopneumonia

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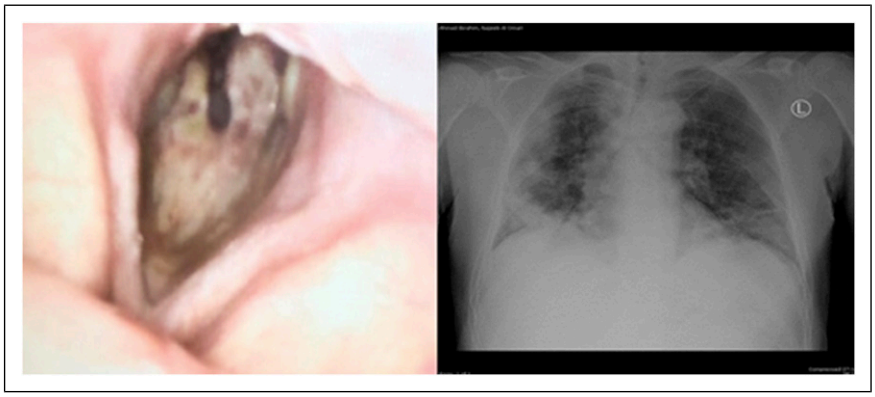

Figure 1. Flexible laryngoscopy showing rigid larynx and extensive calcification with a severely stenosed tracheal lumen mainly involving the anteriolateral wall. Chest x-ray showing bronchopneumonia with significant narrowing of tracheobronchial tree.

requiring IV antibiotics and supportive treatment. The reason for his recurrent chest infections was unknown.

The patient was referred to our otolaryngology department for further evaluation and to rule out an upper respiratory cause for his shortness of breath. He had no significant history of weight loss, and the patient's head and neck examinations were unremarkable. Flexible laryngoscopy revealed rigidity in all the cartilaginous parts of the larynx with thickening in the upper tracheal rings. A simple chest x-ray showed irregular narrowing in his trachea and the major bronchial airway primarily, with some narrowing in the small bronchi and bronchioles **(**Figures 1**).**

CT of his neck and chest revealed abnormal calcifications and irregular nodular thickening in the anterolateral walls of the trachea and major bronchi, sparing the posterior wall **(**Figure 2**).**

Due to the severity of his symptoms, we admitted the patient for diagnostic bronchoscopy and supportive treatment.

Rigid bronchoscopy revealed a diffusely nodular, irregular inner wall of the trachea, sparing the posterior wall, with severe narrowing in a small upper segment of the trachea, leading us to perform limited balloon dilatation of the stenosed segment, with multiple punch biopsies taken from the inner wall of the trachea.

Histopathology showed abnormal metaplasia of the osteocartilaginous elastic tissue of the tracheal mucosa.

Postoperatively, the patient returned to his usual health status and daily activities, with improvement in his shortness of breath and cough. He spent a period of 2 years after his surgery with no deterioration in his condition, no chest infections, and only mild symptoms.

Within the first few months of the COVID-19 pandemic, the patient developed acute exacerbation in his shortness of breath, as well as severe cough, high fever, myalgia, and generalized weakness. The patient presented due to worsening of his symptoms which were similar to those seen with COVID-19. A COVID-19 test was positive. He was admitted for severe COVID-19 with exacerbation of TO symptoms to the intensive care unit for supportive management and close monitoring. 48 hours after admission, his condition worsened, and he was found to have COVID-19 bronchopneumonia with worsening of TO as shown on x-ray **(**Figure 2**).**

After recovering from COVID-19 and being discharged, the patient noticed persistent shortness of breath and cough, with a return to his pre-balloon-dilatation state according to the patient. Soon thereafter, he developed mild stridor with worsening shortness of breath, which led him to seek care at our clinic. On reevaluation, he had no audible stridor or wheeze. However, repeat flexible laryngoscopy showed worsening in tracheal patency and progression of the disease in comparison with his previous findings, the deterioration was felt to be due to the effect of the recent COVID-19 infection on the patient's airway.

Follow-up flexible laryngoscopy 4 months later showed further rapid progression of the disease, with a rigid larynx and extensive calcifications, and a severely stenosed tracheal lumen mainly in the anterolateral wall.

Discussion

Clinical features and presentation

Tracheobronchopathia osteochondroplastica (TO) is a rare, benign disease of the endobronchial system with nonspecific symptoms and various treatment approaches.[2]

The condition was first macroscopically described by Rokitansk in 1855, and microscopically described by Wilks in 1857. Some theories have been formulated about the pathogenesis of this condition, with Dalgaard stating that the elastic tissue suffers metaplasia, with cartilage formation and calcium deposition; Virchows theory was that echondrosis and exostosis promote calcium deposition and ossification of the tracheal rings. Aschoff-Freiburg reported changes to the tracheal elastic tissue, and used the term osteoplastic tracheopathy to describe the condition, and similarly, in 1964, Secrest et al. labeled it osteoplastic tracheobronchopathy.[3]

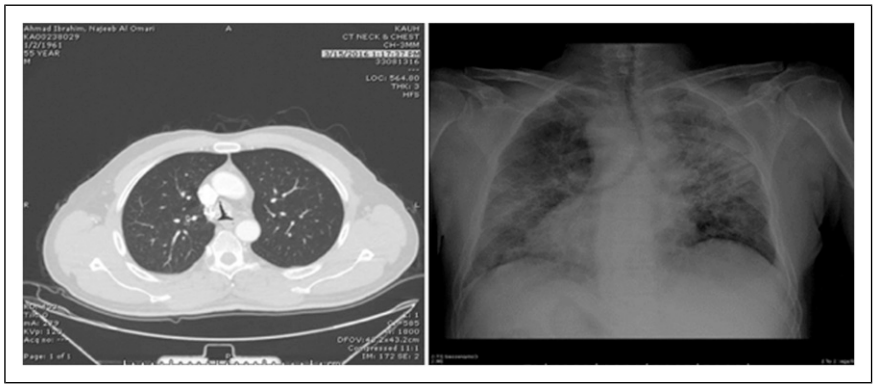

Figure 2. Axial view of CT chest revealed severe narrowing and calcifications in the anterolateral wall of the upper trachea. Chest X-ray of TO patient post-COVID-19 infection.

TO is a very rare disease mostly diagnosed as an incidental finding by bronchoscopy or on autopsy, with most patients diagnosed being between the ages of 50 and 70 years, and patients in the 5th decade of life being the most frequently affected.[4] According to Secrest, it is estimated that only 5% of the cases are diagnosed during the person's life.[3]

While most patients with TO are asymptomatic, when symptomatic, common presenting symptoms are chronic cough, dyspnea, wheeze, hemoptysis, and recurrent respiratory tract infections, which often lead to the misdiagnosis of asthma.[5]

Histopathological findings

Histologically, the mucosal bed may look normal, with areas of inflammation and necrosis, as well as abnormal proliferative cartilaginous or bony formations on the submucosa. Often, you may find squamous metaplasia of the columnar epithelium, calcium deposits, fragments of adipocytes, and active hematopoietic medullar bone tissue are seen.[6]

Comparatively, in our patient, histopathological study of the intraoperative biopsies taken showed fragments of bony tissue lines by respiratory epithelium, as well as the presence of hematopoietic cells.

Radiological findings

Plain chest radiography may show irregularity and narrowing of the affected segments of the tracheobronchial tree, similar to our patient's X-ray findings. CT of the chest may show irregular thickening and nodularity of the tracheal cartilage, sparing the posterior (membranous) tracheal wall.[7] CT of the neck and chest done for our patient revealed evidence of irregular thickening, nodularity, and calcifications of tracheal cartilage involving the anterior and lateral walls while sparing the posterior (membranous) wall extending down to the proximal portions of both main stem bronchi, causing significant luminal narrowing.

Diagnosis and treatment

Definitive diagnosis of the disease is confirmed through a combination of typical laryngoscopic, radiographic, bronchoscopic, and biopsy findings. The typical bronchoscopy findings are often described vividly as a cobblestone, beaded, or stalactite cave or a rock garden appearance.[8,9] During our patient's bronchoscopy, we saw multiple subglottic hard, bony osteoma like masses causing airway stenosis.

There is no specific treatment for TPO. Recurrent infections and atelectasis are seen as complications during the disease course and are treated in conventional fashion. Intubations, if required, may be difficult because of the calcified rings of trachea. Occasionally, tracheostomy is required. Surgical treatment options should be considered when conservative measures have been failed. Resection of the affected tracheal segment, anterior laryngo fissure, partial laryngectomy, removal of lesion with bronchoscopy, and rigid bronchoscopic dilatation are also reported as surgical options as it is difficult to remove bony lesions with a rigid bronchoscope.[10]

In conclusion, tracheobronchopathia osteochondroplastica, while a rare condition, is likely more common than previously thought, and due to the resemblance in presentation to common conditions such as bronchial asthma, it is likely to be under or misdiagnosed. Therefore, it is important for otolaryngologists to be familiar with this condition and its presentation, and respiratory physicians and otolaryngologists alike must be vigilant when dealing with patients with similar presentation.

With the advent of the COVID-19 pandemic, given the rapid progression of our patient's condition after recovery from COVID-19, and taking into account the lack of

further data comparing the relationship between COVID-19 and TO, we believe physicians should be aware of a possible relationship between the 2 entities. However, future studies are recommended to clarify this possible relationship.

As there are no clear guidelines for the management of TO, our plan is to follow-up with our patient regularly with repeat flexible laryngoscopy and with chest x-rays, in collaboration with regular pulmonary evaluation as long as there is no deterioration in his condition. In the event of worsening in our patient's respiratory condition, our team may consider performing long lumen tracheotomy, with possible further dilation or laser ablation to improve airway patency.

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