Management and prognosis of nonpulmonary large arterial disease in patients with Behçet disease

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Objective: **The purpose of this study was to evaluate and report our treatment policies in the management of nonpulmonary arterial aneurysms in Behçet disease and to assess the prognosis in a cohort of 25 patients diagnosed between 1996 and 2007 by formally reassessing their outcome at the present time.**

Methods: **We identified 25 patients (24 men/1 woman) with Behçet disease with nonpulmonary aneurysms (n = 23) or occlusions (n = 2) between 1996 and 2007. All patients fulfilled the International Study Group Criteria for Behçet disease. Aneurysms were demonstrated with contrast-enhanced computed tomography (CT) or magnetic resonance angiography (MRA) after first-line ultrasonography. Standard surgical procedures were carried out in 22 patients. One patient with a nonruptured saccular aortic aneurysm and 2 patients with carotid aneurysms were managed only medically. For the patients with aneurysms located in the aortic bifurcation, we preferred aorto-bi-iliac bypasses; for the six extremity aneurysms, we were able to ligate the arteries; and for the other 10 extremity aneurysms we used polytetrafluoroethylene (PTFE) grafts for bypass procedures. All patients received immunosuppression with cyclophosphamide and corticosteroids before the operation and were continued in the postoperative period. All patients were examined between January and December 2010 paying special attention for new and anastomotic aneurysms and graft patency.**

Results: **There was one death and 1 patient was lost to follow-up. The remaining 23 patients (92%) were under follow-up after a mean of 7.4 ± 2.9 years after their operation. Four PTFE grafts (40%) inserted for extremity aneurysms occluded with no disabling consequences. Also, 6 patients who were treated with ligation postoperatively began to complain of mild to moderate claudication. In 2 patients, aneurysms recurred at the anastomotic site, whereas in 3 patients, new aneurysms developed at other sites.**

Conclusion: **The surgical management of large, nonpulmonary arterial disease of Behçet disease is currently quite satisfactory. When the false aneurysm is in the infrarenal aorta, aorto-bi-iliac bypass is the preferred surgical intervention. Extremity aneurysms can be treated with synthetic graft insertion. In selected cases, ligation can give satisfactory results; however, postoperative claudication is common. In some patients with small intact saccular aneurysms, surgery may not be necessary. Patients must be prescribed immunosuppressive therapy with cyclophosphamide and corticosteroids before and after the surgical intervention in order to avoid Behçet disease activation. (J Vasc Surg 2012;55:157-63.)**

Behçet disease is a multisystem vasculitis characterized mainly by skin mucosa lesions.[1,2] It may also involve the eyes, blood vessels of all size and types, the central nervous system, and joints.[2] Vascular disease develops in up to 25% to 35% of the patients and has a definite male preponderance.[3,4] Venous involvement is more common than arterial disease (75% vs 25%).[2-4] The most common manifestation is deep venous thrombosis (DVT) in a lower extremity, followed frequently by vena cava disease, pulmonary artery disease, aortic or peripheral artery aneurysms, venous sinus thrombosis, and Budd-Chiari syndrome.[4]

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Large artery involvement is seen in 1.5% to 2.2% of all patients with Behçet disease, whereas aneurysmal pathology is more common than occlusions.[2,4,5] The most common sites for aneurysm development are the abdominal aorta, pulmonary, femoral, popliteal, and carotid arteries.[2,4,5] The main pathologic event is a lymphocytic and neutrophilic necrotizing vasculitis leading to saccular aneurysms and rupture.[6] Immunosuppressive therapy with or without vascular surgery is used in disease control.[7] Recurrent aneurysm formation after open surgical and stent graft treatment is a frequent and serious complication.[8-10] Due to its unique pathology, surgical as well as medical management methods are different from atherosclerotic aneurysms. As a dedicated Behçet disease vascular disease referral center, herein we report our experience with 25 patients with Behçet disease referred to us within a decade with emphasis on disease features, management, and prognosis.

METHODS

From 1996 to 2007, 23 patients with Behçet disease with nonpulmonary artery aneurysms and 2 patients with peripheral arterial occlusions had been treated at the Department of Cardiovascular Surgery, Cerrahpasa Medical

Faculty, Istanbul, Turkey. Sixteen of these patients were referred from the multidisciplinary Behçet disease outpatient clinic of the same university hospital, while the remaining 9 patients were referred from other hospitals with the diagnosis of Behçet disease and aneurysm formation. The multidisciplinary Behçet disease outpatient clinic of our hospital has been operative for over 30 years, the staff meets weekly, currently has around 8000 registered patients, and regularly consults our surgical department for any patient with large arterial disease.

All patients fulfilled the International Study Group criteria[11] for Behçet disease at the onset of arterial disease and had been diagnosed as Behçet disease before referral, except for 1 patient. This 37-year-old man was referred by another hospital with a false aneurysm of the aorta. We first suspected that the diagnosis was Behçet disease based on the morphology of the aneurysm during the operation, and then the diagnosis was verified by the Behçet disease research center of our hospital. Arterial disease was diagnosed either by Doppler scan, computed tomography (CT), or by magnetic resonance angiography (MRA).

Medical therapy (cyclophosphamide and corticosteroids), as a rule, was initiated as soon as the patient was hospitalized and prepared for surgery. Details will be given further on in this study.

In this cohort, 22 of 25 patients were treated surgically, whereas 3 patients (1 with an abdominal aorta and 2 with carotid arteries) with small asymptomatic saccular aneurysms received only medical treatment. In 1 among 22 operated cases, large ruptured saccular aneurysm was treated operatively, whereas a small nonruptured saccular aneurysm of the same aorta was followed under medical therapy alone.

During the postoperative period, all patients were seen every 3 to 6 months with routine physical examination, including peripheral pulse examination and blood/urine tests by the multidisciplinary Behçet disease outpatient clinic. CT, MRA, or Doppler scan were obtained yearly.

Between January and December 2010, all patients were called back to the outpatient clinic for formal evaluation to investigate the long-term outcome, including Behçet disease activation, clinical status, graft patency, and new or recurrent aneurysm formation. The outcome information was taken with the help of a telephone interview in only 1 patient. This patient had attended our outpatient clinic for 1 year after his operation and is currently followed by another center.

Statistical analysis. Continuous variables with a more or less normal distribution were expressed as means ± SDs and others as medians and interquartile ranges. Statistical analyses were carried out using statistical software (SPSS, version 13.0 for Windows; SPSS, Chicago, Ill).

RESULTS

A total of 25 patients (24 man/1 woman) with Behçet disease with nonpulmonary aneurysms and occlusions were referred to our vascular surgery department between the years 1996 and 2007. Demographic and clinical characteristics are shown in Table I. The mean age at the diagnosis of vascular lesions was 39 ± 10 and the mean time elapsed between the diagnosis of Behçet disease and emergence of vascular disease was 9.0 ± 7.9 years. Venous thrombosis was present in the majority of the patients (22 of 25; 88%), mainly in the form of lower extremity DVT (17 of 25; 68%). Three of 25 patients (25%) had vena cava thrombosis. None of the patients had pulmonary artery involvement. Nine patients (38%) had eye disease, of which 5 patients (20%) had loss of useful vision. Neurological disease was seen in 2 patients (8%): 1 patient had dural sinus thrombi and the other had parenchymal central nervous system disease.

Table I. Demographic and clinical characteristics of the patients with Behçet disease with nonpulmonary artery disease

	n = 25
Men, n (%)	24 (96)
Age at aneurysm's onset, mean ± SD, years	39 ± 10
Disease duration, mean ± SD, years	9.0 ± 7.9
Manifestation of Behçet disease, n (%)	
Oral ulcers	25 (100)
Genital ulcers	18 (72)
Papulopustular lesions	14 (58)
Erythema nodosum	13 (54)
Venous thrombosis	22 (88)
Eye disease	9 (38)
Terminal eye disease	5 (20)
Neurological disease	2 (8)
Signs and symptoms at presentation	*n (%)*
Pulsatile mass	5 (20)
Painful mass	8 (32)
Back and or abdominal pain	7 (28)
Mass in the neck	3 (12)
Digital gangrene	1 (4)
Transient ischemic attack	1 (4)

Table II. Anatomic localization of 26 aneurysms (initial and new) in 23 patients

	Number
Abdominal aortic	7
Iliac	1
Common femoral	5
Superficial femoral	3
Popliteal	5
Carotid	3
Posterior tibial	1
Iliofemoral saphenous graft	1

Aneurysms or occlusions. As shown in Table II, there were a total of 23 patients with 26 aneurysms and 2 patients with 2 occlusions. Three patients had a second new aneurysm in the popliteal artery - abdominal aorta (n = 1); popliteal artery - superficial femoral artery (n = 1), and posterior tibial artery - superficial femoral artery (n = 1), whereas 2 patients had anastomotic aneurysms. The mean

Table III. Types of surgical procedures

	Number
Aneurysmorrhaphy (patching)	1
Aortic tube graft interposition[a]	3
Iliac artery tube graft interposition[a]	1
Aorto-bi-iliac bypass[a]	3
Common femoral graft interposition[a]	5
SFA graft interposition[a]	1
Femoropopliteal bypass[a]	1
Popliteal graft interposition[a]	3
Carotid graft interposition[a]	1
Ligation	6
Carotid endarterectomy	1

SFA, Superficial femoral artery.
[a]Polytetrafluoroethylene (PTFE) graft.

aneurysm diameter (taking the largest diameter) was 4.4 ± 2.0 SD cm. Apart from the aneurysms, 1 patient was operated on for superficial femoral artery occlusion that led to digital gangrene and another for carotid artery stenosis at the bifurcation level causing transient ischemic attacks. The most common localization for arterial involvement was the infrarenal abdominal aorta followed by the popliteal, common femoral, and carotid arteries.

Surgical procedures. Surgical procedures performed are listed in Table III. We had performed almost the same operations as in our first report[8]: "polytetrafluoroethylene (PTFE) graft interposition" was done in 2 aortic, 1 iliac, 5 common femoral, 1 superficial femoral, 3 popliteal, and 1 common carotid artery aneurysms. We had done "aorto-bi-iliac bypass (PTFE)" in 3 aortic aneurysms, and 1 patch closure of the aortic defect above the superior mesenteric artery. In infrarenal aortic aneurysms, we preferred to use bifurcated grafts instead of tubular grafts. In this way, we excluded the aorta and the bifurcation where recurrence occurred frequently as had been brought up in our earlier manuscript (2 of 9 abdominal aortic aneurysms).[8] From our earlier experience with 24 patients with Behçet disease, we had learned that grafts inserted for extremity aneurysms occluded without disabling ischemia.[8] Based on this, we were able to ligate two superficial femoral, two popliteal, one posterior tibial arteries, and one venous conduit between external iliac and common femoral arteries. Apart from these, we had 1 patient with digital gangrene who required femoro below-knee bypass for occlusion. Also, in another young patient with transient ischemic attacks, internal carotid artery stenosis was treated by endarterectomy and PTFE patching.

Among the patients referred to us, we had operated on all aneurysms except for 3 patients with asymptomatic intact aneurysms. In 1 patient, there were three saccular aneurysms at the aorta (1 cm each; Fig 1). In another patient, there was a 2-cm saccular aneurysm at carotid bifurcation. The remaining patient had presented with bilateral fusiform aneurysms at the common carotid arteries (3 and 2 cm; Fig 2). Among the patients we operated on, there was 1 patient whose symptomatic bifurcation aneurysm (7.5 cm) was treated with a bypass procedure, whereas the nonruptured aneurysm at the level of superior mesenteric artery (1.5) was given only medical treatment.

Pathology. The main macroscopic pathology in all patients was either intact or ruptured but contained saccular pseudoaneurysms. They were found to be surrounded by a thick fibrotic tissue and enlarged lymph glands. Pathologic investigation was available in almost all patients and revealed similar findings to that found in our previous report.[8] Briefly, this was characterized as infiltration of inflammatory cells, especially around the vasa vasorum, disruption of internal elastic membrane, loss of elastic and muscle fibers in the media, periadventitial, and intimal fibrosis along with organized thrombi (Fig 3). In the current group, there was only 1 patient in whom atherosclerotic changes were more prominent than inflammatory vasculitis.

New aneurysms/recurrences. Three patients had new aneurysms at previously uninvolved sites and 2 patients had recurrences. All patients were under immunosuppressive treatment.

One patient who underwent a PTFE interposition for popliteal artery aneurysm, developed a second aneurysm this time in the abdominal aorta 2 years after the first presentation. Aneurysm at the abdominal aorta was treated with aorto-bi-iliac bypass. In another patient whose popliteal artery aneurysm was treated with a PTFE graft interposition which occluded 3 months later, a superficial femoral artery aneurysm developed after 2 years. This aneurysm was ligated. In another patient whose posterior tibial artery was ligated, developed a superficial femoral artery aneurysm which was treated with PTFE interposition.

Recurrence at the initial surgical site occurred in 2 patients. One patient had anastomotic dehiscence which occurred at the aortic patch 6 months after the surgery. This patient died of exsanguination while waiting for an endograft. The other patient had a false aneurysm at the PTFE femoral artery anastomosis. He was treated with ligation. Apart from these, there were no other recurrences or new aneurysm formation.

Graft occlusion. Four extremity grafts of 10 (40%) were occluded. One common and one superficial femoral artery graft occluded at 1 year, one popliteal artery graft occluded 3 months later, and another popliteal artery graft occluded shortly after operation with mild to moderate claudication. Because graft occlusions were well tolerated by the patients due to arterial collaterals, no additional procedures were undertaken.

Apart from suture infection that occurred in 3 patients, no major adverse event was seen in those patients that we operated on.

Survival. By the end of follow-up, only 1 patient had died 6 months after being operated on, as we noted above. Another patient was lost to follow-up after 1 month of the surgery. We had the outcome information in the remaining 23 patients. All 23 patients (92%) were alive after a mean of 7.4 ± 2.9 years. By the end of follow-up, 12 patients (52%) complained of mild to moderate claudication. The majority



Fig 1. a and **b**, A 37-year-old man, diagnosed with Behçet disease for 15 years, was admitted to the outpatient clinic for flank and abdominal pain of 2 months' duration. Computed tomography (CT) angiogram showed three saccular aneurysms each with a diameter of 1 cm at the infrarenal aorta. The patient's symptoms disappeared after medical treatment with cyclophosphamide and corticosteroids.

(16 of 23 patients; 70%) were in complete remission for other stigmata of Behçet disease; whereas the remaining patients had one or more Behçet disease symptoms. There were only 6 patients with oral ulcers, 2 patients with papulopustular lesions, 3 patients with arthralgia, 1 patient with erythema nodosum, and another with stasis ulcer on the tibia due to the chronic DVT. The only patient who had to be interviewed over the telephone had attended our outpatient clinic for the following postoperative year then was followed by another institution. He reported that he had no pain, no claudication, and no Behçet disease symptom.

Medical treatment. Corticosteroids were administered as 3 pulses of 1 gram prednisolone (every other day) and continued at a dose of 1 mg/kg/day for 1 month then tapered by 10 to 20 mg/month but usually continued for at least 1 to 2 years at a dose of 5 to 10 mg/day.

Three patients, including the 1 patient who was lost to follow-up, received only corticosteroids.

A total of 20 patients received cyclophosphamide and 2 patients received azathioprine (AZA) in addition to corticosteroids, making 22 patients (88%) in total who were treated with immunosuppressive agents. Cyclophosphamide (most commonly in the form of monthly 1 gram intravenous boluses) was given for a duration of median 12 months (interquartile range, 12-14 months), then switched to AZA for maintenance treatment. By the end of the follow-up, 15 patients (60%) were still using an immunosuppressive agent (AZA, n = 13; mycophenolate, n = 1; interferon, n = 1). Apart from 1 patient who had died and 1 patient lost to follow-up, 8 patients were drug free.

DISCUSSION

We report here the outcome and treatment results of 25 patients with Behçet disease with nonpulmonary artery aneurysms (23 patients) and occlusions (2 patients) followed in a single center.

The frequency of arterial aneurysms in Behçet disease is reported to be rare.[2,5] In a retrospective survey of 6000 patients with Behçet disease registered in the multidisciplinary Behçet Syndrome Research Center at the Cerrahpasa Medical Faculty, the frequency of aneurysm formation (including pulmonary artery aneurysms) was 2.5%.[12] As has previously been reported, arterial aneurysms are mainly seen among men[2-5] and the same was true with only 1 female patient in the current series.

Major arterial involvement is known to be a late phenomenon developing 5 years after the fulfillment of the diagnostic criteria.[2-5,8,12] Similarly, in the current study, the time elapsed between diagnosis of Behçet disease and emergence of aneurysms was a mean 9.0 ± 7.9 SD years. Another distinguishing feature of the patients with arterial involvement is accompanying severe eye problems and venous thrombosis.[5,13] These were also apparent in our study. One third of the patients had lost useful vision in both eyes and 88% of the patients had venous thrombosis. Venous thrombosis is also known to be strongly associated

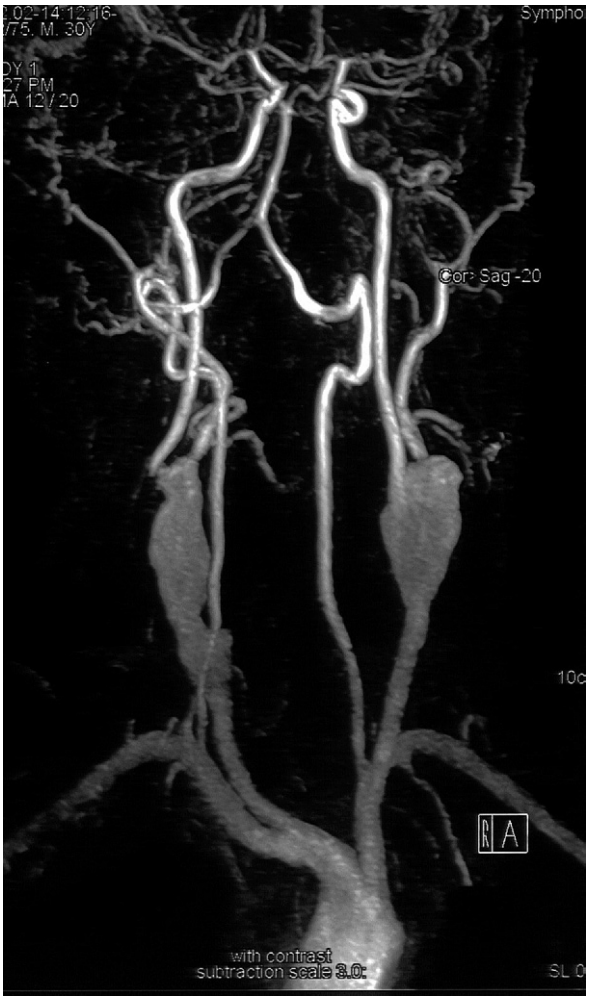


Fig 2. A 30-year-old man, diagnosed with Behçet disease for 3 years, was referred to our department for swelling and pain on both sides of the neck. Magnetic resonance angiogram (MRA) showed bilateral fusiform aneurysms (2 and 3 cm in diameter and 5 and 4 cm in length, for right and left side, respectively) at the carotid arteries. The patient's symptoms gradually disappeared after medical treatment with cyclophosphamide and corticosteroids.

with pulmonary artery disease.[2,4,12,14] However, the association of peripheral artery disease with pulmonary artery involvement seems rather weak[12] and our observation of no patients with pulmonary arterial aneurysms in this group supports this contention.

In Behçet disease, active inflammation of the involved artery destroys the elastic and muscle cells in the media with aneurysmal dilatation. Occlusion of the vasa vasorum speeds this process by transmural necrosis and later with perforation and pseudo aneurysm formation.[6,15] False aneurysms originate from defective areas in the aortic wall. Sometimes there is more than one defective or weakened area. We noticed that dimensions of the abdominal aortas were within the normal range (data not given) with abundant inflammatory fibrotic tissue with or without enlarged lymph nodes around. These features contrast with what is seen in atherosclerotic aneurysms.

Although there are reports of patients who were treated as an abdominal emergency because of free rupture,[16,17] none of our patients with abdominal aorta and extremity artery pathologies were treated on an emergency basis. This was similar in our previous report[8] in which only 1 patient with a common carotid pseudo aneurysm had to have emergency surgery because of airway compression.[8] Behçet disease has an undulating course of exacerbations and relapses. During each exacerbation, dense inflammatory tissue builds up around the involved arteries. In all our abdominal aortic cases, the aorta was found to be surrounded by a thick fibrotic tissue. Presumably, this fibrotic tissue prevents the catastrophic intraperitoneal free rupture.

Another feature of Behçet disease is the arterial occlusion. This distinctly less common pathology has a relatively benign course. Since 1977, we have treated only 2 patients with Behçet disease with symptomatic occlusive pathology, which are reported in the current manuscript. One of them had digital gangrene, and another was a young patient who had transient ischemic attacks due to severe narrowing of the carotid bifurcation. Femoral below-knee bypass with a PTFE graft had been inserted in the former and standard carotid endarterectomy with a PTFE patch was undertaken in the latter.

Aneurysm formation in Behçet disease is rather painful because of its inflammatory nature. For this reason, peripheral aneurysms are easily detected by the patient himself/herself. However, the detection of aneurysms confined to the abdominal aorta needs a great degree of suspicion. Usually patients will complain of back pain, stubborn constipation, and other symptoms due to venous thrombosis.[8] Flank pain will be the only symptom in iliac artery false aneurysms due to constriction of the ureter with dense inflammatory tissue around the aneurysm. A CT scan will give important information about vascular morphology. It is very important to refrain from conventional angiography in that the arterial punctures have the propensity to cause a pathergy reaction and eventually cause localized arteritis and aneurysm.[18,19] We use contrast-enhanced CT, MRA or Doppler scan during the preoperative and postoperative period.

Most of the vascular surgery performed in Behçet disease is for aneurysms. The commonest localization of the aneurysms is the aortic bifurcation.[8,9,16,17] Except for small saccular aneurysms without rupture, we resorted to surgery in all others. During operation, proximal aortic control is secured below the left renal vein without difficulty. Now we prefer to use bifurcated (PTFE) grafts between the aorta and the proximal common iliac arteries. This method excludes the aortic bifurcation and no anastomotic dehiscence is encountered. While thick fibrotic tissue around the aorta makes suturing easy and safe, it prevents proper isolation of the synthetic grafts. For this reason aorto graft-enteric fistula is a serious complication in patients with Behçet disease.[20] In our first report,[8] 2 patients (2 of 9) had died because of massive gastrointestinal bleeding

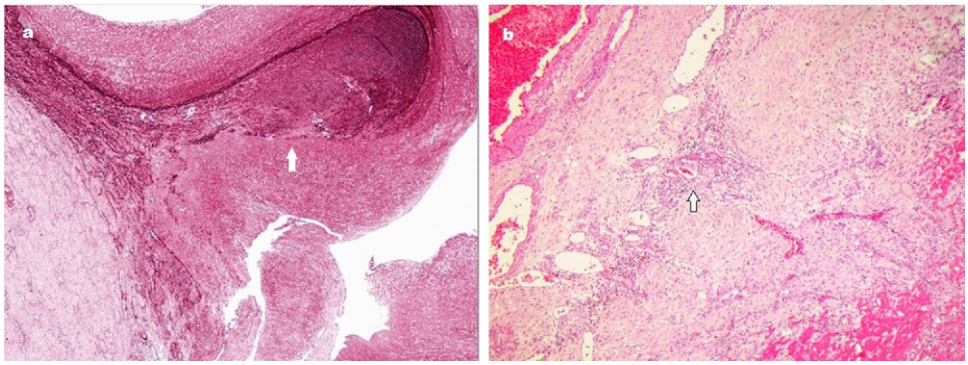


Fig 3. a, Histopathological image of popliteal artery aneurysm. Loss of internal elastic membrane (*arrow*) and loss of elastic and muscle fibers in the media can be seen along with periadventitial and intimal thickening (Elastic Van Gieson, ×100). **b,** Also infiltration of inflammatory cells around the vasa vasorum (*arrow*) is noted (hematoxylin and eosin stain, ×200).

due to aortoenteric fistula. We avoided this complication by wrapping an omental flap around the grafts.[8] We used this technique in the five aortic aneurysms we operated on and saw no gastrointestinal bleeding complication.

Synthetic patch closure of the defects has a high recurrence rate with suture dehiscence.[9] This complication occurred in 1 of our patients with a false aneurysm originating from a defect on the anterior wall of the aorta between the origins of celiac and mesenteric superior arteries. We had repaired the defect with a PTFE patch, but he had a suture dehiscence 6 months later. This patient died of exsanguination while waiting for an endograft.

In Behçet disease, the diseased arteries are also quite friable and we think that this feature might contribute to the high recurrence rate when a bypass procedure is performed. From our earlier experience, we had noticed that graft occlusion was well tolerated in patients with Behçet disease.[8] This time, of 10 PTFE grafts inserted for extremity aneurysms, four grafts occluded in <1 year. Apart from mild to moderate claudication, this complication was again well tolerated, therefore, no further attempt is made for occluded grafts. The same finding is reported by Kwon et al.[9] Also, Hosaka et al[21] reported 35.7% graft occlusion incidence in their patients. So, in our first report, we had suggested ligature in the first instant.[8] We had measured the sump pressures and if they were over 50 mm Hg we ligated the arteries.[8] We were able to ligate two superficial femoral, two popliteal, one posterior tibial arteries, and one iliofemoral venous conduit without any severe disabling ischemia in this series. We think that ongoing inflammation in Behçet disease induces abundant new collaterals. It is known that angiogenesis may develop as a compensatory response to ischemia and inflammatory stimulus and that Behçet disease is one of the few vasculitis that can cause angiogenesis.[22] Also, vascular endothelial growth factor which is a potent stimulator of angiogenesis is reported to be increased in Behçet disease.[23]

We recently had to ligate a common iliac artery for a recurrent saccular aneurysm 10 years after the last operation of iliofemoral venous conduit ligation in a patient in this series. This patient complained of short-distance claudication probably because of ligation at two levels. All other patients either with ligation or with occluded grafts experienced mild to moderate claudication. When the sump pressure is low or back bleeding is poor, we inserted PTFE grafts which are commonly used in our department. These days we never use vein grafts in Behçet disease. Because of the high frequency of venous involvement in Behçet disease, veins immediately degenerate and develop false aneurysms. In the current cohort, we ligated one iliofemoral venous conduit because of the degeneration of the vein graft inserted in another clinic.

With the introduction of endovascular grafts, reports started to appear about treating Behçet disease aneurysms with endovascular techniques.[9,24-27] Kwon et al[9] reported 40% recurrence rate for stent graft insertion in the infrarenal abdominal aorta. When the aneurysm is in the aortic bifurcation, which is the commonest site, there is a risk of new aneurysm formation in the proximal landing zone of the graft just distal to the take-off of the renal arteries.[9] Similar complication of proximal landing zone aneurysm has been reported by Nakai et al.[10] So with the endovascular treatment, the pathology gets even more complicated. Also, there is the risk of aneurysm formation at the femoral cut down sites.[18,19] Thus, none of our patients had a stent inserted.

Interestingly, four saccular aneurysms at the abdominal aorta in 2 patients were followed-up by medical therapy only. In 1 patient, there were three saccular aneurysms at the aorta (Fig 1). In another patient, two aneurysms were detected at the aorta. One was a symptomatic large bifurcation aneurysm which was operated with a PTFE graft. The other lesion was a saccular aneurysm of 1.5-cm diameter which was conservatively managed by immunosuppressive therapy. This aneurysm partly occluded with thrombus and decreased in size under immunosuppressive treatment during a 5-year follow-up. Likewise, 2 patients with carotid aneurysms were treated only by medical

means. One patient had a small saccular aneurysm at the bifurcation and the other had bilateral common carotid fusiform aneurysms (Fig 2). As we were planning an operation for the second patient, neck enlargement diminished with induction immunosuppressive therapy and an MRA showed reduction in the size of the fusiform aneurysm. Hence, we decided to follow the patient under medical therapy. During the follow-up period of 6 years, he had no problems with his carotids but had stasis ulcers on the lower extremities due to chronic DVT.

It is also worth noting here that the overall mortality among our 24 patients operated on before 1997 for large arterial disease was 17% at 4 years.[8] Although strict comparisons between the previous and the cohort might be subject to many biases, the fact that all the reported experience was from one center and the operating surgeon had been the first author in either instance makes us rather confident that we are doing better these days in managing these vascular complications of Behçet disease.

In conclusion, the surgical management of large, nonpulmonary, arterial diseases of Behçet disease is quite satisfactory. When the false aneurysm is in the infrarenal aorta, aorto-bi-iliac bypass is the preferred surgical intervention. For false aneurysms located between the visceral arteries, patching with synthetic material gives unfavorable results. Extremity aneurysms can be treated with synthetic graft insertion. In selected cases, ligation can give satisfactory results; however, postoperative claudication is common. In some patients with small intact saccular aneurysms, surgery may not be necessary. Patients must be prescribed immunosuppressive therapy with cyclophosphamide and corticosteroids before and after the surgical intervention in order to avoid Behçet disease activation.

AUTHOR CONTRIBUTIONS

Conception and design: HT, ES, VH
Analysis and interpretation: HT, ES, VH
Data collection: HT, ES, CA, VH, KB
Writing the article: HT, ES,
Critical revision of the article: HT, ES, VH, HY
Final approval of the article: HT, ES, CA, VH, KB, HY
Statistical analysis: ES
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Overall responsibility: HT, ES

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