Millard-Gubler Syndrome due to Parenchymal Neuro-Behcet's Syndrome Lesion: a Rare Case Report

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Abstract

Behcet's disease; it is a vascular-inflammatory chronic recurrent disease that can affect many systems in the body. Millard Gubler syndrome is one of the brainstem syndromes that occurs due to lesions involving the ventral part of the caudal pons. We wanted to present a case of Millard Gubler syndrome, which developed due to Behçet's syndrome lesion in the pons ventral region in a patient who presented with limitation of outward gaze on the left, inability to close the left eye completely, inability to completely wrinkle the left side of the forehead, and loss of contralateral muscle strength.

Keywords: **Behcet's syndrome, parenchymal, abducens, facial, lesion**

Acta Neurol Taiwan 2024;33:19-22

INTRODUCTION

Behcet's disease; it is a vascular-inflammatory chronic recurrent disease that can affect many systems in the body. Although its etiology is unknown; it can affect many organs and systems. Common symptoms include oral and genital aphthae, papulopustular skin lesions, and eye inflammation. Mucocutaneous lesions, uveitis, vasculopathy, musculoskeletal involvement, pulmonary, cardiac, gastrointestinal and nervous system involvement may be seen [1,2,3,4].

Millard Gubler syndrome is one of the brainstem syndromes that occurs due to lesions involving the ventral part of the caudal pons. Since the pyramidal fibers do not cross yet, paresis occurs on the opposite side of the body due to the lesion. Outward gaze paresis on the same side due to abducens (6th cranial nerve) involvement and peripheral type facial paralysis on the same side due to facial nerve (7th cranial nerve) involvement. Although it is most commonly seen secondary to ischemic stroke, demyelinating lesions involving the ventral part of the pons can also occur due to tumors and infectious causes [5,6,7,8]. In our presentation, we wanted to present a patient who was diagnosed with Parenchymal Neuro-Behcet's and who developed Millard-Gubler syndrome due to pons ventral part involvement. When the literature was reviewed, no Millard Gubler syndrome due to Neuro-Behcet's lesion was found, and we found it valuable to present it because Millar Gubler syndrome often occurs with ischemic stroke.

CASE REPORT

A 33-year-old right-handed male patient presented

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Received October 12, 2022 Revised November 10, 2022
Accepted November 14, 2022
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with complaints of headache, blurred vision, shifting at the corner of the mouth, inability to close his left eye completely, numbness in the right arm and leg, and loss of strength for about two months. The patient had aphthous lesions in his mouth and genital area three to four times a year. He had no known disease or medication. On physical examination, his blood pressure was 130/70 mmHg, heart rate was 80/min, temperature was 36.5, and oxygen saturation was 97% in room air. Electrocardiography (ECG) was in normal sinus rhythm. In the neurological examination of the patient, the left eye did not fully close, the outward gaze was limited in the left eye, he could not wrinkle the left side of his forehead, there was effacement in the left nasolabial groove, right upper and lower extremity strength -5/5, and hypoesthesia on the right. Deep tendon reflexes were normoactive. No significant pathology was detected in the brain computed tomography (CT). In the magnetic resonance imaging (MRI) T2 sequence of the patient, a lesion was detected in the pons ventral region (Figure 1). No vessel occlusion was found in the brain-neck CT angiography. No pathology was detected in MRI venography. The patient was admitted to the Neurology service for further examination and treatment.

In laboratory tests, complete blood count, platelet function tests, coagulation tests, routine biochemistry tests, complete urinalysis, protein electrophoresis, antiphospholipid antibodies, antinuclear antibodies, vitamin b12, thyroid function tests, HbA1c, erythrocyte sedimentation rate, autoantibody screening (anti-SSA, anti-SSB), antithyroid antibodies, syphilis serology (fluorescent treponemal antibody), Schirmer test, homocysteine, anticardiolipin, rheumatoid factor, protein C and S , Anti-streptolysin O (ASO), rheumatoid factor (RF), immunoglobin A,G ,M was normal. Elisa tests (Hepatitis A, HIV, hepatitis B, hepatitis C) were negative. When viral meningitis agents (Herpes simplex virus 1-2 (HSV), varicella zoster virus (VZV), enterovirus, parechovirus, ebstein barr virus (EBV), cytomegalovirus (CMV), adenovirus) were investigated, no agent was found. Brucella tests came back negative. Pathergy test for Behçet's was positive. No significant pathology was found in the results of CSF

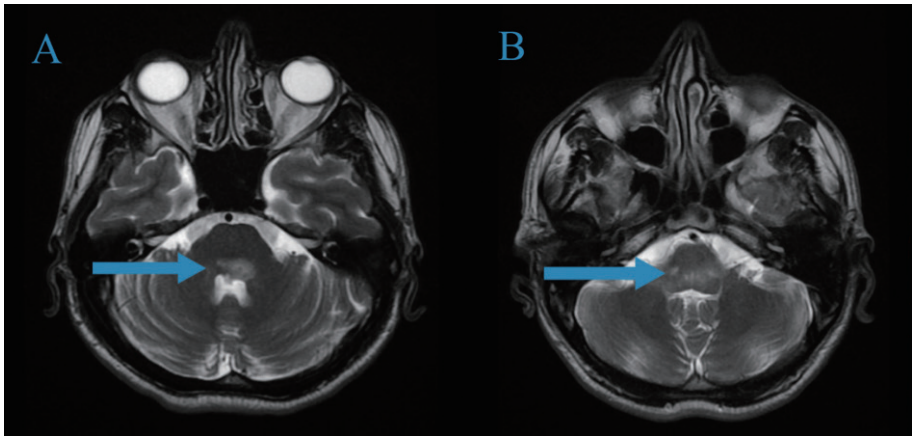

Fig. 1. Lesion in the pons ventral region in MRI T2 sequence

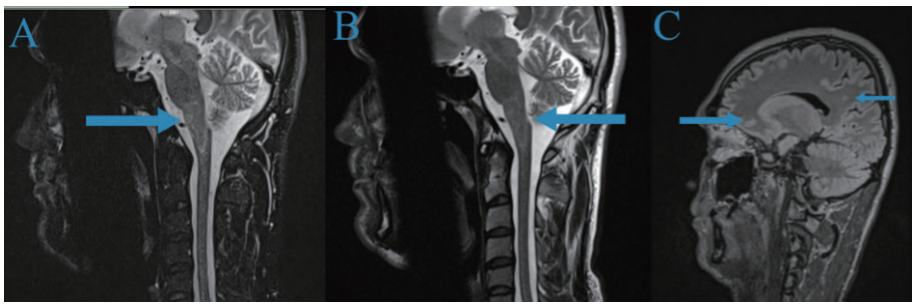

Fig. 2. T2 sequence hyperintense lesions at the craniocervical junction and brain stem

examination. Oligoclonal bands (OCB) and neuromyelitis optica (NMO) were negative. HLA B5 was negative. T2 hyperintense lesions in the craniocervical junction and brain stem were recorded in cervical MRI (Figure 2).

Visual evoked potentials (VEP) were consistent with bilateral visual pathway dysfunction. Neuro-Behçet syndrome was considered in the patient and treatment was planned with metiprednisolone 1gr/day for ten days. Azothiopurine was switched to maintenance therapy.

The follow-up of the patient continues.

DISCUSSION

Behcet's disease; it is a vascular-inflammatory chronic recurrent disease that can affect many systems in the body. The disease was described by Turkish dermatologist Hulusi Behçet in 1937. Although its etiology is unknown; It can affect many organs and systems. Common symptoms include oral and genital aphthae, papulopustular skin lesions, and eye inflammation[1,2]. Mucocutaneous lesions, uveitis, vascular vasculopathy, musculoskeletal involvement, pulmonary, cardiac, gastrointestinal and nervous system involvement can be seen. Neuro-Behcet's disease can be divided into two groups as Parenchymal and non-parenchymal [2,3].

Patients with neurological involvement due to Behcet's disease often have parenchymal central nervous system involvement. Parenchymal Neuro-Behcet's syndrome is most commonly seen with meningoencephalitis. Headache, fever, genital-oral ulcers may occur. If brain stem involvement is present, the patient may have ophthalmoplegia, cranial nerve involvement, cerebellar or pyramidal dysfunction. Depending on cerebral hemisphere involvement, hemiparesis, hemisensory loss, seizures, speech disorders can be seen. Sphincter dysfunction may occur due to medulla spinalis involvement [1,3,4]

Non-Parenchymal Neuro-Behcet's syndrome involvement is in cerebral vessels. Cerebral vein thrombosis (CVT) is common. It can be seen in patients with aneurysm[2,4].

Although there are diagnostic criteria for Behcet's disease, there is no definitive diagnostic criteria or test for Neuro-Behcet's syndrome. HLA-B51 may be helpful in diagnosis. Although the pathergy test is one of the diagnostic criteria, its sensitivity is low. The absence of oligoclonal bands in the cerebrospinal fluid (CSF) can help differentiate it from Multiple Sclerosis (MS). Magnetic resonance imaging (MRI) is the gold standard for the diagnosis of Neuro-Behcet's syndrome. Acute or subacute lesions appear hypointense or isointense on MRI T1 sequence and hyperintense on T2 sequence. Lesions are around the brainstem and sometimes extend into the diencephalon. Our patient also had a lesion in the pons ventral region.

Neuro-Behcet's syndrome is three times more common in males than females[1,2,3,4].

Millard Gubler syndrome is one of the brainstem syndromes that occurs due to lesions involving the ventral part of the caudal pons. Since the pyramidal fibers do not cross yet, paresis occurs on the opposite side of the body due to the lesion. Outward gaze paresis on the same side due to abducens (6th cranial nerve) involvement and peripheral type facial paralysis on the same side due to facial nerve (7th cranial nerve) involvement. Although it is most commonly seen secondary to ischemic stroke, demyelinating lesions involving the ventral part of the pons can also occur due to tumors and infectious causes[5,6,7,8].

In our patient, limitation of outward gaze in the left eye, effacement in the left nasolabial groove, inability to wrinkle the left side of the forehead, complete closure of the left eye, and loss of strength in the contralateral muscle strength examination were found to be significant in terms of Millard Gubler syndrome. As a result of the investigations performed on the etiology of the lesion in the ventral pons in the MRI of the patient, Behçet's syndrome was suggested. Cerebrovascular disease was excluded in our patient. The symptoms of the patient regressed after 1 g/day methylprednisolone treatment given to the patient for ten days. The patient was started on azathioprine in maintenance therapy and was called for controls.

As a result; although Millard Gubler syndrome is most commonly seen to occur secondary to ischemic stroke, it can also be seen in lesions involving the ventral pons. In our case, it was confirmed as Millard Gubler due to Behcet's syndrome lesion. Behcet's disease should be kept in mind after ischemic stroke is ruled out in patients with similar findings.

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