

# Neuro-Behcet's Disease with Ventricular Hemangioblastoma

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## ABSTRACT

*Behçet disease is an inflammatory multisystem vasculitic disease which has a classical triad of oral and genital ulcerations with uveitis. But the neurological involvement may present mild and subclinic symptoms at some patients. Hemangioblastoma is a benign vascular tumor of central neural system. Here in, we aimed to present a 25 years-old man with neuro-Behcet's disease in a patient who was admitted with the complaints of headache, dizziness and vomiting with hemangioblastoma which rarely showing ventricular located.*

**Key words:** Behçet disease, hemangioblastoma, ventricular

## Behçet Hastalığı ve Ventriküler Hemanjioblastoma

### ÖZET

*Behçet hastalığı multisitem özellikle ağız ve genital ülser ile seyreden inflamatuvar vaskülitik bir hastalıktır. Fakat nörolojik tutulum subklinik veya hafif olarak bazı hastalarda bulunabilir. Hemanjioblastoma benign santralsinir sisteminin vasküler tümörüdür. Burada 25 yaşında baş ağrısı, baş dönmesi ve kusma şikayeti olan hemanjioblastomalı bir behçet hastalığı olan erkek hasta sunulmuştur.*

**Anahtar kelimler:** Behçet hastalığı, hemanjioblastoma, ventrikül

## INTRODUCTION

Behcet Disease (BD) is multisystem vasculitis disease with unknown origin which has a classical triad of oral and genital ulcerations with uveitis (1). Neurological involvement of BD is reported to be between 2.2% and 49% in literatures. This status is called as neuro-behçet disease (NBD) (2,3). Hemangioblastomas are the infrequent benign tumors of the central nervous system (CNS). Ventricular location is reported to be very unusual in the literature (4,5). We aimed to present NBD patient with ventricular hemanjioblastom.

## CASE

A twenty five years old male was admitted to emergency service presenting with the complaints of headache, dizziness and vomiting. He had suffered from a history of recurrent oral aphthous lesions and genital ulcer, Behçet's disease. Vital and laboratory investigations were normal. Cranial MRI scan showed a solitary, solid, 21x18 mm size, an ventricular tumor placed at the fourth ventricle. MRI revealed signal changes consistent with edema at bulb. It was isointense on T1 weighted images and contained focal hypointense signal void hyperintense on T2 weighted images. After gadolinium-DTPA injection the mass was markedly enhanced (Figure 1). Magnetic reso-

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nance venography demonstrated corruption in filling, contour irregularities and decreased calibration at sinus rectus (Figure 2). Cerebral angiography was performed for the differential diagnosis which was demonstrated a large vascular tumor showing heavy vascular blush fed by branches of the posterior inferior cerebellar artery of the left vertebral artery (Figure 3). According to these radiological features hemangioblastoma was diagnosed. Patient's general condition deteriorated progressively, and he died despite intensive steroid therapy and general support. Pathological examination of the tumor could not be performed because patient's relatives did not accept post-mortem examination.

## DISCUSSION

Neuro-behçet disease is approximately 10-20% of patients (6). While neurological involvement may be associated with severe symptoms, however some patients may present mild and subclinical symptoms (2). Two different neurological involvement (parenchymal and non parenchymal) have been described in BD. NBD is primarily parenchymal (80%). Radiologically, lesions of NBD are often localized at brain stem, basal ganglia, or diencephalon. Non parenchymal involvement is frequently associated with major vascular structures in dural venous sinus and called as vascular BD (2). Increased intracranial pressure, severe headache, ocular motor paralysis, seizures, and coma may develop due to venous sinus thrombosis. Furthermore, carotid artery and vertebral artery occlusion, intracranial arteritis, and aneurysm cases have been reported (7). Multiple sclerosis, sarcoidosis, inflammatory demyelinating diseases, central nervous system tumors have to be considered in the differential diagnosis of NBD. Also, NBD occasionally manifests as a tumor like lesion with mass effect (1,2,6).

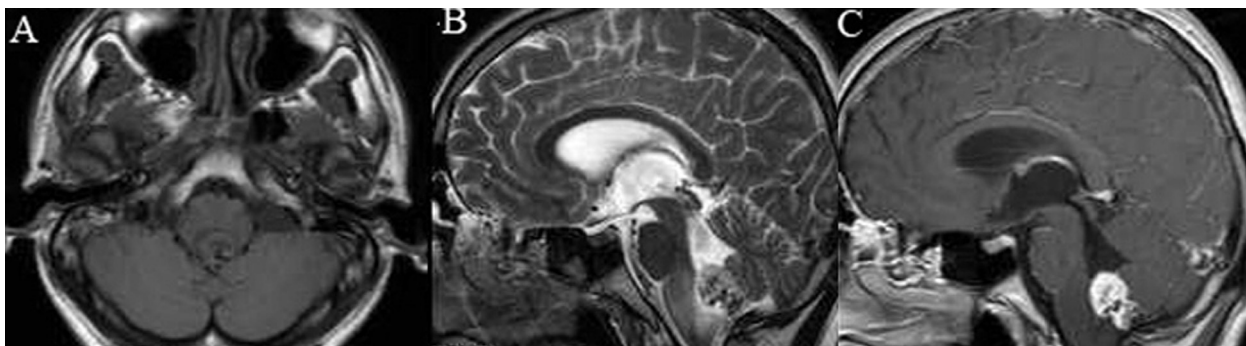
Hemangioblastomas are the infrequent benign tumors of the CNS and incidence is approximately 2% of all intracranial tumors. These tumors are usually placed in cerebellum, spinal cord and brain stem (3). Ventricular location (especially 4th ventricle) and pure solid form of hemangioblastomas are very unusual reported in the literatures (3,4,8). In our case vascular subtype NBD was considered due to diagnoses with sinus rectus involvement at magnetic resonance venography and at same time MRI revealed 4th ventricle localized mass. But the differential diagnosis of mass couldn't been done with MRI. Cerebral angiography was performed for the differential diagno-

sis of the mass which revealed heavy vascular blush. Angiographic findings are specific, and the diagnosis may be more accurate (9). Thus, the mass was accepted to be an incidental ventricular hemanjioblastom without confirming with pathological investigation.

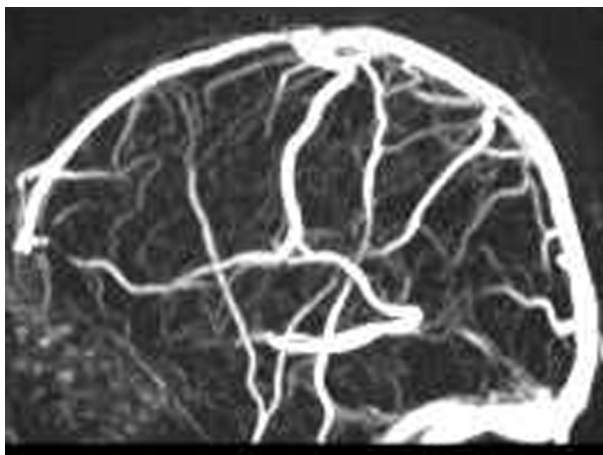
To our knowledge, NBD co-occurrence with hemanjioblastom has not been reported in the previous literature. Hemanjioblastoma should be considered as a differential diagnosis of tumor like appearance in neuro-Behçet's disease patients.

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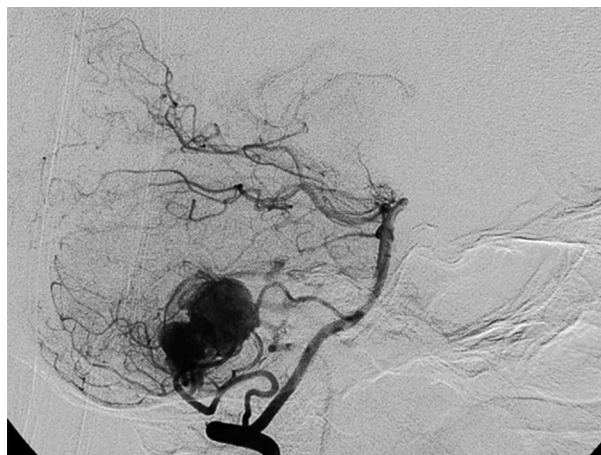
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**Figure 1.** Magnetic resonance imaging. T1- weighted showing isointense mass (A) and T2 - weighted showing a heterogeneous hyperintense mass compared with the normal white matter with central hypointense signal void areas (B). After gadolinium-DTPA injection the mass was markedly enhanced (C).



**Figure 2.** Magnetic resonance venography, decrease in sinus rectus calibration, contour irregularities and corruption in filling



**Figure 3.** Left vertebral angiograms demonstrating a tumor showing heavy vascular blush fed by branches of the left posterior inferior cerebellar artery of the left vertebral artery.