

Behcet's Disease with Involvement of Major Arteries: A Case Report

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Abstract- Vascular involvement is not infrequent in Behcet's disease (BD). It is generally seen in the form of superficial thrombophlebitis or occlusion of major veins. In rare instances, arterial occlusion and aneurysm formation may be seen in BD. We reported a young male with BD, diagnosed at the age of twenty for relapsing and remitting oral ulceration, skin rash, arthralgia and ocular painful redness for three years. At the age of 21, he had recurrent abdominal aortic aneurysm and inconspicuous neurological manifestations including dizziness, tinnitus and transients of blurred vision. The carotid angiography disclosed the occlusion of bilateral common carotid arteries (CCA). A carotid endarterectomy was subsequently performed to reduce the risk of stroke. The pathological examination of the occluded segment of CCA revealed chronic inflammation, which was attributable to BD. There was no atherosclerotic change. To the best of our knowledge, this is the first case report of concurrent bilateral CCA occlusion and relapsing abdominal aortic aneurysm. Even in the absence of specific neurological symptoms, we suggest that cerebrovascular investigation need to take into consideration in BD patients with unexplained cranial symptoms.

Key Words: Behcet's disease, Carotid occlusion, Aortic aneurysm

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INTRODUCTION

Behcet's disease (BD) is a multi-system inflammatory disorder with unknown etiology. It is characterized by recurrent oral and genital ulcerations, uveitis and skin lesions predominantly affecting young adult males. The prevalence of vascular complications in BD ranges from 7% to 38%⁽¹⁾. Among vascular involvement, all types and sizes of blood vessels could be the targets. Nevertheless, arteries are less frequently affected than

veins⁽¹⁾. Moreover, occlusive lesions of carotid arteries are very unusual in BD. Concurrent involvement of carotid arteries and abdominal aorta has not been reported before. Herein, we describe a BD patient with relapsing aneurysms of abdominal aorta and occlusive lesions in bilateral common carotid arteries.

CASE REPORT

At the age of 20, this young man with 3-year history

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of recurrent polyarthralgia, ocular painful redness, oral ulceration, and skin folliculitis was diagnosed as having BD. At the age of 21, due to progressive enlargement of a pulsating abdominal mass, he was admitted to our hospital for comprehensive work-up of the underlying etiologies in September, 2000. He also reported mild dizziness, tinnitus and transients of blurred vision.

On admission, he was alert, fully oriented and afebrile. His vital signs were stable. There was no hepatosplenomegaly nor palpable lymph nodes. The neurological examination revealed intact mentality and cranial nerves. The muscle strength was normal, and the plantar response was the flexor type bilaterally. No cerebellar dysfunction or gait disturbance was detected. However, the physical examination was notable for a pulsating abdominal mass and an area of nodular erythema over right lower leg. A subtle bruit was noticed on auscultation of the left carotid area. The right carotid pulsation was absent. The ophthalmological examination

revealed bilateral uveitis. Except for heavy cigarette smoking, there was neither family history nor traditional cardiovascular risks identified.

Results of routine laboratory tests, including hemogram, biochemistry and lipid profiles were normal except for a high erythrocyte sedimentation rate (ESR: 54 mm/hour) and C-reactive protein (64.8 mg/dL). The level of rheumatoid factor, protein C and S, antithrombin III, homocystein, immunoglobulin, C3 and C4 complement were all within normal limits. Serum antinuclear and anticardiolipin antibodies were negative. A skin biopsy was performed, and perivascular inflammation with lymphocytes predominance was found pathologically.

A computed tomography (CT) of the abdomen demonstrated a huge aneurysm extending from the infrarenal portion of the aorta to the right common iliac artery (CIA) with intraluminal thrombus formation. A carotid duplex examination disclosed high-grade steno-

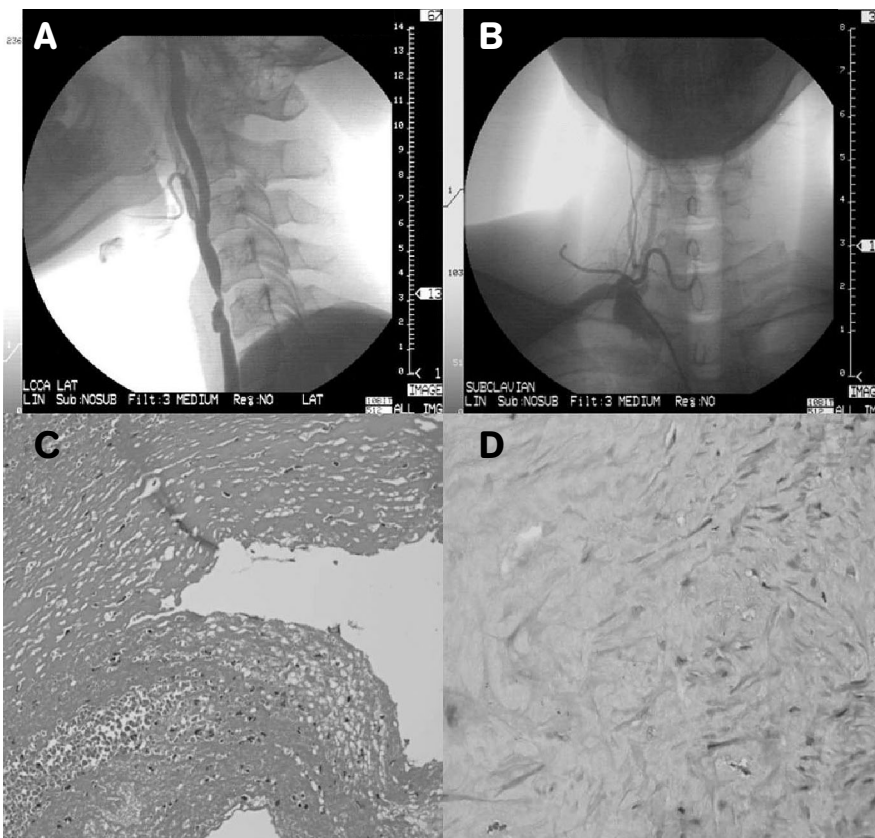


Figure. Cerebral angiography demonstrated: (A) high-grade segmental stenosis of the left common carotid artery (CCA), and (B) complete occlusion of the right CCA with collateral circulation from the right vertebral artery.

Histopathological findings revealed neutrophil-predominant inflammatory infiltration with a focal destructive lesion on the wall of abdominal aorta (C), and myxoid degeneration in the thickened wall of the left carotid artery (D). (H&E stain; original magnification, 40X).

sis of bilateral common carotid arteries (CCAs). The cerebral angiography demonstrated a complete occlusion of the right CCA with collateral circulation from the right vertebral artery, and an up to 90%-degree eccentric stenosis of the left CCA (Figs. A-B). The abdominal aortic aneurysm was resected successfully and a Dacron graft was interposed for reconstruction on September 25, 2000. The patient recovered well postoperatively and was discharged with aspirin (100 mg/day), prednisolone (1 mg/kg/day), colchicines (1 mg/day) and azathioprine (50 mg twice per day).

Eight months later, progressive and predominantly right-sided intermittent claudication developed. Diminished peripheral pulsation and hypothermia on the feet was found on physical examinations. A follow-up abdominal CT scan revealed moderate thrombotic occlusion of the right CIA and pseudoaneurysm formation at the site of the graft anastomosis with the infrarenal abdominal aorta. A digital subtraction angiography showed distal occlusion of the right external iliac artery without demonstration of bilateral superficial femoral arteries. Intravenous prostaglandin EPG1 (PGE1, Alprostadi) was given and anticoagulant was added for prevention of the post-operative arterial thrombosis.

During the following three years, the patient received reparatory surgeries for anastomotic pseudoaneurysms of the abdominal aorta and recurrent aneurysms involving bilateral CIAs, and also for spontaneous rupture of a developing aneurysm of the left CIA. During the period, a Tc-99m HMPAO brain perfusion SPECT demonstrated decrease radioactivity in bilateral occipital lobes, the basal portion of the temporal lobes and the left basal ganglion. These findings were indicative of diffuse ischemic changes involving the whole cerebral parenchyma. Without any new neurological deficit, the patient subsequently received endarterectomy of the left CCA. The histopathological findings were myxoid degeneration with chronic inflammatory infiltrates of the vascular wall (Figs. C-D).

However, a spontaneous rupture of the abdominal aortic aneurysm recurred and was complicated by massive internal bleeding nine months later. Although surgical repair was performed smoothly, the patient finally

expired from septic shock two months after the operation.

DISCUSSION

BD is a chronic, relapsing inflammatory disease of blood vessels. Typical clinical manifestations are oral thrush and genital aphthae. Systemic symptoms, including ocular and skin lesions, neurological disease or arthritis may also occur. According to the diagnostic criteria proposed by the International Study Group⁽²⁾, our patient who had recurrent episodes of oral ulcers, uveitis, polyarthralgia, skin lesions with folliculitis and erythema nodosum, was clearly a BD patient.

Among the vascular involvement of BD, superficial thrombophlebitis and deep venous thrombosis are considered as typical manifestations⁽³⁾. A native series in southern Taiwan disclosed less than 20% of BD patients had thrombophlebitis, which suggests geographic and genetic differences of the clinical characteristics⁽⁴⁾. Although the arterial involvement is less than 5% in the course of BD, it is the major prognostic factor of morbidity. Among arterial lesions, peripheral arteries are more commonly affected⁽⁵⁾, while large artery involvement, including occlusion and aneurysm, is uncommon but potentially life-threatening. For instance, a rupture of abdominal aortic aneurysm is one of the most severe condition⁽⁶⁾.

Occlusive lesions in the bilateral carotid arteries as seen in our patient are extremely rare, and only two cases have been reported^(7,8) (summarized in Table). In both cases, the intervals between the onset of BD and that of arterial manifestation were more than 10 years, and the definitive diagnosis of BD was made at the time of ischemic strokes. No concurrent arterial lesions elsewhere were found. Similar to previous reported cases, our patient did not have hypertension, dyslipidemia or other risk factors of atherosclerosis. Thus CCA occlusions were considered to be secondary to BD with the vascular involvement. Furthermore, we propose that the mild and non-specific neurological symptoms in the disease course were attributable to the gradual development of CCA occlusions as well as marginally adequate collat-

Table. Reported cases of Behcet's disease (BD) with carotid artery occlusion

Reference	Age/Gender	Neurological symptoms	Duration of BD before the initial arterial manifestations	Other major arterial involvement	Treatment	Clinical outcome
(7)	41/male	Blurred vision, acute confusion, right hemiplegia	18 years	(-)	Medical management	Good
(8)	43/male	Headache, right hemiparesis, gait and speech disturbance	8-10 years	(-)	Medical management	Good
The current report	21/male	Dizziness, transient blurred vision, unilateral tinnitus	3-4 years	Abdominal aortic aneurysms	Endarterectomy of the carotid occlusion; resection and repair of aortic aneurysms	Expired

eral circulation from the vertebrobasilar system.

Vasculitis is increasingly recognized as the typical pathogenesis in BD⁽⁵⁾. Biopsies confirm the presence of perivascular inflammation near the lesions of BD, including oral and genital ulcers, erythema nodosum, posterior uveitis, epididymitis, enteritis, and lesions in the central nervous system⁽⁹⁾. Large vessels are affected by a vasculitis of the vasa vasorum, which is responsible for aneurysm or pseudoaneurysm formation^(5,6). The histopathologic features of large arteries in BD have been scarcely documented because of an extremely low incidence of approximately 1.5-2.2%⁽¹⁰⁾. A comparative study had disclosed characteristic pathological findings of aortic aneurysms in BD, the neutrophil-predominant infiltration in the tunica medial and tunica adventitial with preserved internal elastic lamina during active stage, were the differences from Takayasu's arteritis and other inflammatory aneurysms⁽¹⁰⁾. Vascular occlusion associated with BD can also be attributed to vasculitis with intimal thickening and endothelial dysfunction. However, pathological findings of large vessel occlu-

sions in BD have not been reported previously. In our patient, the histopathological examination from the specimen of the endarterectomy of the left CCA revealed focal myxoid and hyalinized degeneration of vascular walls, without atherosclerotic changes. Vascular occlusion was also demonstrated by extensive adherent thrombus formation, without clinical evidence of thromboemboli.

In conclusion, BD should be considered as a possible etiology of carotid artery occlusions. Arterial occlusions or stenoses may be asymptomatic or associated with ischemic symptoms, depending on the adequacy of the collateral circulation. We suggest that cerebrovascular investigation should be considered in BD patients with unexplained cranial symptoms.

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