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Neuro-Behçet's Disease Presenting with Acute Psychosis: A Case Report

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Abstract **Case Report**

Behçet's disease (BD) is a chronic systemic inflammatory vasculitis of unknown etiology characterized by recurrent episodes of oral aphthous ulcers, genital ulcers, skin lesions, ocular lesions, and other manifestations. This disease affects many organs and systems and shows a wide range of clinical manifestations. The prevalence of anxiety, depression, and general psychiatric symptoms is higher among patients with BD compared with healthy individuals. However, syndromes such as psychosis appear to be less frequent. Therefore, we present a case of BD complicated by schizophrenia-like symptoms.

Keywords: Behçet's disease, psychotic symptoms, vasculitis.

INTRODUCTION

Behçet's disease (BD) is a rheumatological disease first described by dermatologist Hulusi Behçet in 1937. The disease has an unknown etiology and a recurrent pattern characterized by oral and genital ulcerations. It has the capacity to involve nearly all body organs such as the gastrointestinal tract, skin, mucosa, ocular and vascular systems, joints, and pulmonary, urogenital, musculoskeletal, heart, and nervous systems and causes significant morbidity and mortality. A relapsing-remitting disorder, inflammatory responses, and positive comeback to immunosuppressive therapy are the main indications of the autoinflammatory–autoimmune nature of the disease.

It is more common in Mediterranean, Middle Eastern, and Far Eastern countries similar to Turkey, Greece, Iraq, Iran, Japan, and China. The higher incidence of the disease in the mentioned regions may be due to some genetic and environmental factors. Age and gender are other characteristics that may increase the risk of BD. Generally, the disease affects people aged 20–40 years and is more severe in men. Common symptoms of the disease include arthritis, oral aphthae, skin lesions, and ulcers in the genital area, which can also develop ocular and vascular complications [2].

These symptoms negatively affect the physical and mental health of patients and reduce their quality of life. It has also been reported that oral ulcers may negatively affect body image in affected individuals and may restrict the feeding and speaking process [4, 5].

In the progressive course of the disease, patients frequently complain of weight loss, depression, and fatigue. However, chronic rheumatological problems in these patients limit their mobility and daily activities, leading to impaired self-esteem, which negatively affects their capability to form constructive relationships with others.

The frequency of psychiatric symptoms in BD is reported as 8–50% in medical literature, although there has not been too much reported about the comorbidity of psychotic symptoms with BD.

Therefore, we present a case of BD complicated by psychotic symptoms.

CASE PRESENTATION

A 35 young man, married, father of 2 children, presented with 2 weeks of bizarre behavior. According to the information received from his father, his complaints started with outbursts of anger, talking to himself, hearing voices, and seeing visionary images.

It was learnt that the patient, who had no known psychiatric history, was diagnosed with BD in 2017 by the rheumatology department due to recurrent oral aphthae, genital ulcer, arthralgia, thrombophlebitis, and uveitis in his past medical history. Although, colchicine, methylprednisolone, immunosuppressants drugs (azathioprine, cyclophosphamide) were recommended to

the patient during this period, he did not use them regularly and did not go to follow-up examinations.

Psychiatric Examination

Self-care ability was reduced he had logorrhea, and his affect was anxious, he was euphoric with multiple projects. He had auditory hallucinations such as human voices speaking to him and visual hallucinations. He has a delirium of persecution towards his wife which included to aggressive behavior. His sleep, appetite, and libido were reduced. He had no insight.

Physical Examination and Laboratory Findings

There were three ulcerated lesions, the largest of which was 4 cm in diameter, converging in the left leg pretibial region, papulopustular lesions located scattered in the extremities, and scarring in the scrotum. In the last eye examination, no ophthalmic finding for Behçet's involvement was detected. No neurological pathology was detected in the neurological examination and magnetic resonance imaging (MRI).

Treatment and Clinical Course

The patient, was hospitalized, was started aripiprazole 5 mg/d for psychotic symptoms in association with a mood stabilizer: sodium of valproate 500mg/jr. During the follow-up period, the aripiprazole dose was increased to 20 mg/d due to the lack of regression in the findings, and the sodium valproate to 1500 mg/jr.

In the 6th month of treatment and is being followed up as an outpatient, in remission with treatment.

DISCUSSION

Central nervous system (CNS) involvement is one of the most important causes of mortality and morbidity in BD. CNS involvement is seen at a rate of 5–10% in BD, and it is called neuro-Behçet syndrome (NBS) [8]. Neurological involvement is seen on average of 3–6 years after the onset of systemic findings [9].

In parenchymal involvement, the clinic is usually in the form of the brain stem or corticospinal tract syndrome, while in nonparenchymal involvement. It is in the form of increased intracranial pressure due to cerebral venous sinus thrombosis (CVST). The prognosis for nonparenchymal involvement is better than that for parenchymal involvement. While NBS progresses with relapse and remission in many patients, it shows a progressive course from the beginning in a minority of patients.

The most common symptom of NBS is headache. Psychiatric symptoms are present in 5–25% of patients in NBS [4]. Behavioral change is more common in parenchymal NBS than in brainstem and pyramid cerebellar syndrome [10].

A diagnosis of psychotic disorder due to the direct effects of BD could not be definitively made, since there was no neurological finding, normal cranial MRI, and no finding suggestive of increased intracranial pressure. Although sometimes no pathology is detected in neuroimaging methods in BD with neurological involvement, it is known that mild cerebrospinal fluid (CSF) changes can progress with various psychiatric symptoms. Because the patient did not have a psychiatric complaint before the diagnosis of BD and the absence of a psychiatric disease in the family other than the diagnosis of depressive disorder in his mother, neurological involvement of BD cannot be definitively excluded.

In conclusion, this case suggests that BD may present with only psychiatric symptoms without clinical or laboratory findings suggesting any neurological involvement. Further studies are needed on the relationship between clinical and laboratory findings of the disease and psychiatric symptoms.

Conflicts of Interest: There are no conflicts of interest.

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