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Case Reports & Case Series

Bilateral trigeminal neuralgia in association with a possible Charcot-Marie-Tooth disease: A case report

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ABSTRACT

Introduction: Trigeminal neuralgia is described by the International Headache Classification 3rd edition as a chronic, painful, and sporadic condition characterized by electric shock-like hemifacial pain. The initial management of trigeminal neuralgia is with medication, if there is persistence of pain, surgical management is the gold standard. Microvascular decompression is a non-destructive technique that improves pain in 98% of cases. In the literature, only 5 cases of CMT-associated trigeminal neuralgia that were surgically managed with microvascular decompression have been published.

Case report: A 58-year-old male patient with a personal pathological history of CMT diagnosed at 34 years of age, initially presented with incapacitating right hemifacial electric shock-like pain. The pain had been tolerated with pharmacological management for 5 years. Since the patient had a partial response to carbamazepine and the pain increased suddenly, a trigeminal nerve decompression was performed. Surgical findings were arachnoid adhesions and contact with superior cerebellar artery, the adhesions were liberated and Teflon was placed in the contact area. The patient had no further pain.

Four years later, the patient presented again with a similar clinical picture characterized by left hemifacial pain with no control of pain with medication therefore, surgical management was decided. This time around, the surgical findings were adhesions between the trigeminal nerve and the anteroinferior cerebellar artery, which were liberated and teflon was placed between the nerve and the artery. After surgery, the patient had no pain.

Discussion: Very few cases of surgical management of trigeminal neuralgia in association with CMT have been reported. The surgical findings, in this case, showed adhesions bilaterally, which could be another factor contributing to the bilateral neuralgia. Microvascular decompression has proven to be a good option for surgical management.

Conclusion: The surgical management for trigeminal neuralgia in association with CMT is still an understudied subject, and yet microvascular decompression is a suitable technique for this condition, because it may resolve the vascular decompression by the placement of teflon and may also liberate adhesions if present.

1. Introduction

Trigeminal neuralgia is described by the International Headache Classification 3rd edition as a chronic, painful, and sporadic condition characterized by electric shock-like hemifacial pain that is triggered by innocuous stimuli in the trigeminal nerve territory in one or more of its three branches. Trigeminal neuralgia has a prevalence of 3 to 5 per 100,000 people [1,2]. The initial management of trigeminal neuralgia is with medication, yet when medical management fails to control pain, surgical management is the gold standard. Microvascular decompression is a non-destructive technique that improves pain in 98% of cases [3].

Charcot-Marie-Tooth (CMT) is the most common hereditary neurological disease, and still, it is a rare entity with less than 200,000 cases reported per year in the United States [4]. It is characterized by motor and sensory neuropathy with diminished nerve conduction. Trigeminal neuralgia has been associated with CMT in previous reports though this association has been attributed to the primary neural dysfunction inherent to the disease, without consideration of a vascular component [5].

The influence of premature atherosclerotic changes in blood vessels as an etiology of trigeminal neuralgia has been associated with

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biochemical changes in CMT; other authors have investigated alteration in calcium channels, a possible association with proenkephalin A, and vascular or skull base abnormalities as the possible causes [3,5,6]. It has also been suggested that a focal demyelination because of the primary dysfunction in CMT, makes a nerve more susceptible to irritation by neurovascular compression [5,7]. In the literature, only 5 cases of CMT-associated trigeminal neuralgia that were surgically managed with microvascular decompression have been published [8,9].

Here, we report a patient who presented with CMT-associated trigeminal neuralgia twice, 4 years apart, and was treated with microvascular decompression.

2. Case report

A 58-year-old male patient initially presented with incapacitating right hemifacial electric shock-like pain. The patient had a 6-year history of the pain, which was triggered by stimuli such as touching, chewing, and brushing teeth. The pain had been tolerated with pharmacological management for 5 years with partial response to carbamazepine 200 mg every 8 h and pregabalin 300 mg every 12 h; however, in the past year the pain had become incapacitating and since the patient had no economical resources to pay for the medical treatment, a surgical approach was decided.

The patient had the following personal pathological history: no known family history of polyneuropathy. CMT was diagnosed at 34 years of age, no molecular diagnosis was possible because of economical reasons, however he begun at 34 years of age with distal muscular atrophy in superior extremities with predominance in thenar, hypothenar, distal extensors and flexors, in lower extremities predominance in anterior tibial muscles, soleum and gastrocneminum, also fasciculations were noted as well as progressive diminishing of strength in all 4 extremities. Because of the chronic disease he developed cavus foot, with a high arched foot and hammer toes, as well as sensitive alterations such as glove and socking pattern paraesthesia, bilateral neurosensitive hearing loss with left predominance, and left eye amaurosis.

He had a hernioplasty performed at 11 years of age, pesticide contact at the age of 11, and chronic alcoholism since the age of 25 years.

During clinical examination, the patient presented no alteration in superior mental function. However, the following were noted: left eye amaurosis with ipsilateral papillary atrophy, bilateral hypoacusia, right maxillary and mandibular hyperesthesia (V2, V3), strength: 5/5 superior extremities and 4/5 inferior extremities, and diminished tropism and tone predominant in anterior tibial muscles, soleus, and gastrocnemius, with hypoesthesia in stocking-glove pattern, and ataxia.

An electromyography and a nervous conduction velocity test were performed with results compatible with a sensory-motor polyneuropathy characterized by axonal degeneration. A head MRI was also performed which sowed no evidence of structural or vascular lesions. Since the patient had a partial response to carbamazepine and couldn't afford because of economical reasons a higher dose of medical treatment and the pain increased suddenly, a trigeminal nerve decompression was considered. Surgical findings were arachnoid adhesions at the pontocerebellar angle and adhesions of the trigeminal nerve to the superior cerebellar artery. The adhesions were liberated and teflon was placed in the contact area. During the postoperative evaluation, the patient had no more pain but presented with V2-V3 paresthesia that diminished with time. No further medication was needed.

Four years later, the patient presented again with a similar clinical picture characterized by left hemifacial pain of V3 predominance, with no control of pain with medication in the past year (gabapentin 300 mg every 12 h and carbamazepine 200 mg every 8 h); therefore, surgical management was decided. This time around, the surgical findings were adhesions between the trigeminal nerve and the anteroinferior cerebellar artery, which were liberated and teflon was placed between the nerve and the artery. After surgery, the patient had no pain and needed no further medication.

Written informed consent was obtained from the participant for the publication of this case report.

3. Discussion

Very few cases of surgical management of trigeminal neuralgia in association with CMT have been reported. Different theories have been suggested on the etiology of this association: a focal demyelination that results in a more susceptible nerve, atherosclerotic changes in the vessels, a possible association with proenkephalin A, and vascular or skull base abnormalities. The surgical findings, in this case, showed adhesions bilaterally, which could be another factor contributing to the bilateral neuralgia. Microvascular decompression has proven to be a good option for surgical management in most cases because, during surgery, liberation of adhesions, as well as vascular decompression, can be achieved. Our patient presented with bilateral trigeminal neuralgia twice, 4 years apart, and in both instances, adhesions were found between the vessel and the nerve. Since this is a rare association, a statistical analysis would be difficult to perform because of the small number of reported cases. Yet, if more cases are published, we can know more about this association and determine the best way to treat it.

4. Concluding remarks

The surgical management for trigeminal neuralgia in association with CMT is still an understudied subject, and yet microvascular decompression is a suitable technique for this condition, because it may resolve the vascular decompression by the placement of teflon and may also liberate adhesions if present. More reports of this rare condition are needed for furthering our knowledge and for identifying the best treatment option.

Author contributions statement

RR-G is the tutor of this project. CC-M was in charge of the project analysis.NP-C and MCB were in charge of data recollection.

Declaration of Competing Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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