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# A POSTERIOR CRANIAL FOSSA LESION MIMICKING MYASTHENIA GRAVIS

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## ABSTRACT

A 41-year-old male with prior diagnosis of seronegative myasthenia gravis presented with complaints of bilateral ptosis, dysphagia, nasal voice and nasal regurgitation. Despite treatment with pyridostigmine, there was lack of significant improvement in the symptoms. Further investigations, including antibody assays and nerve conduction studies did not support the diagnosis. Magnetic resonance imaging revealed a lesion in the posterior cranial fossa, consistent with meningioma, which was compressing surrounding structures. This case highlights the importance of imaging techniques and considering alternative diagnosis in patients with unusual presentations to ensure appropriate management.

## KEYWORDS

Myasthenia gravis, posterior cranial fossa, ptosis, meningioma, neuromuscular junction

## LEARNING POINTS

- This case emphasizes the importance of imaging in patients presenting with symptoms similar to those of neuromuscular junction disorders.
- It also highlights the possible challenges in diagnosis of such patients.

## INTRODUCTION

Myasthenia gravis (MG) is a chronic autoimmune disorder which primarily affects voluntary muscles responsible for controlling the eyes, mouth, throat, and limbs. Common symptoms of MG include drooping eyelids (ptosis), double vision (diplopia) and difficulty in speaking (dysarthria) and swallowing (dysphagia)[1]. Diagnosis of MG typically involves blood tests for anti-acetylcholine receptor antibodies and anti-muscle specific kinase (MuSK) antibodies, as well as electrophysiological testing[2].

We present a unique case of a patient who initially exhibited symptoms resembling neuromuscular junction disorders, leading to a misdiagnosis of MG. However, unresponsiveness to treatment, along with negative results from blood tests and nerve conduction studies, confirmed that MG was not the correct diagnosis. Subsequent investigations, including imaging scans, revealed an unexpected finding: a space-occupying lesion located in the posterior cranial fossa.

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## CASE DESCRIPTION

A 41-year-old male presented to the hospital with primary complaint of bilateral partial ptosis. Associated symptoms included dysphagia in the last 1.5 months, voice changes which included nasal voice and nasal regurgitation for one year, generalized weakness for one month and bilateral upward gaze restriction. Past surgical history was negative, and the patient was mobile without support. Medical history included prior diagnosis of seronegative MG at a previous healthcare facility for which he had been taking pyridostigmine but had minimal response to the medications, so he was admitted for reevaluation.

The patient was vitally stable with a Glasgow coma scale (GCS) score of 15/15. On examination, pupils were equal and bilaterally equally reactive to light, with a positive upward gaze restriction. Dilated fundoscopic examination of the eyes revealed no papilledema. Bilateral ptosis was observed with significant drooping of the eyelids. The patient had no signs of fatigability and there was no proximal muscle weakness. Muscle tone was normal with a 5/5 strength according to the Medical Research Council (MRC) scale. Sensations were intact. Plantar reflexes were normal. No diurnal variation was seen in the symptoms. Blood tests for anti-acetylcholine receptor (AChR) antibodies and anti-MuSK antibodies were performed along with nerve conduction tests. Single-fiber electromyography (EMG), and a Tensilon test were not performed. Chest X-ray was normal. Results of motor nerve conduction studies indicated a normal test to the tested nerve of the right upper and lower limbs and left facial nerve. Results of sensory nerve conduction studies indicated a normal test to the tested nerves of right upper and lower limbs. Repetitive stimulation revealed no significant decrement in muscles innervated by the right ulnar, musculocutaneous and facial nerves. Needle examination revealed a normal EMG pattern in the tested muscles.

The patient exhibited symptoms similar to those seen in diseases affecting the neuromuscular junctions (NMJs). Although MG was considered as a possible diagnosis, the lack of significant improvement in symptoms after treatment with pyridostigmine, as well as the absence of fatigue and lack of symptom improvement with rest, suggested an alternative underlying cause. The nerve conduction studies and the absence of anti-acetylcholine receptor and anti-MuSK antibodies both further confirmed that the patient did not have MG. Further assessment was done by conducting magnetic resonance imaging (MRI) of the brain *(Fig. 1)*. A space-occupying lesion was detected in the posterior cranial fossa of the patient. The lesion was extra axial, well circumscribed and infratentorial in location. It measured 4.73 x 5.76 x 5.14 cm in anteroposterior (AP) x transverse (TV) x craniocaudal (CC). Microscopic examination showed fragments of tissue revealing a lesion composed of clusters of elongated cells with collagen deposits. Occasional psammoma bodies and pseudonuclear inclusions were seen as well. No malignancy, mitosis or invasion was seen. The lesion was consistent with meningioma, causing a mass effect on the surrounding parenchyma and a contralateral shift of the midbrain and pons. Additionally, there was effacement of the fourth ventricle, leading to upstream dilation of the ventricular chain. Sagittal sections demonstrated crowding of cervicomedullary junction with tonsillar herniation with a decent of 2.04 cm from the McRae line. The interhemispheric fissure was centered on the midline. No signs of increased




*Figure 1. Magnetic resonance imaging of head: A) and B) axial views, and C) sagittal view, showing a large posterior fossa meningioma.*

intracranial pressure were seen. The cortex and white matter showed normal development, and no abnormalities were seen with the basal ganglia, internal capsule, corpus callosum or thalamus. The sella, pituitary and parasellar structures were unremarkable. The cerebellopontine angle was normal on each side and normal development was seen in the paranasal sinuses and mastoid air cells. These findings, in conjunction with the exclusion of MG as a diagnosis, led to the conclusion that the lesion was the cause of the patient's symptoms.

Surgical intervention was advised, and the patient underwent retrosigmoid craniectomy with excision of the lesion. However, within 24 hours of the procedure, the patient went into coma. This outcome highlights the importance of early diagnosis and intervention in patients presenting with symptoms similar to NMJ disorders as well as the possible challenges in diagnosis of such individuals.

## DISCUSSION

Seronegative MG is a condition in which patients with MG have no detectable antibodies in their serum which can be due to laboratory test inaccuracies, decreased antibody production, immunosuppressive therapy, immunodeficiencies, antigen depletion, and immune senescence[3]. Our patient displayed typical symptoms of diseases involving NMJs. Blood tests and lack of response to an acetylcholinesterase inhibitor ruled out MG. An MRI of the brain showed a space-occupying lesion in the posterior cranial fossa. It is important to note that this brain lesion may have been compressing the nuclei of the levator palpebrae superioris (LPS) muscle, leading to the symptoms. It is worth mentioning that lesions causing dysfunction of the LPS can also result in upward gaze palsy due to the close proximity of the nuclei of the superior rectus muscle to the LPS.

An extensive literature search revealed cases of brain lesions presenting with symptoms similar to MG. In one study, a 19-year-old man with symptoms of MG showed variable diplopia and facial weakness. A positive Tensilon test and response to pyridostigmine suggested MG, but no AChR antibodies were found. EMG showed minor decremental changes. Imaging revealed a vertebrobasilar aneurysm compressing the brainstem. Following aneurysm occlusion, the patient recovered fully and was asymptomatic after 6 months[4]. In another study, a 54-year-old man initially diagnosed with ocular MG showed an excellent response to pyridostigmine but later developed worsening symptoms, including central neurogenic hyperventilation. Brain MRI revealed a high signal lesion in the brainstem. Despite temporary improvement with corticosteroid therapy, symptoms returned. Further evaluation confirmed a diagnosis of B cell lymphoma. This case highlights the importance of brain MRI in atypical ocular myasthenia and that spontaneous regression or steroid response does not rule out primary central nervous system lymphoma. Early histopathological confirmation is crucial for accurate diagnosis and treatment[5]. One specific study with a presentation similar to the case being discussed has been published Kumar in 2014[6]. A 14- year-old boy with symptoms of bilateral ptosis had grade 1 papilledema. Extraocular movements, pupils and neurological examination were normal. A magnetization transfer led to the diagnosis of a tuberculoma in the brain[6]. A similar study with a lesion in the dorsal part of the midbrain has was published by Sarkar in 2017[7]. In this case, an 11-year-old male presented with drooping of the eyelids with symptoms of headache and ataxia. Extraocular movements were normal with a normal pupillary size. There was no weakness in the limbs and tendon reflexes were normal. Blood counts were normal with a raised erythrocyte sedimentation rate. Subsequent MRI revealed a single ring enhancing lesion in the dorsal midbrain[7]. In both the above cases, the extraocular muscles were intact, whereas upward gaze palsy was detected in our case.

## CONCLUSION

This case underscores the importance of considering alternative diagnoses in patients with NMJ disorder-like symptoms, particularly when standard MG treatments fail. It highlights the critical role of brain MRI in uncovering unexpected conditions, suggesting that imaging should be an essential part of the diagnostic process in such cases.

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