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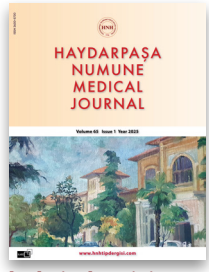

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CASE REPORT

A Case of Myasthenia Gravis Presenting with Pseudo Bilateral 6th Cranial Nerve Palsy

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Abstract

Myasthenia gravis (MG) is an autoimmune disease of the neuromuscular junction caused by autoantibodies to acetylcholine receptor antibodies (AchRs). Patients with myasthenia gravis (MG) usually present with diffuse muscle weakness. Myasthenic weakness typically affects extraocular, bulbar, or proximal limb muscles. Clinical manifestations often begin in the eye muscles and are characterized by ptosis. When MG is isolated to ocular symptoms only, it is referred to as ocular myasthenia gravis (OMG). OMG can also present atypically, mimicking isolated cranial nerve palsies. A 21-year-old male patient with no significant medical history presented to the emergency department with complaints of diplopia, blurred vision, and headache. Neurological examination revealed bilateral outward gaze limitation. The patient, who had no history of chronic disease or regular medication use and had no special features in his family history, was diagnosed with myasthenia gravis after extensive tests. This case highlights that myasthenia gravis in young patients may present with atypical symptoms, deviating from the traditional manifestations, thus providing a valuable educational contribution to clinical practice.

Keywords: Abducens nerve palsy; diplopia; myasthenia gravis.

Normal binocular vision relies on the proper functioning of the coordination between the eyes, eye muscles, and neural structures. Additionally, it requires a complex process that ensures the parallel movement of the eyes and the integrated processing of visual information by the brain. Diplopia may reflect dysfunction at any point within this dynamic system and can sometimes indicate a significant local or systemic disease. The initial step in evaluating diplopia is to determine whether the double vision is monocular (with one eye) or binocular (with two eyes). The next step is to investigate which cranial nerves or eye muscles are affected and at which level the problem lies within the neurological axis.[1]

Some patients may not be aware of their double vision; instead, they may report symptoms such as blurred vision, ghost images, confusion, eye fatigue, or dizziness. When diplopia is detected, the two images may be horizontal, vertical, or diagonal. Diplopia can be present equally in all directions of gaze, or it may vary depending on a specific direction.[2]

In this case, the findings of diplopia developed as a result of myasthenia gravis, mimicking bilateral sixth cranial nerve palsy. Myasthenia gravis is an autoimmune neuromuscular junction disease caused by autoantibodies against acetylcholine receptors. This condition primarily manifests as weakness in skeletal muscles and typically presents initially with ocular symptoms such as ptosis and diplopia.

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When myasthenia gravis is confined to ocular symptoms, it is referred to as ocular myasthenia gravis (OMG). Although sixth nerve palsy may be considered in the differential diagnosis, myasthenia gravis typically presents with a combination of paralysis affecting the III, IV, and VI cranial nerves.[3] In this case, the diagnostic process and clinical features of a patient with myasthenia gravis presenting with pseudo-bilateral sixth cranial nerve palsy will be discussed.

Case Report

A 21-year-old male patient presented to the emergency department with complaints of double vision (diplopia), blurred vision, and headache that had started 5 days ago. The symptoms of double vision, particularly when gazing outward, had developed suddenly, and there was no history of trauma. The diplopia resolved when he closed one eye, and the symptoms did not show a fluctuating course.

The patient reported having a frontal headache for approximately two months, which was described as pressure-like and compressive in nature, occurring mainly in the evenings regardless of position. This headache occasionally woke him from sleep and was relieved with analgesics. There was no history of syncope or seizures. The patient's review of systems was negative for symptoms of rheumatologic, viral, or demyelinating diseases.

On neurological examination, the patient's vital signs were within normal limits, and no pathological findings were noted except for bilateral sixth cranial nerve palsy. The fundoscopic examination was normal. Visual evoked potentials (VEP) were within normal limits. The patient underwent contrast-enhanced brain MRI, MR venography, MR angiography, and MR diffusion, but no significant pathology was identified on cranial imaging. Routine laboratory tests, including complete blood count (CBC), biochemistry, autoimmune panel, thyroid panel, and B12 levels, did not reveal any significant abnormalities.

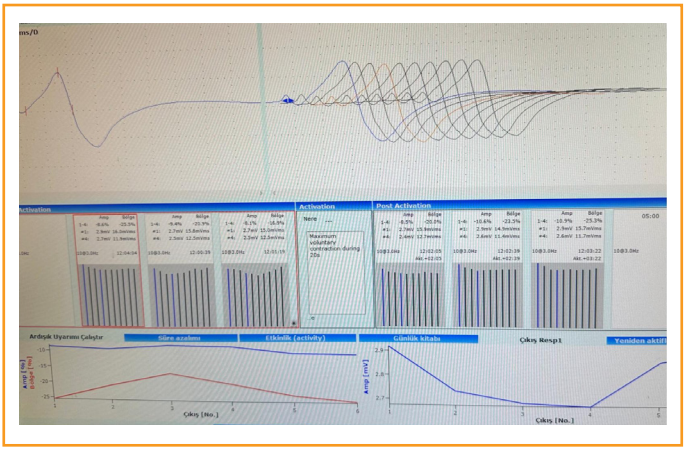

Figure 1. Repetitive nerve stimulation at 3 Hz applied to the right accessory nerve resulted in a significant decrement response of -10.9% following maximal muscle contraction.

After obtaining informed consent, cerebrospinal fluid (CSF) pressure was measured to rule out benign intracranial hypertension. The opening pressure was 18 cm H2O, and the closing pressure was 12 cm H_2O. CSF cell count and biochemistry were within normal limits.

To exclude a rare ophthalmoparesis-induced myasthenia gravis diagnosis, repetitive electromyography (EMG) was performed, and acetylcholine receptor antibodies were sent for testing. The EMG results were consistent with postsynaptic membrane dysfunction (Fig. 1). The acetylcholine receptor antibody test was positive. The patient was started on Mestinon (3x1) and Prednol (1 mg per kg body weight). There was a minimal improvement in the outward gaze of the left eye, and the patient is currently under clinical follow-up.

Discussion

Normal binocular vision is a complex process that requires the functional integrity and collaboration of ocular structures, eye muscles, and the neural system as a whole. Diplopia can indicate pathology at any point within this complex system. It may arise from a local disturbance in one eye or from a systemic disease. The approach to diagnosing diplopia begins with determining whether the double vision is monocular or binocular.[1]

In the presented case, since the diplopia resolved when one eye was closed, we classified the condition as a binocular issue. The next step is to determine the location of the lesion. In the differential diagnosis, life-threatening conditions such as vascular lesions, myasthenia gravis, demyelinating diseases, meningitis, and botulism should be considered. The sixth cranial nerve is the longest nerve in the cranial cavity, making it particularly vulnerable to numerous pathological processes in the posterior and middle cranial fossae. This nerve can be affected by conditions such as tumors, trauma, and ischemic events.[4]

The patient's headache description was evaluated in terms of an intracranial space-occupying lesion. The imaging tests did not reveal any space-occupying lesions. Furthermore, lumbar puncture was performed to rule out idiopathic intracranial hypertension; the opening pressure was measured at 18 cm H_2O, and the CSF examination was normal. These findings, along with the type of headache, led to the exclusion of intracranial hypertension as a diagnosis. The patient reported no significant changes in symptoms during hospitalization.

Ptosis, a common finding in myasthenia, was not observed in the patient. Additionally, there were no clinical signs of fluctuating diplopia or worsening muscle weakness in the evenings. While these findings initially reduced the suspicion for myasthenia gravis, the rare cause of sixth cranial nerve palsy, myasthenia gravis, was investigated, and the diagnosis of ocular myasthenia gravis (OMG) was confirmed. The diagnosis was supported by characteristic EMG findings and a positive acetylcholine receptor antibody test. The three main autoantibodies important in pathogenesis are anti-AChR antibodies, anti-MuSK antibodies, and low-density lipoprotein receptor-related protein-4 (LRP4) antibodies. The sensitivity of anti-AChR and anti-MuSK antibodies in ocular myasthenia is 55% and less than 10%, respectively.[5]

Myasthenia gravis (MG) is an autoimmune neuromuscular junction disorder caused by the production of autoantibodies against acetylcholine receptors.[6] MG affects ocular, bulbar, respiratory, axial, and limb muscles, leading to characteristic fluctuating fatigue and muscle weakness.[7] When MG is limited to ocular symptoms, it is referred to as ocular myasthenia gravis (OMG), and it is more common in men. OMG may present atypically, mimicking isolated cranial nerve palsies. Cleary et al.[8] briefly touched on this, in which EOMs were examined in 49 patients with OMG and determined that 72% had bilateral EOM involvement and that 4% presented with isolated abducens nerve palsy.

In patients presenting with isolated cranial nerve palsies, it is crucial to rule out paralysis and tumors first, and clinical evaluation is essential. However, after excluding stroke and tumors, clinicians should consider OMG in the differential diagnosis and approach it with a high level of suspicion. This case makes a valuable contribution to the literature by demonstrating the atypical presentation of ocular myasthenia gravis (OMG) and emphasizing the importance of a comprehensive and systematic approach in diagnosing diplopia. The patient's symptoms, which initially excluded myasthenia gravis (MG) due to the absence of ptosis and fluctuating diplopia, highlight that OMG can present in a way that mimics isolated cranial nerve palsies. While the clinical presentation initially suggested other possible causes of diplopia, this case underscores the critical importance of considering OMG in the differential diagnosis, even in the absence of classic features such as ptosis and the evening worsening of symptoms.

This case contributes to the literature by reinforcing the idea that OMG can sometimes present in a manner that deviates from the traditional understanding, which necessitates clinicians, especially in young patients with isolated cranial nerve involvement, to approach with a high level of suspicion. Furthermore, it emphasizes the importance of comprehensive diagnostic testing in patients presenting with diplopia and the need for clinicians to consider autoimmune etiologies even in the absence of typical symptoms. Therefore, our case serves as an important example that highlights the significance of a detailed clinical history and careful evaluation in the timely and accurate diagnosis of ocular myasthenia gravis, ultimately leading to better patient outcomes.

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