Papers and Originals

Pain in the Face*

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In the neurological outpatient clinic there are some presenting symptoms that daunt the physician, and others that immediately sharpen his interest. A complaint of dizziness, for example, falls into the first category. It means so many different things, from cataclysmic aural vertigo to a middle-aged woman's vague feelings of anxious insecurity in a crowded shop, while the pathophysiological basis of the symptom is poorly understood. The physician knows that even after a careful history and examination, and the employment of special tests, it is quite likely that he will find himself unable to account for the clinical phenomena in a really satisfactory way. Among the second category of primary symptoms that immediately arouse the neurologist's interest, pride of place must go to episodic attacks of loss of consciousness, usually comprehensible on a careful clinical history alone, and a never-failing source of fascination. Among these another is pain in the face.

A neurologist in a general hospital, and especially one associated with a dental hospital, sees many patients with this complaint. Our dental colleagues have usually dealt with the dental aspect of cases they refer to us, and these are often difficult and unusual. Unfortunately specialists in other fields are sometimes less well informed, and I have seen an old lady with trigeminal neuralgia tube-fed for days in a peripheral hospital because the nature of her agonizing and remediable pain had completely escaped recognition.

The definitive diagnosis of facial pain is not always easy, and indeed, if it were, the subject would lack its clinical interest. My account will begin with a number of more or less well defined syndromes and progress to more confused and problematical categories.

Trigeminal Neuralgia

Of all forms of facial pain trigeminal neuralgia is the most clear-cut and characteristic. It is a disease of the elderly. Curiously enough, in tropical countries the mean age of onset appears to be distinctly lower, but so, of course, is the expectation of life—perhaps this represents nothing more than the telescoping of the patient's life-history. Here, however, it most characteristically affects patients in the sixties, seventies, and eighties. The pain is unilateral and entirely limited to the trigeminal distribution, and practically always first affects either the second or third division of the nerve, spreading only later, if ever, to the first division ; it is most exceptional for trigeminal neuralgia to begin with pain in and around the eye. It comes in a succession of short, sharp, momentary bursts like electric shocks or machine-gun fire. Mild but otherwise typical cases are uncommon, and the pain is usually excruciating, immobilizing the patient's face except perhaps for agonized twitching (hence the name tic douloureux), and sometimes causing him to press his hand to his face with an expression of profound distress. Periods of complete relief are continually interrupted by volleys of pain that are always a reflex response to some form of sensory stimulation—speaking, eating, touching the face, especially at a "trigger zone" such as the nostril or the angle of the mouth. A loud noise or a cold wind may have a similar effect ; since the pain is reflex it rarely disturbs sleep. A series of such episodes may occur many times a day for several weeks, to disappear completely for many months. But the pain returns, extending more widely—though still strictly within the fifth-nerve distribution—in the course of time, and with remissions that become briefer. The word "never" is unknown to medicine, but I have yet to see spontaneous cure of this disorder, and this common experience profoundly influences our approach to treatment.

The age of the patient, the character and distribution of the pain, and the remittent history are invariable features of trigeminal neuralgia, but there are others almost equally striking that must be satisfied for a firm diagnosis. The most important of these is that *there are no physical signs, either sensory or motor, of a structural lesion of the fifth nerve.* If loss of sensation or impairment of the corneal reflex is present the condition is not trigeminal neuralgia: the nerve is affected by structural disease such as the pressure of an eighth-nerve tumour or an aneurysm, or erosion by a nasopharyngeal cancer.

Though most cases of trigeminal neuralgia arise in old age or late middle life, and though the diagnosis in younger patients is usually incorrect, the condition is occasionally seen in its classical form in young people. In such cases the neuralgia is not infrequently accompanied by signs of pathological involvement of the long tracts of the spinal cord, especially in the form of absent abdominal reflexes or extensor plantar responses, and is a symptom of multiple sclerosis. Most often neuralgia develops during the course of the chronic disease and behaves exactly like the idiopathic syndrome. Very occasionally, like facial palsy, it may herald multiple sclerosis. In my experience this chronic nervous disease also furnishes most of the few cases where trigeminal neuralgia is bilateral, affecting either side alternately. Treatment of the condition in multiple sclerosis is exactly the same as in other cases.

Treatment

Except for a handful of enthusiastic experts, alcohol injection of the Gasserian ganglion has lost its vogue—the technique is difficult and the result unpredictable. Nor has division of the sensory tract in the midbrain achieved much popularity in Britain, the operation being regarded as unduly hazardous. Treatment by partial section of the sensory root of the fifth nerve is the most widely practised approach. Attempts to limit the extent of fibre section to spare the first division of the nerve, and hence to avoid corneal analgesia with its attendant risks, imply a small risk of postoperative recurrence.

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Furthermore, the operation leaves a sometimes disagreeable numbness of the face. The patient with severe pain is glad to exchange it for sensory loss, but where the neuralgia has been in remission, or relatively mild, he may complain as bitterly about the postoperative result as he did about the condition for which it was performed. Nor does he share the doctor's awareness that his condition will certainly recur. For this reason the patient with trigeminal neuralgia should not be put on a waiting-list, but advised to report back for operation during an attack, so that he will be fully aware that his severe pain has been cured at the cost of mildly troublesome paraesthesiae.

The introduction of Tegretol (carbamazepine) has transformed the therapeutic situation in this disorder, and already surgical hands are prone to lose their cunning in sensory root section. Carbamazepine is an imipramine-like drug, originally introduced as an anticonvulsant, and it has a remarkably specific effect on the pain of trigeminal neuralgia, which has resisted virtually every previous analgesic. The effect is so specific that in outpatient practice we now use the drug as a diagnostic test. It is toxic, and often causes headache, dizziness, and drowsiness, but there few patients who cannot tolerate it if the dose is gradually built up over the course of a few weeks from half a tablet (100 mg.) to something like six tablets daily.

Early reports of blood dyscrasias as a result of the administration of carbamazepine caused considerable alarm, made neurologists timid in its use, and have so far precluded its introduction into medical practice in the United States. However, the complication is rare and one that must be faced with many effective modern drugs. It constitutes no contraindication to the use of this preparation in trigeminal neuralgia, since the small risk of surgery even in the most skilled hands remains greater than the danger of the very occasional catastrophic complications of carbamazepine. Many of these patients are elderly and decrepit, and operation and anaesthesiae of any kind carry some risk. The tendency to spontaneous remission means that few patients will need the tablets continuously throughout life, and if response is satisfactory, and if a dose effective enough to preclude more than a twinge of pain in the morning (before the first tablet has been taken) can be tolerated, operation can be avoided. In the younger patient the situation is different. The risks of toxic complications increase with the amount and duration of dosage, and in severe cases successful operation will avert the risk of drug toxicity during the many years that lie ahead.

In practice, the introduction of carbamazepine has led to a remarkable reduction in the number of operations undertaken for trigeminal neuralgia: recent reports of its use describe successful treatment of trigeminal neuralgia in about 75% of cases. Today the main indication for surgery is the patient's inability to tolerate effective dosage without uncomfortable minor toxic symptoms. In this connexion, and especially in older patients, there is something of a swing back to less radical surgery, involving perhaps division of the submental nerve when the main trigger zone is in the lower jaw. Since (unlike alcohol injection of the ganglion) these measures in no way jeopardize later resort to root section where this proves necessary, there is nothing against such a limited peripheral approach.

Glossopharyngeal Neuralgia

The only condition other than tic douloureux in which I have found carbamazepine to be effective is its very much rarer analogue affecting the glossopharyngeal nerve. This condition does not share the predilection of trigeminal neuralgia for old age, but it is similarly intermittent and causes agonizing paroxysmal pain of identical quality in the ipsilateral tonsillar bed and often also in the auditory meatus. The condition is not well known, and all my own cases have been subjected to long and ineffective courses of psychotherapy for "hysteria" before somebody—usually a competent psychiatrist—has tumbled to the correct diagnosis. Carbamazepine may be remarkably effective, but, if it is not, the condition can be cured either by avulsion of the glossopharyngeal nerve in the neck or by the rather more major procedure of its division in the posterior fossa.

It is curious that we have no clear idea of the pathology either of trigeminal or of glossopharyngeal neuralgia. In the case of the former its age-incidence suggests a relation with arteriosclerosis, but if the syndrome is ischaemic it is strange that irritative phenomena do not at some stage give place to the objective sensory deficit that would bespeak infarction.

Post-herpetic Neuralgia

Post-herpetic neuralgia is a complication of shingles in the elderly, and hardly occurs before middle-age. Usually seen at its worst where the eruption has been severe, it may of course affect any part of the body, but many of the most distressing cases involve the trigeminal distribution, causing intense facial pain. Diagnosis is not often difficult, except in the occasional instance where the skin eruption has escaped notice and may be evident only as one or two small insensitive tissue-paper scars within a wider area of hyperaesthetic skin.

At its worst the pain may be as severe as tic douloureux, but its behaviour is quite different. Severe pain arises spontaneously, without reflex excitation, and persists steadily for half an hour or more, in contrast to the paroxysmal bursts of trigeminal neuralgia, while it also often disturbs sleep. Some patchy loss of superficial sensation is usually present at the site of the healed eruption. But the most important difference between the two conditions is that post-herpetic neuralgia tends to improve, and indeed practically always does so, even if the process takes 6 to 18 months—except for a handful of cases in which the patient denies improvement over several years.

The natural history of the disorder determines its management. In this connexion it is important to remember that while the maximal incidence of pathological change in zoster infection is seen in the posterior root ganglia, the infection of the nervous system is general, and the sensory pathways especially are involved at every level up to and including the thalamus. This is probably why injection or division of the sensory root of the trigeminal or any other nerve rarely relieves the pain, and is contraindicated. Posterior rhizotomy and even prefrontal leucotomy have proved equally ineffective. Indeed, the only surgical measure that has brought relief in the most obstinate cases is stereotaxic thalamotomy—and even this is all too often temporary.

Whether or not the picturesque hypothesis of reverberating pain circuits following facilitation consequent on initially extreme stimulation is more than a fancy, there is a strong clinical impression that the condition arises most severely when control of the pain of the acute attack has been inadequate—which in a disorder so well known to cause intense pain is in any case unforgivable. The older patient with an acute attack of painful shingles deserves powerful analgesics. The observation that subsequent neuralgia is often most severe where there has been extensive superficial tissue destruction has led some to advise the use of steroids in the acute phase, though by others these are regarded as contraindicated on the theoretical grounds that they might diminish protection against spread of the virus. The employment of an antihistamine drug might represent an acceptable compromise.

By the time the patient with post-herpetic neuralgia presents as a case of facial pain the problem is one of analgesia. A background of 50 to 100 mg. of chlorpromazine three times a day (or even more) potentiates the action of an analgesic. During the day compound codeine tablets or something similar may be enough, constipation being kept at bay by Senokot Even at night 50 mg. of pethidine or 5 mg. of Physeptone

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(methadone hydrochloride) may suffice, but in severer cases I follow Sir Charles Symonds in prescribing a powder comprising ⅛ gr. (8 mg.) of heroin and 10 gr. (0.65 g.) of aspirin, one powder to be taken before retiring and a second kept by the bedside in case of need. This preparation can be virtually guaranteed to give the patient the reasonable night's sleep that will make daytime pain or discomfort at any rate tolerable. The truism that addiction is a function of the addict rather than the drug is supported by long experience of this regimen: the patient herself usually takes the initiative in cutting the dosage and finally abandoning the powders when they are no longer needed. If the choice were put to me I would in any case rather have a cheerful pain-free 80-year-old addict than a miserable ancient dying exhausted by pain, sleeplessness, and malnutrition—but I have never had to make the choice. The attitude of some members of the profession to heroin can only be described as neurotic. The drug has saved some of these unhappy patients' lives, and to dispense with an invaluable preparation because it is abused by a handful of people is as sensible as to prohibit the distillation of whisky on similar grounds.

Migrainous Neuralgia

Among other well-defined causes of facial pain, one of those least familiar to the generality is the condition variously known as migrainous, ciliary, or sphenopalatine neuralgia, "histamine headache" (on the dubiously relevant basis that the injection of histamine may provoke a typical attack in a sufferer), and—perhaps best of all—"cluster headaches." This vivid transatlantic term epitomizes the characteristic tendency of the syndrome to remit completely for many months and to recur in the form of a series of frequent attacks over the course of a few weeks.

The clinical picture is remarkably consistent. The typical patient is a young or middle-aged man, sometimes with a family or past history of true migraine, subject to attacks of fierce facial pain, centred behind the eye, always on the same side, often with running of the eye and stuffiness of the nostril on the side of the headache. Attacks often disturb sleep, sometimes with the regularity of an alarm-clock, and last for 30 minutes or more with an intensity that may make a phlegmatic man weep with pain.

Migrainous neuralgia thus shows features suggestive both of migraine and of tic douloureux, and if they are remembered they will protect many patients from unnecessary angiography and air encephalography—procedures sometimes undertaken in such cases where clinicians have failed either to take a careful history or to appreciate its significance. The effect of ergotamine tartrate on migrainous neuralgia is virtually specific, and more impressive than in the case of migraine itself. Such patients may be severely incapacitated, and it is often wise to admit them to hospital for a few days so that they can be taught to inject subcutaneous Femergin (ergotamine tartrate) (0.5 mg. three times a day, or on retiring in purely nocturnal cases) for a week or so. This almost invariably stops the episode at once, and can be repeated by the patient when the pain recurs—as it almost certainly will—after an interval of many months. A very occasional case is resistant, and in these cases the possible efficacy of stellate ganglionectomy can be assessed by the response to stellate ganglion block during a paroxysm.

Until I discovered that Wilfred Harris carried out this procedure for the same condition nearly half a century ago I regarded it as a departmental contribution to therapeutics. Our own experience with this method has been remarkably successful, but reports from reliable colleagues indicate that we have been lucky, and that something like 50% success is the most that can be expected. The nature of migrainous neuralgia remains uncertain. However, the evidence suggests that it arises from an anomaly in the innervation of the carotid circulation, and that ganglionectomy relieves the pain by interrupting the autonomic pathway.

Costen's Syndrome

Named after an American dental surgeon, this syndrome comprises pain originating in the region of the temporomandibular joint and radiating forward over the cheek and face, brought on by chewing. The pain is sometimes severe, but likely to be confused with trigeminal neuralgia only if history-taking is either slipshod or uninformed. In contrast with tic douloureux the pain is activated only by chewing and not by any of the other familiar "triggers" of the more serious disorder, while the temporomandibular joint on the affected side is often tender on firm pressure.

Costen's syndrome is associated with over-closure of the bite or with the change in shape of an edentulous jaw, and is caused by sliding forward of the mandibular condyle with consequent arthropathy. It may be relieved by "building up the bite" with plastic prostheses placed over the lower molars. The benefit yielded by this artificial limitation in the excessive mobility of the mandible is usually immediate but not always maintained; in a few severe and intractable cases the aid of an oral surgeon must be invoked and an operation carried out on the joint, usually with an excellent result.

Structural Causes of Facial Pain

In facial pain due to a structural lesion involving the trigeminal nerve there is usually a short history and some sensory impairment on examination. In the earliest stage this may amount to no more than some blunting of the corneal reflex on the affected side, but if it is genuine this is highly significant.

Such pain is rarely paroxysmal, though in very occasional cases of acoustic tumour intermittent pressure on the nerve may simulate trigeminal neuralgia, especially if involvement of the seventh nerve also causes hemifacial spasm. This suspicion always arises where either fifth or seventh nerve involvement is associated with ipsilateral deafness; corneal insensitivity is a crucial finding, and where any suspicion arises expert otological assessment should be sought, in addition at least to radiological examination of the internal auditory meatus and measurement of the protein content of the spinal fluid.

Nasopharyngeal carcinoma is an unpleasant disease with which the clinician often becomes familiar only through a series of mistakes. Long before pain or a tell-tale blood-stained nasal discharge appears, and sometimes even before the growth can be seen by the surgeon under general anaesthesia, the lesion may be announced by a painless ophthalmoplegia. However, when pain appears it is often severe and intractable. Again the timidity of doctors in relation to analgesics is baffling. Patients whose outlook is hopeless and whose pain is appalling are referred to neurosurgeons with a view to operative division of sensory pathways when inquiry shows they are having nothing more than a few compound codeine tablets daily, and that—save the mark—the surgeon "has successfully weaned" the sufferer from stronger drugs. Such statements bespeak an attitude that regards pain as ennobling rather than demoralizing, and that endows a mutilating operation with some strange moral superiority over the use of such splendid euphoriant analgesics as heroin and cocaine. In the case of malignant disease certain to kill within months, surgery should be invoked only when pharmacology has failed—and in these circumstances the result will often be euthanasia, even when such an intention was far from the surgeon's mind.

Intracranial aneurysm arising from the posterior communicating artery is a classical cause of first-division trigeminal pain, usually with oculomotor palsy. The lesion can be visualized

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by angiography and the pain relieved by common carotid ligation.

Dental Pain.—The days when every outpatient with trigeminal neuralgia had been rendered edentulous in the pursuit of relief are past, but it is important to recall that dental disease is still the commonest cause of facial pain—and that its provocation by hot, cold, or sweet food is virtually pathognomonic.

Psychosis

In an occasional case facial pain is symptomatic of psychotic illness, usually agitated depression. The painful syndrome lacks the definition of those previously described, and diagnosis depends on recognition of the other features of endogenous depressive illness. The patient is usually middle-aged, agitation is prominent, and sleep is characteristically disturbed, with early waking. Depression itself is less often a presenting feature than admitted in response to direct questioning. As with another even more typically depressive symptom—the complaint of severe and persistent pain in a normal tongue—hypochondriacal fears of cancer are often unspoken.

In some of these patients improvement of the depressive condition with appropriate drug treatment is accompanied by fading of the facial pain. However, in more than half the patients depression improves without amelioration of the painful syndrome. Improvement in mood and general condition makes the pain less distressing, but the grateful patient insists it is still present, unaltered but bearable. Such patients are among those comprised in a larger group of cases of "atypical facial pain."

Atypical Facial Pain

Cases of facial neuralgia that defy inclusion under the rubric of any one of the syndromes already described are unfortunately almost as common as those that lend themselves to classification. They also present a most discouraging prospect for treatment. The cause of the syndrome or syndromes is unknown, and if the patient shows no response to antidepressive drugs the chances are that she will show precious little response to anything else the doctor prescribes.

The typical patient is female, middle-aged, edentulous, haggard, and importunate. The pain is usually maximal in the distribution of the second division of the trigeminal nerve, of long duration, constantly present, sometimes unilateral but more often spreading across the midline or alternating from side to side. It has resisted the attention of a procession of dental surgeons and physicians, and often various forms of nerve block or surgical procedures on the jaws. Holes have been bored in the patient's sinuses on the doubtful premise that chronic pain analogous with the severe and localized variety typical of acute sinusitis is commonly caused by chronic sinus infection. Analgesic drugs yield only brief respite. All other measures have been of no avail, and the patient presents herself again without much hope of relief. All too often she is right, and after a few unconfident and unsuccessful trials of various forms of empirical drug treatment she drifts off to some other doctor or clinic.

All doctors tend to attribute lack of therapeutic success to perversity, neurosis, or exaggeration—thereby excusing our failure, comfortably absolving ourselves from further effort, and placing the blame fairly and squarely on the patient's shoulders. Sometimes it is impossible to resist this temptation or to feel that it is unsupported by suggestive evidence: a good example is the occasional bright and cheerful victim of severe daily migraine who triumphantly announces the complete and utter ineffectiveness of yet another therapeutic routine with the arch smile of a contestant in a parlour game. Such considerations do not in my view apply in most instances of atypical facial pain. The patient looks ill and is often emaciated. She is clearly suffering, whatever the nature of her pain. The persistence of the pain after depression has been relieved is also suggestive, and despite its lack of clear definition I believe it probably arises on the basis of some form of pathophysiological disturbance, and is more than a purely mental entity or idea of pain. Again one is tempted to clutch at the straw of autonomic disturbance, but the emotional state of these patients usually renders them unsatisfactory subjects for surgical experiment, and I have not so far advised ganglionectomy or ganglion blockage even in the occasional wholly unilateral case. To the best of my knowledge I have never cured a case of atypical facial neuralgia, but I have known several who recovered during or after and almost certainly independently of my therapeutic efforts.

Summary

Facial pain is a common symptom in the neurological clinic and presents interesting and difficult diagnostic problems. The remittent paroxysmal pain of trigeminal neuralgia is the best-defined of all the syndromes encountered in this connexion. It is a disease of the elderly, shows considerable remissions, is unassociated with any abnormal physical signs, and always recurs. About three-quarters of all cases can be effectively relieved by Tegretol (carbamazepine), and in the remainder surgical section of the sensory root of the nerve remains the treatment of choice.

Post-herpetic neuralgia on the other hand shows little response to surgery but a marked tendency to spontaneous recovery, and is best treated by powerful analgesics, including heroin in severe cases. Migrainous neuralgia or "cluster headache" is a well-defined syndrome of unilateral first-division pain, remarkably responsive to ergotamine, but often unrecognized. Among structural causes of facial pain temporomandibular arthritis, nasopharyngeal cancer, and intracranial aneurysm should be remembered. It may also be a symptom of agitated depression and may respond to antidepressive drugs. There remains a considerable group of cases of distressing facial pain which lack the clear-cut characteristics of any of the forms described above, occur almost exclusively in middle-aged women, and resist every form of treatment but sometimes clear spontaneously.

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