CASE REPORT

Trigeminal neuralgia in an elderly male patient with serologically proven myasthenia gravis: Co-occurrence or causation?

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Abstract

Trigeminal neuralgia and myasthenia gravis, both are debilitating neurological pathologies causing a significant negative impact on a person's quality of life by virtue of symptoms like unrelenting excruciating pain and muscle weakness respectively. The case was distinct as the elderly male presented with two variable presentations of distinct neurological diseases over a short period. He presented to us with sudden onset ptosis and monocular diplopia followed by sharp stabbing lancinating hemifacial pain around the V1-V2 region two months later. Hereby, trigeminal neuralgia was diagnosed in a case of seropositive ocular myasthenia gravis. Timely and appropriate diagnoses were made with the support of pertinent investigation and imaging and a relevant treatment plan was commenced.

Keywords: Ocular Myasthenia Gravis, Trigeminal Neuralgia, Autoimmune, Facial Pain

Introduction

Myasthenia Gravis (MG) is an autoimmune neuromuscular junction disease characterized by fatigable and fluctuating weakness of skeletal muscles. The triad of ptosis, oculomotor paresis, and orbicularis oculi weakness raises suspicion for the purely Ocular form of Myasthenia Gravis (OMG). Over one-half of patients with MG initially present with isolated ptosis, diplopia or both, and no signs or symptoms of weakness elsewhere [1].

Trigeminal Neuralgia (TN) is a neuropathic pain disorder characterized by sudden, severe excruciating, shock-like pain paroxysms usually on one side of the face at the second and/or third trigeminal branch region [2]. It can be either primary/ idiopathic with unknown precise cause or secondary, which might be associated with some other diseases [3].

Current proposed mechanisms of pain in TN are vascular structure pressing on the trigeminal nerve in its root entry zone or demyelination of trigeminal sensory fibres or compression of the trigeminal nerve by a tumor, a cyst, arteriovenous malformation, physical damage to the nerve due to trauma, a dental or surgical treatment, or infection but these fail to sufficiently explain the total clinical picture [4]. There are not enough studies reporting on the simultaneous occurrence of neuropathic pain disorder and autoimmune diseases or secondary causes of TN.

We hereby report a case of an elderly male who presented to us with sudden onset left-sided ptosis and monocular diplopia followed by sharp stabbing lancinating hemifacial pain around the V1-V2 region two months later.

Our case highlighted a rare and novel co-occurrence or association between the above two pathologies. The case also illustrates how an appropriate and timely diagnosis of these two debilitating conditions was imperative for formulating an optimal management plan and how the patient demonstrated excellent response to first-line treatment of MG and TN.

Case Report

A 66-year-old gentleman, musician by occupation with nil co-morbidities presented with sudden onset left-sided ptosis, a sensation of heaviness around the eyelids and uniocular diplopia. He did not complain of any upper limb, neck or bulbar weakness, fatigability, dysphagia, dysarthria or shortness of breath. On examination, ptosis increased on prolonged up-gaze. Cogan lid twitch and icepack test were positive. No ophthalmoplegia, nystagmus or pupillary abnormalities were noted. Upon further examination, mild Sensorineural Hearing Loss (SNHL) was found in the left ear. Serum anti-Acetylcholine Receptor Antibodies (AChR-Abs) was ordered as MG was high on our index of suspicion.

Serum AChR-Ab was elevated to level of 17.2 nmol/L (normal= 0.4 to 0.5). Thyroid profile was within normal limits. Electrophysiological testing, Electromyography and Nerve Conduction Study (EMG-NCV) and Repetitive Nerve Stimulation (RNS) studies revealed no abnormality. CT scans showed no thymoma or thymic hyperplasia. A diagnosis of OMG was made. The patient was started on oral corticosteroids and pyridostigmine. Patient showed good response on subsequent follow-ups as ptosis was on a resolving trend.

Two months later the patient presented with sharp, electric shock-like and unrelenting facial pain starting at the side of his left nostril, moving up to the forehead and traveling down the nasolabial fold and then back to the angle of the jaw, mostly in the region of V1–V2 distribution for the past 2 weeks increasing in severity and frequency over the last two days. The pain was triggered by washing his face, eating, shaving, paroxysms of cold air and brushing of his teeth.

On examination, mild SNHL was still present and on further questioning, the patient revealed progressive worsening of hearing in left ear for many years. There was no history of dizziness, ataxia, weakness of limbs, fever, chills, rashes on the face, early morning nausea or vomiting, weight loss, jaw claudication, altered taste sensations, lacrimation, conjunctival injection, vision loss or rhinorrhoea.

Before this follow-up visit, he underwent ophthalmological, ENT and dental reviews for his hemifacial pain which were unremarkable. In the context of sudden ptosis, diplopia, insidious onset hearing deficit, ophthalmic complaints followed by new onset facial pain and headache, an MRI angiography and audiogram were ordered.MRI angiography revealed a close association of the left superior cerebellar artery and the trigeminal nerve on the left side and no abnormalities in the brainstem and cerebellum (Fig.1) hence possibilities of migraine, postherpetic neuralgia, Bell's palsy, brain tumour, Multiple Sclerosis (MS), arteriovenous malformation, space-occupying lesion in the brainstem especially cerebellopontine angle tumour which can also be contributing to symptomatic TN were ruled out. An audiogram revealed severe SNHL on the left side. This was attributed to him listening to loud music on

headphones for the majority of his life owing to his occupation. A diagnosis of TN was made and carbamazepine 100 mg twice a day was prescribed, while steroids with pyridostigmine for his OMG were continued.Several weeks later, the patient exhibited significant improvement as diplopia disappeared while ptosis was on a resolving trend. The facial pain though emerging in shorter bouts was gradually improving.

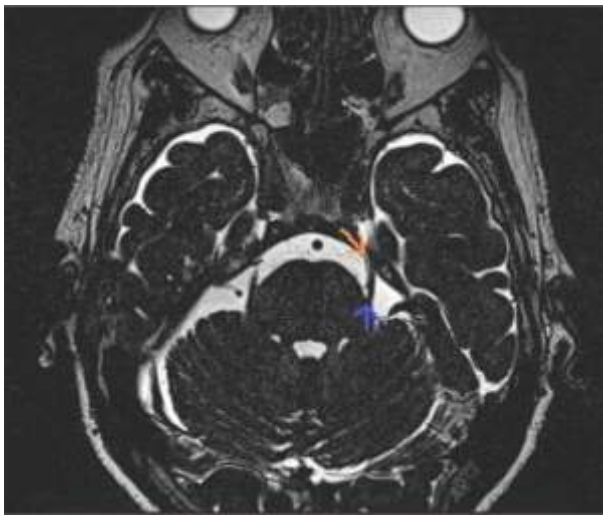

Figure 1: Axial T2 image shows left superior cerebellar artery (orange arrow) abutting the medial aspect of left trigeminal nerve (blue arrow)

Discussion

Several cases and articles have reported co-occurrence of MG and other neurological disorders and association of TN with other autoimmune and connective tissue disorders. A case reported simultaneous findings of serologically proven MGs manifesting as OMG along with pharyngeal-cervical brachial variant of Gullian-Barre syndrome [5].

Hagen *et al*. [6] reported 2 cases of TN in SLE among 81 studied subjects. They also found 26% prevalence of TN in mixed connective tissue diseases like systemic sclerosis, rheumatoid arthritis, dermatomyositis, Sjogren syndrome and 47% prevalence in undifferentiated connective tissue disorder.

Studies have also reported the prevalence of TN in MS patients. Notably, TN affects 15% of individuals before the diagnosis of MS and the most likely postulated mechanism is a demyelinating plaque [7]. OMG affects male more than women and with a unimodal peak in men at around 70 years. Our patient was of nearly the same demographics.

Supplementary testing for thyroid dysfunction is also judicious in patients with MG, since about 4-5% of patients with MG may have concurrent autoimmune thyroid disease [8]. Also, according to Tanovska *et al*. additional testing can be considered in patients with MG to look for concurrent autoimmune disorders including autoimmune thyroid disease, systemic lupus erythematosus and rheumatoid arthritis [9].

The diagnosis of TN is usually based on the characteristic clinical picture, which comprises of key clinical features and physical examination. Several Cochrane systematic reviews and available evidence reveal that carbamazepine is the best-studied treatment and drug of choice for initial and long-term management of TN [10], as prescribed in our patient. There are not enough studies reporting on the simultaneous occurrence of neuropathic pain disorder and autoimmune diseases or secondary causes of TN. To the best of our knowledge and upon searching the literature, co-occurrence of OMG and TN has not been documented before and the prevalence of TN in autoimmune diseases is poorly understood.

Although there was evidence of slight neuro-vascular compression which may act as a mechanism behind TN in our patient, we could not determine if neurovascular compression was the only mechanism or a concurring mechanism secondary to the autoimmune disease.

Conclusion

We highlight a rare case of TN in a patient with recently diagnosed AChR-Ab positive OMG to raise awareness among healthcare practitioners that TN may have an association with autoimmune pathology and it can be manifested before, after or at the time of diagnosis of MG. This report also emphasizes the importance among clinicians to have a thorough awareness and understanding of diverse neurological conditions which can overlap or occur simultaneously to facilitate accurate diagnosis, effective management and avoid unnecessary and possible harmful procedures and delayed treatment.

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