TRIGEMINAL NEURALGIA – A REVIEW

Andrew J Keogh, Consultant. Rewati-Raman Sharma, Registrar,
E. Nanji, Senior House Officer, Department of Neurosurgery, Royal Preston Hospital

INTRODUCTION

In mythology, neurosurgery started in Greece with the delivery of the goddess Athena from the head of her godfather Zeus. Zeus, when pregnant in his forehead with his daughter Athena, suffered from a neuralgia of the first trigeminal branch, surely a symptomatic trigeminal neuralgia, and was cured by removal of the tumour-like girl[1]. However, in 1756, the French surgeon, Nicolaus Andre, named the condition of trigeminal neuralgia as "tic douloureux" in view of its spasmodic nature conceiving the illness in terms of convulsions[2]. The condition had long been described with probably one of the best and most dramatic description given by Dr John Locke in 1677, when he was asked to see the Countess of Northumberland, the wife of the British Ambassador to France – "I found her in a fit of such violent and exquisite torment . . . it forced her to such cries and shrieks as you would expect from one upon the rack . . . when the fit came there was . . . as it were a flash of fire, all of a sudden . . . speaking was apt to put her into these fits . . ."[3]

Trigeminal neuralgia is a condition resulting in paroxysms of pain along the course of the fifth nerve. These paroxysms of pain have been variously described as red hot pokers or electric shocks or as if needles were being inserted into the face. At its worst, the pain can be one of the most severe that can be experienced and the sight of a patient with severe trigeminal neuralgia IS one of the most distressing to witness. The incidence is reported as 4 per 100,000 of the population per year. The majority are idiopathic and occur over the age of 50. Below this age, the condition should be considered symptomatic and may be a presenting feature of multiple sclerosis (M.S.): up to 4% of patients with this neuralgia have M.S.[4] and about 1% of M.S. patients have trigeminal neuralgia. A tumour along the course of the fifth nerve such as a meningioma, a neurofibroMa or an epidermoid, can result in symptomatic pain as can vascular abnormalities such as an ectopic vessel, an aneurysm or an arterio-venous malformation, especially in, or near, the root entry zone at the brain stem.

For many years the hypothesis that a gross anatomical lesion in the form of a vascular loop causing compression on the fifth nerve root has been accepted by many to explain the idiopathic type. Dandy in 1934[4] first proposed vascular compression of the fifth nerve root as the cause, subsequently supported by many others, notably by Gardner and more recently, Jennetta[6]. Some disagree and the contrary arguments to this hypothesis are admirably summarised by Adams[7].

The diagnosis is a clinical one. The characteristics of the condition are summarised in Table 1. Some would advocate that all patients should have some form of neuro imaging – a computerised tomography (CT) scan or preferably, magnetic resonance imaging. In the majority (idiopathic trigeminal neuralgia), this is probably unnecessary, especially in the absence of neurological signs or raised intracranial pressure and in the present economic climate wasteful of limited resources.

CHARACTERISTICS OF VTH NERVE NEURALGIA

1. Paroxysms of pain contained to the 5th nerve distribution
2. Provoked by obvious stimuli with a 'trigger' spot
3. Occurring in the mouth/ear zone (60%), nose/orbit zone (30%), with less than 5% confined to 1st division
4. Rarely at night; very rarely bilaterally (and never at the same time)
5. No sensory loss, no muscle spasm
6. A response to carbamazepine – at least initially
7. Cutting or damage to the nerve giving at least temporary relief

Table 1

Most patients are best treated by carbamazepine (Tegretol) as 80% of patients will respond satisfactorily[8]. Medical management is not, however, without its hazards. Nearly all patients, given a sufficiently large dose of carbamazepine, will develop dose-related side effects of drowsiness, dizziness, blurring of vision, unsteadiness and double vision, whilst idiosyncratic side effects in the form of skin rashes, blood disorders and, rarely, aplastic anaemia can occur. Some limited success is reported with other drugs when carbamazepine is not helpful such as Phenytoin, Epilim or Baclofen, often in combination.

The reasons for initial referral for a surgical opinion are twofold viz. either inadequate or failed medical treatment. Surgical intervention is ultimately the result of the latter, either because of unacceptable side effects in the absence of total pain control or allergic skin reactions. The choice of surgical intervention depends upon the experience and interest of the surgeon.

In 1992 an audit of patients undergoing some form of surgical procedure for trigeminal neuralgia in the Neurosurgical Department at Preston was done for the following reasons:-

1. In 1990 the department was offered a place in an open multicentric drug trial (new carbamazepine analogue) designed to evaluate its efficacy, tolerability and safety in neuralgic patients who had shown a poor response to established treatments.

2. In recent years there has been a re-emergence of the view that vascular compression was the cause of the idiopathic variety with vascular decompression increasingly advocated.

3. The need to formulate a defined treatment protocol for the patients with trigeminal neuralgia, as a wide variety of procedures had been carried out in the Neurosurgery Department over the years with no particular management protocol being adopted.

METHODS

A retrospective review of 271 patients undergoing some form of surgical procedure for trigeminal neuralgia over the period 1965 to 1991 was undertaken. Three further patients presented during the period reviewed with symptomatic neuralgia related to tumour in the cerebello-pontine angle. These 271 patients underwent 395 neurosurgical procedures. No particular policy with regard to surgical management was adopted. A surgical procedure was defined as any intervention and included ganglion injection techniques (alcohol or Phenol in glycerine), radio-frequency thermo-coagulation lesions, simple operative techniques of avulsion or sectioning of peripheral nerves and major surgery in the form of subtemporal fractional root sections and more recently, posterior fossa partial sensory rhizotomies and microvascular decompressions.

RESULTS

The incidence at the time of procedure revealed a female predominance, with the majority over the age of 50 (Table 2), whilst the mean duration of symptoms prior to surgical intervention was 4.4 years, with range of 0.5 to 36 years (Table 3). Of the 271 patients, 50 (19%) (Table 4) had associated significant systemic disease influencing the choice of management.

INCIDENCE OF SURGICAL PROCEDURE FOR VTH NERVE NEURALGIA

AGE	MALES	FEMALES	TOTAL
20-29	0	2	2
30-39	3	7	10
40-49	7	23	30
50-59	27	51	78
60-69	44	88	132
70-79	53	61	114
80-89	8	29	37
TOTALS	**153**	**253**	**403**

Table 2

DURATION OF SYMPTOMS OF VTH NERVE NEURALGIA AT THE TIME OF SURGICAL INTERVENTION

AGE	YEARS	RANGE IN YEARS
20-29	2	1-3
30-39	4.8	2-10
40-49	4.5	2-8
50-59	3.9	2-14
60-69	4.4	1-16
70-79	4.5	1-11
80-89	6.4	0.5-36

Table 3

INCIDENCE OF SERIOUS ASSOCIATED MEDICAL DISEASE IN PATIENTS WITH VTH NERVE NEURALGIA

Recent M.I. or current angina/I.H.D.	16
Arterial hypertension	18
Cerebrovascular disease	5
Chest disease, eg. bronchitis, emphysema	9
Thyrotoxicosis, diabetes	2

Table 4

A. Fifty-eight patients underwent some form of injection technique into the gasserian ganglion through the face. Only t28 of these had a single effective injection whilst the remaining 30 required two or more subsequent procedures at varying intervals. In view of its selectivity in destroying pain fibres, the radio-frequency induced lesions were the method of choice latterly but the technique was abandoned for the following reasons:-

1. It was time consuming of already limited anaesthetic time.

2. It had a high failure rate.

3. There were insufficient numbers of patients to acquire the necessary skill required with this technique.

B. Seventy-eight patients underwent one or two peripheral nerve avulsions depending upon the site of the pain (supra-orbital, supratrochlear, infra-orbital, mental nerves) and 71 of these required a further procedure because of recurrence.

C. In the early years a partial root section via a subtemporal approach was used, then replaced by a posterior fossa approach, this being technically superior. One hundred and seventy patients were submitted to some form of major surgery – mean 6 patients per year, range 0-14 per year. Of these, 38 had one previous procedure, whilst 11 had multiple previous interventions.

Of 44 patients undergoing a major procedure where microvascular decompression was contemplated, only 23 had an obvious arterial abnormality. A further 14 had no obvious vascular compression, and in 7 there was a doubtful compressive loop. In this group of 44, 6% of patients had an early recurrence, that is, within the first two years, although the recurrences were controlled by medication.

DISCUSSION

The management of trigeminal neuralgia for the majority of patients is probably straightforward, their pain being controlled by carbamazepine (Tegretol). Experience shows that, if possible, a small dose should be taken to start with. This should be increased gradually. A sensible initial dose would be 100 mg 3 times a day, avoiding taking it on an empty stomach, increasing by no more than 100 mg daily with no more than 200 mg taken at any one time. At night a larger dose of 300 or 400 mg is worthwhile in order to minimise pain first thing the following morning, a not uncommon problem. In this way pain control will be achieved with a much reduced risk of dose-related side effects. If pain-control is achieved, then it is reasonable to reduce the dose slowly with a view to stopping. A readily available supply must be to hand to restart at the first sign of recurrence. Patients treated with carbamazepine by the

authors are supplied with a list of guidelines summarising the above points.

Where carbamazepine has failed, the next line of management is, with the patient's agreement, entry into the current drug (carbamazepine analogue) trial. Over the past two years some twenty-four patients have been entered into this trial with encouraging results. No definitive conclusions can yet be made as the length of the trial is two years and the local trial numbers are insufficient.

Avulsion or section of a peripheral nerve is a simple operative technique but has very limited application as few patients have pain confined to a peripheral nerve distribution – eg. less than 5% of patients present with neuralgia that is confined to the first division. There is a 100% recurrence rate within a short period of time – average pain relief after supra and infraorbital avulsion is about 33 months[9]. This technique should be confined to the elderly, frail patient who is in acute pain. The number of sections/avulsions carried out over the audit period reflected the workload, at least in part; the technique is simple, quick and gives immediate beneficial results. A high rate of sections is unacceptable in view of the 100% recurrence rate in a short time and the high risk of dysaesthesia.

Three techniques are currently practised which involve the insertion of a needle into the gasserian ganglion through the face. A radio-frequency lesion-making technique (RF thermal coagulation of the gasserian ganglion) is the one performed by most in view of its selectivity. It affects pain fibres whilst preserving touch. The reported incidence of recurrence at 5 years is 25%, or approximately 20% over a 1-10 year follow-up. A lower recurrence rate is associated with a higher dysaesthesia rate – occurring in approximately 5% of patients overall. A large number of other problems have been described, including facial sensory loss (between 1 and 5%) with dangerous corneal anaesthesia in 4% of patients. Others include cranial nerve palsies, carotico-cavernous fistulas, brain abscesses and meningitis and intracerebral haemorrhage. Overall, these complications probably occur in less than 1% of patients[10]. The wide variety of complications reported reflects the large number of patients involved. The technique requires skill and experience. Many patients consider it unfriendly; they may need to be woken several times for testing in the course of one treatment.

The injection of glycerol into the trigeminal ganglia is easier than radio-frequency coagulation. The recurrence rate, however, is higher at 5 years, being 30-45%. There is less dysaesthesia than with RF lesions, no significant facial sensory loss and it does not need expensive equipment, as well as being patient-friendly requiring one anaesthetic without having to wake the patient up for testing. In the best hands, there is a 10% immediate failure rate. In view of the high recurrence rate, and to a lesser extent, the significant, albeit small, dysaesthesia rate, many people have given up this technique[11].

The third injection technique is that devised by Mullen[12] viz. percutaneous microcompression (PMC). This is easily learnt, and has a recurrence rate at 5 years of some 20-25%. The dysaesthesia rate depends upon the compression time (ie. the less compression, the lower the rate.) No facial sensory loss is reported (although it can occur in the authors' experience) and is both patient- and operator-friendly, again requiring one simple and short anaesthetic. Overall, this technique seems to be the one that should be preferred by the occasional operator. It has been the technique adopted following the audit in this department (Fig. 1). This consists of inflating the balloon of a Fogarty catheter at the foramen ovale and maintaining the pressure on the ganglion for one to five minutes. In a small number of patients bradycardia, sudden hypotension and, rarely, a sudden cardiac arrest, are the real risks.

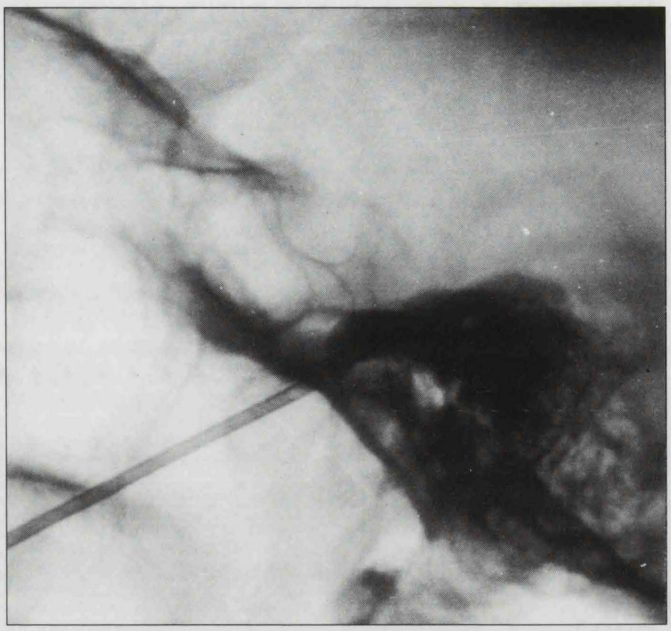

Figure 1 – Lateral skull x-ray showing needle at margin of foramen ovale with the distorted Fogarty catheter balloon compressing the trogeminal ganglion.

The major surgical technique adopted by most operators is that of microvascular decompression (MVD). It should be borne in mind that in view of its nature viz. a post fossa craniotomy, it carries a significant mortality and morbidity of some 1% each. Dysaesthesia is much less likely, although the reported recurrence rate at 5 years varies from 6% to 30% or more[14]. In some patients no vascular compression of the nerve root can be demonstrated at surgery. Under these circumstances, most would carry out a partial rhizotomy although this carries an even higher 5-year recurrence rate.[14] Examination of patients pre-operatively by MRI may identify those with an obvious vascular loop[15] and, therefore, those likely to benefit from a decompression. This is the subject of a current research project within the department.

In those patients undergoing a partial rhizotomy, a higher recurrence rate of 50% over 5 years equivalent to 3.5% per year, is reported[13]. There is clearly a danger of dysaesthesia occurring and, of course, of sensory facial loss dependent on the amount of nerve root sectioned. For both major procedures of MVD and rhizotomy, 85% of patients overall appear to have a good immediate response. There is about a 5% immediate failure rate following MVD.[16]

CONCLUSIONS

1. Patients with the condition need ready access to professional help in the event of recurrence. This may vary from mere advice about medication, to an emergency admission depending upon the severity of the pain.

2. There are many treatments available for patients with trigeminal neuralgia and no one method is ideal.

3. No standard protocol of management has yet been published, although the following is current practice.

a) The first step is carbamazepine therapy. If this fails, patients are entered in the drug trial.

b) Failed medical treatment leads to a percutaneous microcompression.

c) The next step, (bearing in mind that there are only small numbers – perhaps a mean of six patients a year) – is a posterior fossa surgical approach with microvascular decompression or partial rhizotomy dependent on the findings. The current research project utilising MR may help to distinguish those patients most likely to benefit from MVD.

4. The danger of dysaesthesia cannot be over emphasised, as should it develop, it is virtually impossible to treat and may be more disabling than the original neuralgia. It should also be remembered that true neuralgia can occur even in the presence of a dysaesthesia. Injection techniques can be successfully repeated, although a re-exploration to carry out a partial rhizotomy or a further microvascular decompression can be undertaken. Either may be very difficult because of previous surgery.

REFERENCES

1. Loew F. History of cranial nerves surgery. Introductory lecture. In: The cranial nerves. Samii M, Jannetta PJ (Editors), Springer-Verlag Berlin Heidelberg New York 1981 pages 1-5.

2. Andre N. Observations pratiques sur les melodies de urethre. Paris: Delaguette 1756.

3. Dewhurst K. A symposium on trigeminal neuralgia with contributions by Locke, Sydenham and other eminent seventeeth century physicians. Jon Hist Med Allied Sci. 1957; 12: 21-36.

4. Dandy WE. Concerning the cause of trigeminal neuralgia. AM J Surg 1934; 24: 447-455.

5. Gardner WJ. Trigeminal neuralgia. Clin Neurosurg 1968; 15: 1-56.

6. Jennetta PJ. Treatment of trigeminal neuralgia by micro-operative decompression, in Youmans JR (Fd). Neurological Surgery, Ed II Philadelphia; WB Saunders 185; Vol 6 pages 3589-3603.

7. Adams CBT. Microvascular compression: An alternative view and hyopthesis. J Neurosurg 1989; 70: 1-12.

8. Rasmussen P, Ritshede J. Facial pain treated with Carbamazepine (Tegretol). Acutre Neurol Scand 1970; 46: 385-408.

9. Grantham EG, Segerberg LH. An evaluation of palliative surgical procedures in trigeminal neuralgia. J Neurosurg 1952; 9: 390-392.

10. Rovit RL, In Rovit RL, Murali R, Jennetta PJ Percutaneous frequency coagulation. Trigeminal Neuralgia Williams & Williams; Baltimore 1990: 128-134.

11. Young RF. Glycerol rhizolysis for treatment for trigeminal neuralgia. J Neurosurg 1988; 69: 39-45.

12. Mullan S, Lichtor T. Percutaneous microcompression of the trigeminal ganglion for trigeminal neuralgia. J Neurosurg 1983; 59: 1007-1012.

13. Klun B. Microvascular decrompression and partial sensory rhizotomy in the treatment of trigeminal neuralgia: Personal experiences with 220 patients. Neurosurg 1992; 30: 49-52.

14. Burchiel KJ, Clarke H, Haglund M, et al. Long-term efficacy of microvascular decompression in trigeminal neuralgia. J Neurosurg 1988: 69: 35-38

15. Wong BY, Steinberg GK, Rosen L. Magnetic resonance imaging of vascular compression in trigeminal neuralgia. J. Neurosurg 1989; 70: 132-134.

16. Bederson JB, Wilson CB. Evaluation of microvascular decompression and partial sensory rhizotomy in 252 cases of trigeminal neuralgia. J Neurosurg 1989; 71: 359-367.

ACKNOWLEDGMENT

To the Medical Illustration Department of Royal Preston Hospital for the photographs and to Mrs L Freeman for the preparation of this manuscript.