Cerebral Venous Thrombosis in Behçet's Disease

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A 20-year-old Turkish patient presented with superior sagittal and lateral sinus thrombosis. The frequency of cerebral venous thrombosis generally is underestimated and may be the first clinical manifestation. In case of intracranial hypertension, cerebral venous thrombosis and Behçet's disease should be taken into consideration, especially in the high risk population from Eastern Mediterranean countries and Japan. CT with enhancement and MRI study with T1- and T2-weighted images were used to detect the thrombosed veins and to evaluate the thrombosis. Treatment with anticoagulation and corticosteroid therapy in cerebral venous thrombosis is well established today. Clinical course and prognosis in cerebral venous thrombosis are generally benign if adequate management is started in time.

(Key Words: Neuro-Behçet's syndrome, Cerebral venous thrombosis, Magnetic resonance imaging)

INTRODUCTION

Behçet's disease is a clinical syndrome of unknown etiology, originally described in 1937, with the classic triad of iritis and oral and genital ulceration (1). Since then, advances in immunology and anatomic pathology, as well as numerous reports of other manifestations, indicate that Behçet's disease is a systemic vasculitis affecting the skin, joints, eyes, gastrointestinal tract, neural tissues and blood vessels (2, 3). Neurologic manifestations of Behçet's disease (neuro-Behçet) have been reported in 10-25% of patients. These are nonspecific and include loss of vision, diplopia, nystagmus, cranial nerve palsies, speech disorder cerebellar signs, and cerebral and spinal sensory and motor disturbances. In general, three patterns of neurologic manifestations have been observed: a brainstem syndrome, a meningoencephalitic syndrome, and in organic confusion syndrome (4). The cerebral venous thrombosis manifestations as intracranial hypertension in Behçet's disease may be the initial manifestation (5-7). We report a case of Behçet's syndrome with intracranial venous thrombosis.

CASE REPORT

A 20-year-old Turkish man was admitted to our clinic August 14, 1991, because of headache, nausea, vomiting, and diplopia. He had a history of recurrent stomatitis and genital ulcer the past seven years. On January 16, 1991, he had suffered from recurrent arthritis and iridocyclitis. On April, 1991, Behçet's syndrome diagnosed. Physical examination revealed oral aphthae, genital ulcer, and folliculitis on the neck and back. Patergy was positive. Ophthalmological examination showed iridocyclitis in the right eye. The pupil was not reactive to light. Neurologic examination revealed lateral gaze palsy in the right eye and bilateral papilledema. The right side deep tendon reflexes were more brisk than on the left side.

Laboratory data. ESR was 69 mm/hr. The platelet and coagulation test (hematocrit, platelet count, spontaneous platelet aggregation, venous stasis and fibrinogen level) were normal. C-reactive protein was +3. Antinuclear antibodies, rheumatoid factor and the tests for lupus erythematosus cells were negative. Serum TPHA and FTA-ABS test were negative. The C3 value was 101 mg/dl,
(normal: 55-120/dl). C4 value was 43.3 mg/dl, (normal: 20-50 mg/dl). Although the initial pressure of the cerebrospinal fluid (CSF) was high (45 mm Hg), the CSF showed no leukocytes, and normal protein and glucose concentrations. Bacterial and tuberculosis cultures of the CSF were negative. Acute and convalescent viral titers of blood and CSF were negative.

A diagnosis of Behçet's disease was established, on the basis of clinical symptoms and laboratory findings, and it was regarded as being of the complete type according to the criteria of the Behçet's Disease Research Committee of Japan (8).

Brain CT. On the 7th hospital day, an enhanced CT scan revealed empty delta sign. MRI studies. On the 14th hospital day (during the chronic stage) T2-weighted images (TR=1600 ms, TS=90 ms) revealed a high signal area in the superior sagittal sinus (Fig. 1a, b). T1-weighted images (TR=600 ms, TE=45 ms) detected a hyperintense signal in the sigmoid portion of the lateral sinus (Fig. 2).

Clinical course. Dexamethasone therapy (20 mg/day for 4 days, 16 mg/day for 4 days, 12 mg/day for 4 days, and 8 mg/day for 4 days) and small doses of dexamethasone therapy thereafter. Anticoagulant therapy (Heparin) was 20 000 IU/day with corticosteroid, and followed by oral anticoagulant (Warfarin) for two months. Improvement of the neurologic symptoms occurred after one month. The papilledema disappeared after 2 months. Improvement was noted in the arthritic, ocular and skin symptoms. The iridocyclitis was aggravated after withdrawal of systemic corticosteroid therapy. The patient received topical mydriatics and corticosteroids. Ocular symptoms are suppressed, but a gradual improvement of ocular symptoms was not recorded.

DISCUSSION

Behçet's syndrome is a systemic disease of unknown etiology. It has a worldwide distribution, but the majority of the patients are of Mediterranean Middle-Eastern and Japanese origin (1). Cerebral venous thrombosis manifesting as benign intracranial hypertension with papilledema are well-known feature of Behçet's syndrome (5-7). The basic brain lesion, according to Japanese researchers, is a chronic relapsing inflammatory cellular infiltration around venules and capillaries, and occasionally around arteries. It is believed that the inflammatory reaction is brought about by human leukocyte antigens generated by many factors, which might be environmental, genetic, or other. Venous thrombosis occurs in approximately one third of patients with Behçet's disease (8). The vena cava and the portal vein are the most common sites, but involvement of the dural venous sinuses is rare.

Fig. 1a, b T2-weighted MR images showing a hyperintense signal area in the superior sagittal sinus.
Venous thrombosis may be precipitated in some patients by high fibrinogen levels, but in many cases the exact mechanism is unknown (9). The recent development of MRI has brought about a remarkable improvement in the diagnosis of Neuro-Behçet's disease. Our patient had intracranial hypertension diagnosed by elevated cerebrospinal fluid pressure in the presence of normal ventricles, with papilledema and headache. Empty delta sign on brain CT and on MRI, and a hyperintense signal on T1-and T2-weighted images in the sinuses, confirmed the cerebral venous thrombosis. The CSF examination is useful because of its abnormal composition. The increased protein and pleocytosis are present in two thirds of the patients. Mainly seen when focal signs are present, pleocytosis and the presence of red blood cells are found in patients presenting with benign intracranial hypertension, pointing to sinus thrombosis as the possible cause of this syndrome (9-11, 12). The empty delta sign on CT described by Buonanno et al, (13) appears after contrast injection and reflects the opacification of collateral veins in the superior sagittal sinus (SSS) wall, contrasting with noninjection of the clot inside the sinus. It is the most frequent direct sign, present in approximately 30% of published cases (14-16). A variety of MRI findings have been described, mainly related to the evaluation of thrombosis. At a very early stage, there is an absence of flow void and the occluded vessels appear isointense on the T1-weighted images, and hypointense on T2-weighted images. A few days later, the absence of flow void persists but the thrombosis hyperintense, initially on T1-and then on T2-weighted images. MRI diagnosis is particularly easy in the case of superior sagittal sinus thrombosis but convincing images have also been obtained in cases of thrombosis involving the lateral sinus, straight sinus, internal cerebral vein, cavernous sinus, and vein of Galen (17-19). The treatment of choice for the complications of Behçet's syndrome is unclear. Although steroids are useful during acute episodes and decrease the cranial pressure,
their long-term role in treatment is questionable (18). A lumbar peritoneal shunt can be performed for persistently elevated intracranial pressure. The treatment of the thrombotic process is still being debated. The use of anticoagulants has long remained controversial because of the risk of further bleeding into an already hemorrhagic infarct; such a complication has been well documented. The risk of increasing intracranial hemorrhage has probably been overstated, however, an increasing number of observations favor the use of heparin (11, 12, 19-23). Corticosteroid treatment in combination with anticoagulants has been proposed (5, 24). Our patient was successfully managed with corticosteroid associated with anticoagulants. Corticosteroid treatment in combination with anticoagulants should be considered as the first-line therapy for cerebral venous thrombosis in Behçet's disease. Although rebound phenomena after withdrawal of corticosteroids are common. The treatment of the ocular complications of Behçet's disease may require long-term regimens (25).

REFERENCES