Case Report

Tracheobroncopathia Osteochondroplastica: Three Case Reports With Literature Review

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ABSTRACT

Tracheobroncopathia osteochondroplastica (TO) is a benign disease of the large airways seen very rarely. It is characterized by 1-3 mm sized ossified nodular lesions in submucosa. Its etiology is unclear, but it is stated that malignancy, chronic inflammation, amyloidosis, and genetic factors might have an effect on it. It was first described by Wilks in a 38-year-old man diagnosed with tuberculosis in 1857. Generally, patients are asymptomatic and TO is diagnosed incidentally. But symptoms become significant with infections and obstruction in tracheabronchial tree. Generally chest radiography is normal, so thorax computed tomography can be remarkable in diagnosis of TO. Besides, final diagnosis can be established by viewing ossified nodules in trachea and bronchus through the fiberoptic bronchoscopy. Amyloidosis, tuberculosis, sarcoidosis, bronchial carcinoma, and tracheobronchial calcinosis must be remembered in differential diagnosis. Also ossifications in submucosa and proof of bone marrow in histopathological examinations are important in diagnosis of TO. Mostly palliative treatment is performed to the symptoms . We want the clinicians to keep in mind for this very rarely seen tracheal disease with three case reports.

KEYWORDS: *Ossificated nodular lesion, thorax computed tomography, tracheal disease, tracheobroncopathia osteochondroplastica*

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INTRODUCTION

Tracheobroncopathia osteochondroplastica (TO) is a very rare benign disease of the tracheabronchial tree with unknown etiology. It is significant with cartilaginous and ossified nodules located in submucosa.[1] Almost 400 cases have been stated since its first definition.[2] It is suggested that TO is seen after 4th decade with male predominance in literature and because of its asymptomatic nature, it is diagnosed incidentally.[2,3] Treatment is generally palliative and nonspecific.

CASE REPORTS

Case 1

A 52-year-old female patient was admitted to our clinic with complaints of dsypnea, coughing, sputum, and chest pain. She complained of dsypnea for several years and was diagnosed with asthma 10 years ago. There was no exposure with irritant agents and toxic gas inhalation in anamnesis. She is nonsmoker and has no medication except bronchodilatators. In physical examination, stridor and wheezing were identified. Laboratory findings and chest radiography were normal, but pulmonary function tests were concordant with mild obstruction. As her complaints increased , thorax computed tomography was performed and ossified nodules were detected in thorax CT [Figure 1a]. Then fiber optic bronchoscopy (FOB) was performed for hyperdens nodules in thorax CT. Hard, expansible, cartilaginous, and ossified nodular lesions the largest of which was 5 mm were determined in bronchoscopy [Figure 1b]. Nodules located through the whole trachea beginning from vocal cords. Bronchoscopic biopsies were conducted from nodular lesions. Histopathologically, ossification sites in submucosa were detected with normal bronchial mucosa [Figure 1c]. She was treated with antibiotics, water vapor, budenoside, and salbutamol nebulization. She relaxed after palliative treatment and was followed-up.

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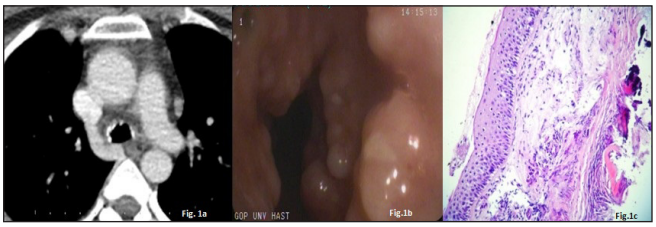

Figure 1: a-c: The 52-year-old female patient. (a): In thorax computed tomography :Multiple calcified nodulary lesions are seen in the anterior wall of trachea. (b): In fiberoptic bronchoscopy: White, bright, and hard cartilagenous nodulary lesions are seen originating from anterior and lateral walls of trachea (" rock-garden" or "cobble stone") sight. (c): In histopathological examination: A few number of mononuclear inflammatory cells in subepithelial stroma and calcified osteoid material are seen (HE × 100)

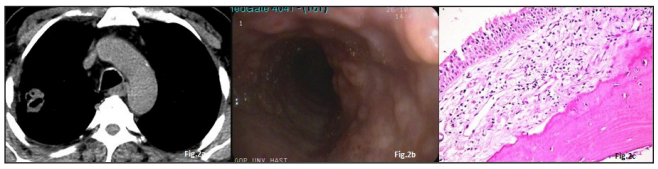

Figure 2: a-c: The 71-year-old male patient. (a): In thorax computed tomography: Multiple calcified and ossified nodules located in the anterolateral wall of trachea and also a cavitary lesion are seen at the right upper lobe. (b): In fiberoptic bronchoscopy: Multiple calcified hard nodules protruded into the airway lumen from the tracheal submucosa and cartilage tissue are seen. (c): In histopathological examination: Mature ossification is seen in subepithelial stroma (HE × 100)

Case 2

A 71-year-old male patient was admitted to our clinic with complaints of hemoptysis and cough . He was diagnosed with pulmonary tuberculosis (TB) and in follow-up about 2 months with the treatment of anti-TB (INH, RIF, ETH, PYR). Also he was diagnosed with diabetes mellitus since 20 years. Because of TB and hemoptysis under the treatment of anti-TB treatment, we decided to examine him with thorax CT. In thorax CT, there were many cavitary lesions in the lungs especially at the right upper lobe . Also we detected calcified nodular lesions in trachea when we evaluated the images of thorax CT carefully [Figure 2a]. We offered the patient FOB to investigate the bronchial tree for hemoptysis and calcified nodular lesions. In FOB, we identified many calcified nodular lesions and conducted biopsies on these lesions [Figure 2b]. Also we detected a little hemorrhage from right upper lobe and cold saline solution with adrenalin was used to stop the hemorrhage. In follow-up, the bleeding stopped and histopathological examination of biopsies from tracheal wall confirmed the submucosal calcification and ossification [Figure 2c].

Case 3

A 69-year-old male patient was admitted to our clinic with complaints of dyspnea, cough, and chest pain. In physical examination wheezing and clubbing were identified. Because of left hilary fullness was seen in chest x-ray, he was examined with thorax CT in detail. In thorax CT besides the diffuse ground glass opacities and hilar lymphadenopathies, a lot of calcified nodular lesions were detected at the anterior trachea wall [Figure 3a]. So we offered FOB to evaluate the radiological findings in thorax CT with bronchoalveolary lavage and bronchoscopic biopsy. The submucosal nodules from the anterolateral wall of trachea were determined in FOB[Figure 3b]. The tissue samples from the nodular lesions were obtained through FOB. The results of histopathological examination demonstrated submucosal ossificiation and cartilage tissue in the biopsy samples[Figure 3c].

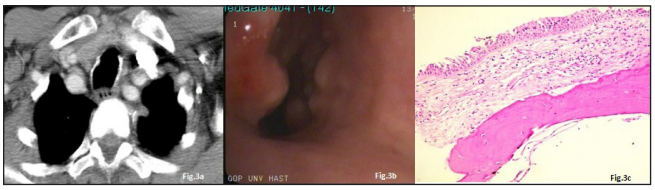

Figure 3: a-c: The 69-year-old male patient. (a): In thorax computed tomography: Calcification and ossification with nodular lesions are seen at the lateral wall of the trachea. (b): In fiberoptic bronchoscopy: Nodules originated from anterolateral wall of trachea are seen and they restricted the tracheal lumen. (c): In histopathological examination: Ossified subepithelial stroma and cartilage tissue is seen (HE × 40)

Discussion

TO is a benign disease of trachea and bronchus with unknown etiology. The disease consists of cartilaginous and ossified nodular lesion under the submucosa. It was first defined by Wilks in a 38-year-old man with TB in 1857.[3] At the same time TO was identified by Rokitansky and Luschka in the middle of 19th century. In literature, less than 400 cases have been reported so far.[2] The incidence is 0.01-4.2% in bronchoscopy. Jabbardarjani *et al*.[4] notified the prevalence as 0.11%.

The etiology of TO has not been identified yet. Although TO is associated with amyloidosis, degenerative and metabolic anomalies, genetic predispositions, congenital anomalies, chronic infections, inflammation, trauma and silicosis , there is no clear proof.[4,5] In literature, association between TO and atrophic rhinitis (ozena) is more distinct than others.[3,4] Generally there are two theories identifying the pathophysiology of TO. The first one was defined by Virchow in 1863. It claims that echondroma is the main lesion and exposure with calcification and ossification lead to nodular formations. The second theory is metaplastic theory, defined by Aschoff–Freiburg and also suggested by Dalgaard. This theory states that nodular lesions occur as a result of metaplasia and ossification of connective tissue. According to these theories, the bone morphogenic protein-2 and transforming growth factor

β1 are important facts for nodular formations in tracheal mucosa.[2,5]

TO is a benign and generally asymptomatic disease. It is identified incidentally in intubation or bronchoscopy. But sometimes it can be symptomatic, then it is diagnosed. In TO, coughing is the most common symptom at initial diagnosis.[2] Leske *et al*.[5] confirmed coughing as the most common symptom (54%) in their study with 41 cases. In a study with 10 patients of Jabbardarjani *et al*.[4] dsypnea was the most common symptom.

The radiological imaging is the most helpful method in diagnosis of TO. Chest radiography is mostly normal in TO, but important in differential diagnosis of other lung diseases imitating TO such as pneumonia, lung cancer, chronic obstructive pulmonary disease, and so on. Thorax CT is the most valuable radiological imaging method in diagnosis. Irregular, hyperdense, calcified nodular lesions extending in tracheal lumen can be determined in thorax CT.[2] The low signal intensity in imaging with magnetic resonance imaging is beneficial for nodular calcifications. But its application in TO is very rare.

Generally the respiratory function test is normal, all kinds of the obstruction pattern can be seen in respiratory function test. Usually restrictive pattern is caused by atelectasis or other restrictive diseases and it is seen rarely.[2]

Bronchoscopy is the gold standard in diagnosis of TO.[2] As the clinical manifestation is generally asymptomatic or the symptoms are nonspecific, it is difficult to diagnosis. Frequently, thorax CT is beneficial in diagnosis of TO and differential diagnosis from other diseases. But its credibility is not as high leveled as bronchoscopy. Especially, bronchoscopy and bronchoscopic biopsy are very important in differential diagnosis of TO with diseases, such as amyloidosis, chronic infection diseases, sarcoidosis, papillomatosis, bronchial carcinoma, and tracheal calcinosis.[2] Besides viewing the calcified and ossified nodular lesions in bronchoscopy, detecting the hardness of these nodular lesions by touching with bronchoscopy and forceps can be helpful in diagnosis.[2,3] This original view in bronchoscopy is called "rock-garden" or "cobble-stoned."[3] Histopathologically, the sight of cartilaginous and ossified sites with or without bone marrow in submucosa is significant for the diagnose of TO.

Usually the treatment is symptomatic and palliative. The general management like antibiotics in recurrent infections, humidification of the room air, and prevention of the mucosal irritation can be performed in treatment. Also, it is supposed that inhalation of steroids can be useful in palliative treatment of TO.[3] In life-threatening and serious cases, the resection of nodular lesions, cryotheraphy and endobronchial stent insertion can be preferred for conservative treatment in surgery.

Conclusion

TO is a benign disease with calcified and ossified nodular lesions originating from anterolateral walls of trachea and bronchus. Because of its silent clinical trial, it is generally diagnosed incidentally during the other evaluations. The malignant transformation in TO has not been reported yet. The serious symptoms and clinical manifestation like stridor, asphyxia, and respiratory failure can arise due to the severe obstruction in tracheal lumen. The clinicians can diagnose this incidental and asymptomatic disease easily and more frequently with the increase in using the bronchoscopy and thorax CT.

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Conflicts of interest

There are no conflicts of interest.

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