

Behçet's Disease Presenting with Multiple Pulmonary Artery Aneurysms and a Pseudoaneurysm

MULTİPL PULMONER ARTER ANEVİRİZMALARI VE PSÖDOANEVİRİZMASI OLAN BİR BEHÇET HASTALIĞI OLGUSU

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Summary

We report a case of Behçet's Disease with multiple pulmonary artery aneurysms and a pseudoaneurysm. We thought that pseudoaneurysm had been due to transthoracic needle aspiration. Pseudoaneurysm and pulmonary artery aneurysms disappeared with immunosuppressive therapy.

Key Words: Behçet's Disease,
Pulmonary artery aneurysms

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Özet

Bu yazıda multipl pulmoner arter anevrizmaları ve psödoanevrizması olan bir Behçet Hastası sunuyoruz. Psödoanevrizmanın transtorasik aspirasyon biopsisi sonrasında geliştiğini düşünüyoruz. Pulmoner arter anevrizmaları ve psödoanevrizma immünsüpresif tedavi ile düzeldi.

Anahtar Kelimeler: Behçet Hastalığı,
Pulmoner arter anevrizmaları

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Behçet's disease is a chronic systemic disease manifesting as recurrent aphthous stomatitis with any two of the following features: recurring genital ulcerations, uveitis or retinal vasculitis, typical defined skin lesions, or a positive pathergy test. Other well-known clinical features are large vessel arteritis, phlebitis, meningoencephalitis, synovitis, and discrete intestinal ulcerations (1-3). Pulmonary symptoms may occur in 5 to 10% of patients with Behçet's disease and are thought to be due to pulmonary vasculitis (4,5).

We report a patient with Behçet's disease complicated with bilateral pulmonary artery aneurysms and a traumatic pseudoaneurysm.

Case Report

Twenty-seven year old man was admitted to the hospital because of hemoptysis, chest pain, arthralgia and fever.

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One year before entry he was admitted to the Immunology Department with recurrent oral and genital ulcers, cutaneous lesions, arthralgia; and was diagnosed as Behçet's disease.

Ten months after diagnosis he was admitted to another hospital because of left sided pleuritic chest pain, cough, hemoptysis and fever. Chest x-ray film revealed bilateral linear opacities in both lower lung fields and 3 cm rounded opacity adjacent to the left hilum. A computed tomographic (CT) scan of the thorax disclosed bilateral pleural thickening, patchy infiltration and 2x3 cm nodular density in the left lower lobe. Corticosteroids and anticoagulation therapy was given with the diagnosis of pulmonary embolism and Behçet's disease. Patient didn't respond to this therapy. Fiberoptic bronchoscopy was not diagnostic; and a needle aspiration biopsy of the density in the left lung was performed. Microscopic examination of the biopsy specimen revealed necrotising, nongranulomatous vasculitis with lymphocyte infiltration. Fever returned to normal, hemoptysis was reduced, and the patient was discharged with corticosteroid and coumadin therapy.

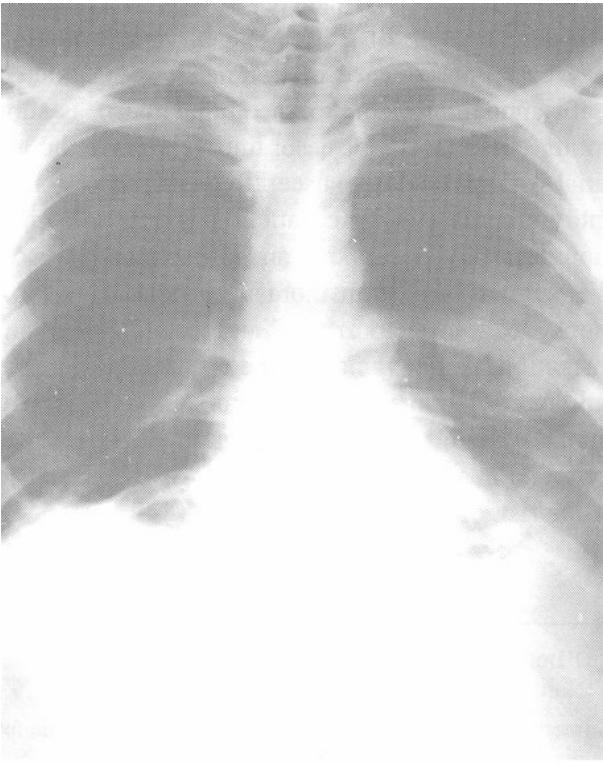


Figure 1. Chest x-ray.

One month later he presented with hemoptysis, pleuritic chest pain, arthralgia and fever again and was referred to our hospital. On physical examination he was febrile; breath sounds were diminished at both lung bases. Chest x-ray revealed a large, well-circumscribed mass lesion in the left middle and lower lung fields, and two ill defined nodules adjacent to the right hilum (Figure 1). CT scan of the chest was reported to show that two nodules, 30 mm and 20 mm, related to left hilum. There was a large area of consolidation in the anterior segment of left upper lobe and superior segment of left lower lobe; and was reported that this area of consolidation could be due to parenchymal hemorrhage secondary to thromboembolism. And CT showed patchy infiltration in the posterobasal segments (Figure 2). Digital Subtraction Angiography (DSA) of pulmonary arteries revealed an aneurysm, 3 cm, in the proximal left lower lobe pulmonary artery and an 8 cm pouch related to this aneurysm. This pouch was thought to be a pseudoaneurysm due to trauma. DSA also showed smaller aneurysmal dilatations in the branches of right main pulmonary

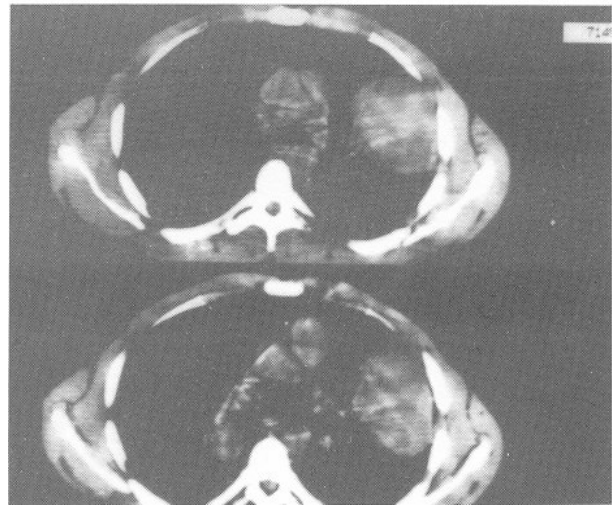


Figure 2. CT scan of the thorax.

artery. Abdominal ultrasonographic study was normal.

The patient consulted by thoracic surgeons and accepted as inoperable because of bilateral pulmonary aneurysms.

Therapy with cyclosporin A and corticosteroids was started. The patient responded well to this therapy, and he was discharged. Two months after his discharge he came for follow-up examination. The patient was readmitted because of hemoptysis. At the time of readmission, CT scan of the chest was reported to show that the aneurysms and pseudoaneurysm became smaller because of thrombosis. Cyclophosphamide was added to therapy. Hemoptysis was ceased.

He was followed with regular intervals. Clinical and radiological improvement was observed (Figure 3). Immunosuppressive therapy was tapered over one year, colchicine was added. At the end of the immunosuppressive therapy. CT scan of the chest showed there were no aneurysmal dilatations, and the pseudoaneurysm had been disappeared. Two years after this admission, he was treated for deep venous thrombosis in the Immunology Department.

We learned that the patient had died with a sudden onset of massive hemoptysis, five years after the diagnosis.

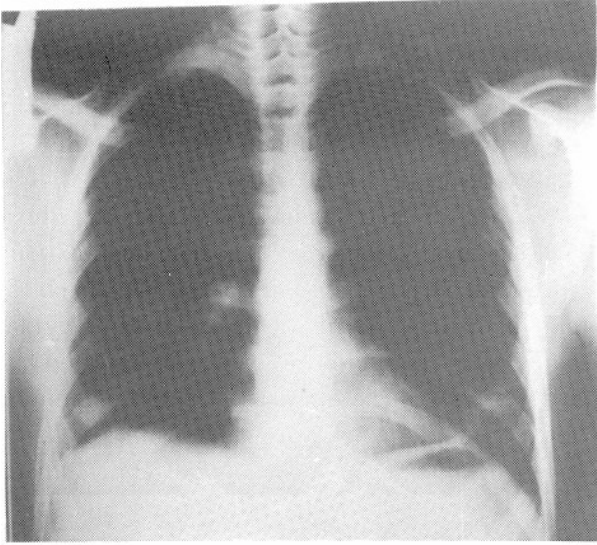


Figure 3. Control chest x-ray after the treatment.

Discussion

Behçet's disease is a chronic multisystem disease affecting many organs. The characteristic pathology is a leukocytoclastic vasculitis (5,6).

Pulmonary involvement is uncommon and can range in severity from fleeting radiographic opacities, pleuritis with effusion, hemorrhage with alveolar infiltrates to diffuse interstitial lung disease. A case of Behçet's disease presenting with a pulmonary mass lesion was also reported; and it was shown that this mass was the radiological image of a large area of infarction with hemorrhage. The primary pathologic finding in the lungs of patients with Behçet's disease is a vasculitis affecting vessels of various sizes. This vasculitis may lead to thrombosis or destruction of elastic lamina, presumably from arteritis of the vasa vasorum, with subsequent dilatation and aneurysm formation; or

perforation and pseudoaneurysm formation (6-8). The development of a pseudoaneurysm formation may be a consequence of trauma. It is speculated that remodeling of the extracellular matrix is suboptimal in Behçet's disease, thus rendering the arterial wall particularly susceptible even to very low entity trauma (9). Our patient has multiple pulmonary artery aneurysms and a pseudoaneurysm. We suggest that transthoracic needle aspiration biopsy caused hemorrhage and pseudoaneurysm formation. Anticoagulation therapy might increase pulmonary hemorrhage. Percutaneous lung biopsy should be avoided when investigating a chest opacity found in a patient with Behçet's disease because of the risk of puncturing aneurysm (4).

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