Abdominal aortic aneurysm and Behçet’s disease

Alex G. Little, M.D., and Christopher K. Zarins, M.D., Chicago, Ill.

A case of abdominal aortic aneurysm with Behçet’s disease and a review of two similar cases previously reported are presented. This case and additional review of other reported major vascular complications of Behçet’s disease lead us to conclude that the pathogenesis of the abdominal aneurysm is related to involvement of the aortic wall or vasa vasorum by the Behçet’s disease process. Surgical treatment of this rare complication of Behçet’s disease should include extra-anatomic bypass if the possibility of infection cannot be definitely excluded.

From the Department of Surgery, The University of Chicago Hospitals and Clinics, Chicago, Ill

Based on the clinical triad of relapsing iridocyclitis and ulcers of the mouth and genitalia, Behçet, in 1937, defined a syndrome that has become identified with his name.1,2 It is now clear that Behçet’s disease is a systemic vasculitis that affects both veins and arteries. Small-vessel arteritis causing multiple organ system involvement is common and frequently affects skin and mucous membranes, joints, eyes, and the central nervous system.3,4,7 Clinical manifestations include ulcerations of the mucous membranes and genitalia, skin rashes, polyarthritis, neurologic deficits, and inflammatory ocular disorders. Other organ systems such as the gastrointestinal tract, pancreas, kidney, and heart are less commonly involved.8,13

Vascular manifestations of Behçet’s disease result from involvement of large vessels. Both phlebitis of peripheral veins and spontaneous thrombosis of deep veins have been reported.2,4,5 Clinical manifestations related to involvement of large arteries are less frequent, but patients with thrombosis and aneurysm formation have been reported.2,4,11 Abdominal aortic aneurysms have been observed in only four patients.3,6 Here we report an additional case of abdominal aortic aneurysm occurring in a patient with Behçet’s disease and review the English literature on this subject for the purpose of discussing pathogenesis and treatment of this unusual complication of Behçet’s disease.

CASE REPORT

C. M., a black man born in 1950, enjoyed good health as a child and adolescent. During 1971 he began to have headaches, neck stiffness, and throat soreness with intraoral aphthae and lost weight as a result of anorexia. In 1972 he developed abdominal pain and at another hospital was found to have a right lower quadrant mass. At surgery an inflammatory mass involving the cecum and terminal ileum was resected, and an ileo-ascending colostomy was constructed. Reoperation was required for evacuation of an abscess in the right paracolic gutter. A mushroom-shaped catheter was placed to provide drainage and was removed 11 days later. Gross examination of material from the initial operation revealed three small perforations of the terminal ileum and a 2 cm perforation of the cecum. Microscopic examination showed both nonspecific diffuse acute inflammation and distinctly focal areas of vasculitis involving arterioles (Fig. 1).

Following discharge, he continued to be anorectic and never really felt well. For 3 years he had intermittent purulent drainage from the drain site. In 1975 the headaches and neck stiffness recurred, and he began to have shaking chills when exposed to coldness. During the next year he developed stiffness in his left knee and noted shallow ulcers of his scrotum and penis as well as a pustular, nonpruritic rash over his trunk and limbs. In 1976 he began to have nocturnal fevers and chills.

In 1977 low-back pain began following a day of lifting heavy boxes, and the patient was referred to our institution. On physical examination, no neurologic deficits were found,
but a pulsatile mass was detected in the right epigastric area. Fully healed incisions were present on the abdominal wall, and there were no areas of purulent drainage. At that time he was afebrile with a pulse rate of 88 beats/min and a blood pressure of 110/70 mm Hg. Aortography demonstrated a false aneurysm posterior and to the right of the infrarenal aorta (Fig. 2). With the presumptive diagnosis of a mycotic aneurysm, blood cultures were drawn and antibiotic therapy was begun. At surgery, an 8 cm mass was found surrounding the aorta and involved the bifurcation. Proximal and distal control was obtained, and, following heparinization, the infrarenal aorta and common iliac arteries were transected and oversewn with 4-0 wire. The aneurysm was opened, and it was found that the back wall of the aorta was entirely eroded, because the vena cava and vertebral bodies were plainly visible. The aneurysm wall was soft and friable, but there was no purulent material present, not was there an old drain tract. Specimens of the wall were obtained for aerobic and anaerobic cultures, and Penrose drains were placed in the aneurysm site and brought out through the abdominal wall. An axillary bifemoral bypass using an 8 mm Gore-Tex graft was then performed.

All blood and intraoperative cultures were sterile, and no organisms were seen on Gram-stained smears of the aneurysm wall. Histologic examination of the false aneurysm wall showed only necrotic tissue. The patient progressed to a regular diet, and antibiotics were stopped on the fifth day. There was minimal drainage from the Penrose drains, and they were accordingly advanced and withdrawn. A small-bowel obstruction that failed to resolve with nasogastric tube decompression necessitated reexploration on the tenth day. Adhesions were lysed, and although no obvious infectious process was present, cultures were taken, and a red rubber catheter was placed in the site of the aneurysm. Postoperatively the patient made good progress and was soon tolerating a regular diet. Again all intraoperative cultures were reported as negative. The patient was able to walk without difficulty and had pedal pulses bilaterally; ankle/arm systolic pressure ratios were 0.8 on the right side and 0.7 on the left. Peripheral vein indwelling catheters had to be changed with unusual frequency because of erythema and tenderness.

After discharge, the patient's appetite returned, and he gained 15 kg. The drainage catheter was advanced and removed 2 months after surgery. During a clinic visit 1 year after operation, his auxiliary bifemoral graft was noted to be clotted. He denies claudication, jogs, and has normal sexual function. In 1979 he developed an iliolumbar deep vein thrombosis and required hospitalization for treatment with heparin. He is presently taking warfarin sodium (Coumadin) and wears support stockings. It is now 9 years since his operation for spontaneous perforation of the iliopectineal area and 4 years since the aortic exclusion and bypass procedure. Since the episode of deep vein thrombosis, the patient has had no major problems. He did have a single, spontaneously appearing pustular lesion on one leg, which required incision and drainage. Histologic examination of the wall of the cavity showed an obliterative type of arteriitis.

**DISCUSSION**

This patient had a history of oral and genital ulcerations—two thirds of Behget's original triad. The further history of skin rashes, episodes of superficial thrombophlebitis in response to indwelling catheters, and occurrence of deep vein thrombosis is typical.
The spontaneous perforations of the ileum and cecum are related to the ulcerative enteritis reported to occur in these patients.\textsuperscript{9,10} He has no indications of central nervous system or eye involvement at this time.

The vasculitis of Beh\c{c}et's disease has the histologic features of endothelial cell swelling, fibrinoid necrosis, and leukocytic perivascular infiltration.\textsuperscript{2,7} As seen in Fig. 1, these features are present in the mesenteric arterioles and also in the specimen from the pustular leg lesion. Involvement of arterioles and venules causes the typical manifestations of a systemic vasculitis. In contrast to other types of vasculitis, however, large vein involvement is common and leads to deep vein thrombosis both centrally and peripherally.\textsuperscript{2,4,5,7} Our patient has had problems with phlebitis of superficial veins and thrombosis of his deep iliofemoral venous system.

Involvement of large arteries has been clearly demonstrated but infrequently reported. Enoch et al.\textsuperscript{4} reported the development of popliteal artery aneurysms in a 16-year-old boy who had been treated with both adrenocorticotropic and oxyphenbutazone. The patient developed ischemia of the left leg, and arteriography revealed a normal aorta and iliac vessels and a left popliteal artery aneurysm. At surgery it was determined that a true aneurysm had ruptured, and a large false aneurysm was excised and replaced with autogenous saphenous vein. Histologic studies were not carried out. During recovery it was noted that the right dorsalis pedis pulse was diminished, and arteriography demonstrated a right popliteal aneurysm. When symptoms developed 3 months later, a saccular aneurysm was excised and replaced with saphenous vein. Histologic studies showed an obliteratorive endarteritis and complete destruction of the arterial wall. At the first presentation a mycotic aneurysm was suspected, presumably because of the history of steroid therapy; however, all blood cultures were negative.

Mounsey described a 24-year-old man with Beh\c{c}et's disease who had been treated with corticosteroids.\textsuperscript{3} The patient developed low-back pain after slipping from a curb, but lumbar spine x-ray films were normal. Three
months later he developed acute abdominal pain, loss of sensation over several lumbar dermatomes on the right side, and an absent right knee jerk, and a pulsatile abdominal mass was discovered. Lumbar spine x-ray films demonstrated erosion of the right side of the body at L4. Surgery was performed, and a large aneurysm was found, resected, and replaced with a Terylene graft. No objective evidence of infection was found, but osteomyelitis of the vertebra was suspected because of the spinal erosion. Twelve days after surgery the patient developed a fecal fistula followed by development of an abscess in the right lower quadrant. Three weeks following this event the patient died of upper gastrointestinal bleeding, and postmortem examination demonstrated an aortoduodenal fistula. Microscopic examination of material from the operation was said to reveal an "old septic osteomyelitis with a polymorphonuclear exudate." In fact, however, no organisms were cultured from the bloodstream or aortic tissue either during or after operation.

Finally, Hills mentioned the description of two aortic aneurysms and three aneurysms of "small arteries" in the Japanese literature. She then reported a case of a young man with Behçet's disease who, during examination of back pain, was found to have an abdominal aortic aneurysm. At surgery the aorta appeared thickened and inflamed, but "no source of infection could be found," and aortoiliac bypass was carried out uneventfully. The aorta was not biopsied, so there was no histologic investigation. The patient had an uneventful course and was doing well at the time of most recent follow-up.

The common thread in these case reports, ours included, is the concern for possible infection. In no instance, however, was infection documented either by Gram stain or culture. Therefore, it seems likely that these arterial aneurysms result from involvement of the arterial wall in the primary Behçet's disease process. The presumed pathogenesis is involvement of the vasa vasorum of the aortic wall leading to dilatation with true aneurysm formation or perforation with false aneurysm formation. That this is the actual sequence of events is suggested by failure to confirm infection as the causative mechanism and by the finding of an obliterative endarteritis of the vasa vasorum in the specimen of popliteal artery aneurysm of Enoch's patient. Supportive data from the patients with abdominal aneurysms are not available: one patient was not biopsied; one was studied after dying of an aortoduodenal fistula, and examination could only show acute inflammation; and our patient's specimen consisted only of a chronic false aneurysm wall.

Many of these patients did poorly because attention was directed to their aneurysm too late. We urge physicians involved in the care of patients with Behçet's disease to be on the alert for signs or symptoms referable to the aorta or its major branches. Any patient with back pain should be suspected of having an aortic aneurysm and should be evaluated and treated accordingly. If an aneurysm is found and surgery performed in a situation where the possibility of infection cannot be discounted, extra-anatomic bypass is the procedure of choice. Subsequent reconstruction with aortofemoral bypass could be performed on an elective basis, although our patient is doing well and there is no reason to advise a further procedure. Of interest is the fact that our patient has had asymptomatic thrombosis of the axillary bifemoral graft despite having a ligated aorta. Discovery of the thrombosis was an incidental finding in a man who is working steadily, jogging daily, and having normal sexual function. This demonstrates the potential, in the younger patient, for development of arterial collaterals sufficient to supply both the pelvis and the lower extremities.

REFERENCES