A CASE OF VASCULO-BEHCET'S SYNDROME

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ABSTRACT

A case of Behcet's syndrome with vascular complications is presented. This case had a
familial occurrence of Behcet's syndrome. His vascular complications were deep vein thrombo-
phlebitis of both legs, aneurysms of the left common iliac artery and the left femoral artery
and the occlusion of the left subclavian artery. He was successfully operated on for the
aneurysms.

This paper discusses the problems accompanying the aneurysm of Behcet's syndrome. The
authors pointed out that the subclavian steal syndrome may be diagnosed as neuro-Behcet's
syndrome and stressed the importance of vascular survey in Behcet's syndrome when the patient
complaints of thrombophlebitis.

INTRODUCTION

Behcet's syndrome is not very rare in this country, but the arterial involvements
in Behcet's syndrome such as aneurysm and arterial occlusion have recently drawn
attention. Aneurysm is one of the life-threatening complications of Behcet's
syndrome and the operation on this presents special problems.

CASE REPORT

First admission: A 46 year-old Japanese man, who had had recurrent oral aphtha
and ulcers in the scrotum since he was 17 years old, was admitted to our hospital
on August 2 in 1975 because of pain and swelling in both legs. About one and half
year before the admission he was attacked by speech disorder and ataxia of the right
side but recovered completely. His 18-year-old son was suffering recurrent oral

aphtha and ulcer in the scrotum. At the
time of admission the diagnosis was vas-
culo-Behcet's syndrome. His erythrocyte
sedimentation rate was 144 mm/hour. The
serum test for syphilis was negative. He
was treated conservatively and discharged.

Second admission: On September 20 in
1975, he was admitted because of the recur-
rence of thrombophlebitis of the legs.

The physical examination revealed a
pulsating tumor in the left inguinal area.
The arteriography showed aneurysms of
the left common iliac artery and left fem-
oral artery (Fig. 1).

He was hypertensive but the renal ar-
terys were normal.

The deep veins of both legs were throm-
boxed (Fig. 2).

The aneurysms were stationary and the
thrombophlebitis subsided. We decided to
follow-up him as an outpatient.

Third admission: Two month after the
second discharge he was admitted again because of an acute increase in the size of the femoral aneurysm and severe pain in that area. The next day, an operation was performed. The femoral aneurysm was resected and the iliac aneurysm was ligated. A dacron tube (8 mm) was grafted from the iliac artery to the femoral artery (Fig. 3). Histological examination of the resected specimen showed thickening of the intima, disappearance of the elastic fiber and abrupt rupture of the whole arterial wall. A cellular infiltration was seen in the media and adventitia (Fig. 4). One month after the operation, the graft was patent (Fig. 5), but the arteriography revealed that the left subclavian artery was occluded and the left brachial artery was fed by the distended left vertebral artery (Fig. 6). The blood pressure of the right arm was 172/94 mmHg and the left arm was 162/102 mmHg. He was discharged on the 40th post-operative day and the graft was patent three months after the operation.

*Fourth admission:* Four months after the operation, he was admitted again because of the occlusion of the graft. The angiography revealed rich collateral vessels anastomosing mainly from the left 4th lumbar artery to the deep femoral artery (Fig. 7). Operation was not performed and the patient was discharged.

**DISCUSSION**

The diagnostic criteria of Behcet’s syndrome used were those set up by Behcet’s Syndrome Research Committee of Japan (Shimizu). Our case fulfilled the major
three criteria; recurrent aphthous ulcerations in the mouth, skin lesions (subcutaneous thrombophlebitis of the legs) and genital ulcerations. The vascular complications in Behcet’s syndrome have been known as thrombophlebitis (Nishiyama and Urayama).

Mishima first reported on the resected abdominal aortic aneurysm in 1961. Urayama called Behcet’s syndrome combined with venous and/or arterial complications as angio-Behcet’s syndrome and Shimizu categorized them as vasculo-Behcet’s syndrome. Aneurysm in Behcet’s syndrome is frequently multiple and apt to rupture (Mishima; Sasaki; Tzubota; and Masuoka), it recurs frequently (Sasaki; Mishima; Masuoka), and its prognosis is rather poor (Tanaka; Mishima; Fukuda; and Clinical Conference). Mishima presented six cases with arterial complications, of which two cases had thrombophlebitis. Tanaka, Sasaki, and Fukuda presented cases with arterial complications but without thrombophlebitis. But recently many authors (Iwabuchi; Urayama; Forman; Hils; Tzubota; and Masuoka) presented cases with both arterial and venous complications.

In our case the aneurysm of the left iliac artery and the occlusion of the left sub-
clavian artery were revealed by arteriography.

So we concluded that Behcet's syndrome with thrombophlebitis requires examination also of the arterial system. In the presented case, the left subclavian artery was occluded and a subclavian steal phenomenon was revealed, and we attributed his central nervous symptoms to the subclavian arterial occlusion, because the side of the brain affected was the left side, the symptoms were mild and temporary and the patient

Fig. 3. After the graft was planted, the femoral aneurysm (7cm×5cm×5cm) was resected.

Fig. 4. The arterial wall is ruptured (arrow). The intima is thickened and thrombosed. Media is necrosed. Cellular infiltration of media and adventitia. H.E. (×10).
recovered completely. Without arteriography we may diagnose him as neuro-vasculo-Behcet’s syndrome. Reivich stated that the subclavian arterial occlusion causes a decrease of 41% of the intracranial arterial flow. Arterial occlusion does not require an operation if the collateral pathway is established. In this case one of his two sons was affected by oral aphtha and scrotal ulcer but his wife is healthy. Mason and Barnes reported four familial cases in 33 cases. Takano presented 12 cases in six families out of 40 cases of Behcet’s syndrome. Sezer found the disease in three brothers and showed it as the evidence of communicable disease. But Shimizu stressed that Behcet’s syndrome does not affect the spouse of the patient.

In Behcet’s syndrome the familial tendency may be one of the clues to the pathogenesis of the syndrome.

REFERENCES

5) Shimizu, T., Yamamoto, K., and Matsumura, N.: On the vascular changes observed in the cases with Behcet’s disease. Miyakkan-gaku, 6:


Fig. 6. The left subclavian artery is occluded and the left brachial artery is fed by the left dilated vertebral artery.

Fig. 7. Selective lumbar arteriography the rich collateral vessels feed the deep femoral artery.