Eur Arch Psychiatry Clin Neurosci (2002) 252 : 44 – 46

CASE REPORT

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Neuro-Behçet's disease involving the pons with initial onset of affective symptoms

Received: 7 May 2001 / Accepted: 29 November 2001

Abstract We report a case with Behçet's disease with initial onset of affective symptoms. This disease most commonly affects the brain stem in the nervous system, but a large lesion is very rare in the pons; initial onset of affective symptoms has not yet been reported in literature. This case was treated successfully with corticosteroids. Total clinical improvement was observed and the lesion was reduced in size within nine months (1.5 × 1.3 cm to 0.2 × 0.4 cm).

Key words Behçet's disease · affective symptoms · pons · corticosteroids

Introduction

Behçet's disease (BD) is a chronic, multisystem inflammatory disorder with vasculitis as the underlying process [18]. Diagnosis depends on clinical criteria of recurrent aphthous stomatitis, genital ulcers, uveitis, cutaneous or large-vessel vasculitis, synovitis, and meningoencephalitis [6, 11, 12]. Involvement of the central nervous system (CNS) occurs usually following systemic manifestations by months to years, but it has also been reported that CNS involvement could be as the initial feature of the disease [17]. Pallis and Fudge [13] attempted to demarcate three main forms of nervous system involvement – a brain stem form, a meningomyelitic form and a variety with predominant mental symptoms. However, mental changes are seen at later stages of the disease.

The most commonly involved brain area is the brain stem. Some neurologic symptoms have been reported in BD, such as facial nerve palsy, gait disturbance, pathological reflexes [15].

The C-reactive protein level is increased. The positivities of the HLA-B5 and pathergy tests are 54 % and 65 %, respectively. Cerebrospinal fluid examination shows elevated protein and lymphocytic pleocytosis [3]. MRI reveals high intensity lesions in the brain stem on T1-weighted images enhanced with Gd-DTPA, reflecting active inflammation [7].

In the present study, we aimed to present a neuro-Behçet case (NBD) who was first diagnosed as hypomania.

Case report

A 35-year old man, admitted to the out-patient psychiatric clinic due to complaints with lingual ulcer, insomnia, disinhibited behavior, excess money spending and euphoria. These complaints had been observed during the last two weeks. He had neither psychiatric nor neuroimmunologic illnesses in his previous history. There was no significant illness in the family history and no positive neurologic findings were detected on the first admission. Cranial computerized tomography (CT) was normal. For these reasons, the diagnosis was considered as hypomania and lithium was prescribed at a dose of 600 mg/day. The treatment had failed and after three months the patient was brought to the emergency department due to quadriparesia, gait disturbances, speech disorder, diplopia, irritability and a depressive state. At neurological examination, the patient was alert

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and cooperative. Findings were Babinski sign, hyperactive reflexes, mild degree abducens and facial nerve paresia, pyramidal dysarthria and nuchal rigidity. Some psychiatric complaints such as excess speech and insomnia were continuing. Oro-genital ulcers were present. Cranial CT was normal. T1-weighted MRI showed a large hypointense edematous lesion, 1.5x1.3 cm in diameter, in the central part of pons (Fig. 1). Increased protein content and pleocytosis were determined in the CSF. The pathergy test was positive. NBD was considered the most likely diagnosis. After the therapy of methylprednisolone (1 mg/kg/day), he showed marked clinical improvement and the lesion gradually reduced in size from 1.5×1.3 cm to 0.2×0.4 cm within nine months (Fig. 2). Clinical improvement was observed earlier than the reduction in the lesion. Improvement was first observed in psychiatric symptoms, then in speech disorder, diplopia, gait disturbance and quadriparesia findings, respectively.

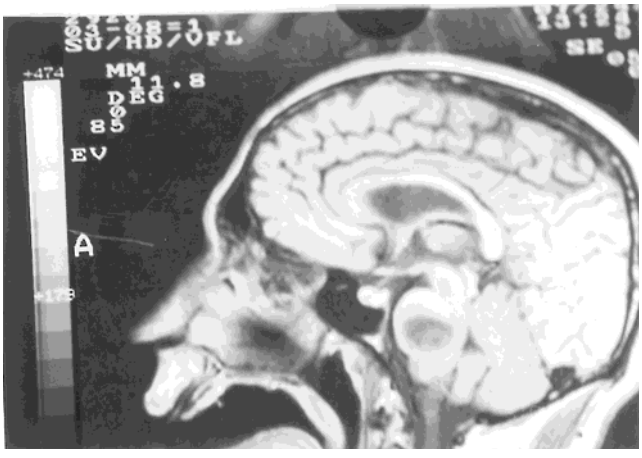

Fig. 1 A large edematous Behçet's lesion is seen in the pons before treatment (T1-weighted and Gd enhancement).

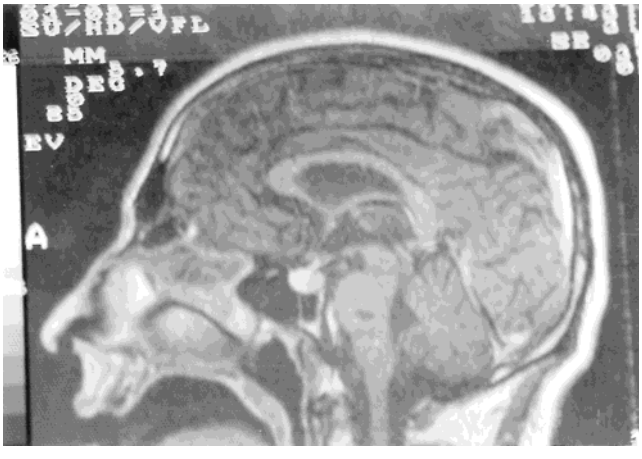

Fig. 2 Lesion is gradually reduced in size from 1.5 x 1.3 cm to 0.2 x 0.4 cm within nine months after treatment (T1-weighted and Gd enhancement).

Discussion

There is no absolutely diagnostic marker for BD. The diagnosis relying solely on the clinical picture. Despite published diagnostic criteria, problems still arise as features may not be present at the same time and incomplete forms of the condition can occur [5].

NBD occurs in approximately 5% of Behçet's diseases. The most commonly involved region of the brain is the brain stem but hemispheres, meninges, and the spinal cord can also be affected either individually or in combination [5]. During the course of the disease, brain atrophy may develop. Steroids have been claimed to help markedly in some NBD cases [3]. Corticosteroid therapy (1 mg/kg/day) had been given for a period of one year. This regimen was highly effective in our patient. Clinical symptoms disappeared totally, and the lesion was reduced in size within nine months.

Clinical presentations in NBD include bilateral pyramidal symptoms, mental changes, cranial nerve palsies, sphincter disturbances, and brain stem symptoms in the majority of patients. Sensory signs are characteristically absent. Isolated psychiatric symptoms and peripheral nerve involvement was observed rarely during the course of the disease [16]. The reported psychiatric symptoms are euphoria, disinhibition, and irritability, apathetic state [3, 4], transient episodes of confusion or in the form of dementia [9]. No case first presenting with psychiatric symptoms has been reported in the literature. Therefore, our case who had an onset with psychiatric complaints is an interesting one.

MRI study with T1- and T2-weighted images are useful to detect the lesions and to evaluate the activity in the NBD. T1-weighted MRI disclosed high signal areas in the cerebral white matter and the brain stem [7]. In this case, T1-weighted MRI enhanced with Gd-DTPA showed a high intensity lesion in the pons, reflecting active inflammation.

Differential diagnosis must be made for some neuroinflammatory disorders. Scattered hypointense lesions, severe demyelination and neuronal loss are seen in the white matter on T1-weighted MRI in multiple sclerosis [8]. Perifocal edema is seen in the pontine tumors. Marked capsule formation and intracavitary iso/hyperintense accumulation is observed at the brain abscess on T1-weighted MRI [14]. Oro-genital ulceration does not occur in multiple sclerosis. Sarcoidosis can present with uveitis, erythema nodosum and arthralgia and the presence of pulmonary nodules on the chest x-ray [10]. Our case did not have erythema nodosum or any pathology on the chest x-ray. According to these clinical, laboratory and MRI findings, NBD was considered the most likely diagnosis.

The rostral pons and caudal mesencephalon are sites for the serotonergic raphe nuclei and noradrenergic locus ceruleus. An increase in norepinephrine and/or serotonin may be a possible inducer of euphoria and hyperactivity. Mania has been reported after brain stem tumor [2].

The reticular formation is distributed throughout the medulla, pons, and midbrain. It plays a role in the rhythmical cycle of sleep and wakefulness. The reticular activating system is also a regulator of the degree of activation allowed to reach the cerebral cortex. The diffuse direct reticulocortical inputs and those from the thalamus are in some way gated so that the cortex does not receive too intense a level of stimulation, which could result in appropriate emotional and cognitive responses to stimuli. This is partly achieved through inputs from the reticular formation to the thalamic reticular nucleus, which monitors and gates the output from the thalamic intralaminar nuclei to the cerebral cortex [1]. Since affective symptoms were improved earlier than neurological findings after corticosteroid therapy, we think that affective symptoms in this case may be explained by a secondary dysfunction of the cortical and thalamic connections due to the damage of the subcortical structures.

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