Part II – Neurological Disorders

CHAPTER 15
HEADACHE AND FACIAL PAIN

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HEADCHE AND FACIAL PAIN

Headache is a very common problem which affects most persons at some point during their lives. The vast majority are due to tension headache, migraine and analgesic overuse. These are termed primary headaches and are mostly benign, but for some they are very debilitating disorders. The diagnosis of primary headache is made entirely from the history as there are no abnormal signs or investigations. A much smaller group consists of secondary or medically serious headaches. These headaches are caused by an underlying disease or disorder. This group includes infections, space occupying lesions, intracranial haemorrhage and usually have signs and abnormal investigations. It is important to separate these two groups clinically as they obviously have different implications for the patient. This chapter reviews the main headache disorders and facial pain and their investigation and treatment. After reading it the student should aim to be able to recognise and distinguish the medically serious headaches and the main primary headache disorders and also facial pain.

Pain

The brain itself is a painless organ. The source of the pain in headache arises from the structures overlying the brain; these include the scalp, skull, meninges and blood vessels. The dura and blood vessels are the most pain sensitive structures in the head. The site of the headache may sometimes be a guide to the origin of the headache. In general pain arising from the anterior and middle cranial fossa is referred to the forehead and front of the head whereas pain arising in the posterior fossa and upper cervical area is referred to the back of the head and neck. Pain arising from the surface of the brain tends to be diffuse, “all over” or occipital or nuchal as in meningitis or spread more focally over the overlying vertex or parietal and temporal areas as in tumours. Pain in the head may also be referred to the face, neck, ears, eyes, teeth and sinuses.

Classification

Headaches can be broadly classified into primary and secondary. The main causes are outlined below in Tables 15.1 & 15.4.

The history

A good history based on the temporal pattern of symptoms is essential in determining the cause of headache. This will include the time course, (onset, duration etc), site, severity, pattern and factors which alter or affect it. It is important to check specifically whether this is the first attack, the onset was sudden or gradual, continuous (daily) or intermittent (periodic), increasing or decreasing and whether the time course is acute (hours and days), sub acute
(days and weeks) or chronic (months years). Determine the main site of the pain, whether it is unilateral or bilateral, frontal, temporal or occipital and its radiation. The character of the pain is also important, whether it is sharp or dull or throbbing in nature. Severity can be scored by the patient and recorded on a scale of 1-10, with 10 being the worst pain ever experienced and 1 the least. There may be some specific exacerbating factors e.g. movement, coughing, bending and relieving factors such as rest and analgesics. Check for any other associated symptoms and in particular whether there is any previous history of a similar headache or chronic analgesia intake.

Examination

Carefully record the vital signs including BP and check particularly for signs of raised intracranial pressure, meningism, tenderness of the temporal arteries, and focal neurological signs. Fundoscopy is essential as papilloedema may indicate raised intracranial pressure. Examine for local causes in the head and neck.

Key points

- headache is a common medical disorder
- vast majority of headaches are primary
- only a small minority are medically serious
- history is the most important part of evaluation of headache
- fundoscopy should be performed in all patients with persistent headaches

**PRIMARY HEADACHES**

The main primary headaches are tension, migraine, analgesic overuse, and cluster (Table 15.1).

<table>
<thead>
<tr>
<th>Primary</th>
<th>Frequency*</th>
<th>Site</th>
<th>Clinical Features</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tension</td>
<td>2-3%</td>
<td>bilateral, frontal,</td>
<td>dull, chronic, band like,</td>
<td>reassurance, relaxation amitriptyline 10-25 mg/nocte 6-12 weeks</td>
</tr>
<tr>
<td>(Chronic &gt; 6/12)</td>
<td></td>
<td>temporal, occipital,</td>
<td>scalp tenderness, episodic</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>neck</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Migraine</td>
<td>5-10%</td>
<td>unilateral, frontal,</td>
<td>throbbing, moderate/ severe,</td>
<td>avoid precipitants, regular sleep, aspirin,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>bifrontal holocephalic,</td>
<td>periodic, nausea or vomiting,</td>
<td>paracetamol, NSAIDs, ergotamine, triptans</td>
</tr>
<tr>
<td></td>
<td></td>
<td>vertex, occipital</td>
<td>photophobia, familial</td>
<td></td>
</tr>
<tr>
<td>Analgesic overuse</td>
<td>2%</td>
<td>bilateral, generalised</td>
<td>mild, bilateral, daily,</td>
<td>stop all analgesics</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>migraine or tension like</td>
<td></td>
</tr>
<tr>
<td>Cluster headache</td>
<td>0.1%</td>
<td>unilateral, retro-orbital</td>
<td>very severe, redness &amp; tearing,</td>
<td>100% oxygen, steroids, verapamil</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Horner’s syndrome, mostly in men</td>
<td></td>
</tr>
</tbody>
</table>

* based on studies in high income countries

**TENSION HEADACHE**

This is the most common form of headache experienced by most people during their lives. Tension headache is characterized by recurring daily attacks of mild to moderate bilateral headaches that may last from hours to weeks. The tension headache becomes chronic when it persists on >15 days per month and lasts for >6 consecutive months. Chronic tension headache
affects 2-3% of adults in high income countries at any one time. A lower prevalence (0.4%) was reported in one study in Tanzania. The pain in tension headache is bilateral mostly occipital, also frontal or temporal in site and often described as a tight band around the head, starting within an hour or so of waking and increasing in severity throughout the day. It frequently radiates to the back of the neck. The severity may fluctuate with the patient going for days or weeks without noticing it. It is distinguished from migraine by its bilaterality, a lack of nausea and lack of discrete episodic attacks. The source of the pain is considered to be secondary to chronic muscle contraction of the scalp, neck and face although this may in itself be a secondary phenomenon. The scalp may be painful and tender on palpation and this may provide a useful clue to the diagnosis. There may be a background of anxiety and worry and a lack of response to ordinary analgesics.

Management
Management is by reassurance, regular exercise, relaxation and local measures. The dose of analgesics may need to be reduced and finally discontinued. Low dose amitriptyline 10-25 mg/po/nocte for 3-6 months may help to break the cycle in chronic tension headache but can take at least 4-6 weeks to work. Benzodiazepines although effective in the short term should be avoided because of the danger of habituation.

Key points
- tension, migraine & analgesic overuse are the main primary headaches
- tension headache is the most common & may persist for months or years
- frequently described as dull “like a tight band around my head”
- scalp may be tender to touch
- treatment includes stopping analgesics, increasing exercise & relaxation
- specific measures include low dose amitriptyline at night for 3-6 months

MIGRAINE
Migraine is a primary headache disorder characterized by periodic attacks that are variably associated with nausea, vomiting, and neurological symptoms. Pain is typically associated with increased sensitivity to light and noise. The attacks last 4-72 hours and their frequency is variable. The majority of patients have one attack or less per week which lasts <24 hours. Migraine affects >10% of the world’s population and its frequency is reported to be >5% in Africa. A family history of migraine is present in most patients and people usually suffer their first attack in their teens or early twenties, or before the age of 40 yrs. Women are affected twice as often as men. There are a number of well known triggers for individual attacks of migraine; these include stress, relaxation, fatigue, hunger, exercise, menstruation and specific foods including cheese, chocolate, red wine, citrus fruits, food additives and caffeine. Spontaneous remission may occur during pregnancy and after the menopause.

Clinical features
In over half of patients with migraine, the onset of headache is preceded in the first 24 hours by a prodrome of warning mood changes, cravings or hunger feelings. A typical migraine may begin with an aura in 10-20% of patients. It occurs characteristically during the hour before the onset of headache. The commonest aura is visual with bright zig zag lines and blurring or
loss of vision. Less commonly, there may be tingling in one limb spreading to ipsilateral tongue and face and dysphasia. Rarely there is a temporary hemiplegia or even complete aphasia. The aura typically lasts a few minutes but sometimes may last longer. These symptoms precede the headache and mostly resolve as the headache starts. When these are present, it is called **classical migraine** or migraine with aura and when absent is called **common migraine**.

The next phase is the headache itself. The headache of migraine is characteristically dull at onset and later becomes throbbing, severe, usually unilateral, frontal, radiating to the neck. It may be bilateral in one third of patients. The headaches last between 4 hours and 3 days with most attacks lasting <24 hours. Normal physical activity usually makes the headache worse. The headache is often accompanied by pallor, nausea or vomiting and the need to lie in a quiet and darkened room. A period of sleep early on may abort the attack. The patient feels completely normal between the attacks but they can recur at variable intervals. Investigations are generally not necessary.

**Key points**

- migraine affects >5% adults in Africa, females >males and clusters in families
- aura is characterized by bright zig zag lines and blurring or loss of vision
- headaches are periodic, mostly unilateral & throbbing lasting 4-72 hours
- associated with nausea/vomiting & visual/neurological disturbance

**PATHOGENESIS**

This involves abnormal nerve impulses originating mainly from the brain stem in the distribution of the trigeminal nerve and spreading to the cortex causing release of inflammatory substances and neuropeptides that stimulate pain fibres on meningeal arterioles. The symptoms of migraine have been attributed to an initial spreading occipital wave of vasoconstriction of cortical blood vessels with decreased blood flow (the aura phase) followed later by regional cortical vasodilatation (the headache phase). Serotonergic neurones occur extensively in the brain and brain stem and their involvement in migraine is suggested by the effectiveness of the 5–hydroxytryptamine (5-HT1) agonists e.g. the triptans in treating the headache phase of an acute attack.

**Treatment**

Identification of trigger factor(s) and avoiding them should be the first line of treatment. Attacks are often infrequent and mild, and resolve with simple analgesics such as aspirin, paracetamol or ibuprofen taken either alone or in combination with an antiemetic, like metoclopramide. Failing that the triptans are of great benefit (Table 15.2). The triptans are the drugs of first choice but are contraindicated in pregnancy, coronary artery disease, vascular disease, and uncontrolled hypertension. They are not recommended in hemiplegic migraine or in migraine with complex auras (sensory, motor or speech disturbances). They are recommended to be taken at the onset of the headache but not before. The main route is oral but if vomiting is a problem then triptans can be given either by nasal spray or subcutaneous injection. A second dose is usually effective for relapse within the first 6-48 hours. The side effects of triptans include chest discomfort and nausea and are more common with parenteral administration. Information on sumatriptan is outlined below, but any of the other triptans are equally efficacious.
An alternative cheaper treatment regime is ergotamine, however it should never be used in pregnancy and regular repeated usage is not recommended because of the danger of ergotism, gangrene, and rarely pulmonary fibrosis. Ergotamine is also contraindicated in peripheral and cardiovascular disease. It is important to note that the combined use together of a triptan and ergotamine is also contraindicated. If the patient is not responding to the combination of analgesics and antiemetics then triptans may be helpful

Table 15.2  Treatment of migraine

<table>
<thead>
<tr>
<th>Acute treatment</th>
<th>Dose/range/frequency</th>
<th>Main side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Non specific treatment</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>aspirin</td>
<td>300 mg tab, 2-3 tab/po/12 hourly</td>
<td>bleeding</td>
</tr>
<tr>
<td>paracetamol</td>
<td>500 mg tab, 2 tab/po/6 hourly</td>
<td>nausea</td>
</tr>
<tr>
<td>ibuprofen</td>
<td>2-400 mg tab, 1-2 tab/po/6-8 hourly</td>
<td>bleeding</td>
</tr>
<tr>
<td>metoclopramide/domperidone</td>
<td>10 mg tab, 1 tab/po/6-8 hourly</td>
<td>dyskinesia</td>
</tr>
</tbody>
</table>

| **Specific treatment** | | |
| Ergot derivatives | | |
| ergotamine tartrate | 1 mg tab, 2 tab/po or suppositories/at onset followed by 1 tab every 30 mins, (max 24 hours dose 6 mg, the total max weekly dose is 10 mg) | nausea, vomiting, ergotism |
| dihydroergotamine* | 0.5-1mg/iv/8 hourly as required, (max total dose 10 mg, supervised) | |

| Triptans | | |
| sumatriptan** | 50 mg tabs, 1 or 2 tab/po/at onset, repeat in 2 hours, (max 24 hour dose 200 mg) | chest tightness, paraesthesiae, fatigue |
| or | 6 mg/sc/at onset, repeat in 2 hours, (max 24 hour dose 12 mg) | |
| or | 5-20 mg/nasal spray at onset, repeat in 2 hours, (max 24 hour dose 40 mg) | |

* used only in intractable migraine in specialist headache units
** other triptans are equally effective

**PROPHYLAXIS**

Dietary triggers for migraine should be avoided and oestrogen containing contraceptives used with caution. Preventative treatment reduces the frequency, severity and duration of the attacks (Table 15.3). If the frequency is weekly or greater or the attacks are disabling, then those patients may benefit from daily prophylaxis. Medications used in prophylaxis and their dosages and main side effects are outlined below, and the initial treatment duration is for 3-6 months. The most commonly used options include amitriptyline, beta-blockers and sodium valproate. The anticonvulsant topiramate can also be very effective in cases resistant to other medications, but it is more expensive.
Table 15.3 Prophylaxis of Migraine

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dosage/range/frequency</th>
<th>Main side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beta blockers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>propranolol</td>
<td>10-80 (160) mg/po/bid</td>
<td>postural hypotension, fatigue</td>
</tr>
<tr>
<td>atenolol</td>
<td>50-200 mg/po/daily</td>
<td></td>
</tr>
<tr>
<td>Tricyclics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>amitriptyline</td>
<td>10-100 mg nocte</td>
<td>dry mouth, sedation, urinary retention</td>
</tr>
<tr>
<td>Anticonvulsants</td>
<td></td>
<td></td>
</tr>
<tr>
<td>sodium valproate</td>
<td>250-750 mg/po/bid</td>
<td>nausea, weight gain, alopecia, tremor, liver dysfunction</td>
</tr>
<tr>
<td>topiramate</td>
<td>25–50 mg bid</td>
<td>renal stones, paraesthesia, weight loss</td>
</tr>
<tr>
<td>Calcium channel blockers</td>
<td>40-160 mg/po/tid</td>
<td>constipation, fatigue, oedema</td>
</tr>
<tr>
<td>5-HT2 antagonists</td>
<td></td>
<td></td>
</tr>
<tr>
<td>pizotifen</td>
<td>0.5-3 mg/po/daily</td>
<td>weight gain</td>
</tr>
</tbody>
</table>

Key points

- majority do not require treatment or respond to simple analgesics & antiemetics
- triptans are the treatment of choice but are expensive and have contraindications
- ergotamine preparations may also be used at onset but have contraindications
- if attacks are severe or >1 per week then daily prophylaxis is recommended
- main prophylaxis includes beta blockers or anticonvulsants in adequate doses

ANALGESIC OVERUSE HEADACHE

These are a group of headaches lasting longer than 4 hours a day which persist for at least 15 days every month for at least 3 months. The term chronic daily headache (CDH) is also used to describe them. These are usually cases of transformed migraine or chronic tension headaches and may affect over 2% of the population in high income countries, most commonly females. These occur mostly as a result of prolonged use of analgesics including simple analgesics, non-steroidal anti-inflammatory drugs (NSAIDs), opiates, ergotamine and triptans. Patients typically complain of daily throbbing bilateral headaches which are only transiently and incompletely relieved by increasing doses of medications. Neurological examination is entirely normal.

Management

The management aim is to decrease the frequency, severity and duration of the headaches by complete withdrawal of medication. The patient will need to be encouraged to have a regular life style and avoid caffeine and be specifically educated about the overuse of analgesics. In particular they will need to understand that there will be persisting symptoms including headaches, nausea, agitation and insomnia, particularly during the first two weeks after stopping. Withdrawal for ordinary analgesics, ergotamine and triptans should be carried out abruptly over a period of 24-48 hours. Withdrawal of opioids may take a period of 2-4 weeks. Remission occurs 2-12 weeks after withdrawal. The patient may need interim symptomatic treatment with an antiemetic, NSAIDs and occasionally a brief course of steroids. Underlying anxiety and depression may also need to be treated. Preventive medications include amitriptyline 10-50 mg/po/nocte.
Key points

- analgesic overuse is a cause of chronic daily headache
- most common sources of CDH are chronic tension headaches & transformed migraine
- management is abrupt withdrawal of all non opioid medication over 24-48 hours
- opioids may take 2-4 weeks to withdraw

CLUSTER HEADACHE

This is an excruciatingly severe, unilateral, headache located around one eye and accompanied by local autonomic dysfunction, redness, swelling and watering of the eye. It occurs in high income countries with a frequency of approximately 1/1000. It has its onset mostly in the 3rd and 4th decade and the male female ratio is about 5:1. It receives its name from its tendency to cluster usually 1-3 times daily (can be up to 8 times) for periods of 3-6 weeks or longer at a time with long intervals, sometimes years completely free of attacks. The attacks are brief, lasting between 30-120 minutes, in contrast to migraine which persists for 4-72 hours. Cluster headaches typically occur at the same time in the 24 hour cycle often waking the patient from sleep.

Management

Stopping the acute attacks of pain quickly is critical. These can be well controlled by inhalation via a mask of 100% oxygen @ 7-10 litres/min for 15-20 minutes. Triptans can be successful if used by injections (e.g. sumatriptan 6 mg/sc) or intranasally (sumatriptan 20 mg). Alternatives include zolmitriptan 5 mg orally. They can be repeated once in 24 hours but should be avoided in patients with multiple attacks because of the danger of overuse. Concomitant use of ergot drugs is absolutely contraindicated because of the danger of a stroke. Prophylaxis during a cluster can be helpful with high dose steroids, prednisolone 60 mg/po/daily for 5 days decreasing by 10 mg every 3 days. Long term prevention is indicated for chronic cluster headaches. This includes verapamil 80 mg bd for 2 weeks increasing by 80 mg every 2 weeks to a maximum of 320 mg bd or tds. Avoiding tobacco and alcohol is also important.

Key points

- cluster headache is severe, unilateral with redness and tearing of the eye
- recognised by its distinctive pain, recurrence and male preponderance
- acute attack can be treated with 100% oxygen inhalation or as for migraine
- prophylaxis is with high dose steroids or verapamil for 1-2 months

SECONDARY HEADACHES

The main causes of secondary headaches are intracranial tumours, infections, intracranial haemorrhage, temporal arteritis and benign intracranial hypertension (Table 15.4). Other causes to be considered include hypertension, arterial dissection, head injury, brain abscess, subdural haematoma and medications. Secondary headaches may lead to serious consequences if the underlying cause is not identified and treated.
Table 15.4  Secondary headaches

<table>
<thead>
<tr>
<th>Main Causes</th>
<th>Onset</th>
<th>Type</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumour</td>
<td>sub acute</td>
<td>severe, generalised, worse on waking, aggravated by coughing &amp; straining</td>
<td>↑ICP &amp; FNDs*</td>
</tr>
<tr>
<td>Meningitis</td>
<td>acute/sub acute</td>
<td>severe, generalised &amp; meningism</td>
<td>fever, neck stiffness &amp; Kernig’s sign</td>
</tr>
<tr>
<td>Subarachnoid haemorrhage</td>
<td>acute</td>
<td>explosive, thunderclap &amp; meningism</td>
<td>neck stiffness, Kernig’s sign, ± FNDs*</td>
</tr>
<tr>
<td>Temporal arteritis</td>
<td>sub acute</td>
<td>non specific/uni/bilateral</td>
<td>tender temporal arteries, blindness, age &gt;50-60 yrs, ESR&gt;60</td>
</tr>
<tr>
<td>Benign intracranial hypertension</td>
<td>sub acute</td>
<td>throbbing, visual obscurations &amp; loss</td>
<td>female, young, overweight, visual loss, papilloedema, 6th nerve palsy, gait ataxia</td>
</tr>
</tbody>
</table>

* focal neurological disorders/deficits

**INTRACRANIAL TUMOUR**

The majority of patients presenting with brain tumour do not have headaches. The headache of an expanding intracranial tumour is produced by raised intracranial pressure and is often described as bursting and severe in nature. The pain is typically continuous and increases in severity over weeks or months. Characteristically it is present on waking in the morning or may wake the patient at night and improves during the day. It is aggravated by measures that increase intracranial pressure; these include lifting, bending, lying down, coughing, sneezing and straining. The distribution of the pain is either generalized or referred to the fronto/temporal area, or vertex/occipital area depending on the site of origin of the tumour. The presence of nausea, vomiting, visual disturbances, altered level of consciousness all suggest raised intracranial pressure.

Neurological examination should check carefully for papilloedema (Chapter 12) and focal neurological findings. A CT scan of the head is indicated in patients when a SOL is suspected. Details on brain tumours, abscess, subdural haematoma and other space occupying lesions are presented elsewhere in this textbook (Chapters 6, 7, 16 & 19).

**Key points**

- majority of patients with headache do not have an intracranial tumour
- clinical features include bursting pain on waking, increasing over time, worse on coughing/straining
- tumours rarely present with headache alone & usually have FNDs or ↑ICP
- head CT scan is indicated in patients with suspected SOL

**MENINGISM**

Meningism results from inflammation or irritation of the meninges. The two major causes are meningitis and subarachnoid haemorrhage (Chapters 5 & 6). The headache in meningitis is usually severe, generalised and associated with fever, photophobia and signs of meningism including neck stiffness and Kernig’s sign. In subarachnoid haemorrhage the headache is
typically sudden or explosive in onset with vomiting and meningism. Diagnosis is confirmed by lumbar puncture. Headaches frequently occur with other CNS infections including encephalitis.

**Key points**

- headache in meningitis is severe & generalised
- headache in SAH is sudden and explosive
- both result in symptoms and signs of meningism

**TEMPORAL ARTERITIS (TA)**

In persons >50 yrs old the extra cranial vessels may become inflamed with an arteritis. The arteritis causes a new onset, severe, throbbing, mostly bilateral headache and exquisite local scalp tenderness. The headache may be non specific at onset later localizing to the temples. Exacerbation of headache with chewing suggests the diagnosis. On palpation, the superficial temporal artery may be hot, tender, swollen and non pulsatile. Other arteries may be similarly affected and blockage of the ophthalmic artery may result in transient blindness which can become permanent in about 25% of cases. Strokes are not uncommon. Females are affected more frequently than males and temporal arteritis is associated with polymyalgia rheumatica. The ESR is characteristically elevated >60 mm/hr. The age of onset is usually >60 yrs. Diagnosis is confirmed by a temporal artery biopsy which ideally should be performed early on but this may not always be practical.

Management is with immediate high dose steroids as soon as the diagnosis is suspected without waiting for investigation results. Prednisolone 60 mg/po/daily is prescribed for a total of 2-3 months and then tapering to a maintenance dose of 5-10 mg on alternate days. The response to steroids is usually dramatic, often within the first 24-48 hours. Steroids may be needed for a total of 18-24 months or until symptoms fully clear. Treatment is guided by monitoring the ESR.

**Key points**

- headache in TA is intense, focal & localised to the temples
- temporal arteries may be tender on palpation
- blindness and strokes are major complications
- both patient's age and the ESR are generally >60
- treatment: high dose daily steroids for 2-3/12 & maintenance dose for 18-24/12

**BENIGN INTRACRANIAL HYPERTENSION (BIH)**

It is an uncommon disorder characterized by headaches with elevated intracranial pressure without any evident cause. It is more widely known in Africa as pseudotumour cerebri. This occurs mostly in young, overweight females in their 20s and 30s. The oral contraceptive pill is a risk factor. Patients typically present with a severe morning throbbing type headache often associated with nausea, vomiting and sometimes visual disturbances. Visual disturbances include transient often postural episodes of loss of vision lasting seconds, as well as more sustained
blurring and sometimes permanent loss of vision. The characteristic finding is bilateral papilloedema. Neurological examination is otherwise normal apart from occasional isolated 6th nerve palsy and a mild gait ataxia. The differential diagnosis includes the other causes of medically serious headaches including cerebral venous sinus thrombosis. Brain imaging with a CT is always normal but should be carried out in order to exclude another organic cause. The diagnosis is confirmed by finding an elevated CSF opening pressure of >250 mm on lumbar puncture.

Management
Acute treatment measures include repeated lumbar punctures in combination with high dose steroids used over 3-5 days. The main long term management includes weight loss, stopping the contraceptive pill and a diuretic, acetazolamide tablets 250 mg tid. Furosemide is an alternative diuretic. BIH remits spontaneously after these measures in many patients but the measures, in particular weight loss, have to be vigorously adhered to. If there is visual loss, (decreased visual acuity, and constricted visual fields) then a lumboperitoneal CSF shunt or optic nerve sheath fenestration should be considered to prevent permanent blindness.

Key points
- BIH presents as headaches, visual disturbances & papilloedema
- typically occurs in young overweight females taking the contraceptive pill
- neurological investigations are normal including neuroimaging
- acute measures include steroids over first 3-5 days and repeated lumbar punctures
- long-term management includes weight loss, diuretics & stopping the pill

Other causes of headache
Other primary headaches include trigeminal neuralgia (see below under facial pain), exertional and coital headaches. These latter headache types are precipitated by exercise or sexual activity. These are benign but can mimic a SAH and this may need to be excluded.

FACIAL PAIN
Facial pain has many local causes including diseases of the eyes, ears, nose, sinuses, throat, teeth and temporomandibular joints and referred pain. Neurologic causes of facial pain need to be distinguished from local causes. Many of the neurological disorders which cause headache may also cause facial pain; these include cluster headache, temporal arteritis and occasionally migraine. However there are some neurological disorders where the pain is restricted to the face, these include trigeminal neuralgia, post herpetic neuralgia and atypical facial pain. The local causes of facial pain are usually determined clinically by their site and associated clinical findings.

TRIGEMINAL NEURALGIA (TN)
This is a very painful disorder affecting the trigeminal nerve. It affects persons of all age groups but mostly those over the age of 40 yrs. It is more common in females than males. Previously no cause was found in most patients with trigeminal neuralgia and these were termed idiopathic. However many of what were previously termed to be idiopathic causes have been shown by MRI to be due to compression of the trigeminal sensory nerve root near the brain stem by an
aberrant arterial branch. Secondary causes include herpetic infection and a cerebellopontine angle tumour compressing the trigeminal nerve during its intracranial course.

Clinical features
Patients with trigeminal neuralgia typically present with unilateral facial pain in the distribution of one or more divisions of the trigeminal nerve. The pains are characteristically sudden, severe, brief, shooting, electric shock like, lancinating or knife like stabs in the distribution of one or more branches of the trigeminal nerve. The attack lasts less than a second usually affecting only one side of the face, typically the corner of the mouth or nose. There are frequently trigger areas on the face where any stimulus however gentle may produce pain. The pain may be triggered by touching, talking, eating, drinking cold or hot liquids or cleaning teeth. The stabs of pains can recur continuously many times per day. Between the stabs there may be a lingering continuous background pain but it is usually much less severe.

The natural history is one of remission but only after weeks or months of facial pain and the neuralgia typically recurs again after a few months. Spontaneous remission can occur. The differential diagnosis includes local causes of facial pain including dental infections, sinusitis, disease of the temporomandibular joint and other atypical forms of facial pain

Management
The pain does not respond to simple analgesics and requires adequate doses of anticonvulsants for control. Carbamazepine is the drug of first choice and a favourable response to treatment is diagnostic of TN. The standard starting dose is 100 mg/po/bd increased as needed over the next few days or weeks. A usual therapeutic and maintenance dose of carbamazepine is 200 to 400 mg twice or three times daily. This may be continued for a period of one or two months depending on the duration of the episode of facial pain and the patient’s tolerance of the drug side effects. Drowsiness and ataxia are the main limiting side effects. Second line drugs include gabapentin, pregabalin, phenytoin and sodium valproate. If the pain becomes intractable or intolerable and an adequate trial of drug treatment fails then the nerve can be considered for interruption either temporarily with an injection of alcohol or more permanently with phenol, glycerol or a destructive procedure.

Key points
- trigeminal neuralgia is very painful & affects branches of trigeminal nerve
- recurring shooting facial pains lasting less than a second are diagnostic
- pain is triggered by touching, eating, hot & cold, cleaning teeth & talking
- natural history is remission but both attacks and remissions last months
- carbamazepine taken in adequate doses daily is the treatment of choice

Selected references
CHAPTER 15  HEADACHE AND FACIAL PAIN