

Tracheobronchopathia Osteoplastica with Behçet's Disease: Case Report

Behçet Hastalığına Eşlik Eden Tracheobronchopathia Osteoplastica

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ABSTRACT Tracheobronchopathia osteoplastica (TO) is a benign osseous metaplasia of the laryngo-tracheobronchial tree. TO is diagnosed more frequently in men over 50 years of age. Symptoms are non-specific, and include dry cough, sore throat, dyspnea, and hemoptysis. Here, we have reported a 49-year-old man who had TO and Behçet's disease (BD). He was admitted with chest pain and productive cough lasting for two weeks. However, his symptoms have been relapsing occasionally for two years. Therefore, we thought that his diagnosis could be BD with lung involvement. Physical examination was normal. There was upper respiratory airway obstruction in pulmonary function tests. Chest computerized tomography demonstrated multiple nodular densities along trachea and main carina. He underwent a fiberoptic bronchoscopic examination and we found multiple white nodules from proximal trachea to the main carina. Bronchoscopic appearance was typical for TO. Our case was interesting, because he had TO which was accompanied with BD.

Key Words: Behçet syndrome; bronchoscopy

ÖZET Tracheobronchopathia osteoplastica (TO) laringotrakeobronşiyal dallanmanın benign ve össeöz bir metaplazisidir. TO tanısı sıklıkla 50 yaş üstü erkeklerde konmaktadır. Semptomlar nonspesifiktir ve bunlar arasında kuru öksürük, boğaz ağrısı, dispne ve hemoptizi sayılabilir. Biz burada 49 yaşında TO ve Behçet hastalığı (BH) tanısı konan bir olguyu sunmaktayız. Olgunun öyküsünde iki hafta süreyle olan göğüs ağrısı ve prodüktif öksürük mevcuttu. Hastada bu semptomlar son iki yıldır aralıklı olarak görülmekteydi. Bu nedenle biz bu olguda akciğer tutulumlu Behçet hastalığı olduğunu düşündük. Fizik muayene normal olarak değerlendirildi. Solunum fonksiyon testi sonucu üst solunum yolu obstrüksiyonu olarak yorumlandı. Toraks bilgisayarlı tomografisinde trakea ve ana karinada multipl nodüller dansiteler gözlemlendi. Hastanın yapılan fiberoptik bronkoskopisinde proksimal trakeadan ana karinaya uzanan multipl beyaz nodüller gözlemlendi. Bronkoskopik görünüm TO yönünden spesifikti. Olgumuz BH ve TO birlikteliği nedeniyle yayınlanmak için ilginç bir olgu olarak değerlendirildi.

Anahtar Kelimeler: Behçet sendromu; bronkoskopi

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Tracheobronchopathia osteoplastica (TO) is an uncommon benign disease of respiratory airway. Its etiology is unknown. TO is characterized by multiple cartilaginous or bony submucosal nodules which project into the tracheobronchial lumen.¹ TO is usually diagnosed in adult men in their sixth or seventh decades.²⁻⁴ The incidence of TO ranges from 1:400 to 3:1000 in autopsies and 1:125 to 1:6000 at in vivo bronchoscopies.⁵⁻⁸ Symptoms may include productive or non-productive cough, hemoptysis,

dyspnea, dryness of the throat and recurrent pulmonary infections.^{2,9} Since many cases are asymptomatic, TO is mainly diagnosed at post mortem examinations.¹⁰⁻¹² However, frequent use of the bronchoscopy has led to an increasing number of antemortem diagnosis. The bronchoscopic appearance is usually diagnostic, and biopsy of the airway lesions is generally not required.¹³

CASE REPORT

A 49-year-old male was under treatment of BD since 1991. He had recurrent oral and genital ulcers, joint pain, eye involvement and positive skin pathergy test. He was using colchicine regularly. Other organ involvement was not detected. After colchicine treatment, joint pain and eye involvement improved, but we have not detected any healing in oral and genital ulcers. He presented with chest pain and productive cough that had been present for two years, but these symptoms were aggravated within last two weeks. Pain was located in the right anterior side of the chest and was aggravated by cough. Previously, he had had cough for two weeks and produced approximately 5 cc of white sputum every 24 hours, but had no hemoptysis. He was treated for lower respiratory tract infection several times in the previous two years. Therefore, we supposed that his symptoms

could be related to lung involvement of BD. Physical examination and chest X-ray were normal. There was upper respiratory airway obstruction in pulmonary function tests: FVC 5.2 liters [125% predicted], FEV₁ 3,65 liters [107% predicted], FEV₁/FVC 70%. Flow volume loops showed fixed airflow obstruction during inspiration, but there was no stridor and inspirational dyspnea in patient. Chest computerized tomography (CT) scan demonstrated multiple nodular densities along trachea and main carina (Figure 1). He underwent a fiberoptic bronchoscopic examination and we found multiple, white, osseous nodules originating from the anterior and lateral walls of trachea from starting from its proximal part to the main carina (Figure 2). Although cobblestone appearance was pathognomonic for TO, various biopsies and bronchial brush specimens were taken for infections. Cytologic brushes and biopsies of the tracheal wall revealed bronchial epithelium with an accumulation of neutrophils and lymphocytes. Smears and cultures for infections were all negative. Since the symptoms and the obstruction were not severe, we decided to perform a close follow up.

DISCUSSION

TO was first described by Wilks in the mid-19th century.^{1,14} Usually, TO is diagnosed at postmortem

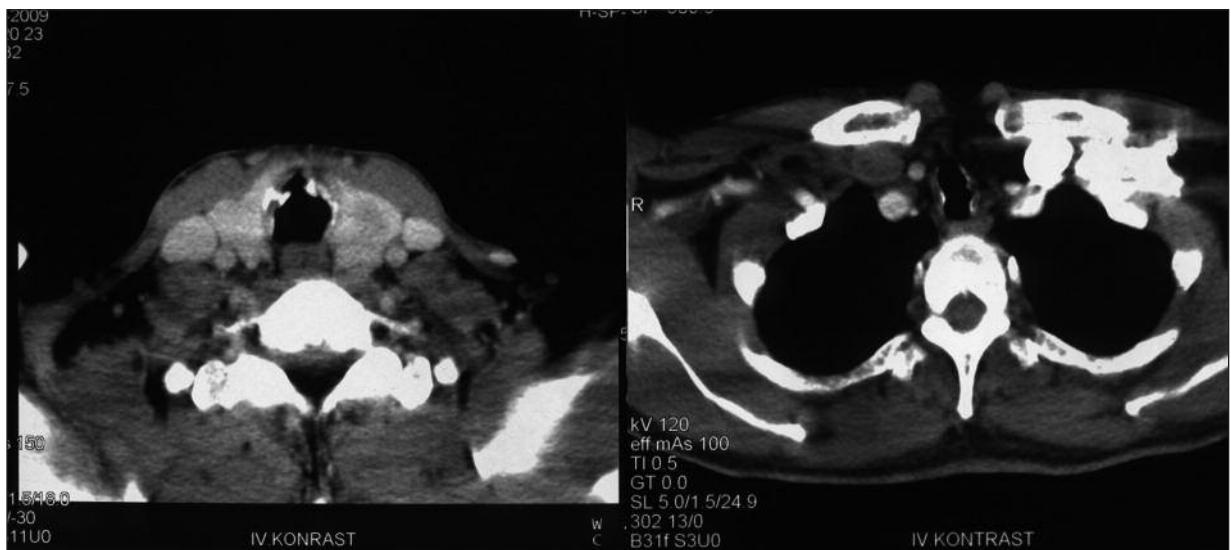


FIGURE 1: Chest CT scan demonstrated multiple nodular densities along trachea and main carina.



FIGURE 2: Multiple, white, osseous nodules originating from the anterior and lateral walls of trachea from proximal to the main carina were seen.

(See for colored from <http://tipbilimleri.turkiyeklinikleri.com/>)

examinations; however; in last two decades, it has been diagnosed using bronchoscopy and CT. The overall incidence of this disease remains unknown. When we searched the national and international literature, we found 22 cases reported from Turkey (Table 1).¹⁵⁻³² Among the 2852 bronchoscopies in last 14 years, this was the first case of TO in our clinic.

It is assumed that the pathological process of the disease progresses over a long period. The diagnosis is not suspected until the patient develops symptoms. Sometimes it is not noticed, because of nonspecific symptoms. Usually, TO is initially misdiagnosed as asthma or chronic bronchitis. When nodules expand into the airway lumen, they cause chronic cough, dyspnea, hemoptysis and recurrent respiratory infections develop.³³ Our patient was treated for lower respiratory tract infection several

TABLE 1: Twenty two cases of tracheobronchopathia osteoplastica reported in the national and international literatures

Age, Gender	Clinic	Diagnosis	Reference
42 F	Dyspnea, cough, wheezing	Thorax CT, rigid bronchoscopy, biopsy	16
56M	Cough, hemoptysis	Thorax CT, FOB, biopsy	Poster presentation Turkish Thoracic Society 12 th Annual Congress-2009
41 M	Chronic cough	Thorax CT, FOB, biopsy	17
41 M	Cough, sputum, dyspnea	HRCT, FOB, biopsy	18
72 M	Postoperative atelectasis	Thorax CT, virtual bronchoscopy, FOB, biopsy	19
64M	Repeated respiratory tract infections	Thorax CT, virtual bronchoscopy, FOB	20
22 M	Cough, sputum, dyspnea	HRCT, FOB, biopsy	Poster presentation Turkish Radiology Society 28 th National Congress-2007
54 F	Cough	Thorax CT, FOB, biopsy	21
44 M	Cough, hemoptysis	Thorax CT, FOB, biopsy	22
49 M	Fever, sweat, cough, sputum	Thorax CT, FOB, biopsy	
9 F	Chronic cough		23
47 M	Cough, fever, chest pain	Thorax CT, FOB	24
85 M	Cough, mild weight loss	Thorax CT, FOB, biopsy	25
77 M	Cough	Thorax CT, FOB, biopsy	26
50 F	Cough, sputum, dyspnea	Tracheobronchography, FOB, biopsy	27
42 M	Cough, sputum, hemoptysis	Thorax CT, FOB, biopsy	
56 M	Chest pain, hemoptysis, dyspnea, cough, fever	Thorax CT, FOB, biopsy	28
40 M	Fever, night sweats, weakness, weight loss	Thorax CT, FOB, biopsy	29
72 M	Fever, cough, sputum	Thorax CT, virtual bronchoscopy, FOB, biopsy	30
67 M	Cough, sputum, hemoptysis	Thorax CT, rigid bronchoscopy, biopsy	31
67 F	Weight loss, cough, chest pain, dyspnea	Thorax CT, MRI, FOB, biopsy	32
54 M	Shortness of breath	Pulmonary function tests, thorax CT, FOB, biopsy	33

CT: Computerized tomography, FOB: Fiberoptic bronchoscopy, HRCT: High resolution computerized tomography.

times in the previous two years. When he was admitted to our clinic, we first supposed that it could be BD's lung involvement.

As known, skin, central nervous system, gastrointestinal system and pulmonary involvements are common in BD. The most common pulmonary vascular problems are pulmonary artery aneurysms, pulmonary emboli/infarct and involvement of small-sized vessels.^{34,35} In addition, bronchial stenosis, mild small airway disease, chronic bronchitis and bronchiectasia, reversible airway obstruction, emphysema and obstructive lung disease were reported.³⁵⁻³⁹ Diagnosis of our patients was completely different from above diseases. Therefore, we thought that our case was independent from BD's involvement. However, there were no supporting literature about association of BD and TO.

Characteristic symptoms of BD are hemoptysis, dyspnea, cough, sputum, fatigue and chest pain.⁴⁰ Our patient had productive cough, chest pain, dyspnea and recurrent pulmonary infections. Because we thought that symptoms of our patient might be due to airway involvement of BD, we performed further investigations. When the patient's flow-volume loop results were assessed, we suspected an airway obstruction and performed a chest CT.

In diagnosis, routine blood tests are not helpful. Results of pulmonary function tests are vari-

able depending on the location of the lesions and the degree of involvement. In symptomatic patients, obstructive pattern is observed. If the tracheal, laryngeal and subglottic involvement is severe, flow-volume loops are more helpful.⁴¹ Restrictive abnormality is usually associated with secondary atelectasis or another restrictive lung disease. In this case, our speculation was a restrictive abnormality, because of presence of BD. However, we found mild upper airway obstruction in respiratory function tests.

Chest CT scan is more sensitive than chest X-ray in the diagnosis of TO. CT can detect the typical features of TO with multiple calcified or noncalcified nodules projecting into the airway lumen.¹³ In addition, thickening or irregularity of the tracheal or bronchial walls, deformed tracheal cartilage rings without an evidence of external compression and submucosal calcification can be determined. Although TO lesions are commonly seen in the upper trachea, whole bronchial system can be involved. There was only total tracheal involvement present in our case.

In conclusion; because TO was accompanied with BD, our case was interesting. However, patient's symptoms were similar to BD's lung involvement. Pulmonary function tests and chest CT are helpful in the differential diagnosis particularly in such a case.

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