Trigeminal neuralgia

Synonym: tic douloureux

Trigeminal neuralgia (TN) can be described as a chronic, debilitating condition resulting in intense and extreme episodes of pain in the face. The episodes are sporadic and sudden and often like 'electric shocks', lasting from a few seconds to several minutes.

TN results from a neuropathic disorder of the Vth cranial nerve (trigeminal nerve). The trigeminal nerve senses mixed modalities including:

- Sensation.
- Nociception.
- Thermoception.
- Motor supply to the muscles of mastication.

Most commonly, the maxillary and/or mandibular branch are involved.

Epidemiology[1]

- TN is the most frequent type of facial pain and the incidence has remained constant ranging from 12.6/100,000/year to 27/100,000/year.
- TN is uncommon in population younger than 40 years (overall incidence of 0.2/100,000/year) and increases in incidence with advancing age, occurring in 25.9/100,000/year in individuals older than 80 years.
- TN appears to be slightly more common among women.
- There may also be a genetic predisposition, as there have been observations of familial clustering. However, the exact method of transmission is unclear, although there is a lack of penetrance[2] .

Aetiology

In nearly all cases, TN is thought to be caused by compression of the trigeminal nerve by a loop of artery or vein; another 5%-10% of cases are attributed to tumours, multiple sclerosis, abnormalities of the skull base, or arteriovenous malformations.

Presentation

TN is a sudden, unilateral, brief, stabbing, recurrent pain in the distribution of one or more branches of the Vth cranial nerve. Pain occurs in paroxysms which last from a few seconds to two minutes. The frequency of the paroxysms ranges from a few to hundreds of attacks a day. Periods of remission can last for months to years, but tend to shorten over time.

- There may be preceding symptoms - eg, tingling or numbness.
- Patients may have certain triggers that set the pain paroxysm off (see 'Diagnostic criteria', below).
- This is followed by sharp, severe, shock-like pains.
- These pains are usually on one side in the cheek or face but pain can involve the eyes, lips, nose and scalp.
- Episodes are intermittent but can last days, weeks or months on end and then not return for months or even years.

Diagnostic criteria

Important information

Site: pain is unilateral in the distribution of the trigeminal nerve, bilateral in only 3% of patients, and rarely is the pain active on both sides at the same time [3] .
Periodicity: episodic and sudden onset of pain, lasting a few seconds to minutes and stopping suddenly, with many attacks a day. There is a refractory period between each attack. Pain might then go into remission for weeks or months; pain-free intervals gradually shorten between episodes.
Character: electric shock-like, sharp, shooting.
Severity: very severe attacks, but attacks can get milder when patients are given drug treatment.
Factors affecting pain: can be provoked by light touch to the face, eating, cold winds, or vibrations.
Associated factors: rarely associated with history of other chronic pain or migraine. Some forms have more continued aching background pain after main attack. Rarely associated with autonomic features.

Triggers

- Vibration.
- Skin contact - eg, shaving, washing.
- Brushing teeth.
- Oral intake.
- Exposure to wind.

Atypical TN

Patients in this subgroup have relentless underlying pain like a migraine associated with superimposed stabbing pains. There may also be an intense burning sensation. This condition is particularly difficult to treat.

Red flags

- Sensory changes, deafness or other ear problems.
- Difficulty achieving pain control, poor response to carbamazepine.
- History of any skin lesions or oral lesions that could lead to perineural spread.

- Ophthalmic division only or bilateral as suggestive of benign or malignant lesions or multiple sclerosis.
- Age of onset under 40 years.
- Optic neuritis.
- Family history of multiple sclerosis.

Differential diagnosis

- Dental pathology.
- Temporomandibular joint dysfunction.
- Migraine.
- Giant cell arteritis (temporal arteritis) - TN rarely affects the forehead alone.
- Cluster headaches.
- Multiple sclerosis and other disorders of myelin.
- Overlying aneurysm of a blood vessel.
- Tumour in the posterior fossa - eg, meningiomas.
- Arachnoid cyst at the cerebellopontine angle.
- Postherpetic neuralgia after shingles.

Investigations

The diagnosis is clinical and it can be difficult to make. No investigations are required initially unless there is uncertainty regarding the diagnosis. Patients who are referred on for specialist review will usually have a brain MRI scan.

MRI scan of the brain is indicated to rule out other potential causes of pain if the diagnosis is uncertain or if red flags are present. MRI may be used to identify:

- Sinusitis.
- Extracranial masses along the course of the trigeminal nerve.

- Pathological enhancement of the trigeminal nerve that could indicate perineural spread of malignancy.
- Cavernous sinus masses.
- Demyelination plaques that might indicate multiple sclerosis.
- Intrinsic brain lesions in the thalamus or trigeminal brain stem pathways such as lacunar infarctions.
- Cerebellopontine angle mass lesions such as tumour, epidermoid, dermoid, or arachnoid cyst, aneurysm, or arteriovenous malformation.

Management

Unfortunately, there is no definitive cure at present (relapses and recurrences occur); however, newer surgical procedures are promising. Management involves three aspects: support and education, medical and surgical.

The National Institute for Health and Care Excellence (NICE) recommends[4] :

- Offer carbamazepine as initial treatment for TN.
- If initial treatment with carbamazepine is not effective, is not tolerated or is contra-indicated, consider seeking expert advice from a specialist and consider early referral to a specialist pain service or a condition-specific service.

Support and education

- Patients need to be made aware that the condition is not life-threatening.
- There is a need, however, also to express empathy towards severity of the condition.
- Education as to the causes and potential therapies.
- Reassurance and support groups.

Referral

Consider referring the person to a specialist pain service or a relevant clinical speciality (for example, neurology, diabetology, or oncology services) at any stage if:

- They have severe pain.
- Their pain significantly limits their participation in daily activities (including self-care, general tasks and demands, interpersonal interactions and relationships, mobility, and sleeping).
- Atypical clinical features (for example, burning pain between paroxysms, loss of sensation, or any abnormal neurological signs) are present.

Medical

- Typical analgesics and opioid analgesics are unfortunately not effective.
- Carbamazepine has the most evidence for efficacy and should be used first-line[3] . It should be tried initially and the dose titrated to achieve pain control. Once patients have been in remission for one month, the drug should be gradually withdrawn. There is consensus that oxcarbazepine is also an effective treatment in TN, although there is a lack of randomised control trial-based data to confirm this.
- Gabapentin combined with regular ropivacaine injections into trigger sites has been shown to improve pain control and quality of life, and pregabalin was found to be effective at one-year follow-up[1] .
- Botulinum toxin type A (BTX-A) in TN produces a response in approximately 70%-100% of patients with mean pain intensity and frequency reduced by approximately 60%-80% with no major adverse events reported[1] .
- There is low-quality evidence that the effect of tizanidine is not significantly different to that of carbamazepine. Pimozide is more effective than carbamazepine, although the evidence is of low quality.
- The data supporting the use of tricyclic antidepressants (eg, low-dose amitriptyline) are lacking at present.

- Other drugs that might be used in a specialist setting include lamotrigine and baclofen, although lamotrigine needs to be titrated over many weeks and has limited value in severe pain.

Failure of these agents should prompt a review of the diagnosis and, if pain control cannot be achieved or drugs cause unacceptable adverse effects, surgical options should be considered.

Surgery

Most of the improvements in the management of TN have occurred because of advances in surgical treatments. Surgery involves either relieving pressure on the trigeminal nerve or damaging it to prevent any pain transmission.

There are various types of surgical procedures that can be used in TN. However, there is little evidence to identify the best surgical procedure. Microvascular decompression seems to be the most effective treatment in terms of patient satisfaction and long-term cost-effectiveness. Newer modalities like stereotactic radiosurgery and botulinum injections have shown promising results.

Rhizotomy

The aim is to damage the trigeminal nerve. This is an alternative to the more invasive decompression. These methods include:

- Percutaneous glycerol rhizotomy (under a local anaesthetic).
- Percutaneous balloon compression rhizotomy (under a general anaesthetic).
- Radiofrequency rhizotomy (performed under sedation).

Editor's note

Dr Sarah Jarvis, 7th February 2022

NICE update - stereotactic radiosurgery for trigeminal neuralgia

NICE has issued a new Technology Appraisal on the use of stereotactic radiosurgery for trigeminal neuralgia. This treatment modality uses precisely focused multiple beams of ionising radiation aimed at the trigeminal nerve where it enters the brainstem, to deliver a high dose in a single treatment session. It does not require open surgery, needle insertion or general anaesthesia. The aim, as with other percutaneous techniques, is to damage the trigeminal nerve and stop the transmission of pain signals.

This follows a systematic review commissioned by NICE concluded that stereotactic radiosurgery is a safe and effective therapy for drug-resistant trigeminal neuralgia [5] .

NICE's guidance confirms that evidence on the safety and efficacy of stereotactic radiosurgery for trigeminal neuralgia is adequate to support using this procedure provided that standard arrangements are in place for clinical governance, consent and audit [6] .

Microvascular decompression

- Microvascular decompression (MVD) aims to decompress the trigeminal nerve, and deals with the cause of TN in the majority of cases not caused by other lesional causes.
- Blood vessels are compressing the trigeminal nerve, and lifting these blood vessels away reduces the pressure. This requires a general anaesthetic. The approach is behind the ear into the posterior fossa on the affected side. Patients are usually assessed by MRI beforehand to look for the presence of compression.
- Over 90% of patients obtain pain relief, with the majority still pain-free at one year [7] .
- This procedure, however, is not without risks and the average mortality associated with the operation is 0.2%. There is a risk of a cerebrovascular event, deafness and even death. The rates of complication depend on the surgeon's expertise.

- Treatment by MVD has a risk of serious complications like cerebrospinal fluid (CSF) leak (3.3%; 95% CI: 0.7%-5.9%), wound infection (1.3%; CI: 0%-3.1%) and cranial venous sinus thrombosis (1.3%; CI: 0%-3.1%) [8] . Mild facial numbness (4%) and dysaesthetic facial pain (11%) were the common complications after MVD. The rate of both complications was significantly higher after gamma knife surgery than following MVD.
- Percutaneous microballoon compression is safe for elderly patients [9] . However, nearly all procedures cause some numbness and, in a few, this can be associated with intense pain obviating the whole point of the surgery ('anaesthesia dolorosa').

Complementary therapies

Due to the lack of curative measures, the use of complementary therapies in TN has evolved quite rapidly. These include the following:

- Transcutaneous electrical nerve stimulation (TENS).
- Acupuncture.
- Biofeedback.
- Vitamin therapies - eg, vitamin B.
- Nutritional therapies - eg, garlic.

There is no evidence available that supports the use of these measures.

Complications

The pain of TN can be so intense that it can lead to a poor quality of life due to mental and physical incapacity. Patients may require psychosocial input - eg, counselling.

Prognosis [3]

- 50% of people with TN experience remissions of at least six months' duration.
- 65% of people newly diagnosed with TN will have a second episode within five years, and 77% within 10 years.

- Periods of remission tend to get shorter with time, and attacks of pain get longer.

Further reading

- Trigeminal Neuralgia; National Institute of Neurological Disorders and Stroke
- International Classification of Headache Disorder (version 3); International Headache Society, 2018

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