

# Basilar Artery Aneurysm Associated with Behcet's Disease: A Case Report

## Behçet Hastalığı ile Birlikte Görülen Baziller Arter Anevrizma Olgusu: Olgu Sunumu

### ABSTRACT

Vascular involvement is common in Behçet's Disease (BD). The venous system is often affected, while arterial system involvement is a rare occurrence. Intracranial aneurysm formation associated with BD is extremely rare. A 38-year-old male patient with BD under the follow-up of the Dermatology department presented with acute headache. The cranial computed tomography revealed subarachnoid bleeding. A basilar artery aneurysm, which was ruptured during the procedure, was found on the cerebral angiography and the patient died the next day. This report adds our case to the literature of posterior cerebral circulation aneurysms associated with BD among the limited number of articles on this topic.

**KEY WORDS:** Basilar artery, Aneurysm, Behçet's Disease

### ÖZ

Behçet Hastalığı (BH)'nda vasküler tutulum sıklıdır. Genellikle venöz sistem etkilenmekte olup arteriyel sistem oldukça az tutulmaktadır. BH'da intrakraniyal anevrizma oluşması çok nadirdir. İntrakraniyal anevrizmalar literatürdeki olguların çoğunda anterior serebral dolaşımında bildirilmiştir. Otuzsekiz yaşında 4 yıldır BH tanısı ile takip edilen erkek hasta akut başağrısı ile başvurdu. Bilgisayarlı beyin tomografisinde spontan subaraknoid kanama görüldü. Serebral anjiyografi sırasında anevrizması rüptüre olan olguda baziller arter anevrizması saptandı ve bir gün sonra öldü. Literatürde BH ile birlikte görülen posterior sirkülasyon anevrizması olgularına eklenmesi amacıyla bu olgu sunulmaktadır.

**ANAHTAR SÖZCÜKLER:** Baziller trunk, Anevrizma, Behçet Hastalığı

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Received: 28.07.2007

Accepted: 23.01.2008

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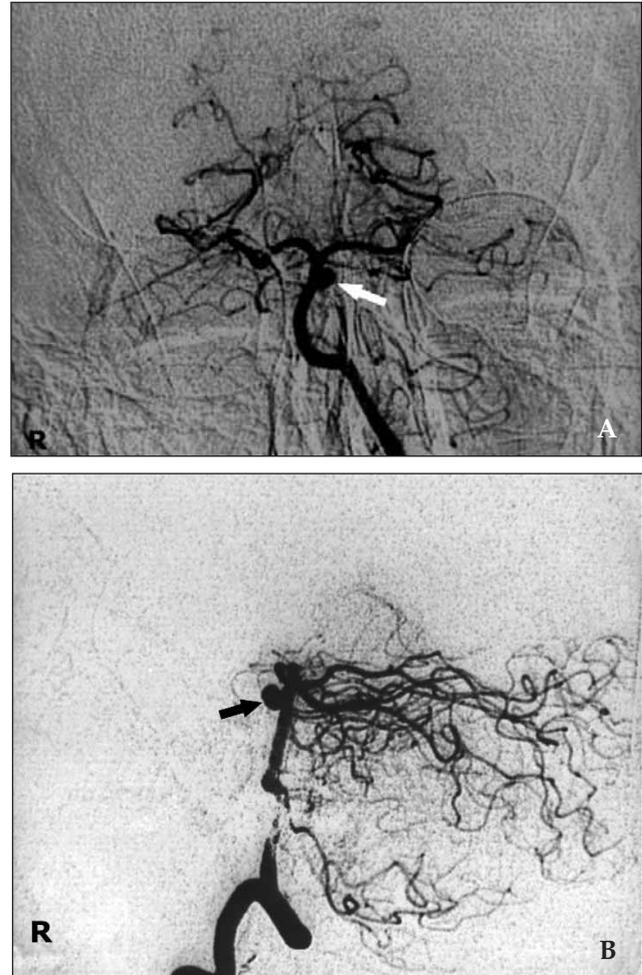
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## INTRODUCTION

Behçet's Disease (BD) is a chronic and recurrent inflammatory disease that may involve multiple systems. It has a wide distribution with higher incidence rates in Mediterranean countries and the Far East. The most common findings of BD are aphthous stomatitis (100%), genital ulceration (75%), and uveitis (60-80%). The rate of vascular involvement in BD has been reported to vary between 23% and 62% (2, 3). The venous system is affected in 95% of cases with vascular involvement. Venous thrombosis usually develops due to vascular involvement. Venous thrombosis is also one of the first findings of the disease (9, 10). Compared to venous involvement, arterial involvement is extremely rare, with an incidence of 2.2-7.7 % (3-7). Arterial involvement is identified as stenosis, thrombosis, or aneurysm. Aneurysm formation in an intracranial vessel is very rare. The aneurysms are mainly located in the anterior circulation in most of the reported cases. The literature reveals few cases with aneurysm formation located in the posterior circulation (1, 6, 8, 12). This report adds our case to the literature of the posterior cerebral circulation aneurysms associated with BD.

## CASE REPORT

A 38-year-old male patient presented to our clinic with the complaints of headache and nausea. Systemic examination revealed oral and genital ulcerations, and neurological evaluation was non-specific except for neck stiffness. The cranial computed tomography of the patient revealed diffuse subarachnoidal hemorrhage. The patient was hospitalized with a Glasgow coma score (GCS) of 15 and World Federation of Neurologic Surgeons subarachnoidal hemorrhage classification of grade I. The patient had been diagnosed and followed-up with BD through the previous four years by the Department of Dermatology. The laboratory findings were as follows: erythrocyte sedimentation rate: 85 mm/h; C reactive protein level: 120mg/L. Antineutrophilic cytoplasmic antibody and lupus erythematosus tests were negative. An aneurysm in the trunk of the basilar artery of the patient was detected upon digital subtraction cerebral angiography (Figure 1). The GCS of the patient decreased to 5 points within a few minutes of the cerebral angiography. The patient was then moved into the intensive care unit, and died on the second day of hospitalization before any surgical



**Figure 1:** A, Anteroposterior view of the digital subtraction angiography (DSA) of the patient shows an aneurysm (arrow) on the basilar artery trunk. B, Lateral view of the vertebral DSA reveals anteriorly enlarged saccular aneurysm (arrow) of the basilar artery trunk.

intervention could be performed. The family has not given us permission to perform an autopsy. The sudden deterioration of the patient led us to believe the reason of death was rebleeding of the aneurysm during the cerebral angiography.

## DISCUSSION

Four types of vascular lesions are encountered in BD: arterial occlusion, arterial aneurysm, venous occlusion, and venous varicosities. (10, 11). Aneurysm formation associated with arterial involvement is observed particularly in the abdominal aorta and in large arteries such as femoral and pulmonary arteries (11). The pathophysiology of aneurysm formation is based on two factors, one of which is rupture of internal and external elastic

lamina due to thinning of tunica media and the other vasculitis developing due to lymphocytic infiltration of vaso vasora (2, 11). Histological studies of aneurysmal areas in BD have shown proliferation of endothelial cells, mononuclear and neutrophilic infiltration, destruction of elastic lamina, fibrinoid necrosis, and thrombus formation (11, 12). Existence of such inflammatory changes may increase the rebleeding tendency of the aneurysm. We believe it may have a role in the rebleeding during the arterial injection of the contrast medium in our case. In contrast to digital subtraction angiography technique through the femoral catheterization, cerebral angiography through the multi-slice computed tomography is a less invasive way to obtain adequate information about the aneurysm (4, 5). Therefore, investigating the aneurysm through multi-slice computed tomography device would probably have prevented the rebleeding of the aneurysm in our case.

Vascular complications developing in BD become evident 3-16 years after presentation (13, 14), and are commonly observed in young adult males (12). Our patient was 38 years old and his vascular findings had become apparent 4 years after the diagnosis.

Review of the 14 reported cases with BD and concomitant cerebral aneurysms showed that the aneurysms defined in these cases were mostly of the saccular type and they were located in the anterior cerebral circulation similar to those in the aneurysm cases without BD. Furthermore, six of these cases had multiple aneurysms (1, 6). Our case, however, is different in that the aneurysm was located in the posterior circulation. Posterior circulation cerebral aneurysms in this group of patients could be related to BD or be incidental; we have found 6 cases (including ours) (Table I) in the English literature.

Treatment of cerebral aneurysms in BD depends on the location and size of the aneurysm as well as whether it has ruptured or not (12). In addition to corticosteroid and immunosuppressive treatment, primary clipping or endovascular treatment might be preferred. Immunosuppressive and corticosteroid treatments are particularly essential to prevent vascular complications. Surgical treatment and endovascular treatment of cerebral aneurysms has not been very effective in BD (12, 14). This has been attributed to the formation of new vascular lesions throughout immunosuppressive and corticosteroid treatments (12, 13).

**Table I:** Posterior circulation cerebral aneurysms reported in BD patients. SCA, superior cerebellar artery; VA, vertebral artery; M, male.

Authors	Age/Se	Location of the aneurysm
Rosensting et al. (12)	36, M	Left SCA
Bahar et al. (1)	40, M	Right VA
Itoh et al. (6)	65, M	Left VA
Kizilkiliç et al. (8)	38, M 55, M	Right SCA Intracranial VA
Ho et al.(6)	30,F	SCA
Present case	38, M	Basillary trunk

## CONCLUSION

Aneurysms of the anterior or rarely posterior circulation might be observed during the course of BD. Considering the BD-related inflammatory changes on the aneurysm wall, three-dimensional multi slice computed tomography angiography might be preferable to digital subtraction angiography through femoral artery catheterization.

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