

NEUROPSYCHIATRIC FEATURES IN BEHÇET'S SYNDROME: NEURO — PSYCHO — BEHÇET

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SUMMARY

It has been reported that the nervous system is involved in 30% of the cases with Behçet's Syndrome and half of these patients also show psychiatric signs.

A neuro-Behçet case with cerebello pyramidal and psychiatric findings (euphoria, depression, emotional lability, logorrhea, paranoid reaction and demential signs) is presented in this article. These psychiatric signs are compared with signs of previously reported cases. Cases of Behçet's syndrome have a lot of psychiatric features which may be specific for Behçet's syndrome. Therefore it is concluded that these type of cases must be called as Neuro-Psycho-Behçet Syndrome.

Key Words: Behçet's Syndrome, Dementia.

INTRODUCTION

The main symptoms of Behçet's Syndrome (BS) which have been known since Hippocrates and were defined by Hulusi Behçet in 1937 are recurring ocular hypopyonic iridocyclitis and ulcers in the mouth and in the genital area. Today, BS is known as a multisystem disease most frequently effecting the skin and the mucosas, the eyes, the joints and the gastrointestinal, urogenital, vascular and nervous systems (1).

Neurological complications of BS was first published by Knapp (2) in 1941. Cavara and D'Ermo (3) introduced the term "Neuro-Behçet" (NB) and later detailed studies on Neuro-Behçet Syndrome were carried out by Mc Menemy and Lawrence (4) in 1957 and by Rubinstein and Urich (5) in 1963. According to Siva (6) the first NB case in Turkey was reported by Polvan in 1956. Since then a number of cases accumulated in Turkish literature (6).

In 1977, Tuncbay et al (7) for the first time in Turkey, emphasized the psychiatric symptoms observed in BS patients. Later Kirbaş pointed out that these psychiatric symptoms could display a psychiatric profile which can be specific for BS. He introduced the term Neuro-Psycho-Behçet (NPB) in 1982 when he presented a case-study during the weekly sessions of the Neurology Department at the Ege University Hospital in İzmir. In 1983, Köksal et al. published a report on a BS case exhibiting psychiatric symptoms. Siva et al (8) published a case in "Recent Advances in

Behçet's Disease" in 1986 using the term Neuro-Psycho-Behçet. A year later, in the "Turkish Journal of Research in Medical Sciences" they published another paper based on neuro-radiological as well as other neuropsychiatric findings observed in NB cases (6).

In fact, psychiatric symptoms in BS are more frequent than generally accepted. Scientists have classified these findings either as mental disorders or as organic confusional syndromes. For example, Pallis and Fudge (9) classified these in 3 groups based on the location of the lesion: 1- brainstem syndrome, 2- meningo-myelitic syndrome and 3- organic confusional syndrome. Later some clinical classifications also appeared. But non of these classifications accepted the psychiatric findings as individual clinical entities.

Neuro-psychiatric symptoms appear 3 - 5 years after the onset of the BS. Neurological symptoms vary from headaches to severe meningoencephalitis which, causes hemiparesis. Papilloedema, speech disorders, urine and feces incontinences and severe psychological disorders, resembling delirium tremens, can be observed (9, 11, 12, 13, 14, 15).

In this paper a NB case with psychiatric features is presented. Comparison of these features with already published data is given in detail.

CASE REPORT

A 37 year old male patient was admitted to our clinic with psychological disorders, weakness in the legs, speech disorders, urinary and fecal incontinence, diplopia, painful ulcerations in the genital area and on the knees. The disease had first manifested itself at the age of 15 with short lasting ulcerations on the scrotum, which did not respond to medical treatment. A year later ulcerations in the mouth and on the tongue were observed. Time to time conjunctivitis, diplopia and blurry vision were also reported.

Although the patient was introverted and apathetic, he succeeded to graduate from a trade school and started to work as a topographer. He was married and had two sons but was indifferent to his family. In 1977 he went to Saudi Arabia for a job from which he returned 11 months later. His character change

was obvious. Although he had been away from his family for a long time, he wasn't enthusiastic about meeting them. His main concern was the money he had earned in Saudi Arabia. Although he seemed enthusiastic about the gifts brought he was still indifferent. A year later his wife noticed that his legs were jerking while he was asleep. Urinary incontinence occurred only once. Then he went to Iraq for three months. When he came back to Turkey he was totally confused. He looked shabby and was dirty. He could not even walk properly. He had dysarthric speech, and urinary incontinence. After a few days, fecal incontinence occurred and he began playing with his stool.

Besides pathologic laughing and crying he had a stereotypic speech and logorrhea. He had hypersexual behavior and impotence. He became aggressive and attacked his wife with a knife and forced her to leave the house. When his abnormal behavior increased, he was referred to the hospital with the diagnosis of "Paranoid Reaction". Due to the neurological findings he was admitted to the department of neurology. Steroid and imipramine treatment was started. But the symptoms persisted with remissions and exacerbations. When we first saw the patient, he was not even able to walk.

There was not any other serious disease neither in his own nor in his family's history. He was a smoker. His blood pressure was 120/80 mm Hg; pulse rate 84/minute and rhythmic. There were painful ulcers on his scrotum and his knees. Crepitation was noticed on his knees.

Neurological examination: Patient was alert and cooperative. Decreased pharyngeal reflex, absence of vomiting reflex, hyperactive deep-tendon reflexes in all extremities, bilateral Achilles clonus, bilateral Hoffmann sign were detected.

Palmomental and Babinski were (+), chin reflex (+++), and the patient had pathologic laughing and crying. Muscle tension was measured to be 0/5 on the left, 2/5 on the right in the lower extremities and 4/5 bilateral in the upper extremities. Abdominal superficial reflexes were absent. His speech was dysarthric and nasal. Ataxia, urine and fecal incontinence, bilateral dysmetria, dysdiadochokinesia were also observed.

Psychiatric Findings: The patient was pyknic, and apathetic, had nasal and dysarthric speech, showed periodic agitations and spoke loudly. His emotional reactions and ability to concentrate were significantly decreased. An obvious decline was noticed in his intellectual skills. He had difficulty in answering very simple questions. His descriptions were poor and his replies were repetitive.

He did not want to speak about his illness and his relatives. He had absurd ideas about returning to work when the lesions on his body healed. He was indifferent, unaffectionate and distant in his relations.

Laziness, weakness and ambivalence especially towards his wife were evident. His daily life was completely dependent on his wife.

Laboratory Findings: Routine blood and urine test results were normal. Liver function tests were normal. Erythrocyte sedimentation rate was 100 mm/hr. Cerebrospinal fluid (CSF) pressure was 240 mm H₂O. CSF protein: 65 mg. Pandy: (+++), cell count: 15/mm³, IgG: 13.2 % ng. Serum : IgG: 2112 % ng, other results were normal. Wassermann and Kolmer: (-). BS - specific skin tests: (+). Electroencephalography, electronistagmography, pneumoencephalography, brain scintigraphy, Rie, cervical and skull x-rays were normal. Electromyography indicated a slight delay in the second component of the eye - closure reflex.

DISCUSSION

The initial symptoms (oral and genital ulcers and conjunctivitis) appeared at the age of 15 in this case. His character which was introverted at the onset of the disease changed remarkably. This was followed first by urine and fecal incontinence, cerebellar findings, dysarthria, cerebellar ataxia and pyramidal irritations. Psychiatric findings initiated with paranoid thoughts and continued with hypo and hyper sexuality.

There was a slight increase in the cerebrospinal fluid (CSF) pressure as well as a definite increase in the number of cells and the protein content of the CSF. Serum and CSF IgG levels were normal. Increased erythrocyte sedimentation rate, slight leukocytosis and pleocytosis in the CSF were similar to the cases already published. Our case showed positive Hoffmann test in the EMG, a positive skin test but had no pyramidal signs in clinical examination which indicated subclinical irritation.

Observed signs were: agitation, euphoria, depression, pathologic laughing and crying, intellectual retardation, emotional lability, stereotypic speech, persecution complex, obsessive-compulsive behavior and sometimes mental confusion and logorrhea. Dementia became evident in the advanced stages of the disease.

The case reported by Siva et al (6) showed a psychiatric picture characterized by euphoria-hypomania, emotional lability, infrequent depression and anxiety, disorientation, persecution complex and psychomotor agitations. The prominent psychiatric symptom of the case which was reported by Tuncbay et al (7) was infrequent disorientation. Besides memory and concentration failure hallucinations and illusions, wrong and irregular speech were also detected. Besides logorrhea, there was also mutism and the patient could only speak when provoked. Extreme behavioral patterns like euphoric and expansive overreactions and depressions were also observed. The patient demonstrated obsessive pathologic behavior as well as behavior like exhibiting his genitalia.

The case reported by Rubinstein and Ulrich (5) was characterized by depression, intellectual collapse, dementia, short lasting complete memory loss, emotional lability, euphoria and pathologic laughing and crying.

Case reported by Miyakawa et al (17) showed a clinical picture of pathologic laughing and crying, apathy and sometimes euphoria, loss of emotional control, disorientation and dementia.

All these features are summarized in Table I. This table demonstrates that most of the psychiatric features are in common.

The overall evaluation of the neuropsychiatric symptoms of five reported Neuro Behçet cases, show that four phases in the clinical evaluation of this Syndrome is possible.

Phase 1: Emotional lability; memory and concentration failures.

Phase 2: Depression

Phase 3: A psychosis like picture with obsessive, compulsive behavior, paranoid thoughts, and hallucinations.

Phase 4: An organic confusional state resembling dementia.

We believe that psychiatric features of Behçet's Syndrome has a characteristic pattern when classified as above. This classification can also be helpful in diagnosis.

As a conclusion we want to stress the fact that Neuro-Psycho - Behçet Syndrome should be an accepted terminology.

Table I: Psychiatric Features of Neuro-Behçet cases.

Psycho-pathologic features	Kırbaş	Siva et al.	Tuncbay et al.	Rubinstein et al.	Miyakawa et al.
Euphoria	+	+	+	+	+
Emotional Lability	+	+	+	+	+
Memory and Concentration Failures	+	+	+	+	+
Disorientation	+	+	+	?	+
Psycho-motor Agitation	+	+	+	?	?
Pathologic Laughing and Crying	+	+	?	+	+
Obsession or Paranoid Reaction	+	+	+	?	?
Demential Signs	+	?	+	+	?

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