

TRIGEMINAL NEURALGIA

Introduction

Also known as ‘tic douloureux’, this is a common type of neuralgia (nerve pain) that is associated with agonizing facial pain that makes it difficult to talk or eat. The pain may be severe enough to cause spasms of the facial muscles (i.e. tic).

Additional extreme distress may be associated with the recurring fear that the pain may return at any time, with resultant reduced quality of life.

Much can be done to help those afflicted with trigeminal neuralgia.

Trigeminal neuralgia is a diagnosis based chiefly upon talking with the patient and obtaining a detailed history, thus good outcomes are based upon expertise in diagnosis. Once correctly diagnosed, treatment options include medication and neurosurgical procedures such as a microvascular decompression (MVD), needle rhizotomy (e.g. glycerol) and stereotactic radiosurgery (SRS).

What are the symptoms of trigeminal neuralgia?

- Brief episodes of pain lasting from less than a second to a couple of minutes.
- Often the attacks come in waves, with episodes lasting days to weeks then settling down for a period before returning.
- Pain is usually stabbing, shooting, sharp or “like electricity”.
- Pain is triggered by light facial touch such as talking, eating, brushing teeth, wind, etc
- Pain is located in the face, most often the cheek, lips or jaw, but may also involve the side of the nose, inside of the mouth, eye or forehead.
- Pain is usually confined to one side of the face, but very occasionally be on both sides.
- Pain usually initially responds to carbamazepine (Tegretol) or oxcarbazepine (Trileptal).

Are there different kinds of trigeminal neuralgia?

The most accepted classification is:

- Trigeminal neuralgia type 1 (TN1): classical form where episodic lancinating pain predominates, also known as “typical TN”.
- Trigeminal neuralgia type 2 (TN2): atypical form where pain is aching, throbbing or burning and constant for >50% of the time, also known as “atypical TN”.
- Trigeminal neuropathic pain (TNP): pain related to unintentional injury to the nerve from facial trauma, ENT surgery, oral surgery, posterior fossa surgery or stroke.
- Trigeminal deafferentation pain (TDP): pain related to intentional injury to the nerve such as rhizotomy, neurectomy, gangliolysis or other denervating procedure.
- Symptomatic trigeminal neuralgia (STN): pain associated with multiple sclerosis.
- Postherpetic neuralgia (PHN): pain following an outbreak of facial herpes zoster.
- Atypical facial pain (AFP): pain predominantly having a psychological rather than physiological origin.

What are the parts (divisions) of the trigeminal nerve?

Hence the name, the trigeminal nerve has three parts or divisions, and is the fifth of twelve pairs of nerves that supply the face and head. The first (ophthalmic or V1) supplies sensation to the forehead, eye and nose. The second (maxillary or V2) supplies the cheek, side of the nose, upper lip / teeth / gums. The third (mandibular or V3) supplies the jaw, lower lip / teeth / gums and tongue. Pain most commonly occurs in the second and third distributions, but may involve any of these. If the first division is involved, it tends to include the second as well. If pain is limited to the first, often it is not trigeminal neuralgia.

What causes trigeminal neuralgia?

The trigeminal nerve is responsible for carrying sensation from the face to the brain. Patients with trigeminal neuralgia have abnormal 'wiring' of the nerve causing 'cross-talk' between the parts of the nerve that convey normal non-painful sensation with those that carry painful stimuli. This is called 'ephaptic' transmission. Often this is caused by a blood vessel (artery or vein) compressing and distorting the trigeminal nerve just as it exits the brainstem, and it is thought that ongoing pulsations from these vessels may be responsible for the pain. In some cases, no vessel is found, even during surgery. Occasionally it may be caused by multiple sclerosis or a brain tumour.

How is trigeminal neuralgia diagnosed?

The most important step is taking a careful history by a consultant physician who is well experienced in the diagnosis and treatment of trigeminal neuralgia. An MRI is a routine test which can help visualise offending vessels near the nerve, but more importantly exclude other causes such as multiple sclerosis or a brain tumour that would require a different treatment.

How is trigeminal neuralgia treated?

Occasionally the pain settles and no treatment is required. More typically medication is the mainstay of treatment, and almost all patients experience significant or complete relief of their pain initially. Sometimes medications may cease working or cause significant side effects. At this point, a neurosurgical procedure should be considered.

What medicines are used in treating trigeminal neuralgia?

Standard pain relief medications such as paracetamol, codeine and opiates do not generally work very well for this condition.

Carbamazepine (Tegretol) and oxcarbamazepine (Trileptal) are the most effective medications and are usually recommended as initial treatments. Gabapentin (Neurontin), pregabalin (Lyrica) and lamotrigine (Lamictal) may also be effective. Whilst these are all anti-seizure medications, trigeminal neuralgia is not a seizure nor is it a form of epilepsy.

Common side effects include drowsiness, confusion, dizziness, nausea and skin rashes. Blood tests are required to monitor for less common problems.

What neurosurgical treatments are available to treat trigeminal neuralgia?

- Percutaneous needle rhizotomy (PNR)
 - Microvascular decompression (MVD)
 - Stereotactic radiosurgery (SRS)
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Who is a candidate for these procedures?

Patients for whom medications are no longer working despite increasing doses or who find the side effects of these medications intolerable.

What is a Percutaneous Needle Rhizotomy?

This is a day procedure and usually the patient goes home later that day or the following morning. It is less invasive than an MVD but more than SRS. It involves sedation (not a general anaesthesia) where a specially designed needle is guided using X-ray and CT navigation to where the trigeminal nerve exits the skull. The nerve is treated with a chemical (glycerol), heat (radiofrequency electrocoagulation, RFE) or a balloon compression. As a balloon requires a larger needle and has higher complications, I usually favour glycerol or occasionally RFE. Most patients (70-80%) get good pain relief after the procedure, however rates of recurrence are significant over the first 5 years. The procedure may be repeated at that time.

Facial numbness is the major drawback to a rhizotomy, but is not usually too troubling. Complications include a small stroke risk (<1%), anaesthesia dolorosa (constant difficult to treat pain despite the face being numb) and an extremely small chance of death.

Percutaneous needle rhizotomy is generally reserved for:

- atypical trigeminal neuralgia
 - trigeminal neuralgia secondary to multiple sclerosis
 - patients unfit for an MVD
 - patients not wishing to undergo an MVD
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What is a Microvascular Decompression (MVD)?

This is a neurosurgical operation that involves admission to hospital usually for 2 to 3 days. General anaesthesia is used thus the patient is asleep for the operation. A 5cm incision behind the ear is made and small craniotomy (hole in the skull) is fashioned. Using a microscope, the brain is gently moved out of the way and the trigeminal nerve is visualised along with all surrounding blood vessels. If the compressive vessel is an artery, it is removed from the nerve and a pillow of shredded Teflon is placed between the two, however when a vein is found it is coagulated and divided.

Most patients wake up with complete or significant pain relief (80 – 90%) and are weaned from their medication over the next one or two weeks. Pain Recurrence of approximately 1-2% per year and is usually treated with a needle rhizotomy or SRS. Sometimes a repeat MVD may be performed.

An MVD offers the greatest chance of long standing pain relief with 70- 80% of patients remaining pain free after 10 years. Stroke risk and death are slightly higher than a needle rhizotomy however still very low (2%), with facial numbness much less common than all other procedures.

What is stereotactic radiosurgery (SRS)?

Stereotactic radiosurgery may also be useful for treatment of trigeminal neuralgia. It is performed by a radiation specialist as a day procedure and avoids surgery in some patients. Results appear satisfactory (comparable with needle rhizotomy) and the technique is very helpful for patients who cannot come off blood thinners for whatever reason or whom the above procedures are deemed too risky. The major drawback to SRS is the delayed onset of pain relief, often requiring 2 – 6 months before benefit is seen.

What is glossopharyngeal neuralgia?

Glossopharyngeal neuralgia is a very similar but much less common condition due to a vessel compressing the glossopharyngeal nerve (9th cranial nerve). Pain appears in the back of the tongue, throat and ear. Causes and treatments are much the same as for trigeminal neuralgia. MVD is the treatment of choice when medication fails, this time targeting the glossopharyngeal nerve. Needle rhizotomy and SRS are not appropriate for this condition.

Helpful Links

- 1) <https://www.tnaaustralia.org.au>
- 2) <https://www.ninds.nih.gov/disorders/patient-caregiver-education/fact-sheets/trigeminal-neuralgia-fact-sheet>
- 3) <https://journals.sagepub.com/doi/pdf/10.1177/0333102416687280>

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