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CURRENT CONCEPTS IN THE MANAGEMENT OF TRIGEMINAL NEURALGIA.

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Abstract

|| Brief Background
The paper describes trigeminal neuralgia a neuropathic disorder of one or both of the facial trigeminal nerves and its management.

|| Materials and Methods
The initial method of treatment of choice before any surgical alternatives is medical therapy using a combination of drugs to suit specific cases. The common surgical techniques employed being Percutaneous neurolysis and Microvascular decompression.

|| Discussion
The discussion focuses on the treatment procedures, indications and their advantages and disadvantages

|| Summary and Conclusions
Trigeminal neuralgia is characterized by stereotypic symptoms and absence of signs that usually allow its clinical diagnosis. Given the various treatment options available, the choice of treatment is dictated by age of the patient, medical comorbidities, and the risks the patient is willing to assume. Highly advanced form of treatment is available only at select centres.

|| Key Words
Trigeminal neuralgia, Percutaneous neurolysis, Microvascular decompression, radiofrequency thermocoagualtion.

Introduction

Trigeminal neuralgia (TN) or tic douloureux (also known as prosopalgia) is a neuropathic disorder of one or both of the facial trigeminal nerves. It causes episodes of intense pain in any or all of the following: the ear, eye, lips, nose, scalp, forehead, teeth or jaw on one side of the face[1]. It is estimated that 1 in 15,000 people suffer from trigeminal neuralgia, although the actual figure may be significantly higher due to frequent misdiagnosis. TN usually develops after the age of 50, more commonly in females, although there have been cases with patients being as young as three years of age[2].

Anatomy

The trigeminal nerve arises from one motor nucleus and three sensory nuclei, which extend throughout most of the length of the brain stem. The principal sensory nucleus is situated within the lateral aspect of the pons. As it exits the pons, the point of change from central to peripheral myelin is known as the root entry zone (REZ). It is at this point that the nerve is thought to be most susceptible to compression by tortuous branches of the posterior circulation vessels, an important cause of TN. The nerve continues anteriorly to the apex of the petrous temporal bone. Here it traverses a defect in the dura to enter Meckel's cave, a CSF-filled space lying immediately lateral to the cavernous sinus. The nerve trunk then expands to form the trigeminal (Gasserian) ganglion, from which the three branches of the trigeminal nerve arise[3]

Pathology

The first detailed description of ultra structural abnormalities in the nerve root in the region of vascular compression was by Hilton and colleagues[4]. The authors observed focal loss of myelin and close apposition of demyelinated axons. The findings in relation to vascular compression of the nerve root were confirmed in a subsequent electron microscope study of trigeminal rhizotomy specimens from two further patients with medically intractable TN in whom, because of the local vascular anatomy, the compressing artery or vein could not safely be repositioned[4].The pathophysiology of TN has been much debated, the pain being ascribed variously to hyperactivity or abnormal discharges arising from the gasserian ganglion, the 'injured' nerve root and the trigeminal nucleus within the brainstem[5].

Diagnosis

The diagnosis of trigeminal nerualgia is solely clinical. Patient typically describes his pain as shooting, "bolt from blue" or shock like. Pain usually lasts for only few seconds, but occurs hundreds of time during the day. Pain is precipitated by triggers like drinking, eating, chewing, washing face or even a breeze of air. Sometimes for the fear of pain patient may avoid eating, talking, brushing or shaving. In between the attacks patient is completely pain free. Pain is usually confined to one or more divisions of the trigeminal nerve. There should not be any neurological deficit and the pain characteristics should not be burning, continuous or irritating. The natural history of trigeminal neuralgia is characterized by exacerbation and remissions. Over a period of time the attacks become more frequent requiring larger doses of medications and possibly surgical interventions. Bilateral pain is uncommon and occurs in around 10% of patients. There is also a variant of TN called atypical trigeminal neuralgia. In some cases of atypical TN, the sufferer experiences a severe, relentless underlying pain similar to a migraine in addition to the stabbing shock-like pains. This variant is often called "trigeminal neuralgia, type 2, based on a recent classification of facial pain[6].

Radiology in TN

For the diagnosis of trigeminal nerualgia investigations are not required. However, in cases where there is a doubt about the diagnosis a CT scan or an MRI can be performed to rule out a CP angle mass lesion, as sometimes this can present with TN. In case surgical intervention especially if microvascular decompression is contemplated MRI with CIS images should be performed. This images may reveal a vascular loop adjacent to the TN root entry. Zone on the affected side. However this is not diagnostic as TN can occur even in absence of vascular Loop's (Fig. 1).

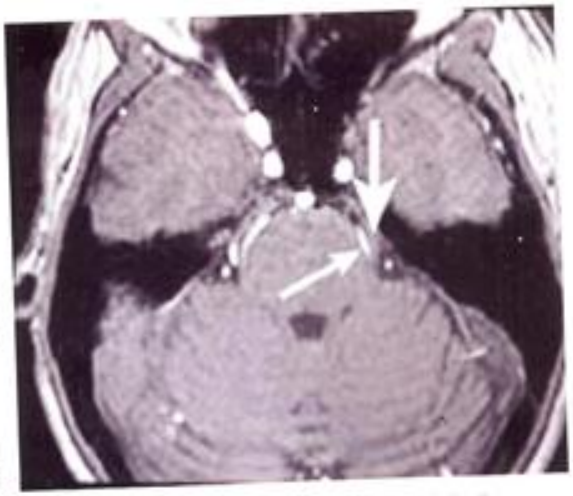

Fig. (01) MRI CIS image showing hyperintense (thin arrow) blood vessel pressing on the nerve(thick arrow).

|| Treatment

Medical therapy

Medical therapy is to be considered the initial treatment of choice before resorting to any surgical alternatives in patients with trigeminal neuralgia. Because of the extreme intensity and brief duration of pain, narcotic analgesics are seldom useful.

Carbamazepine

Carbamazepine and oxcarbazepine are the most effective therapeutic agents[7]. Currently, carbamazepine is the initial drug of choice for the management of TN[8,9] because it controls the pain in approximately 90% of patients. Our recommended dosage is 100 mg twice daily with meals, increasing by 100 mg every other day until pain control is achieved or toxicity develops. We usually continue the medication for three to four weeks after complete pain control. Following this, it is gradually tapered before stopping. If a breakthrough pain occurs, one should go back to larger more effective dose. Twenty to forty percent of patients treated with carbamazepine experience drug-related side effects, including somnolence, dizziness, nausea, and nystagmus.

Oxcarbazepine

Oxcarbazepine, a derivative of carbamazepine, is a newer drug that is reported to have similar clinical effectiveness but fewer side effects than carbamazepine[8,10]

Phenytoin

Phenytoin, although somewhat less effective than carbamazepine, may be useful in many patients because it has lower toxicity[10]. Patients who have obtained effective pain relief while on carbamazepine but can no longer tolerate it seem to be the best candidates for phenytoin. Phenytoin can also be useful when used in conjunction with carbamazepine. The dose required to achieve pain control is usually 5 to 7 mg/kg/d.

Baclofen

Baclofen, a gamma-aminobutyric acid (GABA) agonist, has some efficacy in the treatment of TN[8]. There seems to be a synergism between baclofen and either carbamazepine or phenytoin; therefore, combination therapy in specific cases is a reasonable option[11]. The initial dose is 10 mg three times daily.

|| Surgical therapy

There are two main types of surgical procedures that have proven to be clinically useful; 1) percutaneous neurolysis and 2) microvascular decompression.

Percutaneous neurolysis techniques

The percutaneous techniques for the treatment of trigeminal neuralgia produce a partial destructive lesion in the preganglionic trigeminal rootlets (Fig. 2). Such lesions have been shown to relieve the pain of TN while usually sparing some trigeminal sensory function. The most common method is radiofrequency thermocoagualtion (RFTC).

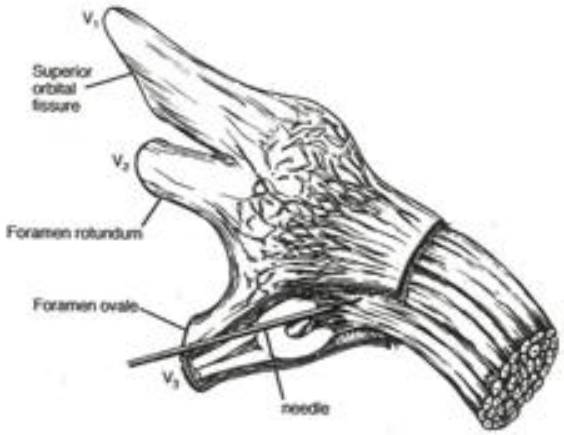

Fig. (02) Trigeminal ganglion, preganglionic rootlets, and postganglionic divisions are shown. The needle is placed through the foramen ovale, through the ganglion, and into the trigeminal cistern containing the preganglionic rootlets.

Fig. 03 OT set up for performing RFTC

RFTC

Radiofrequency rhizotomy is based on temperature dependent selective destruction of pain transmitting C fibres whiles sparing the A-delta fibres[12]. We recommend RFTC for most patients undergoing their first surgical treatment for typical trigeminal neuralgia. The procedure involves retrogasserian needle placement by percutaneous technique (Fig. 3). Needle is introduced through a point 2.5 cm lateral to the

angle of the mouth on side of the lesion. It is passed medial to the mandible and aimed in the direction of the petrous bone and clivus junction, seen on lateral fluoroscopic view (Fig. 4). Lateral part of the foramen ovale is entered. Oblique view localizing the foramen is useful in case of difficult penetration (Fig. 5).

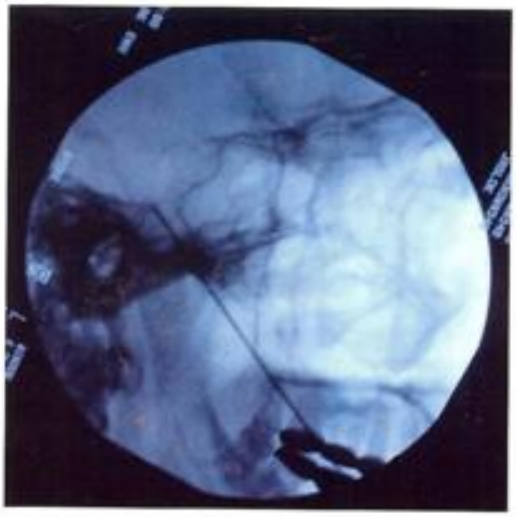

Fig. (04) Lateral radiograph showing the needle at the level of Clivus.

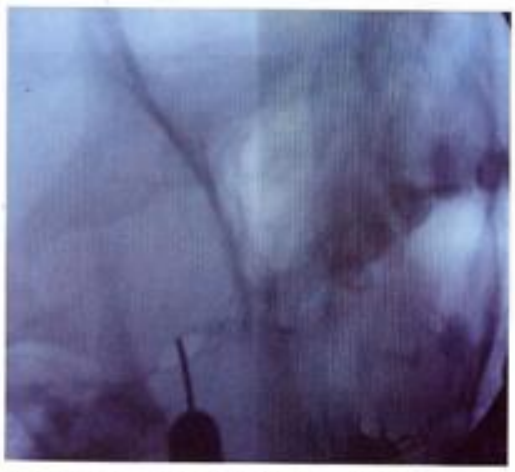

Fig. (05) Oblique view showing needle through the foramen ovale.

Once the needle is in position, the appropriate trigeminal division is stimulated using the current from the radiofrequency lesion generator (Fig. 6). Patient typically experiences paraesthesia in the territory of his pain. In case he does not feel the paraesthesia in the desired territory, the needle position is adjusted. Once the distribution of the paraesthesia is confirmed a short general anaesthetic (IV propofol) is administered and the division is lesioned using 70°C current for 60 sec. The procedure is terminated when the patient develops dense hypalgesia but not anaesthesia in the primarily affected division, especially over the trigger zone, and when touching the trigger zone cannot reproduce trigeminal pain[13,14]. After recovery from anaesthesia, patients may resume full activity and a regular diet. They are usually discharged after an overnight hospital stay. This procedure is well tolerated by elderly or medically debilitated patients. Pain is immediately relieved in 99% of patients[14]. (Table 1) The rate of pain recurrence is approximately 15% to 20% over 10 to 15 years.[15] Patients must be aware that this procedure permanently alters facial sensation, producing significant numbness in 90% of cases, and that it may produce corneal anaesthesia if the first division is affected or the lesion spreads to involve that division. In a review of 500 patients by Taha and Tew,[16] 9% of patients described an intermittent crawling, burning, or itching sensation that did not require treatment, 2% complained of numbness that required treatment ,0.2% developed anaesthesia dolorosa, and less than 1% developed neurogenic keratitis or corneal abrasions. Postoperative dysesthesias are the major adverse effects experienced by patients who have undergone percutaneous RFTC.[16] Patients who suffer from analgesia dolorosa or anaesthesia dolorosa are bothered by constant and severe burning, itching, or crawling sensations, which they may find as intolerable as their initial trigeminal neuralgic pain. Unfortunately, these sensations are often refractory to treatment, although some patients respond to a combination of perphenazine and amitriptyline. The incidence of postoperative dysesthesias has largely

Fig. (06) Radiofrequency lesion generator for RF lesion

declined after the technique modifications described by Tew and Taha.[17] Some of these modifications include continuous sensory examinations during lesion making; asking the patient whether facial numbness is tolerable during the procedure; and quantitating the numbness by asking the patient to compare the pinprick sensation on the treated side with that on the untreated contralateral side[15].

Table 1 Results of RFTC[15]

	Literature review (6205 cases)	Personal Series (395 cases)
Pain relief	98%	92%
Recurrence	23%	20%
Mortality	03	0.0
Keratitis	1	0
Dysesthesia	22	18
Anaesthesia dolorosa	1	1

Microvascular decompression (MVD)

MVD is an other effective treatment for trigeminal neuralgia[18,19]. This operation is based on the observation made by Dandy[20] that the cause of trigeminal neuralgia is compression of the trigeminal nerve at its root entry zone adjacent to the brain stem . The usual cause of this compression is an aberrantly located and elongated arterial loop; however, venous channels and tumours have also been encountered. Jannetta and his colleagues[21,22,23] devised an operative procedure that involves a limited retromastoid craniectomy and microsurgical techniques. This approach allows dissection at the root entry zone of the trigeminal nerve and displacement of the offending vascular structure, usually by the insertion of a small synthetic sponge prosthesis interposed between the nerve and artery. The advantages of MVD over the other percutaneous treatments are that it supposedly, treats the primary etiology of the disease; the trigeminal nerve is preserved and not damaged; deinnervation sequelae, such as facial numbness and dysesthesia, are avoided; and it has a lower rate of recurrence over long-term follow-up. Nevertheless, MVD incurs the risk of an open surgical procedure. MVD also has a higher percentage of serious morbidity and has minimal mortality risks. (Table 2) It involves hospitalization for a period of 5-7 days.

The choice of treatment modality should be made by an informed patient and the ability to tolerate an open surgical procedure under general anaesthesia. The key to this decision should involve consideration of the patient's age, associated illnesses, and assessment of the risks that the patient is willing to assume.

Table 2 Comparison of Results of MVD and RFTC thermocoagulation (RFTC)[14]

10 years F/U	MVD	RFTC
Recurrence	35%	45%
Painful dysesthesia	0	0.9
Morbidity	22%	<2%
Serious morbidity	More common	rare
Mortality	0.8%	Nil
Cost and Hospitalization	More	Significantly small

Stereotactic radiosurgery

Stereotactic radiosurgery is another treatment option for patients with trigeminal neuralgia[24,25]. It has been used as the first procedure in selected patients of advanced age or poor clinical condition, in those receiving anticoagulation therapy, and in those who refuse or are poor candidates for a surgical procedure. Because radiosurgery does not reliably relieve trigeminal neuralgia immediately, patients with acute severe trigeminal pain are not good candidates for the procedure. In this situation, percutaneous procedures should be considered if the patient is not a candidate for microvascular decompression. Patients should be informed of the potential risk of delayed facial numbness after radiosurgical treatment.

Gasserian Ganglion Stimulation

Patients with pain secondary to the damaged nerve or ganglion as occurs after some poorly performed surgery for trigeminal neuralgia or in cases of nerve infiltration by tumors, experience burning or nagging pain in the distribution of the trigeminal nerve. This is known as trigeminal neuropathy. Neuromodulation by the way of gasserian ganglion stimulation or motor cortex stimulation is the only sensible alternative for treating this pain if the medical treatment fails. This involves implantation of the electrode at the target site and delivering current to the target nerve or cortex through a pacemaker. It is presumed that this will either increase their pain threshold or block the painful impulses from the periphery to relieve the pain. This is a highly advanced form of treatment available at only select centres like Jaslok Hospital.

||Summary

When medical treatment for trigeminal neuralgia fails or is limited by significant side effects, neurosurgeons need to inform their patients of all the available treatment options. The best treatment for the patient depends on the age of the patient, medical comorbidities, and the risks the patient is willing to assume.[1]

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