Chapter 20: Headache and facial pain

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Headache

It is probable that nearly everyone has had a headache at some time. It is only when the single attack is exceptional in some way, or if there are repeated attacks, that the patient has to seek the advice of a doctor. There are features in the timing, quality and the situation of the headache which may help in its explanation. The severity of the headache by itself is not a good indicator of its cause.

Pain is such a subjective symptom that, at best, the degree of discomfort which is associated with it is only one feature. If it is of sudden onset, and particularly if associated with neck stiffness, one is justified in considering subarachnoid haemorrhage as a likely cause. A ruptured intracranial aneurysm, or less commonly an arteriovenous malformation, may be the underlying pathology. The circumstances of the pain at the onset can be of material significance. A history of trauma, or unusual effort, or infection may be relevant. Many patients recall an injury, which later proves to be irrelevant. Conversely, many patients with a subdural haematoma and headache do not remember the knock on the head. The drinker who expects a hangover headache, but does not remember the head injury, is a good example of this.

Early morning headache, particularly headache which interrupts sleep, is usually more serious, and typically is a feature of raised intracranial pressure. Aggravation by change in posture, such as bending down, or by coughing and sneezing can be expected. It is, however, when it becomes a particular feature of the headache that it may underline its seriousness. It is worth enquiring as to whether the pain is relieved by lying down, as nearly all patients with cluster headache and many with migraine, find that this aggravates the situation.

A throbbing quality is usually associated with increased vascularity, as in a feverish headache or that associated with infection. A sharp pain may indicate that the cause is local, and affects trigeminal, occipital, or upper cervical roots.

The site of the pain can be of diagnostic help; it may be related to ear infection, dental abscess or sinusitis. The finding of localized tenderness is another pointer to the diagnosis.

There may be unusual circumstances, such as the headache which may commence during sexual intercourse. It is very uncommon but so severe and frightening that a vascular event is suspected. It can be reproduced on subsequent similar occasions - and naturally can cause anxiety. Usually there is no evidence of a subarachnoid haemorrhage, and the condition is self-limiting. The male is the usual sufferer, but on two occasions in the author's experience, the female partner experienced the headache. The explanation seems to be the position of the head and neck during lovemaking.

Single attacks of headache demand an intensive search for the cause. Repeated attacks have the advantage of establishing a pattern, with more clues and information about the incidence.
Examination may reveal evidence of raised intracranial pressure with optic disc swelling, meningeal irritation with neck stiffness, other focal neurological signs, raised blood pressure or tenderness over the superficial temporal artery. One has to be aware that optic neuritis may also show disc swelling and be associated with headache or orbital pain. Usually it is possible to discover the accompanying central scotoma, and of course there is marked impairment of visual acuity. Neck stiffness may accompany acute febrile illness, or any severe occipital and neck pain, and may be unrelated to the presence of blood or inflammatory change in the meninges. It is generally recognized that hypotension is not usually accompanied by headache; it may, however, be the presenting symptom of rapidly rising blood pressure as in toxaemia of pregnancy, renal failure or phaeochromocytoma. Examination of the fundus may show the expected arterial changes with haemorrhages and exudates. Over the age of 60, one always suspects temporal arteritis. There may not be the expected tenderness over the superficial temporal artery and the sedimentation rate may not show the big rise which is customary. Temporal artery biopsy and a trial of steroids are desirable to avoid the very real risk of sudden onset of blindness.

Glaucoma may be responsible for an acute onset of headache and measurement of the intraocular pressure is essential to exclude it.

**Migraine**

It is not unusual for the first attack of migraine to raise alarm and anxiety. The pain may be accompanied by visual disturbance, motor and sensory symptoms, confusion and vomiting. The circumstances and subsequent total recovery may give a clue to the diagnosis. Subsequent attacks will be less alarming, though still requiring management and treatment. The expectation of unilateral pain with visual disturbance and nausea and vomiting is only true for classical migraine. Many more suffer from common migraine, with repeated attacks of headache which need not be unilateral and in some instances may be bilateral with every episode, and not necessarily accompanied by other symptoms (Friedman, 1962; Lance, 1982). Sufferers from classical migraine may also complain at other times of simple headaches with or without nausea. Hemiplegic migraine, associated with unilateral numbness, tingling and perhaps weakness, is usually more incapacitating. Vertebrobasilar migraine is also accompanied by vertigo, ataxia and often slurring of speech. It is rewarding to study the role of trigger factors and the relationship of the headaches to the menstrual cycle, certain foods, stressful situations, and conditions at work. The differential diagnosis from muscle contraction headache can be particularly difficult.

**Ergotamine-dependent headache**

Ergotamine has long had a reputation of being an effective drug for the treatment of acute migraine, preferably taken very early in the attack. It is very tempting for sufferers to take ergotamine as soon as they think an attack is coming on. This may lead to regular medication with ergotamine in tablet or suppository form, amounting to 3-5 mg weekly. What usually follows is that the headaches become more frequent, occurring many times a week. They no longer resemble the original migraine attacks, although an actual migraine may supervene. In the majority, they are simple, severe headaches which apparently respond to ergotamine. The treatment is to stop the ergotamine. Invariably, there is a severe attack of headache within a week of stopping, but provided no recourse to ergotamine is made, the
vicious cycle of headache and ergotamine therapy is finally broken (Rowsell, Neytan and Wilkinson, 1973; Legg, 1976).

**Chronic paroxysmal hemicrania**

This is a relatively rare form of headache with recurrent unilateral attacks only responding to indomethacin. It is much more common in men, and there are no other distinguishing features. After a successful trial of indomethacin, the condition usually improves and it is then possible to withdraw the medication gradually over a period of a few months.

**Cerebral tumour**

It is no exaggeration to say that many sufferers from headaches are seeking reassurance that it is not caused by a cerebral tumour. That a primary or secondary tumour is a rare cause of headache is, in itself, not reassuring to the individual. It is very unusual for a tumour to give rise to headache early in the history, though raised intracranial pressure may cause early morning headache with nausea, vomiting and papilloedema.

Localization of the headache is usually a poor indication of the site of the tumour, with the exception of a rapidly expanding growth. It is generally accepted that the pain is due to stretching and distortion of pain fibres associated with the meninges and larger vessels. The presence of other symptoms such as somnolence, intellectual impairment, disturbances of the senses of smell or vision adds to the significance of the headache. Epilepsy of late onset is another important clue. These symptoms also help with the localization of the tumour. Exacerbation of the headache by change in posture, such as bending, coughing or sneezing is not specific to cerebral tumour or raised intracranial pressure. Localized tenderness is worth looking for, and palpation of the head may be useful aid to the diagnosis. An intensification of the headache may indicate haemorrhage or cystic degeneration. CT scanning has revolutionized the investigation of brain tumours. One word of warning: when a tumour is strongly suspected, a lumbar puncture is inadvisable.

**Benign intracranial hypertension**

This is an uncommon condition frequently associated with headache and usually affecting overweight women. There is a marked rise in the intracranial pressure with papilloedema and visual symptoms and signs. CT scanning of the brain reveals normal or small ventricles, suggesting the diagnosis. Lumbar puncture is safe and gives the only accurate indication of the degree of rise in intracranial pressure, and the subsequent course of the condition (Boddie, Banna and Bradley, 1974).

**Cerebral aneurysm**

A sudden severe pain may be the dramatic presentation of a ruptured cerebral aneurysm with a subarachnoid haemorrhage. It is quite common for a second episode to occur before the significance of the first is appreciated. It is not possible to localize the site of the aneurysm along the circle of Willis or the adjoining vessels, by the situation of the headache. It is often frontal and later may be accompanied by pain in the neck, exacerbated by head and
neck movement. Associated symptoms, or signs, such as a complete third nerve palsy admirably localize the aneurysm, in this instance along the posterior communicating or basilar arteries. The aneurysm may be silent until the moment of rupture though there are many instances when the unilateral pain has been present for weeks or months. It is usually persistent, and localized to one or other orbit and is presumably associated with expansion of the aneurysm. Sadly, CT scanning may fail to reveal the aneurysm and it is the author's usual practice to recommend digital subtraction angiography to try and establish whether an aneurysm is present. It is safer than carotid and vertebral angiography.

**Temporal arteritis**

Although the clinical picture is well recognized, the headache may not be at all severe, and may indeed be absent. It is not uncommon for the patient to appear depressed, or indeed to be so as a consequence of the headache. If the condition is suspected, it is really not safe to await the result of the artery biopsy. Immediate treatment with steroids is recommended. The diagnostic histological criteria persist for a few days after starting steroid therapy.

**Meningitis**

Acute bacterial meningitis is dramatic in onset with high fever, severe headache and generalized symptoms of the infection. Drowsiness and neck stiffness suggest the diagnosis. Subarachnoid haemorrhage may also be associated with fever, but generally not above 38°C. Symptoms and neurological signs, of cerebral involvement usually indicate a meningoencephalitis or cerebral abscess. Viral meningitis is now probably more common than bacterial meningitis - but it is imperative to perform a lumbar puncture to aid the diagnosis. Tuberculosis with or without the presence of cerebral tuberculomata, remains an important cause of meningitis. The onset is often insidious over several weeks, accompanied only by a low grade fever, headache and malaise. CT scanning is not generally available and often it is this examination which demonstrates the presence of a single tuberculoma, frequently in the posterior fossa or there may be multiple lesions. Neck stiffness is still a cardinal sign of any meningeal infection, together with a positive Kernig's sign. Examination of the cerebrospinal fluid should distinguish the various causes of meningitis. Carcinomatous and other malignant invasion of the meninges can also present dramatically with headache and perhaps vomiting and neck stiffness. A low sugar content in the cerebrospinal fluid is still in keeping with the presence of mononuclear cells and a raised protein level. It is the demonstration of malignant cells in the centrifuged deposit which makes the diagnosis.

**Cough headache**

Any headache, from whatever cause, can be expected to be aggravated by coughing. Typically, headache associated with raised intracranial pressure is exacerbated by coughing, sneezing, straining at stool, as well as bending. There is a form of headache which is precipitated by coughing, and with no other accompanying symptoms or signs, which is benign and with a limited natural history of weeks or months. The pain is very severe, if brief, and only comes on with coughing. It improves slowly over the course of many months and, perhaps surprisingly, a more rapid recovery can follow lumbar air encephalography (Symonds, 1956). CT scanning shows a normal ventricular system.
Cluster headache

Described by Harris (1926) as periodic migrainous neuralgia, the clinical syndrome of unilateral severe pain with a blocked nostril and redness of the eye, occurring daily in bouts, has now come to be generally recognized as cluster headache, after the description by Kunkle et al (1952). There is usually a remarkable regularity in the timing of the attacks, by day or night. The pain is intense, and may last an hour or more. Many sufferers admit to knocking their head against the wall, and to feeling suicidal. Attacks occur every six months or a year, and usually last a few weeks. Missing out one year naturally gives hope of a more lasting remission, only to be disappointed. It is very rare in women, and the author’s youngest sufferer was 21 years of age. Rarely, both sides of the head can be affected at different times. Symonds (1952) advocated the use of ergotamine tartrate by injection for the prevention of attacks. Other forms of ergotamine tartrate preparations have also been used including an effervescent tablet and by inhalation. Lithium (Ekbom, 1981) has also been used with success. Inhalation of oxygen may also cut short an attack (Kudrow, 1981).

A comparison between migraine and cluster headache is shown in Table 20.1.

Table 20.1 Comparison of migraine and cluster headache

<table>
<thead>
<tr>
<th></th>
<th>Migraine</th>
<th>Cluster headache</th>
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</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Adolescence</td>
<td>Adult life</td>
</tr>
<tr>
<td>Sex incidence</td>
<td>Female:male, 2:1</td>
<td>Male:female, 10:1</td>
</tr>
<tr>
<td>Family history</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Eyes</td>
<td>Visual aura common</td>
<td>One eye waters</td>
</tr>
<tr>
<td>Nausea and vomiting</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Nature of pain</td>
<td>Pulsating, hemicranial</td>
<td>Constant pain in region of one eye</td>
</tr>
<tr>
<td>Patient's reaction</td>
<td>Lies down</td>
<td>Walks about</td>
</tr>
<tr>
<td>Duration</td>
<td>3-12 hours</td>
<td>Less than 3 hours</td>
</tr>
<tr>
<td>Frequency</td>
<td>1-8 per month</td>
<td>Daily, often at night</td>
</tr>
<tr>
<td>Menstruation</td>
<td>Related</td>
<td>Not related</td>
</tr>
<tr>
<td>Prolonged remissions</td>
<td>Rare</td>
<td>Common</td>
</tr>
</tbody>
</table>

Muscle contraction (tension) headache

The characteristics of headache are never less defined than when associated with muscle tension. Described as 'band-like', 'a tightness', 'a heavy weight on top', it is usually bilateral, insidious in onset, frequent and often associated with other symptoms of tension. Although vomiting is rare, it may be accompanied by nausea, particularly following heavy doses of medication. Although when typical it can easily be distinguished from attacks of classical migraine, it is not unusual to encounter difficulty in differentiating it from common migraine. Both causes can be present, and many sufferers learn to differentiate between the two.
**Temporomandibular joint dysfunction**

Dental malocclusion and arthritic changes in the temporomandibular joints are relatively common. It may present with otalgia and pain radiating up towards the temples, along the jaw and upper neck or localized to the joint. It is not always related to mastication, and can present without localized tenderness and with no indication of a dental problem. It would be relatively easy to diagnose if the pain followed the use of new and ill-fitting dentures, with trismus and pain on chewing. Dental examination and study of jaw movement will help, but more detailed study of the bite and X-rays of the joints may be necessary.

**Cervical spondylosis**

It has been estimated that 60% of people over the age of 40 have some degree of spondylosis. The usual presentation with neck ache, and pain radiating down one or other arm is frequently inconsistent with the severity of the radiological changes. Whilst even relatively moderate changes can be associated with pain, more severe arthritic changes can be completely pain free. Headache is an uncommon presentation and is more likely to be associated with muscle contraction, itself related to the neck symptoms. Occasionally a high cervical lesion at C2 is associated with pain over the occipital region.

**Depression**

Any severe pain, persisting relentlessly, will be associated with depression. Both reactive and endogenous depression can in turn be associated with headache. A detailed history of the onset and which symptom came first, the headache or the depression, is usually inadequate. Personal problems at home or at work are all too common. The headache can be quite persistent and poor sleep, loss of appetite and loss of affect confirm the possible diagnosis. It is essential to carry out a full neurological examination, and to be aware of any change in the characteristics of the pain. Depression can coexist with other causes and antidepressant therapy can be expected to help with the depression and the headache. Certain occupations demand attention to detail and the tasks are necessarily repetitive. Sewing machinists are one such group. They work long hours at home, are paid per item, and develop quite severe headache. They invariably deny any depressive symptoms, but respond quickly to antidepressant treatment. Patients prescribed monoamine oxidase inhibitors should always be warned not to eat foods rich in amines. Blackwell and Mabbitt (1965) identified tyramine in cheese as the cause of the headache and hypertensive reaction which may follow.

**Facial pain**

As with headaches, facial pain may be a manifestation of many and various conditions. The characteristics of the pain, its severity, situation, radiation, precipitation and associated symptoms may give a clue to the cause. Detailed clinical examination may reveal dental tenderness, or impaired facial sensation.
**Infection**

**Sinusitis**

This is considered more fully in Chapter 6. It is an obvious cause if it is associated with localized tenderness, mucopurulent rhinorrhoea and preceded by a history of coryza. It may be associated with a generalized headache.

**Dental infections**

Pain on chewing, with associated localized tenderness, may be the only abnormal finding. Radiological evidence is helpful. A persistent root infection may escape detection for some time. The pain may radiate along the jaw or be referred to the ear.

**Ear and parotid infection**

Otalgia or pain over the mastoid area, associated with otorrhoea, may suggest the diagnosis. Parotitis may present with headache and tenderness in the preauricular region and it may be several days before the swelling of the gland makes the diagnosis more obvious.

**Herpes zoster**

The pain may precede the appearance of the vesicles, usually in the distribution of the ophthalmic or maxillary branches. Ocular complications are common with persistent sensory impairment. Postherpetic neuralgia is much more common in those over the age of 60. Analgesics, antidepressant therapy and transcutaneous nerve stimulation may all be needed in what can be a severe, protracted and painful condition.

**Ocular causes**

Refractive errors and ocular palsies may be associated with pain in the eye as well as headache. Inflammatory lesions such as iritis or uveitis are also painful. The prompt diagnosis and treatment of glaucoma is rewarding in the early relief of pain and prevention of visual impairment.

**Dental occlusion and temporomandibular joint dysfunction**

Despite early orthodontic treatment many still have problems with malocclusion in adult life. Pain and tenderness along the jaw, and the examination of the bite, may suggest the diagnosis. A prosthesis may both improve alignment and relieve pain. Early arthritic changes in the temporomandibular joints may be responsible for unilateral or bilateral pain. It is usually aggravated by tension and anxiety and is often relieved by efforts at relaxation and the use of mild tranquilizers.

**Salivary calculi**

These usually present with pain during and after meals, and there may be a tender swelling of the gland. Commonly involving the submandibular gland and its duct, the pain
can be reproduced by a suitable stimulus such as sucking a lemon. Radiological examination is necessary to demonstrate the offending calculi.

**Trigeminal nerve lesions**

Pain in the face may be accompanied by sensory impairment in one or more of the three branches, ophthalmic, maxillary and mandibular, and motor weakness of the jaw, involving the temporalis, masseter and pterygoid muscles. Involvement of the ophthalmic division is usually accompanied by an impaired or absent corneal reflex. It is usually for the motor defect to accompany sensory changes in the mandibular distribution. Although tumours in the cerebellopontine angle commonly involve the trigeminal nerve, or its central pathway in the brainstem, it is rare for them to present with pain. It is more likely for the patient to complain of unpleasant numbness or altered feeling. The commonest such tumour, an acoustic neuromyosarcoma, usually presents with deafness. As well as sensorineural hearing loss, and a possible complaint of giddiness and tinnitus, there is usually trigeminal involvement, and there may be an early facial weakness and cerebellar symptoms and signs. Other tumours in the region, a fifth nerve neuroma, and a meningioma in the region of the petrous apex, are more likely to present with isolated trigeminal nerve involvement. A nasopharyngeal carcinoma may present with facial pain and sensory impairment. In this instance, there is conductive deafness. Other tumours, in the pituitary region, sphenoid sinus or at the base of the skull, usually present with other cranial nerve lesions. Glioma of the brainstem may involve lower cranial nerves and is accompanied by long tract signs. Paget’s disease of the skull may present with facial pain, as well as headache. Unlike the pain in trigeminal neuralgia, it is usually persistent and without exacerbation by trigger factors. X-rays of the skull, tomography, CT scanning and magnetic resonance imaging may all be necessary to establish the cause.

**Trigeminal neuralgia (tic douloureux)**

The pain is usually described as sharp, needle-like or piercing. It comes on in paroxysms which may be very brief or last many minutes. Any movement of the face, including talking, eating and laughing, or touch as in shaving or applying makeup, may trigger off a paroxysm. The pain is very severe with accompanying tears in the eyes. Once the paroxysm subsides, there is a dull ache, which lingers on and serves as a reminder of the previous pain and a warning of the next one. It may affect one or more of the divisions of the trigeminal nerve, commonly the second and third. A sufferer can always localize the trigger area although reluctant to demonstrate it. The condition is much more frequent over the age of 60. Neurological examination, including motor and sensory function of the trigeminal nerve, is normal. Presentation at an earlier age, and with neurological signs, may be due to multiple sclerosis, acoustic neuroma or meningioma near the petrous bone. Radiological, neurophysiological investigation and examination of the cerebrospinal fluid may be necessary to establish the diagnosis. It is not unusual for sufferers to present to a dentist. Dental extractions are common, and some patients are rendered edentulous. Occasionally dental treatment precedes the onset of pain. Although the cause of trigeminal neuralgia is not established, aberrant vessels in the proximity of the trigeminal nerve have been noted during the course of posterior fossa exploration and root section for the relief of pain. Attacks may last weeks or months at a time, with long periods of freedom. There is usually a progressive increase in the severity and duration of the pain, over the next few years. Carbamazepine
relieves the pain in two-thirds of cases, in doses of 100-200 mg three or four times daily (Campbell, Graham and Zilkha, 1966). Side-effects, particularly drowsiness or giddiness, may restrict the dosage. Poor response or no response, as in a third of cases, justifies a trial of phenytoin, in doses of 100-300 mg daily. It is, however, less effective than carbamazepine. failure of adequate relief of the pain by medication leads to consideration of other measures. The choice lies between phenol injection of the trigeminal nerve, and stereotactic thermocoagulation.

**Trigeminal neuropathy**

The term was first used by Spillane and Wells (1959) to describe persistent sensory disturbance in the trigeminal distribution, with no obvious pathology. Pain may be present. In the author's experience there is usually previous dental treatment and the condition remains localized and non-progressive.

**Glossopharyngeal neuralgia**

The pain is very similar to trigeminal neuralgia, but localized to the lower jaw, in front of the ear, in the ear, or in the throat. It may radiate down the neck. It is much less common than trigeminal neuralgia and typically the pain is triggered by swallowing rather than chewing. Carbamazepine is usually effective, but if side-effects prevent its long-term use, then avulsion of the glossopharyngeal nerve may be necessary.

**Migraine variants**

Usually considered as a unilateral headache, the pain may be periorbital or in the face. The aura may be absent and there may be little or no nausea. Often it is the presence of precipitating factors such as menstruation or dietary triggers, which may suggest the diagnosis.

**Atypical facial pain**

This is not an uncommon presentation of depression. The pain is usually severe and persistent, with only a few of the features of the underlying depression, including interrupted sleep and loss of affect. It responds to antidepressant treatment quite quickly.

**Causalgia**

Typically this is described as a burning or searing pain, which may be accompanied by trophic skin changes. With the increase in head and facial injuries, this condition is becoming more frequent. It may be directly related to the site of the injury or be associated with partial nerve damage. The diagnosis is made on the history. Sympathetic block, or sympathectomy, may be necessary for the relief of pain (Rasmussen and Freedman, 1946).