MEMANTIN RESPONSIVE BEHCET'S DISEASE WITH INITIAL ONSET SEVERE COGNITIVE DECLINE

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Behcet's Disease (BD) is chronic, multisystem inflammatory disorder with vasculitis as the underlying process and most commonly affects the brain stem with a patchy involvement of upper parts in the nervous system. Although predominant mental symptoms are the main forms of nervous system involvement, mental changes and psychiatric problems are seen at later stages of BD. We report a case with Behcet's disease initially presented with depression and dementia. The most prominent findings were depression and cognitive impairement with gait ataxia. This case was treated with steroids, serotonin selective reuptake inhibitors and memantin. The initial presentation of memory problems and depression in neurological involvements of BD have not yet been reported in literature. Therefore, our case who had an onset with psychiatric complaints and memory problems is an interesting one.

Key words: Behcet's disease, depression, dementia, memantin, sertraline.

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INTRODUCTION

Behcet's disease (BD) is a three-symptom complex comprising uveitis, oral aphtae and genital ulcerations. CNS involvement may be rarely as the initial feature of the disease (1-3). Although predominant mental symptoms are the main forms of nervous system involvement, mental changes and psychiatric problems are seen at later stages of BD (2,4). We report a case with BD initial presentation of neurological involvement with different memory and frontal executive dysfunction. The present case was treated with serotonin selective reuptake inhibitors and memantin.

CASE

A 40-year old man, admitted to the out-patient neurology clinic due to complaints with non-structural pancranial headache attacks that started within last 3 months prior to admission and reported as much more occuring in the mornings. His wife also talked about cognition deficits, difficulty in learning and naming. He was forgetting for nearly one year period and denying that. Insomnia, disinhibited behavior, excess money spending and stolling were other parts of the story. There were deficits in problem solving, calculation (affecting skills such as calculation) and judgement. He was a taxi driver and he could not drive for last two months. He had neither psychiatric nor neuroimmunologic illnesses in his previous history. There was a gradual onset and continuing decline of memory function. There was no significant illness in the family history and no positive neurologic findings were detected on the first admission. After systemic examination and evaluation, we found that he had oral aphthae and scrotal ulcerations. Pathergy test was positive. Eye examination showed uveitis. Neuropsychiatric evaluation showed that the memory process was severely affected. He had difficulty in learning and recalling impaired in both verbal and visual modalities. Attention deficits, unsuitable answers and lack of fluency in speaking (severely impaired frontal lobe functions) were seen in the patient. Neurological examination showed mild gate ataxia to the right side. Complete blood count, serum chemistry panel, serum thyroid function tests, serum vitamin B12 levels

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were normal. Evoked potential examinations, electroencephalography and cerebrospinal fluid examinations were normal. Cranial MRI showed cerebellar and brain stem atrophy. The diagnosis of depression was made according to DSM-IV citeria. For these reasons, sertraline 100 mg/day, prednisone 1 mg/kg/d (gradually tapered over 2 to 3 months) and colchicine 1,5 mg/day were prescribed. The treatment was partially successfull and after six months the patient had eye contact while talking, was able to understand what is told to him, had grown insight and had contol of his drives. However he had still difficulty in learning new things and his time orientation was disturbed. On mini mental examination, he scored 21 points and the patient was diagnosed as having dementia according to DSM-IV citeria and memantin was added to the treatment. After 2 months follow-up, cognitive functions were improved and he scored 25 points, orientation and speech problems were solved.

DISCUSSION

Our patient was classified as BD according to the International Criteria (1). Although the problems still arise as features may not be present at the same time and incomplete forms of the condition can ocur, we thought that the earlier oral and genital lesions were probably missed so it was a undetected BD in the past rather than an initial onset of depression and dementia (1,2,5). He denied and said that the skin conditions appeared within last few months. Thus this condition was accepted as an initial onset of neurological involvement with a predominant cognitive syndrome. Cases with headache and psychological problems have mostly neurological involvement. We designated as the nonstructural vascular type headache related to BD, because the type of headache was not full-filling the IHS criteria for migraine (6).

The headache had been resolved in relation to therapy of BD as well. The most commonly involved region of the brain in neurological involvement of BD is the brain stem but hemispheres, meninges, and the spinal cord can also be affected either individually or in combination(4). Secondary dysfunction of the cortical and thalamic connections due to the damage of the subcortical structures may be a cause of affective symptoms in BD (2,3). The patients are presented with ataxia, dysarthria, and dysphagia in brain stem disturbances. During the course of the disease, brain atrophy may develop and is noted in cases with progressive disease(4).

Our patient was physically non-dependent but mentally dependent. We thought that diffuse cerebral and brain stem atrophy found on MRI examination could be a reason of mild gait problems in relation to connections between cortex and brain stem.

The most commonly and severely affected function is memory processes in BD. Attention deficit and impaired frontal lobe functions are the other most common abnormalities(1,5). Although we did not find any organic brain lesion, neuropsychiatric evaluation showed frontal lobe dysfunction. Our patient was first characterised by alterations in personality and social relationhips.

Cognitive deficits occured in the domains of attention, poor capacity of communication and disturbed attentional tasks signed the frontal executive dysfunction. He could not drive anyway as a sign of functional decline probably related to depression and cognitive impairment which are important risk factors in dementia (7).

The relationship between dementia and depression is complex. On the other hand, typical scales and questions used to diagnose depression contain many questions that patients with dementia endorse positively (8). Our patient was firstly denying some behavioral problems and disinhibition symptoms. There was not a lesion or localised atrophy which signed to frontal lobe dysfunction. We accepted firstly that the patient had a psychiatric problem rather than a dementia process. Though we think that it was treated succesfully with selective serotonine reuptake inhibitors, it could be a critism about that this patient's partial respose was secondary to prednisolone therapy rather than to sertralin. Because of this partial response, progressive memory problems, depression, disinhibition, and sleep disturbances were accepted as problems seen in the course of dementia in our patient . There were a cognitive problem and speech problem still after given up steroid which was used for three months. The diagnosis and treatment of speech and language pathologies in dementia is problematic because there are no standard methods. We hypothesized that presence of apathy in depression is associated with poorer frontal executive performance similar to Feil et al. (9).

Despite the absence of a strong scientific support for NMDA blockers (i.e.memantin), the recent successes with these compounds in Alzheimer's Disease have stimulated an interest in their use for frontal lobe dementia

(10). Because of these reasons, we used memantin instead of cholinesterase inhibitors and the cognitive and speech problems was improved in our patient.

In summary, we report a case with Behcet's disease with initial onset of depression and dementia. This case was treated with steroids, serotonin selective reuptake inhibitors and memantin. The initial onset of memory problems and depression in neurological involvements of BD have not yet been reported in literature. Therefore, our case who had an onset with psychiatric complaints and memory problems is an interesting one.

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