

## A case of Behçet's disease with multiple recurrent visceral artery aneurysms

*Tekrarlayan multipl viseral arter anevrizması ile seyreden Behçet hastalığı olgusu*

Ersan Özbudak, Ali Ahmet Arkan, Şadan Yavuz, Muhip Kanko, Turan Berki

Department of Cardiovascular Surgery, Medical Faculty of Kocaeli University, Kocaeli, Turkey

Behçet's disease is a chronic, recurrent systemic vasculitis which can involve all visceral arteries. In this article, we present a 37-year-old male case with Behçet's disease with recurrent visceral artery aneurysm and the surgical treatment.

*Key words:* Aneurysm; Behçet's disease; visceral artery.

Behçet's disease (BD) is a chronic, systemic inflammatory disease that presents with problems in the ocular, genital, vascular, gastrointestinal, and central nervous systems as well as mucocutaneous lesions.

The best known vascular complication of BD is thrombophlebitis in which giant aneurysms are seen in the peripheral and visceral arteries, for example the pulmonary and carotid arteries.<sup>[1]</sup> Herein, we present a rare case in which a patient with BD was operated on twice due to a rupture of a visceral artery aneurysm (VAA).

### CASE REPORT

A 37-year-old male patient was admitted to the emergency unit with abdominal pain, nausea, and fatigue. He had been diagnosed with BD three years earlier at another clinic where he had been admitted with arthritis and uveitis. The patient also underwent a splenectomy one year prior to being admitted to our facility due to a splenic artery aneurysm (SAA). In addition, the patient had been admitted to a neurology clinic with a headache three months earlier and was found to have lacunar infarctions in the pons and left thalamus. He was then diagnosed with neuro-Behçet's disease. The patient had been taking oral methylprednisolone 4 mg once a day and colchicine 0.5 mg three times a day for three years.

Behçet hastalığı tüm viseral arterleri tutabilen kronik ve tekrarlayan sistemik vaskülitir. Bu yazıda Behçet hastalığı olan 37 yaşında erkek bir olgu, tekrarlayan viseral arter anevrizması ve cerrahi tedavi ile birlikte sunuldu.

*Anahtar sözcükler:* Anevrizma; Behçet hastalığı; viseral arter.

Computed tomography (CT) detected a ruptured gastroduodenal artery (GDA) aneurysm with a diameter of 35 mm along with half-thrombosed right hepatic and celiac truncus artery aneurysms measuring 15 mm and 10 mm in diameter, respectively. The patient consulted with the interventional radiology team, and emergency endovascular intervention was carried out. However, the anatomic localization of the aneurysms was technically challenging. The hemodynamic status of the patient deteriorated, and there was also a sharp decrease in the hematocrit levels. Therefore, the patient underwent an emergency operation.

The abdominal aorta and celiac artery branches were explored through a paramedian abdominal incision. The ruptured GDA was found, and a cross-clamp was applied. The aneurysmatic artery was then divided and ligated, which stopped the bleeding. The operation was challenging due to the previous adhesions from the splenectomy; thus, we left the aneurysm sac in place and did not obtain a specimen for a pathological examination.

The patient stayed in the intensive care unit (ICU) for two days and was discharged home on the postoperative fourth day. At the postoperative first month follow-up, the patient was in good condition. After being discharged, we recommended continuing



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Correspondence: Ersan Özbudak, M.D. Kocaeli Üniversitesi Tıp Fakültesi Kalp ve Damar Cerrahisi Anabilim Dalı, 41380 Umuttepe, Kocaeli, Turkey.

Tel: +90 262 - 303 72 64 e-mail: ersanozbudak@yahoo.com

the previous medical therapy and referred the patient to our hospital's rheumatology clinic where chemotherapy with cyclophosphamide and methylprednisolone was initiated two months after the surgery.

## DISCUSSION

Behçet's disease, which was first described by Turkish dermatologist Hulusi Behçet (1889-1948) in 1937,<sup>[2]</sup> is a chronic, inflammatory disorder characterized by multisystem involvement.

Vascular pathologies are observed in 30% of the cases, but the venous system is more frequently affected (88%). The most common vascular pathology is thrombophlebitis.<sup>[3]</sup> Behçet's disease can affect all types of arteries, with the most common being the aorta along with the pulmonary, femoral, popliteal, subclavian, and carotid arteries.<sup>[4]</sup> Visceral artery aneurysms are seen more often in cases with BD than in the normal population and are usually observed in patients between the ages of 50 and 58. The mean diameter is above 3.5 cm, and the incidence rate is four times higher in men.<sup>[5]</sup> What makes our case peculiar is that our patient was only 37-years-old, and the aneurysms were detected in all of the visceral arteries. In addition, no additional aneurysms were found at the time of the previous surgery for the SAA.

Although superior (SMA) and inferior mesenteric artery (IMA) aneurysms associated with BD have been reported in the literature, aneurysms of the celiac artery and its branches are rare<sup>[6]</sup> and are usually asymptomatic. Furthermore, there is normally localized abdominal pain and discomfort in the epigastric area, and the nausea and vomiting which accompany the abdominal discomfort can lead to misdiagnoses, such as pancreatitis or other acute abdominal pathologies. Moreover, it should be kept in mind that upper gastrointestinal bleeding might be the first symptom in SAAs as well as aneurysms in the GDA. The rupture of these aneurysms is one of the most feared complications, with rupture rates ranging from 50-75% in the celiac artery and its branches having been reported in the literature. Furthermore, a mortality rate of approximately 20% in cases of rupture has also been documented.<sup>[7]</sup>

Abdominal Doppler ultrasound (US), CT, and angiography are normally used in the diagnosis of these patients, but the gold standard is angiography. Visceral

angiography has 100% sensitivity compared with 67% for CT and 50% for Doppler US. Endovascular transcatheter embolization is also a useful treatment option for GDA aneurysms. However, when the transcatheter approach fails or when the hemodynamics of the patient are unstable due to rupture, emergency surgery is required.<sup>[8]</sup>

Behçet's disease is an autoimmune vasculitis that can affect all types of arteries. When compared with the normal population, the formation rates for aneurysms and pseudoaneurysms are higher with this disorder, but the time to aneurysmal formation is shorter in patients with BD. This tendency towards aneurysmatic changes in the arteries may lead to life-threatening ruptures; therefore, these patients should be monitored closely via CT for aneurysmal formation and growth.

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