Management of Trigeminal Neuralgia

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Symptoms and signs

Trigeminal neuralgia is a relatively common pain condition. The diagnosis is based on the typical history of episodic electric pain in the distribution of the trigeminal nerve. Touching the face or eating can sometimes provoke an attack. There is commonly a trigger area on the face or within the mouth that stimulation of it will provoke a neuralgic attack. Because of the pain and apprehension, many patients do not clean their face or shave their beard on the affected side. One can often make a spot diagnosis by noticing the dirt and beard distribution over the face after hearing the history.

Causes of trigeminal neuralgia

In most patients, the cause of the pain is thought to be related to the vascular compression syndrome. Peter Jannetta (Jannetta, McLaughlin, and Casey) theorised the vascular compression syndrome that a vessel loop compressing on the root entrance zone of the trigeminal nerve can cause trigeminal neuralgia. With chronic vessel compression, segmental demyelination (Love and Coakham) occurs in the trigeminal nerve. Incoming wave of sensory input from receptors in the face travelling through the trigeminal nerve partially reflects back at the area of demyelination. When the frequency of the incoming wave and the reflected wave comes into resonance, a big volley of wave is generated and causes a sudden surge of pain. Although the vascular compression theory is popular, it cannot account for all phenomena associated with trigeminal neuralgia. Many patients with trigeminal neuralgia do not have a culprit vessel.

Apart from vascular compression, other pathologies such as tumour compression or intrinsic demyelination problems like multiple sclerosis can also produce trigeminal neuralgia.

Clinical workup

Although the diagnosis of trigeminal neuralgia is largely a clinical diagnosis, I believe some kind of imaging study is warranted in the present era. MRI is commonly used to examine the trigeminal nerve. The main use of imaging study is to exclude unusual pathologies, such as tumour. In the MRI, one needs to examine the whole course of the trigeminal nerve from the brainstem to the skull base. With modern MR technique, one often can identify the culprit vessel loop.(Akimoto et al.)

Medical treatment

In about 80% of patients, the pain can be readily managed with medication. The most useful drug is carbamazepine (Tegretol). Its effect is so specific that it can be used as a therapeutic test. If the pain can be relieved by carbamazepine, one can be quite certain that the diagnosis is trigeminal neuralgia. When one starts treatment with carbamazapine, one needs to build up the dose slowly. Among other side effects, the common problems of dizziness and unsteadiness in the beginning of treatment often scare patients off the medication and claim themselves intolerable to this most useful medication for trigeminal neuralgia. I often warn patients about this particular side effect upfront and teach them to titrate the dose themselves. I ask my patients to start with 100mg a day. If they feel well the next day, they add on another 100mg a day. If there is symptom of unsteadiness, they should stop increasing the dose or back down to the previous dose for a few days before stepping up again. I usually ask them to step up the dose till the pain is under control, or the dose has reached 600mg a day. Then I will review them again for the need of a higher dose or second medication. One can push carbamazapine to 1200mg a day.

There are two other useful medications for trigeminal neuralgia, viz. gabapentin (Neurontin) and pregabalin (Lyrica). If the patient fails or is intolerable to these medications, that will be the time to consider surgery for the condition.

Options of surgery

Surgery can be divided into two types, viz. restorative and destructive. The best surgical treatment is to remove the cause of pain. If it is due to vessel loop or tumour compression, surgical decompression should be the procedure of choice.

When no cause is identified, or the cause is difficult to deal with, one can interrupt the nerve so that it cannot conduct the pain signal to the brain.

There are three common surgical options for trigeminal neuralgia.
1. microvascular decompression
2. percutaneous ablative procedure
3. radiosurgery
Microvascular decompression and selective rhizotomy

In the treatment of pain conditions, the best option is to remove the cause of pain and preserve all sensations. This is the attractive point about microvascular decompression. In a young patient who presents with typical trigeminal neuralgia and MRI showing an arterial loop on the corresponding trigeminal nerve, the best option is to decompress the trigeminal nerve and cushion it off the culprit artery. The result of microvascular decompression is generally very good, with excellent pain control rate of about 70-90%. The risk of mortality associated with this operation is less than 1%. The commonest side effect is hearing loss related to retraction injury of VIII nerve. With the use of intra-operative evoked potential monitor, such complication can now be controlled at 1% (Barker et al.).

However, pre-operative MRI sometimes does not show an offending vessel. There is then an uncertainty about the efficacy of microvascular decompression. There may be nothing to decompress or at the time of operation, if proponents of this operation, they will argue that veins and small arteries not easily discernable on MRI can also cause trigeminal neuralgia. They emphasise the need of careful exploration under the microscope. Having said that, I must admit that I have encountered cases without a culprit vessel on exploration.

I now conceptually look upon the operation as an exploratory procedure. I tell my patients that I start the operation with examination of the whole course of trigeminal nerve in the subarachnoid space. If I am convinced that a culprit vessel is compressing on the trigeminal nerve, usually with indentation on the nerve, I will cushion the artery off the nerve. Or if the culprit vessel is a vein, I will coagulate and remove a segment of the vein. If I am not convinced of a culprit vessel, I will do a selective rhizotomy at the root entrance zone. At this level, the trigeminal nerve has a constant topographic pattern. The ophthalmic division (V1) is located at the most superior and medial aspect. The mandibular division (V3) is located at the most inferior and lateral aspect. The maxillary division (V2) lies in the middle. For most patients, trigeminal neuralgia involves the V2 and V3 portions of the nerve. I normally divide the nerve by 50% starting from the lateral inferior aspect and therefore sparing the ophthalmic division.

Percutaneous ablative procedure

There are three common percutaneous techniques in lesioning the trigeminal nerve. All of them involve placement of a cannula through the foramen ovale to access the trigeminal ganglion. Among the three, viz. radiofrequency lesioning, chemical lesioning with glycerol, and balloon compression, I prefer the radiofrequency lesioning technique for its selective and controlled characteristics. It also offers the highest rates of complete pain relief (Lopez, Hamlyn, and Zakrzewska).

Selective rhizotomy of trigeminal nerve with radiofrequency

The procedure can be done as a day case. A cannula is placed through the foramen ovale based on anatomical landmarks and then confirmed with X-ray. The procedure requires intermittent short-lived deep sedation during placement of the needle and lesioning. The position of the needle is adjusted according to the patient’s response during electrical stimulation. With fine-tuning of temperature and duration of lesioning, one can burn off the thinly myelinated pain fibre and preserve the thickly myelinated fibres that subserve touch sensation and motor function. Being an ablative procedure, the drawback of it is loss of sensation after the procedure. Most of the time, if the neuralgia is severe pre-operatively, patients will usually accept the hypeaesthesia after the operation. In order not to over lesion a nerve, patients have to accept a possible recurrence after the procedure. However, it is not a problem to re-lesion the nerve when the pain recurs. The worst outcome from a lesioning procedure is the creation of analgesia dolorosa, a deafferentation syndrome without very effective treatment.

Radiosurgery

Radiosurgery has recently been established as an effective treatment for trigeminal neuralgia (Gorgulho and De Salles). The largest body of literature is related to gamma knife radiosurgery (Regis et al.). Linac based X-knife system (Richards et al.) and cyberknife (Lim et al.) have also been used to treat trigeminal neuralgia effectively.

A high dose of 70-90 Gy focused radiation is delivered to the subarachnoid portion of the trigeminal nerve. The success of radiosurgery demands a very accurate stereotactic system because the target is small. In a recent report by Regis (Regis et al.), the control rate is up to 85%, with a complication rate of 6% facial paresthesia and 4% hypesthesia.

Summary

Trigeminal neuralgia is one of the readily treatable pain condition. Most patients can be managed with medications (carbamazepine, gabapentin and pregabalin). However, if the pain is not controlled well with medication or patients cannot tolerate the side effect of medication, surgical management should be employed early. Microvascular decompression remains the best approach for treatment of trigeminal neuralgia especially if one can spot a culprit vessel loop on MRI. Percutaneous ablative procedure is very useful to control the pain if the pain is at the V2, V3 region. It is particularly attractive for old patients who cannot tolerate a craniotomy. Radiosurgery is a promising option. I believe it is a good choice for V1 neuralgia in patients who are too frail for MVD.

References