Tracheobronchopathia osteochondroplastica: two cases and review of literature

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ÖZET

Tracheobronchopathia osteochondroplastica: İki olgu ve literatürün incelenmesi

Tracheobronchopathia osteochondroplastica (TO) nadir görülen bir hastalıktır. Genellikle 50 yaş üstü insanları etkiler ve klinik belirtiler obstrüktif ve infeksiyöz komplikasyonlar olunca ortaya çıkar. Bu makalede, kronik öksürük nedeniyle başvuran 50 yaşında bir kadın hasta ile hemoptizi ve kronik öksürük nedeniyle başvuran 42 yaşında bir erkek hastayı inceledik. İki olguda da bronkoskopide TO'nun tipik görünümü vardı. Alınan bronşiyal biyopsi örneklerinde kemik yapılaşması dikkati çekiyordu. Nedeni açıklanamayan kronik öksürük, hemoptizi, atelektazi ve tekrarlayan infeksiyonlarda TO mutlaka düşünülmelidir.

Anahtar Kelimeler: *Tracheobronchopathia osteochondroplastica, kronik öksürük, hemoptizi.*

SUMMARY

Tracheobronchopathia osteochondroplastica: two cases and review of literature

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Tracheobronchopathia osteochondroplastica (TO) is an unusual disorder. It mainly affects men over 50 years old and clinical manifestations are observed when obstructive or infectious complications occur. A 50-year old woman was investigated because of productive cough and 42 years old man was investigated because of haemoptysis. In two cases, at bronchoscopy, the typical picture of TO was observed. Microscopic examination of the biopsy material revealed bone formation. TO should be considered in the differential diagnosis as an unusual cause of chronic persistent chough, haemoptysis, persistent atelectasis, and recurrent segmental or lobar infection.

Key Words: *Tracheobronchopathia osteochondroplastica, chronic chough, haemoptysis.*

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Tracheobronchopathia osteochondroplastica (TO) is an unusual disorder, which nodules of metaplastic bone and cartilage develop in the submucosa of the trachea (1). This condition is usually asymptomatic and has a benign course (2).

It mainly affects men over 50 years old and clinical manifestations are observed when obstructive or infectious complications occur. To date more than 300 cases have been reported as TO. TO can be associated with various metabolic, inflammatory and neoplastic disorders, but its pathogenesis remains unknown. A metaplastic origin is actually the main hypothesis proposed. One of our cases has bronchial carcinoid tumor and other has calcified pleural plaques coincidental.

CASE REPORTS

Case 1

A 50-years old woman was investigated because of productive cough. She had had this cough for more then 10 years and produced approximately 10 cc of purulent sputum every 24 hours. Her symptoms became worse during the winter months. She had also had increasing dyspnoea at exercise during the last five years. The patient had had no previous significant diseases earlier. She had never smoked. She lived in a village in central Anatolia that is known for endemic occurrence of pleural plaques, resulting from the use of asbestos for white-washing walls. Her general condition was good and nothing remarkable was noted at clinical examination, except for some rales over the base of the lungs. Routine blood tests were normal. The reaction to 2 TU of PPD was 25 mm, but no tubercular bacilli were found in the sputum. The chest roentgenogram was normal except for distinct calcified plaques on the left diaphragmatic dome. Computerized tomography (CT) of the trachea and the large bronchi was performed twice but led to no diagnosis. Tracheobronchography, however, showed an uneven trachea. At bronchoscopy, the typical picture of TO was observed. Microscopic examination of the biopsy material revealed bone formation.

Case 2

A 42-years old man was investigated because of haemoptysis and productive cough for three mounts. The patient has a 20 pack-year history of smoking. No history of tuberculosis and chronic lung diseases. Physical examination was unremarkable. There was no clubbing or lymphadenopathy. Results of laboratory studies were normal. Chest roentgenogram was normal. A chest CT scan without contrast demonstrated multiple nodular densities in the tracheal and bronchial walls. He underwent a fiberoptic bronchoscopic evaluation, at which 3 to 4 mm nodules were scattered throughout the airways to the level of the segmental bronchi. The overlying mucosa appeared normal in trachea, without evidence of inflammation. In left lingular bronchus entrance was obstructed by bronchogenic mass. The biopsy specimen of this site revealed a glistening, pale white surface with minimal bleeding. Multiple biopsies were performed, resulting in more than 50% patency of the right middle lobe bronchus. Microscopic examination of the biopsy material revealed fragments of normal cartilage and bone formation with normal mucosa. Bronchoscopy was performed and a biopsy of the lesions was taken with some difficulty because of their bony nature. Histology reported a diagnosis of tracheobronchopathia osteoplastica with bone formation and carcinoid tumour of bronchus.

DISCUSSION

Wilks, first described this disease in 1857 in a 38-years old man who died of tuberculosis. At autopsy, he found the larynx, trachea and bronchi to be covered with a number of bony plates (3). He also noted that these bony deposits were predominantly anterior to the trachea and lay between the cartilaginous rings. A review of the literature by Martin in 1974 brought the number of reported cases to 245; however, the number of cases of TO reported in Turkey is very small (4). In our two cases this led to unexpected diagnosis of TO and prompted a review of the literature on this rare disorder.

The cause of the condition is unknown, but several theories have been postulated. Ecchondro-

ses and endochondroses from the tracheal rings were first suggested by Virchow in 1863 (5). In 1910, Aschoff suggested that metaplasia of the elastic tissue may be the cause (6). More recently, Sakula put forward the theory that tracheobronchopathia might be a form of primary localized amyloidosis of the lower respiratory tract that has undergone ossification (7). However, histology findings do not support this suggestion. A possible association with atrophic rhinitis and pharyngitis was found by Harma and Suurkari, who suggested that the lesions were due to a build-up of calcium salts within the tracheal mucosa that led to the development of local bony nodules (8). Exposure to silica could be one cause, since one of the patients of Ashley had pneumoconiosis and another published patient also had silicosis (9,10). However, since silica has been implicated as an etiological agent in TO, it is quite possible that other inhaled noxious substances also can cause the disease. Exposure to mineral fibres has not previously been described in connection with TO. The patient described here had calcified plaques, indicating environmental exposure to asbestos or some other mineral fibres (11). TO is a rare disease, but endemic plaques are a fairly common occurrence in Turkey, so they might have been only a coincidental finding (12).

The lesions most commonly occur in the lower two-thirds of the trachea but may extend anywhere from the larynx to the bronchi. Lobar bronchi are occasionally involved and rarely the process extends into segmental bronchi (13). Histological findings are typically of bony and cartilaginous nodules (some even demonstrating haemopoiesis) situated in the submucosa, invariably with connections of bone, cartilage or connective tissue to the perichondrium of the tracheal rings (14). These findings were similarly with our patients.

Most cases are asymptomatic, so it is difficult to estimate the incidence of the disease accurately. Pounder and Pieterse reported the incidental finding at autopsy to be as high as 1 in 400 (15). Primer reported the disease, again as an incidental finding, in four out of 550 bronchoscopies (16). Males and females are equally affected and age at the time of diagnosis is usually above 50-years, although some cases have been reported in young adults and children (17,18). Those who do present with symptoms typically complain of progressive dyspnoea associated with a chronic cough. This may be associated with intermittent hoarseness, the production of blood-streaked crusts and repeated chest infections (19).

Diagnosis is confirmed by bronchoscopy and the lesions have been described as having the appearance of cobblestones or a rock garden. Radiological diagnosis on plain chest X-ray is rare, but CT of the chest and neck has detected even very mild cases (20). In our second case, the diagnosis was suggested by chest CT which showed multiple nodular densities of the trachea and main bronchial walls. First case has not any CT findings. In contrast to the marked tracheal and bronchial abnormalities seen at bronchoscopy, the trachea and bronchus appeared normal on CT. The presence of calcifications in the nodules could not be conclusively demonstrated. Functional alterations induced by the lesions are variable and depend upon the severity of the disease and the site of the lesion. Spirometry is often normal, but patients with more extensive disease show a mainly obstructive pattern (21,22). We have not found any literature reference to tracheobronchography, which was quite helpful in the diagnosis. However, bronchoscopy is the simplest way of diagnosis in TO.

Prognosis of TO is generally good. In our literature search, the case described by Birzgalis is the only record of the disease requiring surgical intervention (23). The lesions do not appear to have a malignant potential. TO can be associated with various metabolic, inflammatory and neoplastic disorders, but its pathogenesis remains unknown. A metaplastic origin is actually the main hypothesis proposed (24). Our second case has bronchial carcinoid tumour. It can be associated with TO.

There were a few cases of TO in the Turkish literature. Some of the cases were diagnosed incidentally, some cases were suspected for lung tumour on chest X-ray and one case that under-

went bronchoscopy for a difficult previous intubation. Chronic cough and haemoptysis were most common symptoms (25-28).

In conclusion, TO should be considered in the differential diagnosis as an unusual cause of chronic persistent chough, haemoptysis, persistent atelectasis, and recurrent segmental or lobar infection. The bronchoscopic appearance of TO is highly suggestive and confirmed by the appropriate pathologic findings. While the CT can be highly suggestive, it is not always diagnostic. TO also can be associated with environmental exposure and neoplastic disorders.

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