

## Coexistence of tracheobronchopathia osteochondroplastica and lung cancer: a case report

*Trakeobronkopatia osteokondroplastika ve akciğer kanserinin birlikteliği: Olgu sunumu*

Ahmet İlvan,<sup>1</sup> Erhan Ayan,<sup>2</sup> Tuba Karabacak,<sup>3</sup> Oğuz Köksel,<sup>2</sup> Eylem Sercan Özgür<sup>1</sup>

*Institution where the research was done:*

Department of Chest Diseases, Medical Faculty of Mersin University, Mersin, Turkey

*Author Affiliations:*

Departments of <sup>1</sup>Chest Diseases, <sup>2</sup>Thoracic Surgery and <sup>3</sup>Pathology, Medical Faculty of Mersin University, Mersin, Turkey

Tracheobronchopathia osteochondroplastica (TO) is a rare disorder characterized by the projection of numerous bony or cartilaginous spurs into the tracheal lumen. This article reports a case in whom TO was detected accompanied by adenocarcinoma. A 56-year-old male presented with complaints of chronic cough and bloody sputum. Chest X-ray showed homogeneous opacity starting from the upper part of right hilus and extending to the lateral part. Thoracic computed tomography and bronchoscopy were performed. Tracheal lesion specimens were reported as TO and the lesion in the anterior segment of the right upper lobe was reported as adenocarcinoma.

**Key words:** Adenocarcinoma; bronchoscopy; cough; hemoptysis; osteochondrodysplasia.

Tracheobronchopathia osteochondroplastica (TO) is a rare, benign disease, with the earliest cases having been defined in the 1850s by Rokitansky and Wilks. The disease is characterized by nodular structures that involve bone and cartilage formations in the tracheal and bronchial mucosa. The etiology of TO is unknown, and it usually presents with a cough and hemoptysis. It is usually detected either incidentally or during examinations associated with the symptoms it causes, such as intubation difficulty, obstructive pneumonia, or accompanying lung disease, but sometimes it is not found until an autopsy is performed. This report describes an unusual case of TO accompanied by lung adenocarcinoma.

Trakeobronkopatia osteokondroplastika (TO) trakea lümenine doğru uzanım gösteren kemik veya kıkırdak dokusu ile karakterize nadir bir hastalıktır. Bu makalede adenokarsinomun eşlik ettiği bir TO olgusu sunuldu. Elli altı yaşındaki erkek hastanın kronik öksürük ve kanlı balgam tükürme yakınmaları vardı. Akciğer filminde sağ hilusun üst tarafından başlayan ve lateral kısma doğru genişleyen homojen opasite izlendi. Toraks bilgisayarlı tomografi ve bronkoskopi yapıldı. Örneklenen trakeal lezyonlar TO olarak, sağ üst lobun ön segmentindeki lezyon ise adenokarsinom olarak bildirildi.

**Anahtar sözcükler:** Adenokarsinom; bronkoskopi; öksürük; hemoptizi; osteokondrodysplazi.

### CASE REPORT

Our patient presented with a nine-month history of an occasional cough and sputum containing streaks of blood. He had been smoking cigarettes for 60 pack/years and also had hypertension and diabetes mellitus (DM). His physical examination was unremarkable, but a chest X-ray revealed a partially homogeneous opacity measuring 2x4 cm that started on the upper part of the right hilus and extended towards the lateral chest wall (Figure 1). The patient's hemogram was normal, and he had an erythrocyte sedimentation rate (ESR) of 44 mm/hour and a C-reactive protein (CRP) level of 2.83 mg/L. Routine blood biochemistry and urine



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Correspondence: Eylem Sercan Özgür, M.D. Mersin Üniversitesi Tıp Fakültesi Göğüs Hastalıkları Anabilim Dalı, Mersin Üniversitesi Çiftlikköy Kampüsü, 33343 Yenışehir, Mersin, Turkey.

Tel: +90 324 - 241 00 00 / 1264 e-mail: eylemozgur@yahoo.com



**Figure 1.** Chest X-ray showing the perihilar opacity in the right middle zone.

analyses were normal, and a sputum smear was negative for acid-fast bacilli. Thoracic computed tomography (CT) revealed calcifications on the anterior and lateral walls of the trachea that extended from the origin of the trachea to the main carina and an increase in density that was consistent with a soft tissue lesion with smooth borders and an oval configuration which was not clearly distinguishable from the adjacent vascular structures. This extended towards the anterior part of the pleura, the parahilar level of the anterior segment of the right upper lobe, and the neighboring right main bronchus anteriorly (Figure 2). A flexible bronchoscopy was performed after the necessary arrangements, and it revealed a multiple nodular lesion that was pale in color and swollen relative to the mucosa on the anterior and lateral walls which originated from the tracheal entry and extended towards the main carina (Figure 3). In addition, it showed a mass lesion that

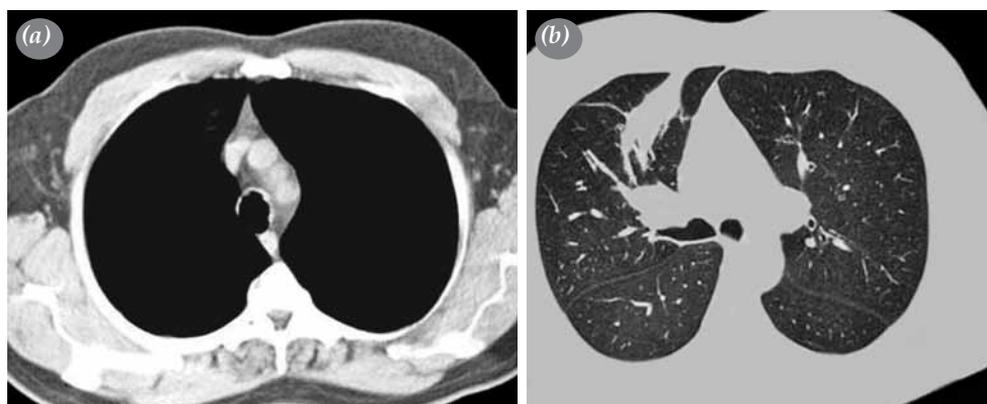
was obstructing the aperture of the right upper lobe segment. Since the biopsies and bronchial lavage were not diagnostic, the flexible bronchoscopy was repeated under general anesthesia, and the lesions that were observed in the first bronchoscopy were seen again. Biopsies were then taken from both of the lesions in the trachea and the mass at the bronchial aperture of the right upper lobe segment, and a microscopic examination of the bronchial biopsy revealed solid tumor nests consisting of cells with an abundant clear cytoplasm and irregular hyperchromatic nuclei with conspicuous nucleoli infiltrating the desmoplastic stroma. Furthermore, the tracheal biopsy showed bone trabecules within the submucosa with a normal overlying respiratory epithelium (Figure 4).

Tracheobronchopathia osteochondroplastica requires treatment if it causes severe symptoms of airway stenosis, but this was not necessary in our case. However, surgical treatment was recommended due to the patient's malignancy, but he wanted to be treated at another center and was discharged. Unfortunately, we were unable to obtain any information regarding his further treatment.

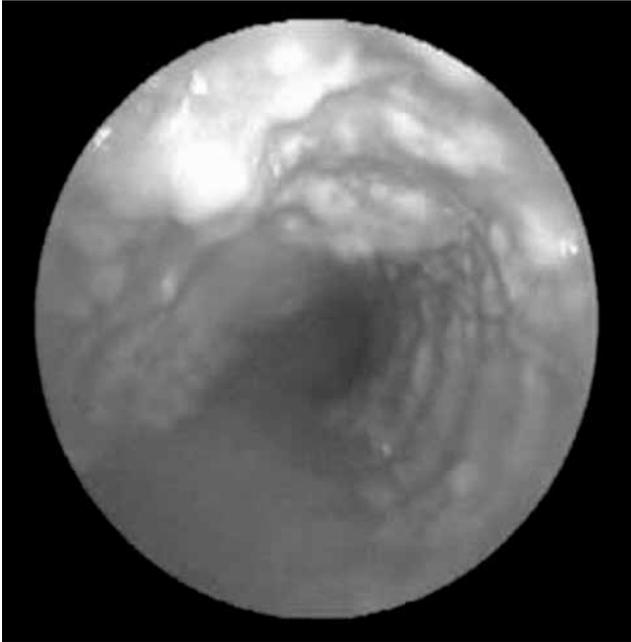
## DISCUSSION

The incidence rate of TO, a very rare disease of the trachea and main bronchi, is not exactly known. One autopsy series identified a rate between 0.2 and 0.3%, and a bronchoscopy series determined a rate of 0.4%.<sup>[1]</sup> In addition, a study composed of patients with a chronic cough had an incidence rate of between 8 and 8.5%.<sup>[1]</sup> In Turkey eight cases, one of whom is a nine-year-old girl, have been reported.<sup>[2]</sup>

Tracheobronchopathia osteochondroplastica lesions are located on the anterior and lateral walls of airways but are not present on the membranous area that makes



**Figure 2.** Thoracic computed tomography showing the tracheal calcification and increase in density consistent with a soft tissue lesion at the parahilar level of the anterior segment of right upper lobe.



**Figure 3.** Bronchoscopic view of the tracheobronchopathia osteochondroplastica nodules in the trachea.

up the posterior wall. Different theories have been proposed to explain the pathogenesis, but as of yet, nothing definitive has been established.

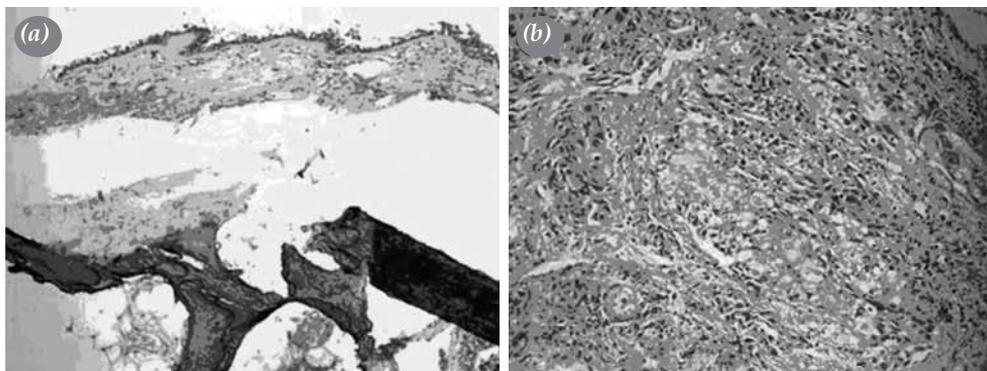
The most common symptoms associated with TO are a cough, sputum, dyspnea, and hemoptysis. It is normally detected incidentally but it can also be diagnosed in examinations conducted for other problems or by investigations aimed at explaining the airway obstruction it causes. Our patient complained of a cough, sputum, and hemoptysis, but these complaints were probably related to the his accompanying malignancy.

Besides our patient, there have also been other case reports of TO accompanied by lung cancer in

the literature. The first case was reported in 1977 by Kissler et al.,<sup>[3]</sup> but we could not obtain satisfactory information about that patient. In addition, a TO case with middle lobe syndrome (Brock's syndrome) caused by mucoepidermoid carcinoma was reported by de Wall et al.,<sup>[4]</sup> and a similar case was also reported by Roggenbuck et al.<sup>[5]</sup> The fourth case in the literature was a three-year-old girl, but for this patient, middle lobe atelectasis was caused by mucoepidermoid carcinoma.<sup>[6]</sup> Thus, the three cases with available data all featured TO together with mucoepidermoid carcinoma. However, to the best of our knowledge, no published reports of coexistent TO and adenocarcinoma exist, which makes our case unique. Although metaplasia is common in TO, there is also no data in the literature indicating malignant transformation. A chronic cough and irritation are believed to cause the metaplasia and coexistence of malignancy while the TO is believed to be coincidental. In our case, the lesion, which caused consolidation in and atelectasis of the right upper lobe, was first thought to be a large TO lesion, but it turned out to be an adenocarcinoma mass.

Chest X-rays in these cases are usually normal or the presenting pathological signs are generally overlooked. However, sometimes chest X-rays can detect irregularity, the narrowing and calcification of the trachea and main bronchi, and complications such as pneumonia, atelectasis, and bronchiectasis. In addition, thoracic CT may show numerous calcified or non-calcified lesions protruding into the airway lumen since these are present in nearly all cases of TO.

A typical bronchoscopic image of TO frequently shows numerous hard and sessile nodules measuring 1-10 mm in size on the anterior and lateral walls of the airway.<sup>[1]</sup> These lesions may then become enlarged and cause airway obstruction. Additionally, calcification, ossification, and cartilage formation at the submucosal



**Figure 4.** (a) Benign bony growths in the submucosa of the trachea (H-E x 100). (b) Solid tumor islands within the desmoplastic stroma (H-E x 200).

area in the bronchoscopic biopsy material are diagnostic features of TO.<sup>[1,7,8]</sup> In patients with severe symptoms and airway obstruction, treatment options include the use of a bronchoscopic laser, cryotherapy, or surgical intervention.

Valid treatment protocols should be used to treat an accompanying malignancy that is associated with TO. However, careful attention should be paid to the possibility of intubation and postoperative complications.

In conclusion, TO should be considered in patients who complain of a chronic cough and hemoptysis, especially when there is a lack of findings on the chest X-ray. Furthermore, it should be kept in mind that in rare cases like ours, these patients may have a coexistent malignancy.

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