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A Case of Myasthenia Gravis Post-COVID Vaccination with Bilateral Neurovascular Conflict

ABSTRACT

Myasthenia Gravis is an autoimmune postsynaptic neuromuscular junction disorder. The body produces auto antibodies against the postsynaptic nicotinic acetylcholine receptors. The disease is characterized by muscle weakness that fluctuates , worsening with exertion, and improving with rest. In about two thirds of the patients, the involvement of extrinsic ocular muscles presents as the initial symptom, usually progressing to involve other bulbar muscles and limb and respiratory musculature, resulting in generalized myasthenia gravis patients.1 The ocular symptoms include drooping of the upper lid, binocular horizontal diplopia, typically more in the evening and improving on rest. On examination, the patients have Ptosis and extraocular muscle limitation, that improve with sleep and ice pack test. An important differential diagnosis of Myasthenia Gravis is Oculomotor Nerve Palsy. Third Cranial Nerve supplies the levator palpebrae superioris, and the four extraocular muscles (superior, medial, and inferior rectus and inferior oblique) and ciliary and sphincter pupillae. Signs include Ptosis, extraocular muscle limitation except for abduction and depression, and anisocoria with a sluggishly reacting pupil. One of the causes of the Oculomotor Nerve Palsy is an abnormal contact of artery or vein with the nerve, so called neurovascular conflict. A Neurovascular Conflict results in an active nerve dysfunction. We present the case of a young adult male who presented in myasthenic crisis following vaccination for COVID 19 infection with not so classical signs of ocular myasthenia but also had features of bilateral neurovascular conflict on neuroimaging. The etiological role of neurovascular conflict in causing third nerve compression was difficult to establish and could also be a incidental finding. There are also no reported cases of precipitation of myasthenia following covid vaccination.

Key words: Disorder, Myasthenia gravis, Neuromuscular

INTRODUCTION

Myasthenia gravis (MG) is an autoimmune post-synaptic neuromuscular junction disorder. The body produces autoantibodies against the post-synaptic nicotinic acetylcholine receptors. The disease is characterized by muscle weakness that fluctuates, worsening with exertion, and improving with rest. In about two-thirds of the patients, the involvement of extrinsic ocular muscles presents as the initial symptom, usually progressing to involve other bulbar muscles and limb and respiratory musculature, and resulting in generalized MG patients.[1] The ocular symptoms include drooping of the upper lid, binocular horizontal diplopia, typically more in the evening, and improving on rest. On examination, the patients have Ptosis and extraocular muscle limitation that improves with sleep and ice pack test.

One important differential diagnosis of MG is oculomotor nerve palsy. Third cranial nerve supplies the levator palpebrae superioris, and the four extraocular muscles (superior, medial, inferior rectus, and inferior oblique), and ciliary and sphincter pupillae. Signs include Ptosis, extraocular muscle limitation except for abduction and depression, and anisocoria with a sluggishly reacting pupil. One of the causes of the oculomotor nerve palsy is an abnormal contact of artery or vein with the nerve, so called neurovascular conflict. A neurovascular conflict results in an active nerve dysfunction. We present the case of a young adult male who presented in myasthenic crisis following vaccination for COVID-19 infection with not so classical signs of ocular myasthenia but also had features of bilateral neurovascular conflict on neuroimaging. The etiological role of neurovascular conflict in causing third nerve compression was difficult to establish and could be a incidental finding. There are also no reported cases of precipitation of myasthenia following COVID vaccination.

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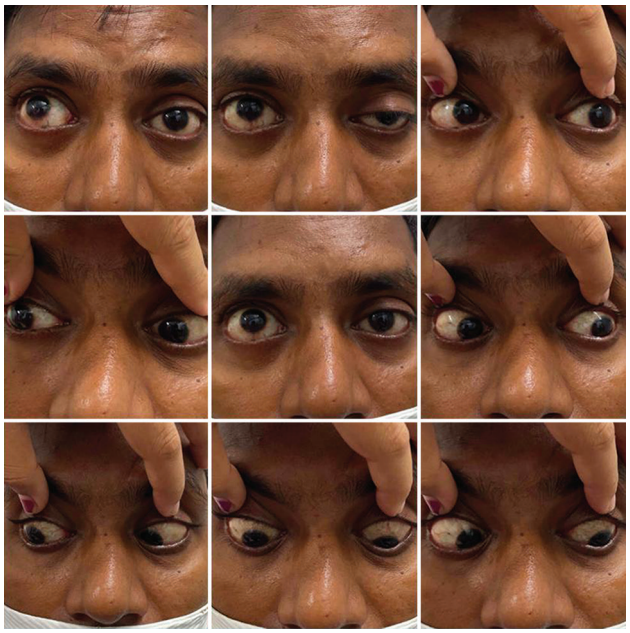
CASE REPORT

A 35-year-old male patient came to the ophthalmology outpatient department with a history of binocular diplopia and drooping of the left upper lid, which was sudden in onset,



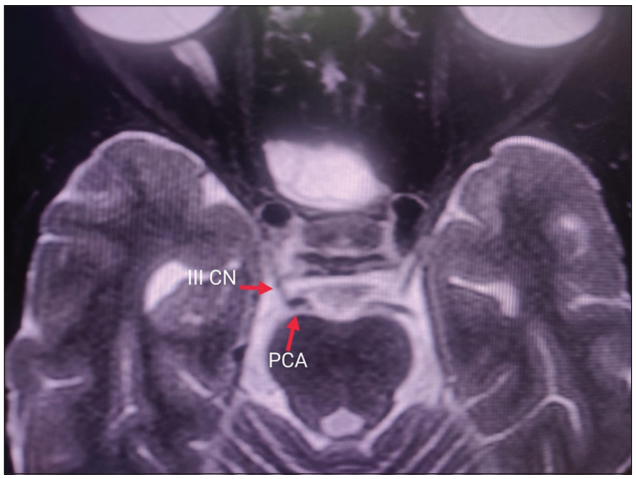
noted 6 weeks ago. The onset was a week after receiving the first dose of covishield vaccine. The patient also complained of dysphagia, slurring of speech, and difficult neck holding. There was no history of any diurnal variation of symptoms, prior squinting, any focal deficits or convulsions, and prior COVID-19 infection. The patient did not report any systemic illness. On ophthalmic examination, the patient's unaided visual acuity was 6/6. Color vision with Ishihara chart was normal in both eyes. Anterior and posterior segment evaluation was unremarkable. Pupils were central, circular, and reacting briskly to light with no relative afferent pupillary defect that there was no anisocoria. Fundus evaluation was normal in both eyes. On diplopia charting, diplopia was found to be more in primary and upgaze.

On strabismus evaluation, Hirschberg's test showed left hypotropia of 15°. The left eye ptosis completely disappeared when the left eye took up fixation on cover test, suggestive of the left eye pseudo ptosis. On an alternate cover test, there was 30 pd hypotropia.

Park's three steps were inconclusive, not pointing to any particular cyclovertical muscle.

Fatigability and Ice Pack Test were equivocal. Extraocular muscles' examination showed both eyes elevation limitation of −4 in the left eye and −2 in the right eye, both eyes adduction and abduction limitation of −1.

On systemic examination, single breath count was <10. Neurological examination revealed a normal sensory and motor system with no cerebellar signs. We had differentials of myasthenia gravis and third nerve palsy. Since the clinical picture and tests were indecisive, we went ahead with neuroimaging. Magnetic resonance imaging brain with orbit showed a bilateral grade 2 neurovascular conflict between posterior cerebral arteries and the third cranial nerve. Constructive interference steady state (CISS) sequences showed indentation and displacement of bilateral 3rd cranial nerve roots by P1 segments of respective posterior cerebral arteries.



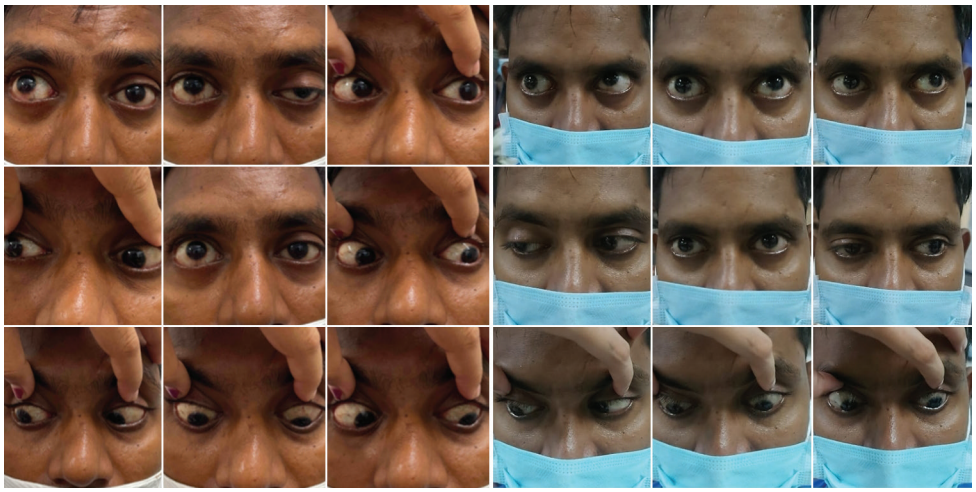
The results of further investigations were as follows. The hemogram was within normal limits. Acetylcholine receptor antibodies were 6.41 nmol/L (<0.4-negative). RNST was again suggestive of post-synaptic neuromuscular junction disorder. HRCT also showed a well-defined soft-tissue density lesion in the prevascular region contiguous with Thymus tissue constant with thymoma again pointing toward MG.

The patient was started on IV immunoglobulins 5 g (16 doses given), oral pyridostigmine 60 mg, oral prednisolone 60 mg in tapering doses, and injection thiamine. The patient was, further, maintained on oral immunosuppressants.

On 2 months follow-up, the patient had undergone thymectomy and HPE biopsy specimen reported Thymoma type B2. The patient showed symptomatic improvement, but had persistent vertical diplopia. Motility examination showed marked improvement in ptosis, elevation, and adduction limitation in the left eye, whereas there was mild ptosis noted in the right eye and no improvement in elevation limitation of the right eye.

DISCUSSION

MG is an autoimmune disease, in which antibodies destroy neuromuscular connections resulting in muscle fatiguability and weakness throughout the day. The ophthalmologist encounters symptoms that mainly include ptosis, diplopia, variable extraocular muscle palsies or incomitant strabismus, and external ophthalmoplegia. It is called a great masquerader due to its variable clinical presentations. Very often, a patient of MG may present to the ophthalmologist with ocular presentation at first progressing toward systemic involvement in the later course of the disease.[5] Depending on the type of



clinical features and the type of antibodies involved, MG can be classified into various subgroups. Each group responds differently to treatment and, hence, carries a prognostic value.[3]

- Early-onset MG: Age at onset <50 years with thymic hyperplasia
- Late-onset MG: Age at onset >50 years with thymic atrophy
- Thymoma-associated MG
- MG with anti-muscle specific kinase (MuSK) antibodies
- Ocular MG: Symptoms only from periocular muscles
- MG with no detectable acetylcholine receptor (AChR) and MuSK antibodies

The diagnosis of MG is mostly clinical. Edrophonium or neostigmine test and ice pack test are diagnostic by observing the improvement in ptosis by 2 mm. The laboratory investigations, such as ACh receptor antibodies, MUSK antibodies, computed tomography thorax to rule out any thymoma, and RNST, aid the clinician in the diagnosis.[4]

Oculomotor nerve palsy is frequently caused by a brainstem infarct, cavernous sinus tumors, and other intracranial lesions. However, neurovascular conflict causing oculomotor nerve palsy is rare.[5]

Non-aneurysmal compression of the oculomotor nerve is a condition rarely reported. The arteries causing vascular compression of the oculomotor nerve causing ophthalmoplegia could be posterior cerebral artery, superior cerebral artery, and posterior communicating artery.[5] Direct vascular compression should be considered in patients with isolated cranial neuropathies.[6]

Different sequences including fast imaging employing steady state, CISS, and balanced-fast field echo are mode of imaging especially sensitive for visualizing the cranial nerves due to high contrast resolution between the cerebrospinal fluid cisterns and nerves. The diagnosis on imaging requires changes in the nerve after contact with the offending vessel.[7]

Thus, addressing the dilemmas in our case of role of COVID vaccination in precipitating MG and whether the neurovascular conflict was causing compression and had a role to play in the relatively suboptimal improvement of the right eye or if it was just an incidental finding which was difficult to discern.

One case has been reported of myasthenic crisis post-vaccination. The proposed pathogenesis was that the patient could not generate an appropriate response to the COVID vaccine as his regulatory cells are defective and COVID vaccine induced a mechanism similar to the cytokine storm.[8]

CONCLUSION

It is necessary to be more pertinent in administering COVID vaccination in known myasthenia patients. It needs more research to validate the role of vaccination in precipitating myasthenia crisis. Neurovascular conflict can be a coincidental finding but definitely confuses the clinician whether they need to be treated is definitely a neurosurgeon prerogative beyond the scope of an ophthalmologist.

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