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Secondary aortoenteric fistula in Behçet's disease

Behçet hastalığında sekonder tip aortaenterik fistül

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Vascular manifestations of Behçet's disease include venous and arterial occlusions, arterial aneurysm and pseudo-aneurysm formation. The main problem of the surgical treatment of vascular lesions in Behçet's disease is the high incidence of complications such as recurrent aneurysms, thrombosis and fistulization to the adjacent organs. Here we present a case of Behçet's disease with multiple complications after aortic reconstructive surgery, including perigraft infection, abscess distal to the graft, occlusion of arteries of the lower extremities, aortoenteric fistula and distal anastomotic site aneurysm rupture.

Key words: Behçet's disease, aneurysm, pseudo-aneurysm, aortoenteric fistula

Behçet hastalığındaki damarsal lezyonların cerrahi tedavisinin ana sorunu, rekürren anevrizma oluşumu, tromboz ve komşu organlara fistülizasyon gibi komplikasyonların sık görülmesidir. Burada, aortic rekonstrüktif cerrahi sonrası mültipl komplikasyon geliştiren bir Behçet hastasını sunuyoruz. Bu komplikasyonlar, oluş sırasıyla, greft çevresi enfeksiyon, greft distalindeki organlarda abse gelişimi, alt ekstremite arterlerinin tıkanması, aortaenterik fistül gelişmesi ve distal anastomoz hattında anevrizma gelişmesi ve rüptürüdür.

Anahtar kelimeler: Behçet hastalığı, anevrizma, psödoanevrizma, aortoenterik fistül

INTRODUCTION

Aortoenteric fistula (AEF) is a rare but deadly medical emergency. Primary AEFs occur between the native aorta and the intestinal tract. Secondary AEFs occur between an aortic graft and the gastrointestinal tract. Patients often present with a "herald bleed", followed by massive gastrointestinal hemorrhage or with sepsis and abdominal pain (1-3).

In 1937, the Turkish dermatologist Hulusi Behçet described the triad of relapsing iridocyclitis and orogenital ulcerations that now bears his name (4). In addition, Behçet's disease commonly involves cardiovascular, pulmonary, neurologic, articular and gastrointestinal systems (5-7). Vascular manifestations include venous and arterial occlusions, arterial aneurysm and pseudoaneurysm formation. The main problem in surgical treatment of vascular lesions in Behçet's disease is the high incidence of complications such as recurrent aneurysms, thrombosis and fistulization to the adjacent organs (5-9).

Here we present a case of Behçet's disease with multiple complications after aortic reconstructive surgery.

CASE REPORT

A Turkish male patient was first diagnosed as having Behçet's disease at the age of 35 years in 1995. At that time he had the classical triad of the disease. In March 1997, he was admitted to our hospital gastroenterology clinic, for the first time, with a complaint of recurrent, dull epigastric pain. Based on clinical and laboratory findings, he was considered as having chronic alcoholic pancreatitis. In March 1998, he was readmitted with newly onset very severe right lumbar and inguinal pain. A diagnosis of ruptured abdominal aortic pseudoaneurysm was made and a tubular hemo-shield graft was inserted with aneurysmorrhaphy. An enlarged, rock hard pancreatic gland was seen during the operation. A possible relation between the

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pancreatitis and Behçet's disease was suggested and the case was reported as the ninth pancreatitis case in Behçet's disease (10).

In November 1998, nine months after the operation, he was readmitted to our gastroenterology clinic with a history of back and left leg pain for one and a half months and increasing fever with chills for the last 15 days. There was a tender and hyperemic swelling over the gastrocnemius muscle of his left leg. Bleeding history was negative. The white blood cell count (WBC) was 32000/γl, hemoglobin 7.7 g/dl, hematocrit 24%, platelets 389,000/γl, erythrocyte sedimentation rate (ESR) 70 mm/h, fibrinogen 8.1 g/L, C-reactive protein (CRP) 18.5 mg/dl, albumin 2.6 g/dl, serum iron 26 γg/dl, serum iron binding capacity 298 γg/dl, transferrin saturation 9%, ferritin 234 ng/ml, and haptoglobin 4.6 g/L. Fecal occult blood was negative. On Doppler ultrasonographic imaging of the aorta, the graft was reported as patent, but there was a tortuosity just distal to the renal arteries (Figure 1). The maximal diameter of the aorta was measured as 18 mm. Vena cava inferior and lower extremity arteries and veins were reported as normal sonographically. Sonographic evaluation of the swelling in the left leg revealed a mass with heterogeneous and mobile echo pattern and not well-demarcated borders that were reported as compatible with abscess or infected hematoma. The mass lesion was incised and a purulent discharge was noted. Both the cultures of the blood and abscess yielded enterococcus. Upper gastrointestinal endoscopic examination was reported as normal. The patient's complaints resolved with drainage, antibiotic therapy of four weeks and other supportive therapies. His anemia was corrected with transfusion of four units packed red blood cells. He was consulted with cardiovascular surgeons for a possible graft complication, especially for perigraft infection, but they believed that the graft was functioning well and that the existing findings were inadequate for reoperation of a patient with Behçet's disease. He was discharged with colchicine, ciprofloxacin, Coumadin, famotidine and vitamin B complexes.

In March 1999, he was readmitted in poor condition with very severe back and leg pain, increasing fever with chills, fatigue and weight loss. He was well for the first 15 days after discharge while taking ciprofloxacin. After discontinuing the drug, his pain and fever reappeared with a gradual increase in severity. Again there was no bleeding in his history. The laboratory values were as follows: hemoglobin 8.6 g/dl, hematocrit 26.5%, platelets 256000/γl, WBC count 10300/γl, ESR 76 mm/h, fibrinogen 7.8 g/L and CRP 17.25 mg/dl. Fecal occult blood was negative again. Both urinary and blood cultures yielded enterococcus. Abdominal ultrasonography demonstrated that the abdominal aortic graft was bending anteriorly together with a minimal splenomegaly and peripancreatic lymph-adenomegaly. On the abdominal computed tomography, the presence of a saccular aneurysmal dilation between the renal arteries and the iliac bifurcation, 5 cm in length, with a maximal diameter of 32 mm was reported (Figure 2). Peripheric angiog-

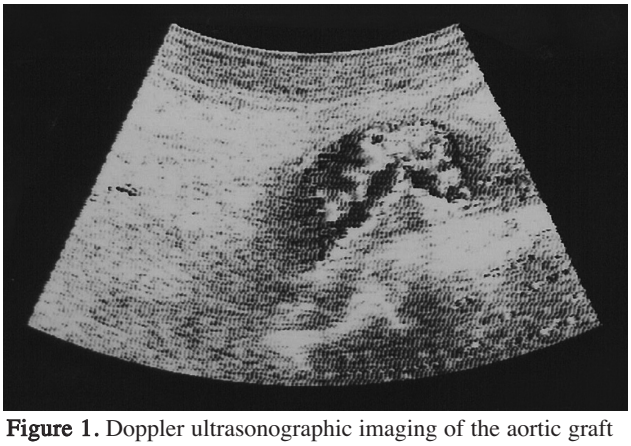

Figure 1. Doppler ultrasonographic imaging of the aortic graft demonstrating anterior bending of the graft, taken at the postoperative first admission of the patient.

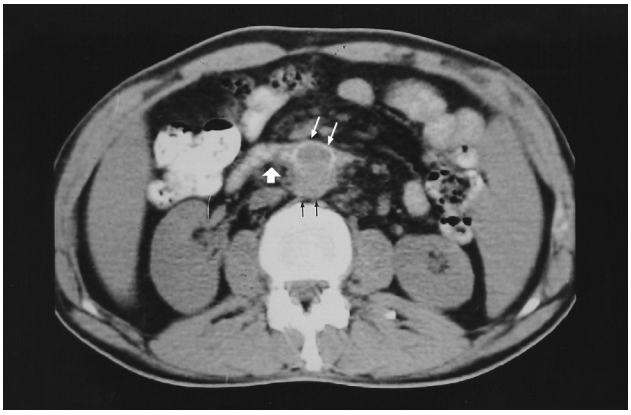

Figure 2. Tomographic film of the abdomen taken at the postoperative second admission of the patient. The first interpretation of this film was the presence of an aneurysm, but when we reviewed the films after the demonstration of aortoenteric fistula, graft invasion of the duodenum was noted. Short arrows point to the aneurysmatic aorta, long arrows to the aortic graft and bold arrow to the duodenum. A close relation between the anteriorly displaced aortic graft and transverse (third) portion of the duodenum can easily be seen.

raphy demonstrated the occlusion of posterior and anterior tibial and peroneal arteries of both legs with collateral circulation. The patient experienced an upper gastrointestinal bleeding during hospitalization. Emergency upper endoscopy showed diffuse mucosal erosions at the antrum, bulbus and descending duodenum. He was reconsulted with cardiovascular surgeons with a suggestion of perigraft infection. The surgeons wished to confirm the suspicion with indium-labeled leukocytes scintigraphy, which was reported as normal. Despite the presence of aneurysm on the tomography, the negative scintigraphy result encouraged the surgeons to follow the patient without operation and the patient was discharged on 15 April 1999.

On 8 June 1999, he was readmitted with a history of massive upper gastrointestinal bleeding that required 10 units of packed red blood cells transfusions. On the sonography, there was an anterior bending of the aortic graft that seemed to have migrated into the duodenum. On the upper endoscopy, a pulsatile foreign body (graft), with a mesh appearance filling all the bowel lumen, was seen between the second and third parts of the duodenum (Figure 3). He was taken for emergency surgery. A duodenal defect 10 cm in length and 2 cm in width was seen and closed primarily with Prolene sutures. The graft was excised and a new tubular hemo-shield graft was inserted with omentoplasty. The cultures taken from the graft and perigraft tissue yielded enterococcus and *Escherichia coli*. Postoperative recovery was uneventful and he was discharged with omeprazole, colchicine, Coumadin and vitamin B complexes.

During the routine follow-up on 6 September 1999, an aneurysmal dilation at the distal end of the graft just proximal to the iliac bifurcation was seen on sonography. He had no complaints. While waiting for the operation, a hypotensive crisis developed. He was reoperated on an emergency basis with distal anastomotic site aneurysm rupture and an axillobifemoral extra-anatomic bypass with total graft excision and aortic closure was done. He was well at the last follow-up examination in September 2005.

DISCUSSION

Perigraft infection without fistulization occurs in 2-6% of patients, while AEF is a complication occurring in 0.6-2% of aortic graft operations. Bacterial infection of a prosthetic graft is caused by intraoperative contamination from bowel injury or the environment, postoperative pressure necrosis of the bowel or transient bacteremia from a satellite source. AEFs occur as two basic types: a fistula from the prosthetic graft suture line to the intestine, which commonly develops as a consequence of suture line pseudoaneurysm eroding the bowel wall (graft enteric fistula), and a paraprosthetic enteric fistula in which the body of the graft erodes into the adjacent bowel (midgraft enteric erosion). The third and fourth portions of the duodenum are most frequently involved. The incidence increases when the initial surgery was necessary to manage aortic rupture (1-3). The case presented here had a paraprosthetic type enteric fistula that eroded the third portion of the duodenum.

Paraprosthetic enteric fistulas form when the mechanical pulsations or infection of an aortic graft erodes the bowel wall. Unexplained fever and malaise frequently develop. Septic emboli that cause abscesses on the lower extremities, septic arthritis, multicentric osteomyelitis and hypertrophic osteoarthropathy have been described (1-3). There was an abscess over the left gastrocnemius muscle,

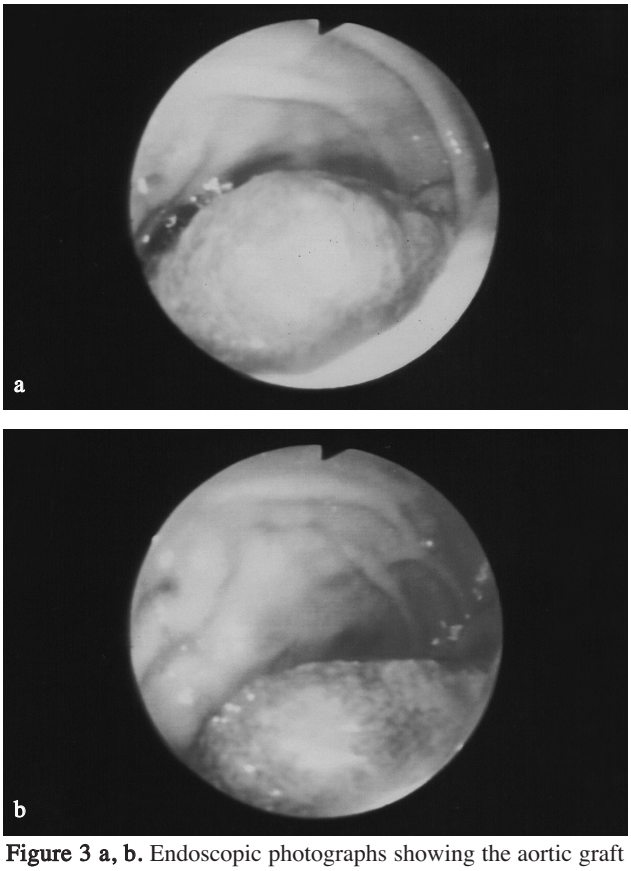

Figure 3 a, b. Endoscopic photographs showing the aortic graft within the duodenal lumen. Of particular note is the absence of blood within the duodenal lumen.

increasing fever with chills, leg and back pain, unexplained anemia and sepsis during the first hospitalization of the case. All these can be explained by the presence of perigraft infection and/or paraprosthetic AEF. In fact, all his findings were "heralding" the presence of perigraft infection and/or AEF. However, this could not be proven until the demonstration of the direct involvement of the duodenum with endoscopy.

The incidence of vascular involvement in Behçet's disease is 10-80% in the literature. Ten percent of vascular lesions are severe or life-threatening. Approximately two- thirds of the arterial lesions are aneurysms that develop most commonly in the aorta. They are most often saccular, punched-out pseudoaneurysms that are prone to infection, rapid enlargement and rupture. Obliterative endarteritis of the vasa vasorum is the etiological factor thought to be responsible for aneurysm formation. Since the wall of the artery is weakened by inflammation, surgical treatment of aneurysms is often complicated (up to 25%) with recurrent aneurysms (especially anastomotic), thrombosis, infection and fistulization to adjacent structures such as vein and intestine (5-9). Surgical results are not satisfactory in Behçet's disease, because of progressive graft thrombosis and formation of new aneurysms at the anastomotic sites (10, 11). In a recent study from our unit, the 10-year complication-free survival rate of patients operated for arterial manifestations of Behçet's disease was found as low as 13%, and the five-year reoperation-free survival rate was only 26% (12).

The first vascular manifestation of Behçet's disease in our case was a ruptured abdominal aortic pseudoaneurysm that had been treated with in situ graft placement and aneurysmorrhaphy. Infection of the prosthetic material, fistulization to the intestine and occlusion of the arteries of the lower extremities were all seen after the first operation. After the second operation, in which in situ replacement of the infected graft with omentoplasty was done, an aneurysm at the distal suture line developed within three months. In case of inflammatory aneurysms, extraanatomic bypass technique is preferred by most authors (1-3, 14).

Tomography is the technique of choice for examining patients with possible aortic graft infection, but its sensitivity may be as low as 33%. Air within the graft, presence of perigraft soft tissue or fluid, new aneurysm formation and persistent separation of the graft and native aortic wrap are the findings compatible with infection. Tomographic manifestations of AEF are similar to those of perigraft infection. In addition, extravasation of contrast materials used may be seen rarely. Some authors report high sensitivity for indium labeled leukocytes scintigraphy in the diagnosis of graft infection, but others have found tomography to be more sensitive (15-17). In our case, the failure of the scintigraphy was apparent; however, tomography reported that there was an aneurysm at the level of the aortic graft. In fact, when we reviewed the tomography films, we saw that the graft was displaced anteriorly and migrated into the duodenum (Figure 2). In addition, the sonographic findings (tortuosity and anterior bending of the graft), which were seen in our case, may be important clues compatible with graft complications. It must be mentioned that these ultrasonographic findings were present since his first admission after the first operation. Tomography and ultrasonography were indeed giving valuable clues about the graft complications in this case, but the correct identification of these findings is not always possible.

In summary, when a patient with known previous prosthetic aortic reconstruction presents with pain, anemia or sepsis, a high index of suspicion must be maintained in order to not miss an underlying graft complication. From the gastroenterologists' standpoint, an upper endoscopy up to the third or if possible fourth portion of the duodenum must be performed in order to demonstrate different clinical presentations of graft complications.

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