

Review Article

Management of Trigeminal Neuralgia- An Update

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ABSTRACT:

Trigeminal neuralgia is an inflammation of the trigeminal nerve causing extreme pain and muscle spasms in the face. In general, the first line of treatment for trigeminal neuralgia is medical. For the majority of patients, these medications are quite effective. If the patient fails or is intolerable to these medications, that will be the time to consider surgery for the condition. This article aims to provide overview about the plans available for the treatment of trigeminal neuralgia.

Keywords: Trigeminal Neuralgia, Atypical Facial Pain, Decompression

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Introduction: Trigeminal neuralgia is a very peculiar disease. The pain, also known as “tic douloureux”, is paroxystic and very severe. It is characterized by brief paroxysms of pain, limited to the facial distribution of the trigeminal nerve and precipitated by stimuli to sensory endings in the trigeminal receptive area.¹ It is an inflammation of the trigeminal nerve causing extreme pain and muscle spasms in the face.²

Pathogenesis: The etiology of trigeminal neuralgia has been established over the last century. Observations by early neurosurgeons Cushing, Dandy and Gardner introduced the idea that trigeminal neuralgia may be caused by blood vessels contacting with the trigeminal nerve root, as seen at operation seeking to relieve trigeminal neuralgia pain by cutting the nerve. The formalization of this

neurovascular compression concept and introduction of effective neurosurgical alleviation of the culprit irritation by microvascular decompression surgery was pioneered by Peter J. Janetta.³ About two percent of trigeminal neuralgia patients harbor a tumor or some other sort of mass compressing the trigeminal nerve. Some of the most common causes are believed to be: **Vascular compression** in which a blood vessel causes compression on the trigeminal nerve in the region very close to the brain stem called the **root entry zone**.

Multiple sclerosis (MS): While trigeminal neuralgia is usually found in patients over the age of 50, multiple sclerosis patients are usually younger than the average TN patient and may have bilateral (two sided) facial pain. Every young person with trigeminal neuralgia does not necessarily have MS.⁴

Diagnostic criteria: Trigeminal neuralgia is a clinical diagnosis. The key feature is a sudden and severe lancinating pain, which usually lasts from a few seconds to two minutes, within the trigeminal nerve distribution, typically the maxillary or mandibular branches. The pain is often evoked by trivial stimulation of appropriately named “trigger zones.”

Occasionally the pain is so severe that it prevents eating or drinking. The nerves affected are usually stereotyped for a particular patient and lie within the sensory distribution of the trigeminal nerve. Box 1 lists the diagnostic criteria for the classic form of the disease.

In many cases the pain does not fit these criteria exactly because of a persistent ache between paroxysms or mild sensory loss. Such disease has been labelled as “atypical”⁵ or “mixed” trigeminal neuralgia.⁶ Patients with atypical disease are more likely to have symptomatic rather than idiopathic disease, and they are often more refractory to treatment than those with classic trigeminal neuralgia.⁷ Atypical trigeminal neuralgia should not be confused with atypical facial pain.⁸

There is no specific test that can identify trigeminal neuralgia, so the best method of diagnosis is to obtain a good history from the patient. A physician experienced in the management of trigeminal neuralgia can often make the appropriate diagnosis in a single interview.⁴

Box 1: Diagnostic criteria for classical trigeminal neuralgia⁸

- Paroxysmal attacks of pain lasting from a fraction of a second to two minutes that affect one or more divisions of the trigeminal nerve
- Pain has at least one of the following characteristics: intense, sharp, superficial, or stabbing; precipitated from trigger areas or by trigger factors
- Attacks are similar in individual patients
- No neurological deficit is clinically evident
- Not attributed to another disorder

Treatment

In general, the first line of treatment for trigeminal neuralgia is medical.

Anticonvulsants are frequently used for medical treatment of trigeminal neuralgia. For the majority of patients, these medications are quite effective.⁴

Box 2: Evidence that vascular compression commonly causes trigeminal neuralgia⁸

- An aberrant loop of artery, or less commonly vein, is found to be compressing the root entry zone of the trigeminal nerve in 80-90% of patients at surgery
- The trigeminal nerve is demyelinated next to the compressing vessel
- Eliminating the compression by surgery provides long term relief in most patients
- Intraoperative assessments report immediate improvement in trigeminal conduction on decompression
- Sensory function recovers after decompression
- Other causes, such as compression by tumours or the demyelinating plaques of multiple sclerosis, produce similar lesions of the root entry zone of the trigeminal nerve

If the patient fails or is intolerable to these medications, that will be the time to consider surgery for the condition. 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.⁹ Treatment begins with 100 to 200 mg two or three times daily. Doses should be increased very progressively and titrated to the severity of the patient’s pain. Serum level is a useful way of monitoring treatment (6 to 12.5 ug/ml). In some cases a maintenance dosage of 200 mg or 400 mg per day is sufficient to keep the patient pain-free. With appropriate adjustments of dosage, pain can be controlled initially in about 75% of patients.¹ If carbamazepine has adverse effects, oxcarbazepine is a prodrug of carbamazepine that is often better tolerated; it provides a logical,¹⁰ if largely unproved,¹¹ alternative when carbamazepine has provided pain relief but has had unacceptable adverse effects. The risk of allergic crossreactivity between carbamazepine and oxcarbazepine is about

25%, so oxcarbazepine is best avoided in carbamazepine allergy.⁸ There are two other useful medications for trigeminal neuralgia, viz. gabapentin (Neurontin) and pregabalin (Lyrica).⁹

Options of Surgery: Two types of surgical procedure are available:

- Microvascular decompression, where the posterior fossa is explored and the compressing vessel and trigeminal nerve root are separated
- Ablative treatments that lesion the trigeminal nerve in different ways⁸

Microvascular Decompression: 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.⁹ It is the surgery to gently reroute the blood vessel from compressing the trigeminal nerve by padding the vessel with a sponge. A 1 inch opening is made in the skull behind the ear, called craniotomy. This opening exposes the trigeminal nerve at its connection with the brainstem. A blood vessel (occasionally a tumor) is often found compressing the nerve. After the nerve is freed from compression, it is protected with a small teflon sponge.² 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.^{9,12} Microvascular decompression has the best chance of long term pain relief, with a very low risk of facial sensory loss and other minor complications; however, it has a small risk of death (around 0.4%).¹³

Trigeminal Glycerol Rhizolysis (TGR): A procedure done in the hospital operating room under monitored anesthesia. After being properly sedated, patient will be placed on the operating table and a special x-ray device will be used to help the neurosurgeon insert the needle. The needle is advanced until it reaches a small pocket

of fluid surrounding the trigeminal nerve. A special contrast (dye) is then injected which can be easily seen on the x-ray, and this determines the volume of fluid around the trigeminal nerve. In this way, the surgeon can calculate the exact amount of glycerol that needs to be given. This is an important part of the procedure and neurosurgeons find it an extremely important part of the process to avoid giving an overdose of the glycerol. Once the glycerol is measured and injected into this pocket, the needle is removed.⁴

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 hypaesthesia 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.⁹

Gamma Knife Stereotactic Radiosurgery: Gamma knife stereotactic radiosurgery has recently been approved by the UK's National Institute for Health and Clinical Excellence for treatment of trigeminal neuralgia, but access to this treatment is limited.¹⁴ Gamma Knife treatment is done under the leadership of a

neurosurgeon, a team of physicists - doctors specializing in radiation and nurses. Gamma Knife was pioneered in Stockholm, Sweden by Professor Lars Leksell and has been in use for more than thirty-five years. It is actually not a knife at all but is a term used for 201 individual beams of focused radiation that all converge on a selected target. While each individual beam is not very effective, the combination of all the beams causes a biological and structural change in the targeted tissue. Trigeminal neuralgia is just one of the indications for Gamma Knife, which is also used for brain tumors.⁴

Summary: The best choice for any individual patient should provide the maximum potential for long-term pain relief with the lowest risk of procedure-related side effects and complications. Medical treatment remains the first step and we can rely on very efficient drugs. However, these drugs are not devoid of side-effects and often lose their efficacy with time. Microvascular decompression has the best chance of long term pain relief, with a very low risk of facial sensory loss and other minor complications. It 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.

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