

Headache and facial pain

Richard Peatfield

Abstract

Headache is one of the most common symptoms seen in both General Practice and Neurology clinics. The Physician's first duty is to exclude and treat any specific structural cause – especially subarachnoid haemorrhage and meningitis if the history is short, and temporal arteritis and cervical spondylosis if it is longer. It is unusual for a cerebral tumour to present with a headache for more than three months without physical signs being present. This review discusses all the common causes of acute, chronic, recurrent and progressive headache as seen in out-patient practice. The majority have migraine, a few have cluster headache and rarer trigemino-autonomic cephalalgias. Headaches are a major cause of distress and impaired productivity in the general population, and much can be reversed with appropriate medication given in adequate doses.

Keywords analgesic; cephalalgia; cluster; headache; hemicrania; migraine; triptan

Headache is among the most common symptoms seen in neurology clinics. It is often incapacitating, but is seldom caused by potentially serious illness. A recent epidemiological study showed that 86% of women and 63% of men suffered at least one tension-type headache in the previous year.¹ The annual prevalence of migraine is 15% in women and 6% in men. Patients are often reassured if told that only a minority of the population do not have at least an occasional headache.

Diagnosis

Few patients have physical signs, and clinical assessment depends almost entirely on the history. The most important questions are listed in Table 1. It is usually possible to make a confident clinical diagnosis without many investigations. Extensive series, many undertaken in the USA, have shown that routine brain imaging of patients with non-progressive recurrent headache and no physical signs is unrewarding.²

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What's new?

- Increased recognition of the morbidity of recurrent headache
- Changes in MRI brain scans of migraine sufferers
- Genetic analysis of the different types of familial migraine
- Very large-scale trials of topiramate in migraine prophylaxis
- Availability of triptans over the counter, and as generic drugs

Types of headache

A classification of the likely causes of headache, based on the history at presentation, is shown in Table 2.

Migraine^{3,4}

Migraine is the most common cause of recurrent disabling headache in the general population, and particularly in patients seeking medical care. It is about twice as common in women as in men. It often starts at puberty, and sometimes earlier in boys. The pain is usually throbbing and is usually felt predominantly on one side of the head, though it can occur on different sides in different attacks. The pain usually lasts for 4–72 hours, but attacks can be shorter in children. The frequency varies widely. Some patients suffer three or four attacks each week, and others only one or two in a lifetime; the former tend to predominate in neurological clinics. Nausea and vomiting are common during the headache phase, and many patients are confined to bed for 1–3 days.

Less than a fifth of the patients have attacks preceded by an aura, and many of these auras are ill-defined episodes of blurring of vision and/or photophobia. A few patients suffer more characteristic disturbances, which may include a bright zigzag migrating across the visual field from the centre to the periphery over about 30 minutes often with transient loss of vision, paraesthesiae in the fingertips or the side of the face, and disturbances of speech. Management of migraine is discussed below.

Tension-type headache

Most individuals suffer attacks of bilateral, non-throbbing headache. This is seldom as incapacitating as migraine, though about

Questions for patients presenting with headache

- Age, occupation, general history
- How old was the patient when the headaches began?
- Do the headaches occur in attacks?
- How frequent are the headaches?
- How long do they last if untreated?
- Where in the head is the pain?
- Is it throbbing? Is it worse on exercise?
- Is there nausea or vomiting?
- Are any focal symptoms (e.g. visual, sensory or speech disturbances) related to the attacks?
- What acute treatments have been tried?
- What long-term treatments have been tried?
- Is the patient taking the oral contraceptive pill or other hormones?

Table 1

Patterns of headache and their causes

Acute single headache

- Febrile illness, sinusitis
- First attack of migraine
- Following a head injury
- Subarachnoid haemorrhage, meningitis

Recurrent headaches

- Migraine
- Cluster headache
- Episodic tension headache
- Trigeminal or post-herpetic neuralgia

Triggered headache

- Coughing, straining, exertion
- Coitus
- Food or drink

Dull headache, increasing in severity

- Usually benign
- Overuse of medication (e.g. codeine)
- Contraceptive pill, hormone replacement therapy
- Neck disease
- Temporal arteritis
- Benign intracranial hypertension
- Cerebral tumour

Dull headache, unchanged over months

- Chronic tension headache
- Depressive, atypical facial pain

Table 2

5% of those affected have continuous, chronic headache. Most never seek medical advice and depend on over-the-counter medication. In those who seek medical care, few investigations can be justified if the history is non-progressive and there are no physical signs on examination.

Many of these patients (perhaps more than 50% of those attending hospital clinics) are taking analgesics (e.g. ergotamine, codeine, caffeine) on a daily basis, and it is now well established that their headaches often settle when the analgesics are withdrawn, suggesting that the analgesics may perpetuate the pain. In some patients, headache can be attributed to the contraceptive pill or hormone replacement therapy (HRT). Others (particularly those with chronic headache) are overtly depressed, though the success of tricyclic antidepressants does not seem to depend on the presence of clinical depression. At present, there is no trial evidence to support the use of triptan medication in patients with a long history of pure tension headache.

Management should focus on reassurance of the patient that no life-threatening disease is present. Once potential precipitating factors have been addressed, many patients respond to tricyclic antidepressants or non-steroidal anti-inflammatory drugs.^{5,6}

Cluster headache^{7,8}

Cluster headache syndrome (previously termed 'migrainous neuralgia') is less common than migraine. Most patients are men. Sufferers describe episodes of exceptionally severe, steady

unilateral pain, usually in the eye, cheek or temple, in episodes lasting between 30 minutes and 3 hours, often several times daily. Characteristically, patients are woken from sleep by the pain. The episodes are often accompanied by autonomic disturbances, including ptosis, a bloodshot eye, watering of the eye, and watering or blockage of the nostril on that side. In about 90% of patients, the pains occur daily for 6–12 weeks, after which there is a remission that may last from 3 months to 2 years.

Few investigations are justified in typical patients. However, the syndrome is occasionally mimicked by structural disturbances in the pituitary gland or cavernous sinus, and atypical patients should undergo CT or MRI of the brain.

Analgesia is difficult, though trials support inhalation of 100% oxygen, subcutaneous sumatriptan, or sumatriptan or zolmitriptan administered by nasal spray. The mainstay of management is regular medication to prevent attacks; verapamil (120 mg t.d.s. or more) is the treatment of choice, though patients also respond to lithium carbonate, methysergide and gabapentin. Corticosteroids (e.g. prednisolone 40 mg daily) are effective, but perhaps to be recommended only in patients in whom previous experience suggests that the cluster is likely to end within 2 or 3 weeks. A few patients with intractable chronic cluster headache may require surgical procedures.

Cervical spondylosis

Cervical spondylosis is the most common cause of new headache in older patients. The pain is usually in the neck, but can be felt on the forehead, perhaps because of the overlap between the descending trigeminal pain fibres and the ascending cervical fibres in the upper cervical cord. In most patients, the pain is bilateral and is worsened on neck movement; it is seldom accompanied by significant vomiting or visual disturbances. Measurement of the erythrocyte sedimentation rate (ESR) is prudent to exclude temporal arteritis. Most patients respond to an anti-inflammatory drug such as ibuprofen or diclofenac.

Temporal arteritis (giant cell arteritis)

Temporal arteritis is less common than cervical spondylosis, but is the most important cause of headache in older patients because of the risk of blindness. The pain may be unilateral or bilateral and is often accompanied by systemic myalgia (polymyalgia rheumatica). ESR is elevated in most patients; a few patients with a normal ESR require temporal artery biopsy if the history seems typical. Characteristically, patients respond, usually overnight, to very high-dose prednisolone. Some authorities recommend as much as 80 mg for a few days; this is reduced once the ESR is within normal limits.⁹ A lower dose may have to be continued for 2–4 years before the condition subsides spontaneously. Temporal arteritis is discussed further in MEDICINE 34:11.

Cerebrovascular disease

Headache is common in cerebrovascular disturbances; 20–40% of patients with transient ischaemic attacks or ischaemic stroke have a mild or moderate headache contralateral to the neurological symptoms. Ipsilateral headache is common for a few days after carotid endarterectomy, and headache is often a symptom of venous sinus thrombosis. Dissection of the carotid artery may be extremely painful, leading to discomfort in the neck or the head, and any patient in whom this is suspected should undergo urgent vascular investigations with a view to anticoagulation if the diagnosis is confirmed.

Sinusitis

Headache and facial pain are common features of febrile illnesses such as influenza. A single, severe headache following a common cold, however, may be caused by secondary bacterial infection in the frontal or maxillary sinuses. This can be confirmed by CT scanning, and usually resolves with conventional antibiotics.

Trigeminal neuralgia

Most patients with trigeminal neuralgia are elderly. The pain is thought to be caused by compression of the trigeminal nerve by an aberrant artery as it leaves the brain stem. In a few patients with multiple sclerosis, a similar pain seems to be generated by a plaque involving the trigeminal nerve within the brain stem. The pain is knife-like, often of great intensity, lasts only 1 or 2 seconds, and is confined to the distribution of the trigeminal nerve on one side, most commonly the maxillary or mandibular divisions. Attacks are often triggered by chewing, eating, speaking or touching the face, or even by the wind. Most patients respond to anticonvulsants such as carbamazepine or gabapentin¹⁰, but destructive procedures on the trigeminal nerve, or even corrective operations on the aberrant artery in the posterior fossa, should be considered when the pain cannot be controlled without unacceptable side effects.

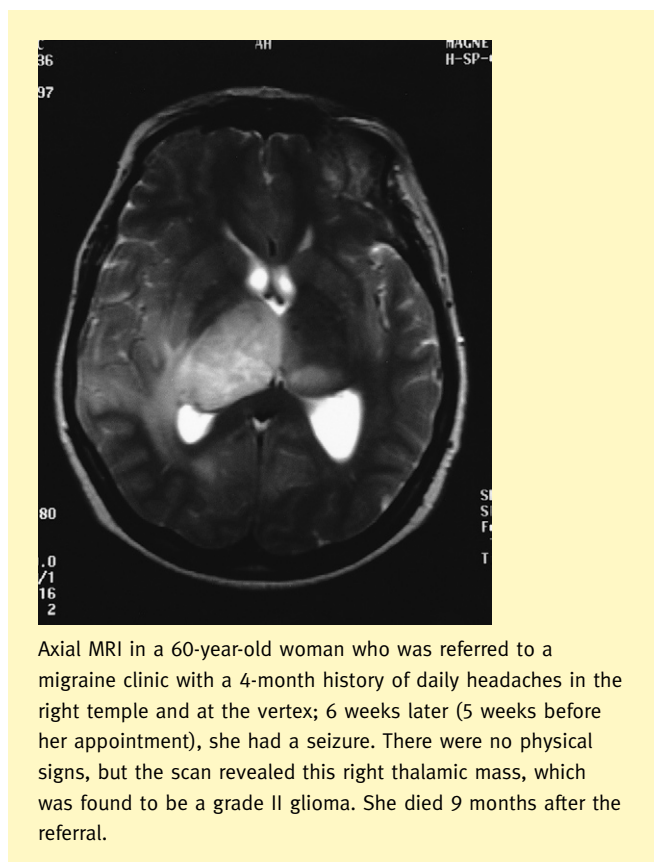
Atypical facial pain

In contrast to trigeminal neuralgia, atypical facial pain is less well localized in the face and the pain may be continuous. It is essential to ensure that all potential structural causes of pain have been excluded; scans may be appropriate, and the opinion of a dentist, ENT surgeon or ophthalmologist should be sought when necessary. Many patients are depressed, and most respond to courses of tricyclic antidepressants.

Raised intracranial pressure

Raised intracranial pressure is an uncommon cause of headache in the absence of physical abnormalities on careful examination; most patients with cerebral tumour present with epilepsy or with focal neurological disturbances relating to the site of the tumour. Tumours usually must be very large to distort cerebrospinal fluid pathways or pain-sensitive vascular structures in the base of the brain, and patients seldom have headache without other features (Figure 1). Possible causes include primary intracerebral glioma and secondary deposits, meningioma, cerebral abscess, and haematoma within or outside the brain. Intraventricular tumours (e.g. colloid cysts) can cause headaches triggered by moving the head, and headache is triggered by coughing in patients with Arnold–Chiari malformations. Patients with a progressive history of headache should be investigated. Indications for scanning are listed in Table 3.

Idiopathic intracranial hypertension is a rare cause of progressive headache that is usually seen in obese young women. Typical patients present with overt papilloedema, though patients without papilloedema have been reported. Once a focal disturbance of brain function has been excluded by scanning, it is safe to undertake lumbar puncture. CSF pressure is found to be greatly elevated and removal of CSF often relieves the headache, if only for a few days. Acetazolamide may be helpful, but some patients require permanent shunting procedures.



Axial MRI in a 60-year-old woman who was referred to a migraine clinic with a 4-month history of daily headaches in the right temple and at the vertex; 6 weeks later (5 weeks before her appointment), she had a seizure. There were no physical signs, but the scan revealed this right thalamic mass, which was found to be a grade II glioma. She died 9 months after the referral.

Figure 1

Subarachnoid haemorrhage

Leakage of blood from an aneurysm, or occasionally from an arteriovenous malformation, causes headache of catastrophically sudden onset, often accompanied by transient or even permanent disturbances of consciousness. Neck stiffness may take hours to develop, if at all, but in most patients the haemorrhage can be seen on CT, particularly when this is undertaken soon after the ictus. If the history is typical and CT negative, lumbar puncture is mandatory. Many of these patients have normal CSF (so-called ‘thunderclap headache’); they do not need formal angiography, though a magnetic resonance venogram should be considered. These headaches are usually attributed to sudden ‘crash’ migraine, though cerebral venous thrombosis, arterial dissection and pituitary apoplexy can also mimic subarachnoid haemorrhage. If the diagnosis of subarachnoid haemorrhage is confirmed, the patient should be investigated and managed by a neurosurgeon.

When to scan a patient with headache

- First or worst headache, particularly if of sudden onset
- Headaches of increasing frequency or severity
- Increased frequency of vomiting and headache on waking
- Headache triggered by coughing, straining or postural changes
- Persistent physical symptoms or signs after attacks (neurological or endocrine)
- Meningism, confusion, impairment of consciousness or seizures

Table 3

Unruptured arteriovenous malformations sometimes present with migrainous headaches.

Meningitis

Patients with meningitis may present with an acute single headache, usually of more gradual onset and accompanied by fever and malaise. Neck stiffness is often prominent. Meningitis is discussed in *MEDICINE* 33:4. Meningococcal meningitis is uncommon, but can develop so quickly that it is essential to give antibiotic treatment without delay. It is then prudent to undertake CT of the brain (and mandatory if the patient is obtunded) before lumbar puncture. Patients with normal CSF glucose usually have viral meningitis. Granulocytes predominate in bacterial meningitis caused by *Meningococcus* or *Pneumococcus*. Lymphocytes predominate in tuberculous meningitis, which should be considered in all patients from areas where tuberculosis is common and who have a headache that has developed over 2–3 weeks.

Other causes of headache

Paget's disease of the skull is an uncommon cause of headache in older patients. It is usually unwise to attribute headache to chronic hypertension unless diastolic blood pressure exceeds 140 mmHg; another cause of the headache should be sought. In contrast, paroxysmal hypertension, which may be caused by pheochromocytoma, may present with severe, episodic headache.

Management of migraine

Careful clinical assessment can usually be followed by reassurance that the patient's symptoms, though distressing and often incapacitating, are benign. Do not overlook modern treatment options to ameliorate attacks, and do not undertake unnecessary investigations that only postpone treatment. The physician must always acknowledge the extent to which the patient's social life and employment can be disrupted by migraine. Reassurance that the long-term prognosis and life expectancy are good should be coupled with adequate management of attacks using the most appropriate drug. Some treatments are expensive, but the cost is significantly less than that of the patient being absent from work, or (perhaps more importantly) going to work and being substantially less effective while there.

Non-drug management

About 20% of patients report that headaches can be triggered by dietary items such as cheese, chocolate, citrus fruit, or some or many types of alcoholic drink. Most of these patients have already eliminated these items from their diet, and additional dietetic advice is seldom justified. There is no satisfactory evidence to support the view that, in most patients, migraine is triggered by an occult response to a foodstuff, and no evidence that these processes are immunologically mediated.

In women seeking advice about their headaches, it is often appropriate to recommend that the contraceptive pill (and perhaps even HRT in post-menopausal women) be discontinued, at least on a trial basis. There is epidemiological evidence that patients taking oestrogen-containing contraceptive pills (particularly the higher-dose types, and if the patient smokes) are at substantially greater risk of stroke, and many authorities believe that these patients should not continue taking such preparations.

Preventive treatment

It is conventional to offer daily medication to patients who experience two or more attacks per month. Several drugs are used (Table 4). Most authorities regard propranolol or atenolol as the treatment of choice in patients without a history of asthma. Doses greater than those required for cardiological indications may be necessary. Most β -blockers without partial agonist activity seem to be helpful, whether they penetrate the brain easily or not. Partial agonists (particularly oxprenolol) seem to be less effective.

The serotonin 5-HT₂ receptor antagonist pizotifen is valuable in the prophylaxis of migraine but is relatively expensive, and many patients find that it increases their appetite. Methysergide probably has a similar mode of action, though it is now recognized that a metabolite of this drug is an agonist at 5-HT_{1B} and 5-HT_{1D} receptors (as is sumatriptan). There is now good evidence that sodium valproate, topiramate¹¹, tricyclic antidepressants and even non-steroidal anti-inflammatory drugs (e.g. naproxen) are also helpful in the suppression of migraine when administered regularly. Lisinopril and candesartan¹² have also been shown to be of value, in smaller trials.

Acute treatment

Drugs for acute treatment of migraine are listed in increasing order of potency in Table 5. Most patients have tried aspirin, paracetamol and ibuprofen before seeking medical care; such patients should be given more potent anti-inflammatory drugs,

Prophylactic medication for migraine

	Usual starting dose (mg)	Dose used in trials (mg)	Cost per month (£ sterling)	Likelihood of 50% improvement compared with placebo (%)
• Propranolol	80	240	3.48	34
• Atenolol	50	100	1.62	33
• Pizotifen	0.5	1.5	5.31	28
• Methysergide	0.5, increasing every 2–4 days	6	37.69	30
• Valproate	400	1000	14.15	34
• Amitriptyline	10	100	3.58	32
• Topiramate	12.5, increasing every 7 days	100	25.89	31

Table 4

Cost of acute medication for migraine

Drug	Dose	Cost (£ sterling)
• Aspirin	600 mg	0.02
• Ibuprofen	400 mg	0.08
• Naproxen	500 mg	0.16
• Ergotamine (<i>Cafergot</i>)	1 mg	0.17
• Sumatriptan (generic)	100 mg p.o.	4.10
• Zolmitriptan	2.5 mg p.o.	4.00
• Naratriptan	2.5 mg	4.09
• Rizatriptan	10 mg	4.46
• Almotriptan	12.5 mg	3.02
• Eletriptan	40 mg	3.75
• Frovatriptan	2.5 mg	2.78
• Sumatriptan	20 mg nasal	6.15
• Zolmitriptan	5 mg nasal	6.75
• Sumatriptan	6 mg s.c.	22.10

Table 5

analgesics with anti-emetics, and then triptans, first orally and then parenterally. It is logical to try each drug in this hierarchy in successive attacks, though not within a single attack; treatment should be changed only when it is clear that the headache has failed to respond. Occasional doses of compound preparations including paracetamol and codeine are safe, but codeine and caffeine tend to perpetuate headache if taken daily.

Triptans: since the introduction of sumatriptan in 1991, the triptans have revolutionized acute management of migraine. The beneficial effects of ergotamine are thought to be mediated via its agonist properties at 5-HT_{1B} and 5-HT_{1D} receptors, and these drugs are pure agonists at these receptors without the other pharmacological properties (e.g. agonism at 5-HT₂ receptors) that seem to contribute to the adverse side effects of ergotamine. As a consequence, most authorities now consider ergotamine to have been almost entirely superseded.¹³ The pharmacological differences between sumatriptan, zolmitriptan, rizatriptan and, probably, eletriptan seem relatively small, and this is reflected in modest clinical differences, even in direct comparative trials. In contrast, naratriptan, almotriptan and, perhaps, frovatriptan have longer durations of action (with delayed clinical benefit in some cases), but there is a lower incidence of short-term side effects and a lower recurrence rate. These agents should, therefore, be recommended for patients in whom these problems are important when they are given one of the first-line drugs. Triptans are contraindicated in patients with ischaemic heart disease.

Side effects – a few patients complain of generalized paraesthesiae or muscular aches that pass quickly. Recurrence of the headache (usually 12–16 hours after administration) occurs in about one-third of patients treated with sumatriptan, zolmitriptan or rizatriptan, but this usually responds to a second dose.

Sumatriptan and zolmitriptan are available as nasal sprays, and sumatriptan is also available as a self-administered subcutaneous injection, and as suppositories in some countries. These preparations are suitable for patients in whom vomiting is a major clinical feature. Sumatriptan seems to be effective only when the patient is

already in pain, and patients whose headaches are preceded by an aura should wait until the headache phase of their attack starts.

Follow-up and prognosis

Migrainous attacks tend to fluctuate in severity over months, and patients who have sought advice about their attacks when they are worsening often improve subsequently, independently of medical intervention. It is realistic to expect patients to improve and to withdraw prophylactic treatment after 6–9 months, re-starting only if the headaches recur. This advice is particularly important for methysergide, because it is believed that discontinuing treatment after 6 months reduces the risk of retroperitoneal fibrosis. ♦

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