

## Spontaneous Multiple Pseudoaneurysms Of The Abdominal Aorta And Right Subclavian Artery, In A Patient With Behçet's Disease

Behçet Hastasında Abdominal Aorta Ve Sağ Subklavian Arterde Görülen Spontan Birden Fazla Yalancı Anevrizma

Aydemir Koçarslan<sup>1</sup>, Mehmet Salih Aydın<sup>1</sup>, Mert Ürküp<sup>1</sup>, Abdussamed Hazar<sup>1</sup>, Mustafa Göz<sup>1</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Harran University, Faculty of Medicine, Şanlıurfa, Turkey

### Corresponding author:

Yrd.Doç.Dr.Aydemir KOÇARSLAN

Harran Üniversitesi Tıp Fakültesi Dekanlığı Morfoloji Binası Diyarbakır Yolu Üzeri Yenişehir Kampüsü  
63300 Haliliye / ŞANLIURFA

Tlf:05058125538 Faks: 0 (414) 318 31 92

Email: drakocarslan@gmail.com

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### Abstract

Spontaneous aortic pseudoaneurysm is a rare vascular pathology related to Behçet's disease. We report a 30 year old refugee woman case of spontaneous subclavian and abdominal aorta pseudoaneurysm who presented with 3 months' history of a painful, pulsatile mass on the right clavicle and abdomen. Aort aneurysm was suspected magnetic resonance angiography revealed 53x50 mm size right subclavian artery pseudoaneurysm and 78x64 mm size abdominal aorta pseudoaneurysm. Patient underwent subclavian artery pseudoaneurysm resection.

**Key Words:** Behçet Disease, Abdominal Aorta, Pseudoaneurysm

### Öz

Kendiliğinden gelişen aort yalancı anevrizması Behçet hastalığında görülen nadir bir damar patolojisidir. 3 aydır sağ klavikula ve batın hizasında olan pulsatil kitle ve ağrı şikayeti ile başvuran subklavian arter ve abdominal aortada spontan gelişen yalancı anevrizması olan 30 yaşında mülteci bayanı sunmayı amaçladık. Magnetik Rezonans anjiografi görüntülemesinde subklavian arterde 53x 50 mm genişliğinde , abdominal aorta 78x64 mm genişliğinde yalancı anevrizma mevcuttu. Hasta subklavaian arter yalancı anevrizması rezeksiyon ameliyatına alındı.

**Anahtar Kelimeler:** Behçet Hastalığı, Abdominal Aort, Yalancı Anevrizma

### Introduction

Behçet disease (BD) is an autoimmune disorder affecting gastrointestinal, cardiovascular, and central nervous systems. Vascular lesions are aneurysms and pseudoaneurysms of the aorta or major branches. Complication of this part with a

higher mortality. Rupture is the main cause of mortality in BD with aorta involvement. Open surgical repair is main treatment, but fatal such as bleeding, recurrence pseudoaneurysm of native aort or graft occlusions are not rare. Behçet's disease sometimes attended by aneurysms of the thoracic

aorta, abdominal aorta, and peripheral arteries (1). Aortic pseudoaneurysm is the most lethal lesion in Behçet's Disease. Aortic pseudoaneurysm has high risk of rupture and death. We aimed to present a 30 years old woman who had two spontaneous abdominal aort and right subclavian artery aneurysm with Behçet's disease.

### Case:

A 30-year-old woman presented with 3 months history of a painful, pulsatile mass on the right clavicle and abdomen. Image diagnosis showed that she had pseudoaneurysms of right subclavian artery, and abdominal aorta at the celiac artery level in Magnetic Resonance Angiography (Figure 1). She had no family history of familial vascular disorder. Patient was operated one day after admission the hospital due to pseudoaneurys rupture risk. We treated first the right subclavian arterial aneurysm with operation. Aneurysm was 78x65 mm size and adherent to the surrounding tissue. Therefore pseudoaneurysm was resected under cardiopulmonary bypass because of lesion was enclosed with right common carotid artery. Median sternotomy was preferred to reach the lesion. CPB was performed with ascendan aorta and venous two stage cannulation. Patient was cooled to 28 degrees for brain protection, right subclavian artery and right carotis artery cross clamped and subclavian artery divided. Polytetrafluoroethylene (PTFE) graft interposition was performed between axillary artery and aorta. Due to hypotension abdominal pseudoaneurysm surgery was postponed second session. The pathological examination of the subclavian artery specimen was consistent with vasculitis. Behçet's disease diagnosis was founded after surgery. Because of this reason immunosuppressive therapy was delayed. She had skin eruption and genital ulcer. The pathergy test was positive. After surgery the patient was

admitted to the intensive care unit. Patients were separated from the ventilator twice but due to respiratory failure again intubated. After a week of surgery, abdominal pseudoaneurysm was ruptured and patient died in intensive care unit.

### Discussion:

This disease was reported by Hulusi Behçet, Istanbul, in 1937 (2). Behçet's disease commonly seen around the Mediterranean Middle and Far Eastern countries therefore referred as the Silk Road disease (3). This disease characterised by relapsing uveitis and recurrent oral and genital ulcers. Our patient was from the Middle East country. Multiple aneurysms are common in Behçet's disease (4). The arterial lesions are primarily caused by necrosis in the intimal and medial layers of the artery due to microvasculitis-induced ischemia that affects the vasa vasorum(5). Japanese meta-analysis reported that multiple aneurysms were in 36% of patients with Behçet's disease (3). Uveitis, recurrent oral and genital ulcers are Behçet's diseases symptoms. Our patient had oral afts and genital ulcers history.

Spontaneous aortic pseudoaneurysm due to Behçet's disease can be seen (6). All clinical manifestations of Behçet's disease are depends on vasculitis. Behçet's disease is most seen in Turkey (80 to 370 cases per 100,000) (3). Behçet's disease can affect the small, medium and large-sized arteries (6). In this disease, the most important predictor of morbidity and mortality is the vascular complications (7). Cardiovascular involvement is the most important cause of mortality in Behçet's disease. Large vessel involvement is seen in approximately one third of patients with Behçet's disease (7).

The most common aneurysm site in BD is the aorta, followed by femoral and pulmonary arteries. Thoracoabdominal aneurysms are uncommon with aortic involvement of BD. Recurrent pseudoaneurysms more than five years after graft replacement with total visceral artery reconstruction

is rare. Iscan et al. reported two cases of TAAA involving BD in 20 patients. Follow-up periods were 11 and 12 months. Hosaka et al. reported one case of TAAA repair in 10 patients, and Dacron graft replacement with visceral artery reconstruction was performed. The follow-up was 160 months, without complications. They also reported a cumulative incidence of anastomotic pseudoaneurysms of 12.9% at five and 10 years postsurgery (8).

Surgical treatment should be performed after the immune suppressive therapy remission achieved with Behçet's disease (7). Our patient's Behçet's

disease diagnosis was founded after surgery so that immunosuppressive therapy was delayed. Endovascular graft placement is a treatment option for the aortic pseudoaneurysm (9). Long-term immunosuppressive therapy is important to limit the pseudoaneurysm recurrence after graft placement (7). In our case subclavian pseudoaneurysm was near the carotis and aneurysm neck was short and abdominal pseudoaneurysm was near the celiac artery so pseudoaneurysms were not suitable for endovascular graft placement. Vascular surgeons dealing with young adults with aortic pseudoaneurysms must be aware of Behçet's disease.



Figure-1:MR angiogram showing the pseudoaneurysm of right subclavian artery and pseudoaneurysm of abdominal aorta at the level of the celiac artery

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