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GIANT RUPTURED ABDOMINAL AORTIC ANEURYSM IN A PATIENT WITH BEHCET'S DISEASE: CASE REPORT

Case Report

BİR BEHÇET HASTASINDA DEV RÜPTÜRE ABDOMİNAL AORT ANEVRİZMASI: OLGU SUNUMU

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ABSTRACT

In 1937, Hulusi Behçet, a Turkish dermatologist, reported a disease associated with iritis and ulcerations of the mucous membranes of the oral cavity and genitalia. The main symptoms of Behçet's disease occur not only in the skin, mucosa and eyes, but also in the joints, digestive tract, vascular system, and nervous system. For many years, vascular complications in Behçet's disease have been attributed to thrombophlebitis, aneurysm and arterial occlusion. Aneurysmal changes develop at relatively earlier ages in this condition. In this case, we report a giant ruptured abdominal aortic aneurysm in a patient with Behcet's disease.

Key Words: *Behcet's disease;abdominal aorta; ruptured aneurysm.*

ÖZET

Türk dermatolog Hulusi Behçet, 1937 yılında iritis, oral kavite ve genital mukoza membranlarında ülserasyon bulgularının varlığıyla ilişkili bir hastalık tanımlamıştır. Behçet hastalığı ana belirtileri cilt, mukoza ve göz ile ilgili olup, aynı zamanda eklemler, sindirim sistemi, vasküler sistem ve sinir sistemi de tutabilmektedir. Yıllardır Behçet hastalığının vasküler komplikasyonları tromboflebit, anevrizma ve arteriyel oklüzyonlar ile ilişkilendirilmektedir. Bu hastalıkta anevrizmal değişiklikler göreceli olarak erken yaşlarda gelişmektedir. Bu çalışmada, dev rüptüre abdominal aort anevrizması ile başvuran bir Behçet hastası sunuldu.

Anahtar Kelimeler: *Behçet hastalığı; abdominal aort; rüptüre anevrizma.*

INTRODUCTION

Behçet disease (BD) is a chronic, relapsing, systemic inflammatory disease. Although several immunological abnormalities have been demonstrated, the exact mechanism of the inflammatory changes occurring remains to be elucidated. The most probable hypothesis is that of an inflammatory reaction set off by infectious agents such as herpes simplex virus 1 or Streptococcus spp. or by an autoantigen such as heat schock proteins in genetically predisposed individuals. Association of HLA-B51 is known as the strongest genetic susceptibility factor for BD. Environmental factors, especially infectious agents (S. sangius etc.) are considered to play important roles in the development of BD. Current data suggest that high microbial load and associated stress proteins found in dental tissues and oral ulceration of patients with BD may iniate immunological crossreaction with the heat shock proteins (HSP) and subsequently the development of autoreactive T cells clones (1).

Behçet's disease is a systemic vasculitis which affects both arteries and veins with ocular, skin, joint, central nervous system, respiratory and gastrointestinal system involvement. This disease affects commonly young males (3-5 times more), than women. Vascular involvement occurs in 2-18 years after onset of BD and is affected in the 25% of these patients. This percentage differs between 2-46 % according to the study's method or population. Arterial involvement can be seen either as aneurysm formation or occassionally as obstruction. In BD, peripheral artery aneurysms are usually seen in the carotid, popliteal, and femoral artery locations (2-5). In this case a patient who had BD was admitted to our emergency department with diarrhea and pulseless femoral arteries and magnetic resonance (MR) angiography revealed the exact anatomy of this giant aneurysm.

CASE REPORT

A 44-years-old male patient who had Behçet's disease for 12 years applied to our emergency department with complaints of diarrhea for a duration of one week and difficulty in movements of his left leg for the last 3 days. On physical examination, his left femoral, popliteal and distal arterial pulses were not palpable. Palpation of the abdomen revealed a huge pulsatile mass. Abdominal computerized tomography (CT) showed an infrarenal aneurysm of the abdominal aorta. MR angiography showed a 10x12 cm sized saccular aortic aneurysm which was located 4 cm distal to the renal artery with 3 cm neck. It was seen that the aneurysmal sac had turned to left side with occlusion of the left iliac artery (**Figure 1**).

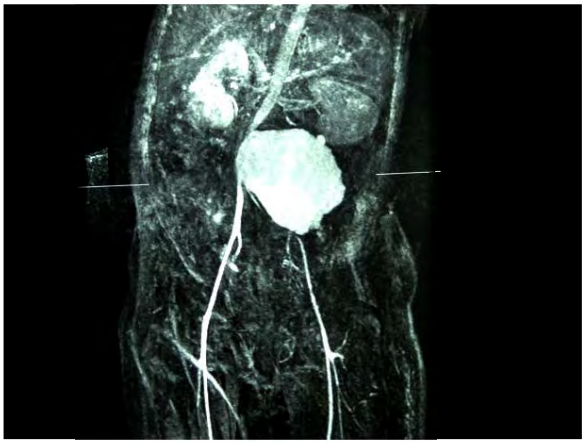

Figure 1: *Magnetic resonance angiography image demonstrating a 10x12 cm sized giant saccular aortic aneurysm of abdominal aorta above the iliac bifurcation which is also compressing the lumen of left common iliac artery.*

The patient was urgently taken to the operation room. During the operation, it was seen that giant aneurysm of the abdominal aorta was ruptured from the left side and rupture side was tamponated by attachment of mesenterium and bowel anses. Upon the dissection, the aneurysmal sac ruptured from this area and bleeding was controlled immediately.

Two clamps were placed over the superior and inferior segments of the aneurysmal sac. The aneurismal sac was opened longitudinally, and both sides of the aneurysmal sac were sutured over and over in order to perform 2 layers over the neck of the aneurysmal sac. After controlling the bleeding, abdomen was closed and he was transferred to the intensive care unit (ICU). He stayed for 3 days in the ICU and on the postoperative 8th day he was discharged from the hospital without any problem. One month later, the control MR angiography showed a 50% narrowing of lumen of the abdominal aorta. The left common iliac artery was normal (**Figure 2**).

Figure 2: *Magnetic resonance angiography one month after the operation showing 50% narrowing of abdominal aorta at the operation side with normal iliac arteries.*

Left femoral and distal arteries of the leg were pulsatile, and he was able to move his leg without any restriction.

DISCUSSION

Abdominal aortic aneurysm (AAA) is usually seen in the population over 60 years of age with a frequency of 4-11 %, and treatment is performed generally by surgery. Most of these aneurysms are localized below the level of renal artery and progress to the iliac artery bifurcation. The most important cause of aneurysm formation in the infrarenal location is high pressure load that is forced by returning pressure waves because of the aortic bifurcation and the deficiency of the lamina elastica interna in this location (6).

In BD, the vasculitis process starts commonly at vasa vasorum. Inflammatory cells are formed by the mononuclear cells and are placed in the perivascular area. Neovascularization can accompany to these mononuclear cells. Changes in the vasa vasorum cause ischemia at the vessel wall and results with ischemia, reduction and degradation of the elastic fibrils and muscle cells and finally this situation prepares a background for the aneurysm formation (7).

Aneurysms due to BD are regarded as special conditions in the vascular surgery. They constitute big problems for cardiac and vascular surgeons because they cause anastomotic site aneurysms and graft obliterations with high percentages. The main rule is to transact aneurysm, control and revascularization in the area of the lesion. Some of the researchers recommend bypassing with healthy arteries as far from the aneurysm as possible, or extra anatomic bypass (8). In another research, ordinary aneury-smectomy and covering of arterial defect with direct sutures or graft are recommended. It is denominated that this technique can be applied in patients of BD with pseudo and saccular aneurysms (9). Particularly, in the surgery of AAA, absence of enough tissue for covering the

graft after fixing the graft in place is cardinal problem, therefore after the surgery of AAA due to BD, enteric fistula into aorta is seen frequently (10). To prevent this complication, Tuzun et al. have surrounded the graft with omentum majus that has been passed meso of transverse colon. This omentum majus is also an ideal tool to fill the pseudoaneurysm pocket that is behind the graft (11).

In conclusion that, MR angiography shows accurate location of AAAs and provides urgent diagnosis, especially in emergency cases. While resection and suturing of aneurysms in BD is fairly difficult due to panvasculitis, our opinion is to suture the aneurysm sac as two-fold over the suture line which makes the suture line more confidential.

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