An association between TGN and CMT disease has already been reported in the literature. In patients with CMT disease, it is possible that TGN may be causally related to the presence of an intrinsic neural abnormality, but relevant vascular compression has not been previously considered. In the present report, three cases of TGN and coexistent CMT disease are described in which microvascular decompression was used as part of the treatment.

It seems vitally important that the possibility of vascular compression of the trigeminal nerve be actively excluded in patients with TGN and a coexistent neurological condition that might otherwise explain the cause of the pain. This is necessary to expand the options for treatment. Magnetic resonance tomographic angiography provides an accurate means for making this assessment preoperatively. If vascular compression is found, decompression is possible, providing pain relief without resulting in neurological deficit.

Case Reports

Case 1

History. This 47-year-old man with CMT disease presented with a 6-year history of episodic left-sided TGN, which had become intolerable over the previous 4 months. The attacks of pain were triggered by talking and touching the left lip. Radiofrequency coagulation, peripheral nerve blocks, and increasing doses of carbamazepine had proven useful; however, his current regimen of 1000 mg carbamazepine per day had failed to control the pain. Since late childhood, the patient had been noted to suffer from CMT disease (peroneal muscular dystrophy). The disease had appeared to be slowly progressive, and he had remained otherwise asymptomatic from the condition.

Examination. Previously the patient had successfully undergone exploratory surgery of the right posterior fossa and decompression for right-sided TGN. Examination revealed anesthesia in the distribution of the ophthalmic and mandibular divisions of the left trigeminal nerve, which was consistent with the fact that the man had undergone previous ablative procedures. No other cranial nerve deficit was evident. There was marked symmetrical distal muscle wasting associated with generalized areflexia, and a reduction in both superficial sensation and proprioception.

Magnetic resonance tomographic angiography, a technique developed in our unit for demonstrating neurovascular compression in TGN, was performed. Gadolinium-enhanced imaging revealed that there was a large vein compressing the nerve at the REZ.

Operation and Postoperative Course. During exploration of the posterior fossa, the MRT angiographic findings were confirmed. The vein was dissected from the nerve, coagulated, and divided. On recovery from the anesthetic,
the patient experienced complete pain relief. He remains pain free 4 years later.

Case 2

History. This 28-year-old man with CMT disease presented with bilateral TGN. He is the son of the patient described in Case 1. On the right side, his pain was located in the distribution of the maxillary and mandibular divisions of the trigeminal nerve and had been present for 4 years. The left-sided symptoms had been present for only 1 year and involved the mandibular division alone. Symptom management consisted of a regimen of 600 mg carbamazepine daily, although at the time of presentation this was proving ineffective.

Examination. On examination the patient demonstrated muscle wasting below both knees, loss of ankle and knee reflexes, and altered sensation in response to both pinprick and light touch on his feet. He displayed no neurological deficits of the trigeminal nerve. Magnetic resonance tomographic angiography was performed and demonstrated bilateral trigeminal compression. On the patient’s left side, the compression was caused by the superior cerebellar artery, whereas on the right side it was due to a large vein (Fig. 1).

Operation. The patient underwent simultaneous bilateral posterior fossa surgical explorations, which confirmed the findings of the MRT angiography. On the right side, the vein was coagulated and divided. On the left side, the superior cerebellar artery was displaced off the nerve by using a sponge.

Postoperative Course and Treatment. The patient experienced immediate postoperative pain relief following the procedures; however, he experienced a recurrence of symptoms on the right side 2 years postsurgery. Repeated MRT angiography demonstrated no evidence of continuing vascular compression. He underwent an additional exploratory procedure on the right side; after a partial neurotomy was performed, he obtained complete pain relief. The patient has since suffered from transient mild TGN on the left side, the symptoms of which have been easily controlled with small doses of carbamazepine.

Case 3

History. This 31-year-old man with CMT disease presented with a 9-year history of right-sided TGN in the maxillary distribution. The patient’s episodes of pain were exacerbated by eating, brushing his teeth, and cold wind. He experienced periods of full resolution of symptoms for up to 8 months at a time. Carbamazepine was partially effective in relieving the pain; however, the maximum dose the patient could tolerate without experiencing excessive drowsiness was 1200 mg. The pain was greatly reduced by induction of a local anesthetic block. A diagnosis of CMT disease had been made when the patient was 10 years of age. He had scoliosis and experienced gradually progressive weakness in his legs with unsteadiness of gait, which prevented him from running.

Examination. On examination all cranial nerve functions were normal. The patient clearly demonstrated distal muscle wasting of the legs, with bilateral pes cavus, bilateral foot drop, and absence of lower-limb reflexes. All sensory modalities were reduced below the groin. Magnetic resonance tomographic angiography revealed vascular compression of the right trigeminal nerve at the REZ, which following gadolinium enhancement was identified as being caused by a vein.

A surgical exploration of the right posterior fossa confirmed venous compression of the trigeminal nerve at the REZ. The vein was coagulated and divided.

Postoperative Course. Immediate postoperative pain relief was experienced by the patient and has been maintained for 2 years. It was possible to demonstrate a mild deficit of pinprick sensation in the left mandibular nerve distribution, presumably due to heating at the time of coagulation of the vein.

Discussion

The cause of TGN must be multifactorial, although in the majority of cases, there is vascular compression of the nerve at the REZ.\textsuperscript{1,8,10,12,16} There is a well-known association between TGN and other neurological conditions, notably MS.\textsuperscript{14} There have been 15 previously recorded cases of TGN occurring in patients with CMT disease.\textsuperscript{4,13} In most of these cases, it was assumed that the pain was caused by the underlying neural abnormality.
Trigeminal neuralgia in Charcot-Marie-Tooth disease

Fromm and associates⁶ have emphasized the possibility of both peripheral and central mechanisms for the pain. They suggest that chronic irritation of the nerve leads to ectopic action potentials within the nerve and failure of segmental inhibitions within the trigeminal nucleus. This theory satisfactorily explains the cause of pain in patients with neural compression that is either vascular in origin or due to tumor. It also explains pain in cases of MS, in which a plaque affecting the trigeminal nerve centrally could lead to spontaneous discharges of pain fiber in the trigeminothalamic tract. The causative plaque is usually seen to lie in the area of the REZ of the trigeminal nerve on magnetic resonance imaging.¹⁴ At our institution, observations involving seven patients who had both TGN and MS revealed that six of the patients had demonstrable neurovascular compression of the trigeminal nerve. We believe that vascular compression, although not uncommon in an asymptomatic population reported in the study (8%),¹³ is perhaps more likely to cause TGN in the presence of a neural abnormality such as demyelination, and this might even increase the incidence of TGN in patients with MS.¹⁴

The reason for the increased incidence of TGN observed in patients with CMT disease remains unclear. Electrophysiological studies and clinical observations have revealed lesions suggestive of focal demyelination.⁵,⁶ It is suggested that TGN is associated with CMT disease for the same reasons as is MS; that is, partial demyelination resulting in myelin sheath disruption, making the neuron more susceptible to vascular compression and irritation.

Most surgical techniques for treating TGN are directed at producing symptomatic relief by damaging the trigeminal nerve.⁸ As a result of these procedures, there is obligatory sensory loss and the risk of motor deficit. Microvascular decompression produces symptomatic relief without resulting in this neurological deficit⁸,¹¹ and offers at least the possibility of cure. Until recently, surgery was performed on an exploratory basis without preoperative identification of neurovascular compression. Understandably, few neurosurgeons were prepared to perform exploratory surgery in patients with TGN and a coexistent neurological abnormality. Magnetic resonance tomographic angiography provides the means to demonstrate neurovascular compression preoperatively and, therefore, raises the option of microvascular decompression.

Conclusions

Patients with TGN who also have an established neurological disease such as CMT disease may still have vascular compression at the REZ and may benefit from microvascular decompression. When they suffer from bilateral TGN, this nonablative option becomes even more important. Magnetic resonance tomographic angiography provides a reliable preoperative method of identifying this option.

References


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