Current Practice

PRACTICAL NEUROLOGY

Facial Pain

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Accurate neuropathological rather than topographical diagnosis must always depend on an adequate and searching history. It is always rewarding to demonstrate abnormal signs which indicate the site of the lesion and confirm the existence of true structural disease of the nervous system when the history has suggested this.

There are, however, several symptoms which a patient may describe in a neurological clinic where no such abnormalities are to be found, and where the disease process must be assessed from the patient's description of symptoms alone and the environment in which they arise. This is often true in the differential diagnosis of episodes of unconsciousness and in the assessment of migraine (which is the subject of a separate article in this series), and it is especially true of patients complaining of pain in and about the face. In the majority of these there will be no abnormal signs to suggest the situation and nature of a pathological process. The family doctor is the first to see and assess such a patient, and on his appreciation of the nature of the complaint will largely depend the early relief obtained through the administration of drugs or from surgical or other technical procedures.

Associated Lesions

When pain in the face is associated with evidence of a physical defect—for example, accompanying numbness or double vision—examination may reveal signs indicating the presence of nasopharyngeal carcinoma—blood-stained discharge, proptosis, and ophthalmoparesis. An acoustic neuroma is suggested by perceptive deafness and blunting of the corneal reflex, with sensory impairment in the trigeminal distribution on the same side, and, perhaps at a very late stage, facial palsy and signs of cerebellar dysfunction. A bruit over the neck and orbit may suggest that the pain is due to compression of the trigeminal nerve by a large carotid aneurysm. More simply, examination of the tongue may show a carcinomatous ulcer responsible for pain in the mouth and lower jaw.

The more difficult cases are those in which facial pain is not associated with detectable abnormality. Pain in the face is a common cause of referral to neurological clinics. Among these patients there are some whose pain is clearly identified with one or other of the defined neurological syndromes—for example, trigeminal and glossopharyngeal neuralgia, migrainous neuralgia, and the pain after herpes zoster affecting the first division of the trigeminal nerve. In another large group the pain is atypical of these conditions and has been so designated. Drug-responsive depressive psychosis is evident in some patients with this atypical pain, but the majority cannot be helped and continue to complain whatever treatment is advised.

Trigeminal Neuralgia

Essential to the management of this condition is its accurate diagnosis. Surgical treatment of other forms of facial pain

may lead to so-called anaesthesia dolorosa, which may prove much more troublesome than the original atypical facial pain for which surgical treatment was advised. Recognition of trigeminal neuralgia depends on an accurate history of pain, usually confined to the second or third division of the trigeminal distribution in the face, strictly unilateral, and occurring in sharp paroxysms which cause the patient to wince and retract from any noxious stimulus. This may be trivial, such as touching the face, shaving, brushing the teeth, chewing, or swallowing; in severe cases even a draught on the face may trigger a short, sharp paroxysm of pain. The typical sufferer from trigeminal neuralgia cannot be missed, for he will resent examination, since this will produce further paroxysms; he may well have lost weight because of his reluctance to chew and swallow, and in the severe case there may be neglect of the toilet of the affected half of the face, which remains unshaven and dirty. This may be accompanied by unilateral furring of the tongue. On examination there is no evidence of trigeminal nerve damage; the corneal reflex will be intact, and there will be no evidence of sensory or motor impairment of the nerve.

Occasionally pain similar to trigeminal neuralgia will be seen in the course of tabes dorsalis, occasionally a diabetic may have facial pain, and occasionally, also, trigeminal neuralgia typical in other respects may be a presenting feature or part of an exacerbation of demyelination in the brain stem—in multiple sclerosis, for example. However, these are unusual presentations occurring in the young; the majority of cases arise in old age or late middle life, when the condition is not easily mistaken.

The natural history of trigeminal neuralgia is for a short series of paroxysms to abate and allow the patient a period of freedom from pain. Unfortunately and inevitably the pain returns, and the periods of relief become shorter, until the patient suffers from persisting repeated paroxysms. It is generally accepted that the condition does not remit spontaneously, and hence treatment is essential in the early and later stages.

Frequently the patient is seen during a remission, and, indeed, the majority of patients by the time they are referred to hospital have entered such a phase. Treatment is then unnecessary, but on the first return of the tic treatment must be instituted immediately.

Treatment

Until recent years the treatment of this condition was essentially surgical: injection of the gasserian ganglion by alcohol or phenol or section of the trigeminal sensory root. However, since the introduction of carbamazepine (Tegretol) the majority of patients can be relieved by medical means. Carbamazepine,

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an anticonvulsant and an analogue of imipramine, has proved to be more effective than anticonvulsants previously used in the treatment of trigeminal neuralgia, such as phenytoin and its derivatives. The drug is made up as a white tablet containing 200 mg. of the active principle and is quite specific for tic douloureux. It can be used almost as a diagnostic test, since if a patient suffers true tic douloureux he is likely to be relieved within hours of administration of the drug. The usual practice is to begin with a small dose, 100 mg. t.d.s., and increase this to a maximum of 200 mg. 6 times a day. After relief it is possible to tailor the dose to the patient's individual requirements and to maintain freedom from pain during a period of exacerbation.

Some 70–80% of patients will be relieved of trigeminal neuralgia by carbamazepine, but unfortunately side-effects are not infrequent, and patients may complain of giddiness, drowsiness, dryness of the mouth, diarrhoea, nausea, and vomiting. These side-effects can be minimized by introducing the drug gradually, as suggested, and in some patients tolerance to the side-effects may be successfully established. To date, two fatal cases and one non-fatal case of aplastic anaemia have been reported in elderly women, but this extremely unusual and infrequent complication should not detract from the drug's usefulness in the management of a complaint which carries a very high suicide risk.

Carbamazepine should not be used when monoamine oxidase inhibitor drugs have been used recently, perhaps in a patient whose pain has brought about a reactive depressive illness. The advent of this drug and the relief patients obtain from it have rendered surgical treatment of trigeminal neuralgia necessary only in a minority.

Surgical Treatment

Prior to the introduction of carbamazepine, injection of the gasserian ganglion was often the preferred form of treatment, but there are now fewer neurologists and neurosurgeons in Britain skilled in this difficult technique. So, unless a patient is referred to centres specializing in gasserian injection, he will probably be best served by trigeminal root section, the sensory root being divided, usually in the middle fossa and occasionally in the posterior fossa. Division of the descending tract of the fifth nerve in the brain stem—intramedullary trigeminal tractotomy—is very rarely performed today. Root section is thus confined to those patients who are unable to tolerate carbamazepine or the small proportion in whom classical trigeminal neuralgia is non-responsive to the drug.

In the past, and indeed recently, there have been advocates of local peripheral neurectomy. Pain confined to the first or second divisions may be relieved by supra-orbital or infra-orbital neurectomy, and pain in the third division may be relieved by submental nerve division or division of nerve filaments in the alveolus. Undoubtedly patients have obtained considerable relief by these local procedures, but it is the experience of most neurologists and neurosurgeons that the relief is transient and very soon the pain recurs. Local neurectomy may have its place in patients who are too old or very rarely who constitute too severe an anaesthetic risk for complete root section, but there should be no doubt in the practitioner's mind that the effect is temporary and that the true tic will return within 9 to 18 months of peripheral neurectomy.

Whatever the operative procedure advised, whether it be avulsion of a peripheral branch or alcohol injection of a peripheral branch, or indeed injection of the gasserian ganglion with alcohol or elective partial or total root section, there is always a certain risk of complications. Frequently after operation herpes simplex eruptions occur in the anaesthetic area. Facial paralysis is occasionally seen. Persistent paraesthesiae may be troublesome, but in every case there will be an area of partial or total hemifacial anaesthesia of which the patient must

be made aware before the operative procedure is undertaken. All patients willingly accept the prospect of a numb face in exchange for trigeminal neuralgia, but they must understand that it will be permanent and what this implies in terms of disability. Operations are planned to spare the ophthalmic division of the trigeminal nerve so as to prevent corneal anaesthesia and possible keratitis. After a procedure in which the corneal reflex is lost great care must be taken of the anaesthetic cornea, since infection is very readily introduced and spreads rapidly. It has been estimated that some 5% of cases subjected to root section develop anaesthesia dolorosa, an unpleasant burning, tingling sensation in the anaesthetic face. This condition is intractable, and its development is therefore disastrous for the patient, since he is subjected to a constant painful numbness of the face in exchange for his trigeminal neuralgia.

Fortunately, in skilled surgical hands trigeminal root section is effective, and only a minority of patients who have required surgical treatment suffer any significant complication from the operation.

Glossopharyngeal Neuralgia

Very similar in character to tic douloureux, but differing from it in location and in having a preponderance in the male, is glossopharyngeal neuralgia, which was first described by Weisenburg in 1910. Though the first case was associated with a tumour of the cerebello-pontine angle, most patients have no recognizable structural abnormality. This disorder is not confined to the elderly, but it is much commoner in later life. The pain, which is similar to that of trigeminal neuralgia in character—sharp and lancinating and momentary only—occurs in the base of the tongue and fauces on one side of the mouth and may also radiate deep into the ear. The pain is frequently severe enough to prevent a patient from swallowing and chewing, and these two actions, together with coughing, often precipitate the pain. Recognition is difficult only because of the condition's rarity.

Unlike true tic, glossopharyngeal neuralgia is not always relieved by carbamazepine, though some cases do respond. In the absence of a structural cause for the condition the treatment is surgical, and consists in avulsion of the glossopharyngeal nerve in the neck, or, more effectively, intracranial section of the ninth nerve and upper two rootlets of the vagus nerve. This more complete operation is not associated with the recurrence of the pain in the ear which sometimes occurs after cervical avulsion.

Post-herpetic Neuralgia

It is estimated that one patient in every 10 infected by herpes zoster who has clinical shingles will suffer from post-herpetic neuralgia. When the herpes has affected the first division of the trigeminal nerve neuralgia may be experienced in its most severe form. Because of the structural change induced in the affected skin, the diagnosis is not usually difficult, unless there has been little permanent scarring of the skin. The pain may precede the rash and reach its height when the rash is in its vesicular stage.

It is accepted that post-herpetic neuralgia, especially after herpes ophthalmicus, is much more commonly seen and is more persistent in the elderly than in the young, and, indeed, the approximate natural history of the condition can be estimated from the age of the patient. It may at times be so severe and so intractable as to lead to suicide. Unfortunately local surgical measures are not effective.

Zoster affects not only the peripheral part of the nervous system but also central structures, so that the lesion should probably be regarded as "central" in type. Many procedures, such as undercutting the affected skin, peripheral neurectomy,

rhizotomy, and cordotomy have been used for post-herpetic neuralgia without effect, and even leucotomy and thalamotomy have proved ineffective in some severe cases. The treatment is essentially medical. It has been suggested by some authors that the early use of A.C.T.H. by injection or steroids by mouth in large doses may prevent the onset of severe post-herpetic neuralgia. This, however, is by no means certain, and unfortunately the degree of neuralgia to be anticipated cannot be estimated early in the clinical course of the inflammatory disorder.

After the establishment of the pain a background of chlorpromazine 25 to 100 mg. six-hourly may potentiate the effect of simple analgesics such as paracetamol, physopetone, and codeine phosphate. In the severe case these often prove ineffective, and more potent analgesics are often prescribed despite the risk of habituation. This factor is of no consequence in the elderly, but must be borne in mind when a younger person or a person in late middle life suffers from the disorder. Symonds and more recently Miller¹ have advocated the use of a powder containing 8 mg. of heroin and 650 mg. of aspirin. The powder is taken before retiring and later in the night if necessary. This useful analgesic may allow the patient a good night's sleep, though he will continue to suffer severe diurnal pain.

Undoubtedly one of the most successful forms of treatment is that originally advocated by Ritchie Russell.² The vulcanite end piece of a vibrator (Pifco massager) is applied to the affected area. Hyperaesthesia can be abolished by the repeated use of this vibrator, and, though the pathophysiological basis of this treatment is uncertain, certainly some patients are relieved. They should be advised to use the instrument hourly for five to ten minutes every day. Central pain, built up because of central facilitation, can be relieved, it is suggested, by persistent bombardment of the sensory pathways by means of the vibrator. Whatever the rationale, the treatment can prove very effective, and in those patients in whom the area is extremely sensitive prior local analgesia can be obtained with a spray of a local cooling agent.

Migrainous Neuralgia

A certain type of headache which has been eponymously described and given alternative descriptive terms such as ciliary or sphenopalatine neuralgia, histamine headache, cluster headache, or migrainous neuralgia, is readily recognized from the history of its development and progression. The patient is usually middle-aged and male, without a family history of true migraine. He suffers episodes of headache, the pain of which frequently wakens him from sleep in the early hours of the morning. He describes pain centred above and behind the eye, of a deep boring character, often associated with lachrymation and a feeling of stuffiness in the nostril on the same side. The pain usually does not respond to simple analgesics and persists for some two to three hours, after which it abates, only to return the following night and so to continue for two to three weeks, producing a cluster of typical attacks. The paroxysms may be seasonal, occurring in the spring or autumn, or more frequently they occur at random, and the periods of freedom become shorter. The pain is extremely severe and is quite unlike the neuralgias, tic douloureux and glossopharyngeal neuralgia, in that it is constant and boring. It does not have the throbbing character of true migraine, nor its concomitants, a visual or systemic aura, nausea, and vomiting. As in the other neuralgias, there is no abnormality on examination.

This form of facial neuralgia is frequently misdiagnosed, and the patient subjected to unwarranted dental treatment, ill-advised antral toilet, and even to carotid angiography before the history is accepted as that of migrainous neuralgia. Simple analgesics are of little value, and by far the most effective regimen is to inject the patient or to teach the patient to inject himself with intramuscular ergotamine tartrate, 0.5 mg. three

times per day, for a period of 10 days. The only contra-indications to this are pregnancy, which is extremely unlikely in the type of patient suffering from this disorder, and vascular disease. This treatment may prove very helpful and can be used by the patient in his successive episodes. If ergotamine tartrate fails, methysergide 1–2 mg. t.d.s. may give relief. Side-effects are quite frequent, and there is always the danger of vascular complications. Though this compound is extremely useful in the long term prophylaxis of true migraine in younger subjects it should be used with caution in middle-aged and elderly males suffering from migrainous neuralgia. Our experience with other forms of treatment is limited. Stellate ganglion block with a local anaesthetic may interrupt a series of attacks of migrainous neuralgia, suggesting that stellate ganglionectomy may prove more permanently effective. There are some patients who have been relieved by this procedure at the price of a ptosis and small pupil on the affected side.

Temporo-mandibular Neuralgia

Seen most commonly in middle-aged women, this form of neuralgia is thought to be due to overclosure of the bite in the edentulous patient. It is produced and aggravated by chewing, and consists of severe exacerbations of deep pain in and about the temporo-mandibular joint passing down the distribution of the third division of the trigeminal nerve. The patient often comments on the association between the onset of pain and movement of the jaw, and there may be some tenderness over the joint. Dental treatment is required, and various prostheses have been devised to readjust the degree of movement allowed at the joint to prevent overstrain and thus to relieve this type of neuralgia.

Atypical Pain

In a large proportion of patients suffering from pain in the face the above clear-cut entities can be readily identified. The remainder form a mixed collection of patients whose pain has proved intractable to analgesics and other therapeutic procedures. These constitute the patients suffering from atypical facial pain. Among them there will be fairly characteristic sufferers from depressive psychosis, recognizable by the evident depressive setting in which the pain has arisen. They must be distinguished from those patients whose pain has produced a reactive depression.

Undoubtedly these patients do respond to monoamine oxidase inhibitors, and for some years patients with atypical facial pain have been given 15 mg. of phenelzine together with trifluoperazine 1 mg. t.d.s. with evident relief. More recently it has been the practice, albeit without the more sophisticated rationale demanded by our psychiatric colleagues, to use tricyclic compounds such as imipramine 25 mg. t.d.s. or amitriptyline 25 mg. t.d.s.

Unfortunately there remains a group of patients, usually women, who have used successively a series of analgesics, anti-depressives, family doctors, the dental department, the department of psychiatry and neurology, yet continue to complain of intractable, severe, constant facial pain and for whom we are unable to offer effective treatment. With these patients the best we can do is to spend time with them, offering them sympathy and freely discussing their complaint, its nature and difficulties. This support, which implies frequent attendance, may carry them along and enable them to function in their particular domestic environment. It is in this group of patients that injection or surgical intervention may prove disastrous, and in whom induced anaesthesia dolorosa becomes the focal point of their miserable lives.

REFERENCES

- ¹ Miller, H. G., *British Medical Journal*, 1968, 2, 577.
- ² Russell, W. Ritchie, *Lancet*, 1957, 1, 242.