Case Report

A Case of Behçet's Disease with a Rapidly Enlarging Aneurysm in the Common Carotid Artery

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A 16-year-old boy had incomplete Behçet's disease with common carotid artery aneurysm and an ulcer in the transverse colon. To our knowledge, this complication in the carotid artery has been previously reported in only 6 cases. Arterial repair and high-dose corticosteroid therapy (pulse therapy) were successful.

Key words: Corticosteroid therapy

Behçet's disease is an inflammatory disorder of unknown etiology characterized by recurrent oral and genital aphthous ulcers, ocular inflammation, and skin lesions. It also frequently involves the central nervous system, the vascular system, and the gastrointestinal tract. In the vascular system 7–37% of patients with Behçet's disease have been reported to suffer at some time from vascular lesions (1–3). Though vascular involvement can include both arteries and veins, the arterial system is rarely affected. In this paper we present an extremely rare case of Behçet's disease which was complicated by the spontaneous development of a left common carotid artery aneurysm and an ulcer in the transverse colon.

CASE REPORT

A 16-year-old boy was admitted to our department because of a mass in the left lateral cervical area and headache. One month prior to the admission he noticed the mass. He visited the outpatient clinic because of the complaints on June 20, 1988. Then the surface of the mass was smooth and painless. Laboratory data from the outpatient clinic were as follows: Erythrocyte sedimentation rate 3 mm/h, white blood cell count 8,300, CRP 1.5+.

The patient was empirically given antibiotics with no improvement in the complaints or laboratory findings. As the mass gradually enlarged, he was admitted on July 6, 1988.

Physical examination data on admission were as follows: his ht was 160 cm, and wt 43 kg. Blood pressure was 120/60 mmHg in the right brachial artery and pulse was 72/min and regular. Body temperature was 36.3°C. On the buccal mucosa there was aphthous stomatitis and the lingual mucosa was covered with yellowish patches. In the left lateral cervical area there was a smooth surface mass, approximately 5×7 cm, and systolic bruit was audible (Fig. 1). Multiple acne covered the face and trunk.

Laboratory data on admission are shown in Table 1: Erythrocyte sedimentation rate was 48 mm/h. The hematocrit was 44.8% and the white blood cell count was 12,600, with 51% neutrophils, 3% band forms, and 37% lymphocytes. CRP was 4+. Serological tests for syphilis and antinuclear factor were negative. Fibrinogen was 583 mg/dl. Ultrasound examination showed a hypoechoic and pulsating mass.

Digital subtraction angiography of left common carotid artery (Fig. 2) disclosed a huge aneurysm just

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Fig. 1. The mass in the left cervical area.

proximal to the bifurcation and the internal carotid artery was not clear.

On the second hospital day he noticed hoarseness and paralysis of the recurrent nerve. On the 8th hospital day there was left ptosis and the left pupil

Fig. 2. A digital subtraction arteriogram shows the huge aneurysm of the left common carotid artery. Note complete absence of blood flow to the internal carotid artery.

Table 1. Laboratory findings on admission

ESR (mm/h)	48	ALP (IU/l)	412
Urinalysis		T.Ch. (mg/dl)	91
protein	(-)	BUN (mg/dl)	6
sugar	(-)	Cr. (mg/dl)	0.4
occult blood	(-)	UA (mg/dl)	4.4
Stool		Na (mEq/l)	138
occult blood	(-)	K (mEq/l)	4.2
Peripheral blood		Cl (mEq/l)	99
WBC (/mm ³)	12,600	FBS (mg/dl)	94
RBC ($\times 10^4$ /mm ³)	508	IgA (mg/dl)	220
Hb (g/dl)	15.2	IgG (mg/dl)	1,340
Ht (%)	44.8	IgM (mg/dl)	232
Plt ($\times 10^4$ /mm ³)	33.0	C ₃ (mg/dl)	120
Coagulation test		C ₄ (mg/dl)	23
APTT (s)	30.8	Serological test	
PT (s)	11.7	CRP	(4+)
Fibrinogen (mg/dl)	583	RA test	(-)
FDP (μ g/ml)	<10	LE test	(-)
Blood chemistry		Anti-nuclear	
T.P. (g/dl)	7.2	antibody	(-)
Alb (%)	61.4	STS	(-)
α_1 -gl (%)	4.6	HLA	
α_2 -gl (%)	12.7	A2 A24, Bw52 B39	
β -gl (%)	8.1	Cw7, DR2 DR4, DRw53	
γ -gl (%)	16.0	DQw1, DQw3	
T.Bil. (mg/dl)	0.7		
GOT (IU/l)	14		
GPT (IU/l)	7		
LDH (IU/l)	318		

was mitotic. As the aneurysm progressively enlarged, his complaints were aggravated. In addition, the follow-up laboratory data became worse; the erythrocyte sedimentation rate was 64 mm/h, the white blood cell count 21,000 and CRP 6+. Because rupture of the aneurysm was quite likely, the patient underwent an emergency operation. The aneurysm was excised and the area was bypassed with Gore-Tex (W.L. Gore & Associates, Maryland, USA). At the time of catheterization to void, a penile ulcer was incidentally discovered. Ophthalmological examination after the operation did not show any abnormal findings. A prick test was negative. A clinical diagnosis of incomplete Behçet's disease was made from the diagnostic criteria (4).

Histological studies (Fig. 3) of the resected aneurysm showed thickening of the adventitia, destruction of the elastic fibers of the media and an active inflammation with an infiltration of lymphocytes and polymorph leucocytes. There was no granuloma or giant cell formation and no foreign material, bacterial or fungi could be demonstrated.

Postoperatively (Fig. 4) the inflammatory reaction became progressively worse and the suture did not hold. On the fifteenth postoperative day, bloody stool developed abruptly and complication of the digestive tract was strongly suspected. The patient was given 1,000 mg of methylprednisolone intravenously. The bloody stool stopped the next day. Colonoscopic examination showed a shallow ulcer in the transverse colon without any lesions in the cecum. After the corticosteroid administration the wound healed and there was a striking clinical improvement. Subsequently, the steroid treatment was tapered off.

DISCUSSION

This is an extremely rare case of Behçet's disease with a rapidly enlarged common carotid artery aneurysm and an ulcer in the transverse colon. The diagnosis of Behçet's disease was made on the basis of the criteria determined by the Behçet's Disease Research Committee of Japan in 1987 (4). As major criteria, the present case had recurrent aphthous stomatitis, a genital ulcer and he had noticeable erythema nodosum-like eruptions of the leg, as minor criteria, he had vascular involvement. These is a strong association of HLA-B51 with Behçet's

Fig. 3. Microphotograph of the aneurysm wall in the resected common carotid artery. Note the thickening of the adventitia and marked splitting of elastic fibers of media (hematoxylin and eosin, original magnification, $\times 40$).

Fig. 4. Clinical course.

disease in Japan (5), but our patient was negative for this particular locus.

Arterial involvement in Behçet's disease is characterized by saccular aneurysm or occlusion of large vessels in young men. The most common site of aneurysm is the abdominal aorta or femoral artery (6, 7). Only 6 cases in which the common carotid artery was affected have been recorded in the literature (6, 8–11). Although some cases of multiple aneurysm have been reported, the arteriography did not reveal another aneurysm in the present case.

On microscopic examination of the aneurysm, there was fragmentation and splitting of the medial elastic fibrosis and degeneration of the vasa vasorum

(6, 12). These findings were identified in our case. Whether the pathogenesis of arterial aneurysm is obliterative endarteritis of the vaso vasorum or primary inflammation in media is unclear (7, 8, 12–14).

Corticosteroids, immunosuppressive agents and other drugs have been used for the treatment of Behçet's disease, but there is no curative therapy. Highdoses of corticosteroid (pulse therapy) that may be administered in cases when general conditions are poor during the acute stage or operations give rise to excessive reaction due to irritability (4). In the present case, 1,000 mg/day of methylprednisolone was administered for 3 days because the suture did not hold after the operation and there was melena. Corticosteroid therapy was very effective in accelerating wound healing and in improving the patient's general condition.

Although the overall mortality rate for Behçet's disease is 3 to 4%, the leading cause of death is rupture of large artery aneurysms (6). In general, vascular involvement appears several years after the onset of dermal, mucosal and ophthalmic symptom (6). In some cases of incomplete Behçet's disease, where arterial involvement is occasionally found, diagnosis may be delayed because the characteristic symptoms are not present or associated symptoms rapidly aggravate (12, 15). It is important to consider the possible complication of aneurysm in young males with Behçet's disease.

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