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Tracheobronchopathia osteochondroplastica limited to the trachea: a case report and rewiew of literature

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ABSTRACT

Tracheobronchopathia Osteochondroplastica (TO) is a rare and benign disease with a progressive course. Its clinical manifestation is observed on computed tomography by the presence of diffuse osteocartilage submucosal nodules invading the lumen of the trachea and usually the main bronchi. Bronchoscopic evaluation is necessary for differential diagnosis. Standard treatment has not been determined as few patients have been reported in the literature. A 58-year-old male patient was admitted to our hospital with complaints of cough and dyspnea increasing with exertion. A fiberoptic bronchoscopy demonstrated many protruding lesions along the trachea with no involvement of the bronchial system. The patient was stable at a two-year follow-up.

Keywords: Tracheobronchopathia osteochondroplastica, tracheal stenosis, benign tracheal lesions, bronchoscopy, trachea

Tracheobronchopathia Osteochondroplastica (TO) is an uncommon, progressive, and benign disease of undetected etiology. TO can be distinguished by diffuse osteocartilage submucosal nodules that invade through the lumens of both the trachea and usually the main bronchi [1, 2]. The natural course of the disease is progressive. TO can be distinguished by the presence of diffuse osteocartilage submucosal nodules which can invade both the trachea and main bronchial lumen; occasionally nodules are present only in the lumen of the trachea. In practice, a careful radiological examination is essential for diagnostic purposes, and bronchoscopy helps to eliminate differential diagnoses. There is no consensus on standard treatment as a consequence of few patients being reported in the literature [1-7]. Here, we present a case of TO who was involved with submucosal nodules only in the tracheal lumen and no more progress in the two-year follow-up.

CASE PRESENTATION

A 58-year-old male patient was admitted to our hospital with complaints of cough and exertional dyspnea that had been persisting for the last two years. TO was diagnosed one year ago by tomography, and a follow-up without treatment was recommended. He had no other underlying diseases or history of smoking. Symptoms were stable at the final presentation, and a thorax CT showed no difference from the previous series (Fig. 1A and 1B).

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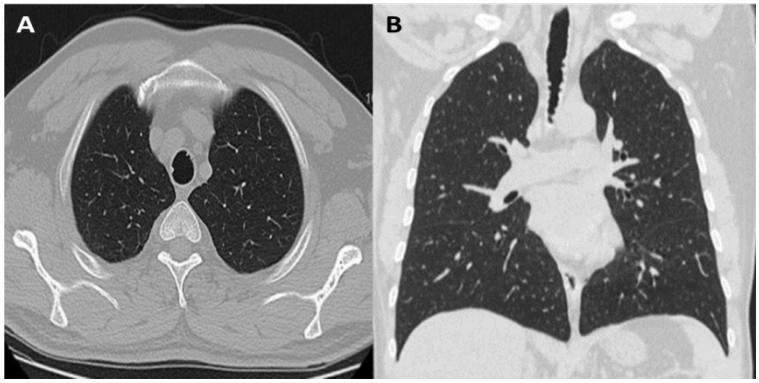

Fig. 1. (A) Axial and (B) coronal CT images showed irregular thickening and nodularity of tracheal cartilage.

A fiberoptic bronchoscopy was performed to evaluate the latest situation under general anesthesia. Several protrusions of lesions were observed throughout the trachea, narrowing down the lumen. The bronchial system was not involved (Fig. 2). Random biopsies were taken from plaques. The patient was discharged on the same day after the procedure, without medication, to come for an annual follow-up. The histopathologic consideration of the biopsies demonstrated osteocartilaginous structures (Fig. 3). There was no evidence of malignancy.

DISCUSSION

TO is an uncommon benign disease that is distinguished by submucosal nodules in the osteocartilaginous structure protruding towards the lumen in the trachea and usually in the main bronchi. The disease was described by Rokintansk in 1855 macroscopically. Wilks investigated microscopically by post-mortem a 38-year-old male who died of pulmonary tuberculosis in 1857 [1]. Von Schroetter proved the diagnosis by using a laryngeal mirror in vivo for the first time in an alive patient in 1896 [1, 2].

Although TO mostly has unknown etiology, some etiopathogenic causes have been identified. However, no genetic transmission has been demonstrated yet. Infections, metabolic diseases, inflammatory disorders, chronic irritation of chemicals, and smoking could be playing a role [2,3,4,7]. Some studies have reported that it may coexist with some malignant diseases [6]. It is more frequently seen in men over 50 years old [1, 2, 7, 8]. Our patient is a 58-year-old male. There were no underlying diseases or any other factors that could alter the etiology.

TO mostly remains asymptomatic [1-3]. Common symptoms are chronic cough, expectoration, dyspnea, recurrent respiratory infection, and wheezing; stridor may be added as the stenosis progresses [1, 2, 6]. He-

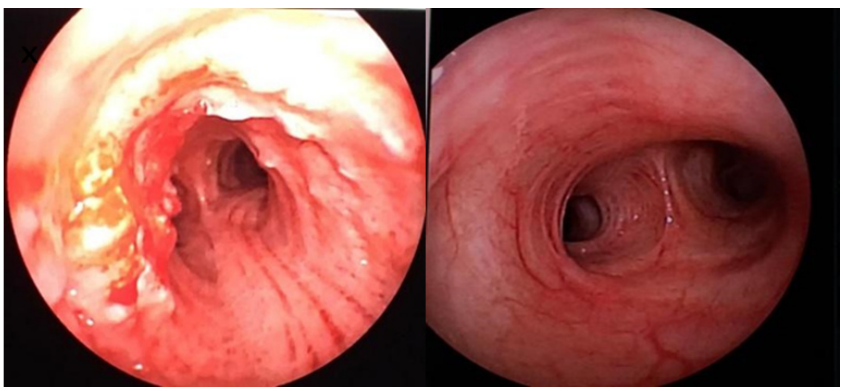

Fig. 2. Bronchoscopic view of the narrowed trachea with protruding lesions on three sides. The bronchial system is not involved by the lesions.

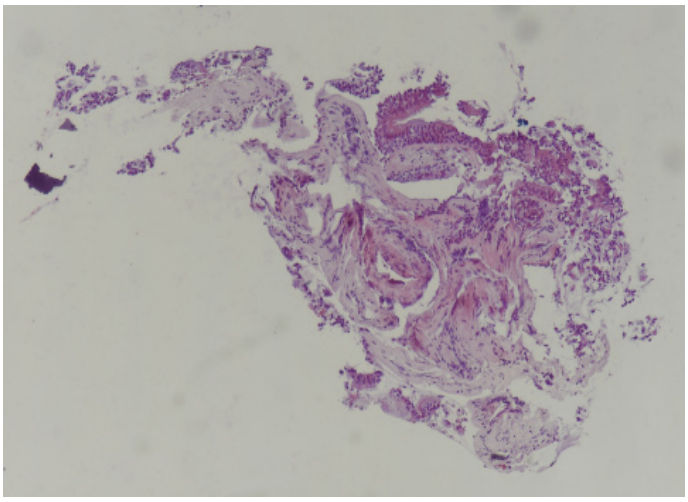

Fig. 3. View by Hematoxylin-eosin staining at 40x magnification. Among the ciliary pseudostratified columnar epithelium of the trachea, osteocartilaginous structures are distinguished.

moptysis is seen in 60% of cases with nodule ulceration or acute infections [9]. Physical examinations of the cases are generally normal [2, 7]. Only a small part of the patients had ventilation anomalies, such as bronchial obstruction findings. But most of the pulmonary function tests are within normal limits [1]. In our patient, there are no specific symptoms and no physical examination findings.

Abnormality and constricting of affected parts of the bronchi and trachea may be observed in a plain chest X-Ray. Calcium deposits can make trachea and bronchial borders as remarkable as lines. CT and bronchoscopy are both considered fundamental in the diagnosis of TO [3, 5, 8]. Radiological findings are fluctuating; however, they are usually overlooked unless observed by an attentive eye. Radiological findings are variable but generally missed unless a focused investigation. Even though CT images are not always diagnostic, CT can be used to detect nodules with irregular calcifications on the lateral and anterior walls of the airway "beaded appearance". Involvement of the posterior wall is not observed [2, 8].

Virtual bronchoscopy, which was started to be used with software added to the CT, provides the opportunity to evaluate the tracheobronchial lumen structures in cases where we cannot image due to stenosis and obstruction with fiberoptic bronchoscopies. Virtual bronchoscopy can also be used to detect possible extensions of pathology outside the lumen. Although tomographic images are sufficient for the diagnosis of TO, bronchoscopy is required. The bronchoscopic appearance is quite distinctive; the 'string of beads' appearance is caused by the submucosal cartilage surface containing abnormal mineralization deposition [2, 3]. In the differential diagnosis, amyloidosis, endobronchial sarcoidosis, calcified tuberculosis lesions, papillomatosis, tracheobronchial calcinosis, and primary or secondary malignant tumors of the trachea should be considered [2, 3]. For this reason, a bronchoscopic biopsy is recommended.

Histopathologically, a calcified protein matrix devoid of cells and submucosal nodules containing cartilage, bone, and blood elements, extending into the bronchial lumen is pathognomonic [1]. The nodules are often followed by squamous metaplasia, which may occur in the mucous columnar epithelium. Microscopic echondrosis may also be observed [1].

TO is a benign condition, so it can be followed without intervention in asymptomatic cases. There is no consensus on treatment because few patients have been reported in the literature. If the lumen narrows over time, interventions such as bougie or stent may be required. Conservative treatments should be applied for the patient's complaints. It has been reported that inhaled corticosteroids can reverse progression in some patients [8]. Reports of successful recovery with dapsone and NSAIDs have been confirmed in some symptomatic cases. The overall prognosis is generally good and the number of patients undergoing further intervention (such as a tracheostomy) is rare. Recommended treatments for severe airway obstruction or recurrent obstructive infection include laser ablation, cryotherapy, surgical resection, stent, and radiotherapy. Neodymium-doped yttrium aluminum garnet (Nd-YAG) laser treatment is considered for patients who develop tracheal stenosis. In case of respiratory tract infection, antibiotics can be added to the treatment [2, 7, 8]. Since our patient has not had any complaints yet, we did not recommend any medication.

CONCLUSION

TO is a rare benign but progressive disease. Although the underlying etiological causes cannot be determined exactly, some factors are thought to play a role. Bron-

choscopy is necessary for a definitive diagnosis. While the trachea and main bronchi are involved in most cases, it can sometimes be seen as isolated tracheal involvement, as in the case we presented. Although definitive treatment has not yet been defined, the approach is generally conservative; with close follow-up and relieving symptoms.

Authors' Contribution

Study Conception: ABK, EA; Study Design: ABK, EA; Supervision: ABK; Funding: ABK, EA; Materials: ABK, EA; Data Collection and/or Processing: ABK, EA, BK, PB; Statistical Analysis and/or Data Interpretation: ABK, BK, PB; Literature Review: BK, BK, PB; Manuscript Preparation: ABK and Critical Review: ABK, EA, BK, PB.

Informed Consent

Written informed consent was obtained from the patient for publication of this case and any accompanying pictures or data.

Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

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