

Behçet's disease presenting with massive hemoptysis related to bronchovascular fistula: A case report

Bronkovasküler fistüle bağlı masif hemoptizi ile seyreden Behçet hastalığı: Olgu sunumu

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ABSTRACT

A 37-year-old male patient was admitted to our hospital with recurrent hemoptysis, 50 mL per day. Thoracic computed tomography showed no pathology responsible for hemoptysis. Bronchoscopy revealed mucosal infiltrations and 2 to 3-mm blotch in the lateral wall of the right lower lobe. After punch biopsy of the suspected area, massive bleeding occurred. Right lower lobectomy was performed urgently. A bronchovascular fistula was noticed at the specimen. Pathological examination result was compatible with clinically suspected Behçet's disease. The patient was given high-dose steroid and cyclophosphamide treatment and received azathioprine maintenance treatment for 18 months. He has been symptom-free for three-year follow-up.

Keywords: Behçet's disease, bronchovascular fistula, hemoptysis.

Behçet's disease is a multisystemic inflammatory disease of unknown etiology with a chronic course and unpredictable exacerbations.^[1] It was first described by Hulusi Behçet,^[2] MD, the Turkish dermatologist, in 1937 with a triple-symptom complex and is mainly characterized by recurrent episodes of mucocutaneous, ocular, joint, vascular, and central nervous system involvement. Recurrent oral and/or genital aphthosis, ocular involvement in terms of uveitis and, retinal vasculitis in combination with variable skin lesions are the cardinal signs of Behçet's disease.^[3] It is episodic and progressive in nature. Kidney and lung presentations are rare.^[4,5] Pulmonary presentation

ÖZ

Otuz yedi yaşında erkek hasta günde 50 mL'lik tekrarlayan hemoptizi nedeniyle hastanemize başvurdu. Toraks bilgisayarlı tomografide hemoptiziden sorumlu herhangi bir patoloji izlenmedi. Bronkoskopide sağ alt lobun lateral duvarında mukozal infiltrasyonlar ve 2-3 mm'lik kabartılar görüldü. Şüpheli bölgede yapılan punch biyopsi sonrasında, masif kanama gelişti. Acilen sağ alt lobektomi uygulandı. Alınan örnekte bronkovasküler fistül saptandı. Patolojik inceleme sonucu, Behçet hastalığı ile uyumlu idi. Hastaya yüksek doz steroid ve siklofosfamid tedavisi başlandı ve 18 ay süreyle azatiyoprin idame tedavisi uygulandı. Hasta üç yıldır semptomsuz olarak takip altındadır.

Anahtar sözcükler: Behçet hastalığı, bronkovasküler fistül, hemoptizi.

draw a great attention in 1960s. Cadman^[6] published 12 pulmonary manifestations found in the literature, and one of them was his own case. Currently, there are case reports available with a few number of patients.

In this article, we present a rare case of Behçet's disease presenting with massive hemoptysis related to bronchovascular fistula which was successfully treated with surgery.

CASE REPORT

A 37-year-old male immigrant patient was admitted to our hospital with recurrent hemoptysis about 50 mL

Received: February 10, 2020 Accepted: April 10, 2020 Published online: July 26, 2021

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Cite this article as:

Bedirhan MA, Arda N, Tanrıverdi E, Yaran V, Sansar D, Cansever L. Behçet's disease presenting with massive hemoptysis related to bronchovascular fistula: A case report. Turk Gogus Kalp Dama 2021;29(3):408-411

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per day. His thoracic computed tomography (CT) scan showed lymphadenopathy around right lower lobe bronchus; however, there was no explanation for hemoptysis on CT and conservative treatment was initiated (Figure 1). After few days, he presented again with the same symptoms and his detailed medical history revealed two massive hemoptysis episodes previously in his country. Thoracic CT scan was repeated and the same images were obtained. Fiberoptic bronchoscopy was planned to find the possible pathology. A written informed consent was obtained from the patient.

Fiberoptic bronchoscopy was performed under local anesthesia. Larynx, trachea, carina, and left bronchial system were all found to be normal. Bronchoscopy showed mucosal infiltrations and a 2 to 3-mm ridge in the lateral wall of the right lower lobe. Also, a clot was observed inside the right lower lobe, suggesting that the reason for hemoptysis was mucosal irregularity. Therefore, punch biopsy was performed. Following punch biopsy, massive bleeding occurred. Early intervention could not control the bleeding, and intubation was performed urgently. After 1,500 mL bleeding and 30-min cardiopulmonary resuscitation, bleeding was taken under control and intubation tube was gently replaced by a double-lumen tube. Meanwhile, CT scans were retrospectively checked and suspected that it was a bronchial system pathology. Thus, the patient was operated. During thoracotomy, the first, right main bronchus was taken under control and later exploration was started. There was no mass around the intermediate lobe and its distal segments. Since the ridge which bled was located at the orifice of right lower lobe, bilobectomy was performed in a



Figure 1. Thoracic computed tomography prior to bronchoscopy showing a normal bronchovascular structure, except for several lymph nodes around.

usual manner. Specimens were examined for bleeding site, *ex vivo*. In the mucosal ridge, a 1-mm ostium opening to the lower lobe artery was observed. Using a mosquito clamp, the bronchovascular fistula was detected (Figure 2).

In the early postoperative period, the patient reported ocular symptoms, genital ulcerations, and arthralgia within the previous year, suggestive of Behçet's disease. On postoperative Day 4, high-dose steroid and cyclophosphamide treatment were prescribed to the patient to prevent Behçet's disease exacerbation, in addition to azathioprine maintenance treatment for 18 months. He was uneventfully discharged on postoperative Day 6.

Pathological examination revealed no specific agents with Ehrlich-Ziehl-Neelsen, Gram, and Grocott staining. No lesions were seen, except for a yellow, irregular area of 22×18×5.5 cm in size, 2 cm away from the bronchial surgical margin in the lower lobe of the right lower lobectomy specimen. The intraparenchymal contours were unable to be fully distinguished and a hepatized appearance was visible due to intense bleeding in the middle and lower lobes. Microscopic examination revealed necrotic nodules and vasculitic changes, compatible with clinically suspected Behçet's disease (Figure 3).



Figure 2. Surgical clamp showing bronchovascular communications at surgical specimen.

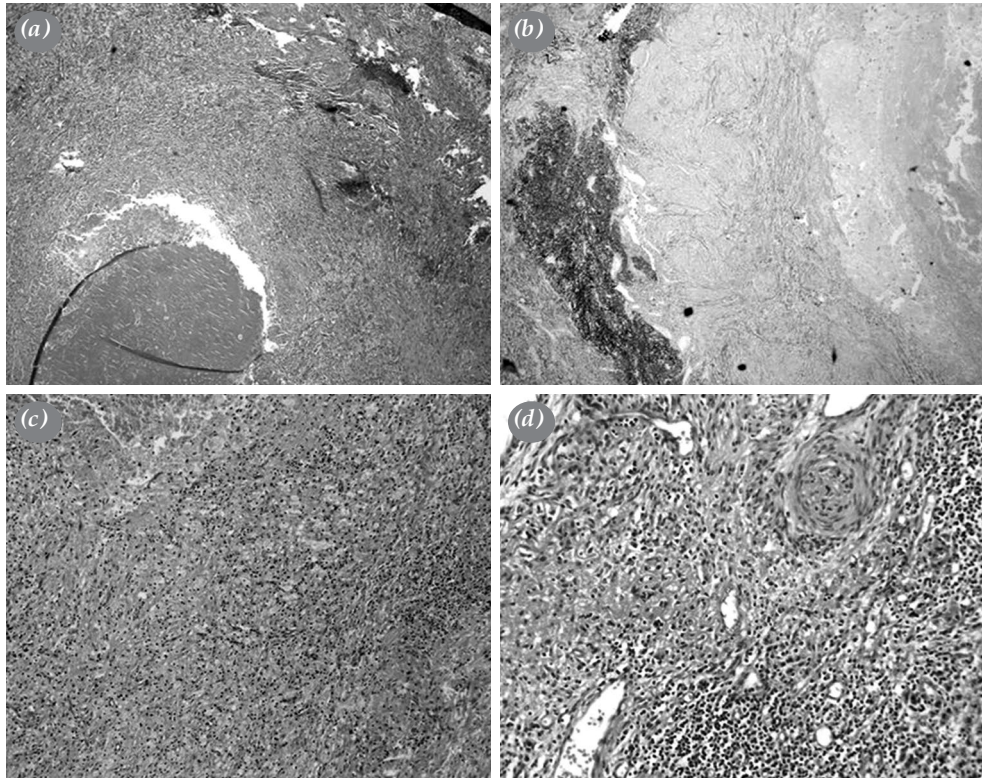


Figure 3. (a) Microscopic examination showing a necrotic nodule due to infarction (H-E, $\times 10$), (b) dissected vessel aneurysm and vasculitic changes in middle- and small-sized arteries (Elastica-Van Gieson $\times 10$), (c) clinical and the pathological findings including foamy macrophages, cholesterol crystals, foreign giant cells, dissected vessel aneurysm, and vasculitic changes (H-E, $\times 20$), (d) necrotic nodule compatible with clinically suspected Behçet's disease (H-E, $\times 40$).

DISCUSSION

The first-line treatment of pulmonary aneurysms in Behçet's disease is non-surgical.^[7] Surgical treatment is usually implemented to those who have not benefited from embolization or immunosuppression therapy with a high mortality risk.^[8] Surgery is indicated for ruptured aneurysms with abundant hemorrhage and enlarged aneurysms, despite immunosuppressive treatment. Despite fragility of vascular tissues in Behçet's disease which brings more surgical risks, lobectomies can be done with favorable outcomes.^[9] Due to the fragility of connective tissues, arteries can be tied with tapes and, then, reinforced with transfixion sutures.

In conclusion, as in our case, hemoptysis without an aneurysm related to Behçet's disease may present with normal CT findings. Therefore, clinicians should keep in mind that hemoptysis in Behçet's disease may occur without an aneurysm as evidenced by CT.

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

The authors received no financial support for the research and/or authorship of this article.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Caso F, Costa L, Rigante D, Lucherini OM, Caso P, Bascherini V, et al. Biological treatments in Behçet's disease: Beyond anti-TNF therapy. *Mediators Inflamm* 2014;2014:107421.
2. Behçet, H. Über rezidivierende aphtösedurch ein virus verursachte geschwüre am mund, am auge und an der genitalien. *Dermatologische Wochenschrift*, 1937;105:1152-63.

3. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. *Lancet* 1990;335:1078-80.
4. Zheng W, Li G, Zhou M, Chen L, Tian X, Zhang F. Renal involvement in Chinese patients with Behçet's disease: A report of 16 cases. *Int J Rheum Dis* 2015;18:892-7.
5. Seyahi E, Yazici H. Behçet's syndrome: Pulmonary vascular disease. *Curr Opin Rheumatol* 2015;27:18-23.
6. Cadman EC, Lundberg WB, Mitchell MS. Pulmonary manifestations in Behçet syndrome. Case report and review of the literature. *Arch Intern Med* 1976;136:944-7.
7. Hamuryudan V, Oz B, Tüzün H, Yazici H. The menacing pulmonary artery aneurysms of Behçet's syndrome. *Clin Exp Rheumatol* 2004;22(4 Suppl 34):S1-3.
8. Yoon W, Kim JK, Kim YH, Chung TW, Kang HK. Bronchial and nonbronchial systemic artery embolization for life-threatening hemoptysis: A comprehensive review. *Radiographics* 2002;22:1395-409.
9. Tuzun H, Seyahi E, Guzelant G, Oz B, Batur S, Demirhan O, et al. Surgical treatment of pulmonary complications in Behçet's syndrome. *Semin Thorac Cardiovasc Surg* 2018;30:369-78.