Neurosurgery and facial pain

PR Eldridge

This article reviews the diagnosis, differential diagnosis and management of trigeminal neuralgia, the commonest facial pain condition treated by the neurosurgeon. The advantages offered by microvascular decompression as a treatment are reviewed and compared with medical treatment and percutaneous techniques.

Mr PR Eldridge is Consultant Neurosurgeon in the Walton Centre for Neurology and Neurosurgery, Fazakerley, Liverpool L9 7LJ

There are many causes of facial pain but only trigeminal neuralgia (TGN) is predominantly managed neurosurgically. Consequently, the majority of this article covers this condition. TGN has become famous because of its association with arterial compression at the root entry zone and the observation that the pain is apparently 'cured' by moving this vessel away. Thus, it is an unusual example of a chronic pain corrected by a surgical procedure designed to remove the cause. TGN is the best understood of a number of neurovascular compression syndromes; of the other conditions only glossopharyngeal neuralgia and possibly geniculate neuralgia present with pain, although the next commonest condition treated is that of hemifacial spasm.

TRIGEMINAL NEURALGIA

Incidence and prevalence

The incidence of TGN is estimated to be 50/million/year and the prevalence at 155/100 000. Frequency increases with age. Between 2 and 4% of multiple sclerosis (MS) cases suffer from TGN, while 5% of cases of TGN are in association with MS (Nurmikko and Eldridge, 2001).

Cause of trigeminal neuralgia

Just after the trigeminal nerve leaves the pons, there is a junctional area between the central and peripheral myelin, measuring about half a centimetre, which is termed the root entry zone (*Figure 1*). Four observations argue that contact by a vessel at the root entry zone is a major cause of TGN (Miles et al, 1997; Leandri et al, 1998):

- In over 90% of cases a vessel, usually an artery (rarely a vein), is found in contact with or grooving the trigeminal nerve as it exits the pons
- The usual outcome of microvascular decompression (MVD) is instantaneous pain relief
- Intraoperative recordings demonstrate recovery of nerve conduction and far-field evoked potentials
- Sensory function improves following decompression, although this takes up to 6 months.

The mechanism by which vascular compression or contact causes TGN is unclear, including the reason why the root entry zone is sensitive to the effects of vascular contact. Demyelination of the nerve at the position of vascular contact has been reported and linked with the observation that where plaques occur in the trigeminal system, TGN is found in sufferers of MS (Nurmikko and Eldridge, 2001). However, this does not explain the intermittent nature of the condition, its paroxysmal quality and neither the phenomenon of triggering, nor that not all individuals with contacts experience pain. Hence, in order to explain these features, it is further proposed that in TGN spontaneous discharges occur within a hyperexcitable trigeminal ganglion (Rappaport and Devor, 1994).

Clinical features

Unilateral, sharp, stabbing or 'electric shock-like' paroxysmal pain is felt in the distribution of one or more branches of the trigeminal nerve.

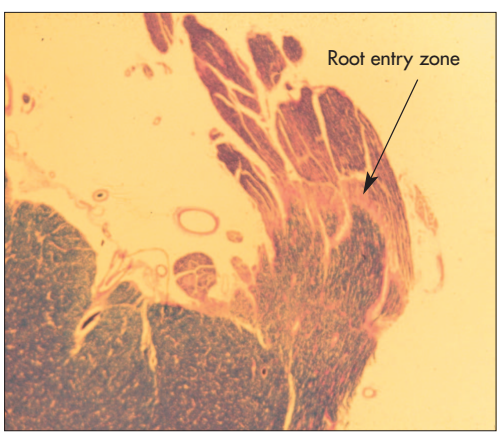

Figure 1. Axial section of trigeminal nerve at the pons showing the root entry zone.

The pain may be triggered by touching the face, chewing, talking or by wind on the face, leading to a tendency for the condition to be worse in winter. The pain is intermittent in nature, with remissions lasting several weeks or months. The response to carbamazepine is so typical that its absence should question the diagnosis, being careful not to confuse lack of response with intolerance of the drug. The pain is refractory to aspirin, paracetamol, non-steroidal anti-inflammatory agents and opioids. In the majority of cases, the pain involves the second or third divisions of the trigeminal nerve, and only relatively rarely is the pain purely in the first division.

Signs

In classical teaching, there are no signs, although a decreased corneal reflex may occur. Therefore, any neurological deficit found on bedside testing should raise the suspicion of a structural lesion or of idiopathic trigeminal neuropathy. However, quantitative sensory testing carried out under laboratory conditions will reveal sensory deficits (Bowsher et al, 1997).

Differential diagnosis

This includes a spectrum of other facial pain and headache syndromes but rarely temporal arteritis or pain radiating from the neck. Perhaps surprisingly, patients rarely present with dental pain to the neurosurgical clinic, although a large number of patients with TGN has first passed through a dental practice.

The conditions which cause most difficulty are atypical facial pain, cluster headache, painful trigeminal neuropathy and temporomandibular joint (TMJ) dysfunction.

Atypical facial pain is a variant of depression, and other features of depression will be present. The pain is continuous, not paroxysmal. The patient is often unshakably convinced of an organic cause. Pain descriptors are often modified to fit one or more of the other syndromes described below, about which, in the Internet age, the patient may be well informed, ascribing great importance to matters of minute detail. This can be an extremely difficult condition to manage.

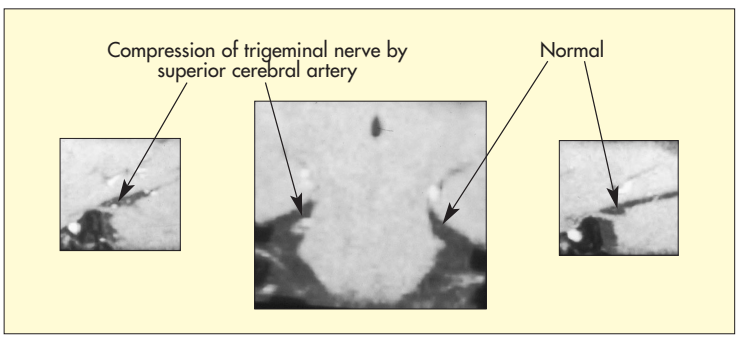

Figure 2. Magnetic resonance imaging demonstrating unilateral arterial compression in the coronal plane and in sagittal reformats along the line of the nerve.

Cluster headache is regarded as a variant of migraine and to be a vascular headache. Attacks are stereotyped and are clustered in time and space. Autonomic features, such as a congested nose or watering eyes, may be present. The condition is provoked by alcohol and maybe relieved by triptans (which may need intramuscular administration because of the time course of the condition).

TMJ dysfunction is perhaps better viewed as a myofascial pain involving the temporalis muscle. It is provoked and aggravated by chewing but not triggered. True disorders of the TMJ do occur but are much rarer.

Postherpetic neuralgia is rare, usually involves the forehead and follows an attack of herpes. Allodynia is often present in the affected area.

Dysaesthetic facial pain is a neuropathic pain resulting from nerve injury and is most often seen as a side effect of lesioning techniques for TGN — such as the foramen ovale methods or alcohol peripheral neurectomy. It is a continuous burning pain — at its worst termed anaesthesia dolorosa, when it is virtually untreatable.

Trigeminal neuropathy describes pain from structural lesions, such as the extremely rare trigeminal schwannoma or lesions in the region of the cavernous sinus. Although classically described as TGN, the pain is different being continuous, progressive and associated with neurological deficit. Idiopathic painful trigeminal neuropathy describes these features when no structural lesion can be identified.

Differential diagnosis can be difficult and relies most of all on the clinical history. Leading questions by the doctor are a hazard; also the patient's description of the pain may not be consistent. There is great merit in seeing the patient on more than one occasion to avoid this problem.

Investigations

No specific test exists for the condition. Magnetic resonance imaging (MRI) detects the presence or absence of neurovascular contact at the root entry zone with a sensitivity of 100% and specificity of 96% validated in a series of 55 cases (Meaney et al, 1995) (*Figure 2*). Since neurovascular contact occurs in 10% of controls, MRI cannot be sensitive or specific for the diagnosis of TGN, and the role of MRI is to aid choices for management. Attempts have been made to demonstrate preoperatively neurophysiological abnormalities, particularly far-field evoked potentials, but these have not proven to be either sufficiently sensitive or specific.

MANAGEMENT

Natural history

The natural history of the condition is for the severity of attacks to worsen and for the periods of remission to shorten. Symptomatically, the syndrome may evolve from its typical form to an atypical form in which there is a constant background burning element to the pain, in addition to the classical features (Burchiel and Slavin, 2000; Nurmikko and Eldridge, 2001).

Medical management

The drug of first choice, as mentioned above, is carbamazepine. While effective, this treatment is not without problems. It is difficult to estimate accurately the numbers of patients who 'fail' medical treatment. Reasons for failure are idiosyncratic reactions to carbamazepine (rash being typical) and dose-related problems. At high dose — to which the patient is driven by the severity of the pain — there may be ataxia and severe drowsiness. However, the cognitive impairment produced by the drug is underestimated and may be the principal reason why some sources have found failure rates with carbamazepine reaching 75% of cases treated. One long-term study exists where either loss of effect or intolerance occurred in over 50% of patients over 10 years (Taylor at al, 1981).

Effective alternatives include lamotrigine, phenytoin, sodium valproate and gabapentin. All these are used in the author's practice and found to be effective in combination, although there is no published study comparing monotherapy and polytherapy.

Some reports, but not in the author's practice, have found baclofen to be effective.

Medication should be withdrawn gradually because of the risk of provoking a seizure (although this can occur, it is rare). Sodium valproate and phenytoin find application in the acute situation, as they can be given intravenously.

Foramen ovale methods

Radiofrequency lesioning: A needle is introduced via the foramen ovale, and electrical stimulation is performed to confirm correct placement of the needle by inducing paraesthesiae in that area of the face in which the TGN is occurring. The patient is then anaesthetized, and a thermocoagulation is performed (60–90 °C for approximately 45 seconds). It was originally suggested that heat preferentially destroyed thin pain fibres, but formal sensory testing finds that all fibres are affected approximately equally. It is difficult to target the ophthalmic division, and this runs the risk of corneal anaesthesia. Masseter weakness is a further risk.

Over 90% will experience immediate relief of pain. However, with time there is a gradual recurrence. Recurrence rates vary with series, ranging from as poor as 25% pain free at 2 years up to 80% pain free (Taha et al, 1995; Yoon et al, 1999), and seem to be related to the technique. If deep hypalgesia is produced during the procedure, essentially by creating a larger lesion, the recurrence rate is lower. Unfortunately, the larger the lesion, the greater the risk of creating dysaesthesias and the feared complication of anaesthesia dolorosa. In series with low recurrence rates, a higher dysaesthesia rate is found; 25% of cases exhibit dysaesthesia which the patient regards as unpleasant but tolerable. However, in 8% quality of life assessment after treatment has been instituted indicates that although pain is controlled, overall quality of life is unchanged. In 1% there is severe disabling anaesthesia dolorosa. The procedure is regarded as safe but not completely risk free. It involves intermittent anaesthesia in an elderly patient, and reported complications include meningitis and caroticocavernous fistulae (Taha et al, 1995; Tronnier et al, 2001).

Glycerol: A needle is again placed into the foramen ovale, and glycerol is introduced under fluoroscopic control. Accurate positioning of the needle is important. The technique has caused much controversy. In the hands of its proponents, excellent results are obtained (best reports being 90% pain free at 1 year). However, others report poor results (only 17% pain free at 5 years at worst) with significant dysaesthesia rates, reaching 44% in some series (Nurmikko and Eldridge, 2001).

Balloon microcompression: This is performed under general anaesthesia. A Fogarty catheter is passed into Meckel's cave and inflated for between 1–6 minutes. Sensory impairment is produced, although this is mild. The best reported results indicate a recurrence of around one third of cases, with a dysaesthesia rate of around 10%. However, some find a much higher recurrence rate and regard it as a temporary treatment with the advantage of producing almost no dysaesthesia. In a comparative study, 44% of patients were pain free at 2 years following balloon microcompression, while similar figures for radiofrequency (RF) lesioning were 58% and 75% for MVD, so that it was concluded that the latter is the treatment of choice (Meglio et al, 1990).

Surgical management

Operation: This is performed via a retromastoid craniectomy (*Figure 3*). The subsequent approach is over the surface of the cerebellum until the nerve is identified (*Figure 4*). Arteries must be dissected free and held clear from the nerve using

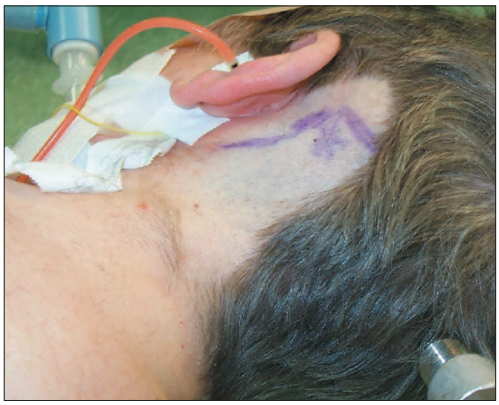

Figure 3. Operative approach and landmarks. The lateral and sigmoid sinus are marked. The star marks the position of the craniectomy.

a small piece of Ivalon sponge or Teflon. If a vein is the cause, it may be coagulated and divided. If no vessel is found, a partial sensory rhizotomy gives good relief — being a centrally placed lesion it seems less likely to cause anaesthesia dolorosa.

Risks: In most published series, the serious morbidity (death or major stroke) is significantly below 1% (Barker et al, 1996). The operation, being performed near the acoustic nerve, also carries a risk of hearing impairment, possibly as a result of traction on the nerve while retracting structures to gain access to the deeper trigeminal nerve. Since brainstem auditory evoked responses have been used as a monitoring device peroperatively, the risk of unintentional hearing deficit has been reduced to about 2–3% from historical series where risks might be as high as 13%.

Outcomes: Overall, of those with clear arterial compression, excellent long-term results are obtained in about 90%, of whom some 70% are completely pain free and off medication up to 20 years postoperatively. Results of venous compression are less good as are the outcomes following partial rhizotomy — approximately 60–70% at 2-year follow-up. Case selection is important, as poor outcomes from MVD are found in patients with atypical pain or in whom the diagnosis is not TGN. Prior RF lesioning may leave the patient with dysaesthesia which is revealed by successful MVD (Hunn et al, 1998).

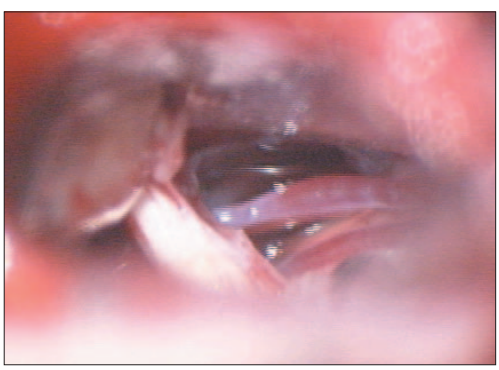

Figure 4. Operative view showing trigeminal nerve with superior cerebellar artery lying in typical position medial to nerve in the angle between it and the pons.

Stereotactic radiosurgery

Some controversy surrounds the results of this technique in which focused radiation is applied to the root entry zone using the gamma knife, although there is no reason why a suitably equipped linear accelerator stereotactic system should not be used. Unfortunately, the tendency of units is to report results as 'good' and not to specify the true 'pain-free off medication' outcome rate used in other techniques. When this stricter criterion is used, results are less impressive: only 65% of patients experience pain relief at 6 months, rising to 75% at 33 months. From this, it will be seen that the effect is not immediate. Furthermore, only 56% of those achieving relief continued to have complete or partial relief at 5 years follow-up, implying a high recurrence rate. There is a dysaesthesia rate and also a dose-related post-radiation numbness (Maesawa et al, 2001).

Choice of technique

The author considers MVD to be the treatment of choice in most instances of severe TGN while recognizing that other views are held.

MVD corrects a structural abnormality and is the only technique that effects a cure. Neurophysiologically, the patient is returned to normal, whereas other methods depart from normal, being destructive lesions. The risk of all destructive lesions is neuropathic pain, at its worst anaesthesia dolorosa. The habit of performing peripheral nerve avulsions or alcohol injections is associated with an almost universal recurrence and a high incidence of late dysaesthetic pains.

The following points support the use of MVD:

- The natural history is for the condition to worsen
- Medical management has significant side effects, and most patients fail this treatment
- MVD is demonstrated to be effective and of low risk, including in the elderly population
- In no series does the efficacy of RF lesioning surpass that of MVD, and in the case of radiosurgery the efficacy is poorer
- Results from MVD are poorer after failed percutaneous procedures
- All destructive lesions (peripheral, foramen ovale, radiosurgery) are associated with a significant recurrence rate, incidence of sensory deficit and production of unpleasant dysaesthetic symptoms or even anaesthesia dolorosa
- The sensitivity and specificity of MRI to detect neurovascular compression is such that patients with arterial compression can be advised of the likelihood of an excellent outcome, and patients can be offered an alternative technique if the scan is negative

- Although no randomized controlled studies exist, the authors of two comparative studies concluded that MVD is the treatment of choice (Meglio et al, 1990; Tronnier et al, 2001).

Fitness for surgery

Because MVD involves craniectomy and TGN occurs in elderly people, fitness for surgery is an issue. However, series exist in which there is little difference in outcome or morbidity when comparing an over 70-year-old age group with a younger age group (Javadpour et al, 2000). With modern anaesthesia, very few patients are unsuitable for this procedure, and the choice between percutaneous RF lesioning and MVD can be based solely on the outcomes of the procedures. It is worth remembering that RF lesioning and balloon microcompression both require general anaesthesia.

Trigeminal neuralgia as an emergency

Occasionally, patients present as an emergency with severe pain. The patient is drowsy and ataxic, having by this point taken so much carbamazepine as to suffer severe toxic effects, and is furthermore dehydrated, being unable to swallow fluids because this action is triggering the neuralgia.

Management requires admission and bed rest for the ataxia, intravenous fluids to correct dehydration and finally measures to treat the pain. An intravenous loading dose of either phenytoin or sodium valproate is usually effective in treating the pain. This may be followed by emergency MVD or, if MRI is negative for compression, one of the foramen ovale methods.

GLOSSOPHARYNGEAL NEURALGIA

In reality, the anatomy of the lower cranial nerves is such that the glossopharyngeal nerve may be considered as the upper part of a complex which includes itself, the vagus and the cranial part of the accessory nerve.

The syndrome may be considered as identical to TGN except in two aspects. First it is considerably rarer, and second the pain is distributed in the area of the glossopharyngeal nerve. Thus, the shooting pain is more within the throat, and triggering is also from the back of the throat. Treatment is identical, with the first-line drug being carbamazepine and surgical decompression providing much the same results as for TGN. There is no equivalent to RF lesioning in this condition.

VII/VIII COMPLEX

Hemifacial spasm, tinnitus and nervus intermedius neuralgia arise from this complex. MVD is effective for hemifacial spasm, which is the most common manifestation of compression at this level. The procedure for tinnitus is rarely performed and controversial, while pure nervus intermedius neuralgia is extremely rare. It can present with otalgia or throat pain (Rupa et al, 1991). HM

Conflict of interest: none.

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KEY POINTS

- Neurovascular compression of the trigeminal nerve at the pons is a major causative factor in the genesis of trigeminal neuralgia.
- Diagnosis is made from the history.
- Magnetic resonance imaging allows reliable preoperative detection of neurovascular compression.
- Approximately 50% of patients fail medical management.
- Microvascular decompression is a safe and efficacious treatment which gives long-term pain relief and returns the neurophysiology of the nerve to normal.
- Percutaneous methods which destroy the nerve can cause neuropathic pain and appear to recur at a higher rate than microvascular decompression.