

Chapter 5

133

Headaches

- Headaches 134
- Hydrocephalus 138
- Shunts and shunt complications 140
- Subarachnoid haemorrhage 142
- Intra-cerebral haemorrhage 144
- Cavernous sinus thrombosis 146
- Sagittal sinus thrombosis 148
- Cerebral tumours 150

⊕ Headaches

'Facial pain' conditions may also present as a headache. This can be a clinical entity in its own right or the manifestation of extra-cranial disease.

Take headaches seriously. Although the majority have a benign cause, if not assessed thoroughly, serious pathology may be missed.

Headache is a common presenting symptom and may be attributable to an obvious diagnosis (e.g. meningitis, subarachnoid haemorrhage), or described as a feature of a less obvious local cause (e.g. TMJ dysfunction).

Most headaches are benign and self-limiting, but a few headaches are due to serious pathology, which may prove fatal or result in significant disability without prompt treatment.

Headaches persisting for longer than 6 weeks with abnormal physical signs should be thoroughly investigated, including:

- FBC with ESR (to exclude temporal arteritis);
- chest X-ray for bronchial carcinoma;
- CT scan to exclude space occupying lesion.

In all cases consider:

- **intra-cranial infections:**
 - meningitis
 - brain abscess
 - subdural empyema
- **mastoiditis**
- **intra-cranial haemorrhage:**
 - subarachnoid haemorrhage
 - intra-cerebral haemorrhage
- **brain tumours**
- **hydrocephalus (see Emergencies)**
- **headache due to raised intra-cranial pressure**
- **dural venous thrombosis:**
 - cavernous sinus thrombosis
 - sagittal sinus thrombosis
- **post-traumatic haematomas (see Emergencies):**
 - extradural haematomas (small)
 - subdural haematomas (acute and chronic)
- **cerebral contusions (see Emergencies)**
- **migraine**
- **cluster headaches (migrainous neuralgia)**
- **tension headache**
- **post-concussion headache**
- **temporal (giant cell) arteritis**
- **glaucoma.**

Temporal arteritis

Temporal arteritis (giant cell arteritis) is a vasculitic disease predominantly affecting the over-sixties. It is an important diagnosis in the elderly patient who presents with severe headache because of the potential for blindness if left untreated.

Aetiology and pathology

The aetiology of temporal arteritis is unknown; however, many patients report a prodromal flu-like illness. It may be that the systemic manifestations of the disease have a viral infection as their cause, although no distinct virus has been implicated.

Histologically there is a panarteritis with giant cell granuloma formation in a disrupted internal elastic lamina. The intimal thickening causes reduced vessel calibre or obliteration, and the vessels are enlarged and nodular. Involvement of the arteries is often patchy, with the description of 'skip lesions' often been made—this is an important feature when undertaking temporal artery biopsy for diagnostic purposes.

Clinical features

Patients present with a headache that may be a generalized 'tension' type or severe and well-localized over the region of the temporal arteries, often with burning or tenderness of the scalp. Jaw claudication with pain on chewing is another common feature and is thought to be due to involvement of the facial artery. Patients may also complain of an altered sensation of the oro-pharynx and a loss of taste. Systemic problems are also common and often made manifest with weight loss, arthralgia and fever.

Of great importance is the particular danger of the **sudden irreversible loss of sight** and optic complications may occur within weeks of the onset of systemic features of the disease. Often the presenting feature is of a visual field disturbance, which becomes progressively worse. Blindness is thought to occur as a result of ischaemic optic neuritis caused by arteritis of the ophthalmic arteries.

Temporal arteritis generally affects medium- and large-sized arteries with an internal elastic component. Branches of the carotid arteries are the commonest sites of involvement, but the vertebral, meningeal, and intra-cerebral vessels can be involved leading to hemiplegia, epilepsy, or local lobe lesions.

Investigations

The clinician has to be astute to the possible diagnosis of temporal arteritis, particularly in the patients with impending visual problems.

ESR—erythrocyte sedimentation rate, is usually markedly raised in excess of 90 mm/h in these patients.

Temporal artery biopsy—will help to confirm the diagnosis on the basis of the pathological findings. However, it must be remembered that the disease generally shows 'skip lesions' and therefore a negative biopsy does not exclude a diagnosis, particularly in the presence of a raised ESR.

Management

The aims of management are to reduce the pain and distressing symptoms of the condition and the prevention of complications, particularly blindness.

Steroids—usually high dose (40–60 mg daily) given urgently to suppress symptoms. Dosage can be titrated against the ESR, and affect on symptoms, but it is often necessary to continue treatment for 2–3 years with a gradually reducing dose until there is no evidence of the disease.

Polymyalgia rheumatica (PMR)

Is a clinical condition of middle-aged and elderly patients which has a recognized association with temporal/giant cell arteritis and is characterized by:

- systemic upset—weight loss, fever, fatigue;
- severe arthralgia—pain and stiffness, usually bilateral and symmetrical;
- ESR—elevated;
- clinically, a rapid response to small doses of corticosteroids.

Around 50% of patients with temporal arteritis have symptoms of PMR, whereas a range of 15–50% of patients with PMR has giant cell arteritis. The clinical entities are more common in the elderly, particularly women, with a reported incidence in the region of 1 in 10 000.

Glaucoma

Patients may complain of facial pain in and around the eyes and it is important to consider ophthalmic conditions as a possible cause especially glaucoma.

Migraine

Migraine is a severe headache that may present as a facial pain affecting the cheek, orbit, or forehead. However, classical migraine with preceding visual disturbances and an aura rarely affects the face. 'Common' migraine is ten times more frequent and is described as a severe pulsatile headache invariably associated with nausea.

Migraine is episodic in nature and is thought to affect approximately 10% of the population. It is more common in females (3:1), usually begins around puberty, and continues into middle-age, and there may also be a family history.

Classic migraine is described as starting with an impending sense of ill health and a visual aura (e.g. flashing lights). The throbbing, severe, sharp, unilateral headache is associated with anorexia, nausea and vomiting, photophobia, and withdrawal—the patient often wants to just go into a darkened room and sleep.

Associations have been made with such trigger factors as stress, diet (chocolate, cheese, red wine), hormonal state (pre-menstrual, OCP), emotions (anger, excitement), and barometric changes.

Management

Recognizing and removing precipitating causes, with simple analgesics in the first instance. Anti-emetics may also be used to reduce nausea. If attacks are frequent and affect routine daily activities, then prophylactic treatment can be considered with, for example, oral pizotifen at night, or daily beta blockers. In severe cases, patients may be prescribed sumatriptan to use in the prodrome state.

Cluster headaches (migrainous neuralgia)

So called because attacks generally occur in clusters, usually at night for 1–3 weeks, every 12–18 months. More common in men between 20 and 40 years, it may be precipitated by alcohol. Typically the patient is woken at night by a severe unilateral stabbing or burning pain, which may be frontal temporal, around the eye, or over the cheek. Nausea is not a common

feature but there is frequently rhinorrhoea, unilateral nasal obstruction, and the eye may be red (conjunctival injection) with lacrimation.

Cluster headaches often respond to ergotamine.

Tension headache

Tension headaches are described as a feeling of pressure, or a 'band-like' tightness that varies in intensity, frequency, and duration. It is often felt bilaterally over the forehead or temples but may affect the vertex, occiput, or eyes. Commonest in middle-aged women with associated stress or depression, it may be chronic or episodic and is only occasionally helped with simple analgesics (NSAIDs). Definite reassurance and a thorough normal physical examination are often therapeutic.

Post-concussion headache

May have features of a tension type headache but is often associated with dizziness and loss of concentration.

Headache due to raised intra-cranial pressure

Usually due to tumour, haematoma, or abscess, the headache is often associated with vomiting, is worse on waking, and improves a few hours after rising. Straining, coughing, or sneezing may exacerbate the headache, although simple analgesics are often useful.

! Hydrocephalus

Hydrocephalus is classified as:

- **Communicating hydrocephalus:** there is free flow of CSF from the ventricular system to the subarachnoid space. The hydrocephalus is usually due to failure of CSF absorption. It is safe to perform a lumbar puncture. A shunt is the only treatment option available.
- **Non-communicating hydrocephalus:** there is an obstruction within the ventricular system so that the CSF cannot reach the subarachnoid space. It is **not safe to do a lumbar puncture in this group**. Treatment options include a third ventriculostomy to bypass the obstruction internally.

Clinical features

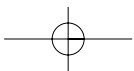
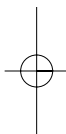
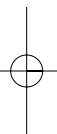
- Headache.
- Vomiting.
- Visual disturbance.
- Deterioration in consciousness.

Investigations

- A **CT scan** will show ventricular dilatation. The fourth ventricle is usually dilated in communicating hydrocephalus, but is small in non-communicating hydrocephalus. The cause of the obstruction might also be visible.
- An **MRI scan** might be necessary, particularly if a third ventriculostomy is being considered, to visualise the anatomy of the basal cisterns.

Management

- **Shunts:** divert CSF into the peritoneum, or less commonly the right atrium.
- **Third ventriculostomy:** creates an internal bypass by forming a stoma between the floor of the third ventricle and the basal cisterns through the lamina terminalis.
- **External ventricular drainage:** the CSF drains via a manometer to an external collecting system. This is usually only performed if there is infection or bloodstained CSF preventing shunt insertion, or in an emergency when there is insufficient time to insert a shunt (it can be done at the bedside with the appropriate equipment).



! Shunts and shunt complications

Shunts consist of:

- ventricular catheter;
- subcutaneous reservoir—for sampling CSF;
- valve—this might have a pumping chamber, depending upon the type;
- distal catheter—most commonly to the peritoneum (VP shunt), but occasionally to the right atrium via the internal jugular vein, etc (VA shunt).

Shunt assessment

- **CT scan:** to look at the ventricular size. It is most useful to compare the scan with a previous scan taken when the shunt was known to be functioning. However, in patients who have had multiple-shunt revisions, the ventricular wall can become stiff and might not dilate.
- **Shunt series:** plain X-rays of the whole of the shunt to look for breakages, disconnections, or migration of the shunt from its usual location.
- **Shunt palpation:** if the pumping chamber can be emptied, but does not refill, it suggests the ventricular catheter is blocked. If the pumping chamber cannot easily be emptied, the distal catheter is blocked. However, if the shunt is old and the materials have lost their compliance this is unreliable.
- **Shunt tap:** a needle is inserted into the subcutaneous reservoir (not the valve pumping chamber) to:
 - measure CSF pressure using an LP manometer. If there is no flow the ventricular catheter is blocked;
 - sample CSF for microbiology.

Shunt obstruction

The commonest site of a blocked shunt is the ventricular catheter (due to choroid plexus), followed by the valve (due to CSF debris) and the distal catheter (due to omentum in VP shunts and clot in VA shunts). A blocked shunt usually presents with similar symptoms to the patients original hydrocephalus, but the **symptoms often progress more rapidly**. CT scan usually confirms the diagnosis, but a shunt tap might be necessary in symptomatic patients with small ventricles. **All symptomatic patients should be admitted** for observation until their symptoms have settled. The obstructed component, or preferably the whole shunt, will need to be replaced. Attempts to clear the obstruction usually fail.

Shunt infection

Shunt infections usually develop within a few weeks of the last shunt operation and are due to contamination from skin bacteria. An infected VP shunt will usually become obstructed by omentum localizing the infection. Patients therefore present with **symptoms of a blocked shunt accompanied by a fever**. They usually do *not* have meningism. An infected VA shunt will usually not block and so the infection may continue undetected for a long period. The symptoms of an infected VA shunt usually consist of **vague ill health and a low-grade temperature**. Diagnosis is by a shunt tap, with CSF microscopy and culture. The CSF white cell count might be

normal, as CSF flow flushes the bacteria away from the ventricles. Antibiotics alone are insufficient to clear a shunt infection. Removal of the shunt and external ventricular drainage are also necessary, with a new shunt being inserted when the CSF is sterile.

Prophylactic antibiotics have not been shown to prevent shunt infections.

Shunt overdrainage

Occasionally a shunt will drain excessive CSF, so that the patient develops low-pressure headaches, which are **worse when upright and are eased by lying down**. If the ventricles are very large, the low pressure can cause them to collapse, tearing cortical bridging veins and causing subdural haematomas. These patients usually have symptoms of raised intra-cranial pressure with a hemiparesis.

Low-pressure headaches are treated with reassurance and advising a high fluid intake. The shunt can be revised if the symptoms persist.

! Subarachnoid haemorrhage

Aetiology

- 70% intra-cranial saccular (Berry) aneurysm (often remain asymptomatic—2% finding in routine post-mortems)
- 15% no identifiable cause;
- 5–15% arteriovenous malformation;
- Rare causes, for example:
 - inflammatory vascular disease;
 - cerebral amyloid angiopathy;
 - drug abuse;
 - scorpion bite!

One-third of aneurysmal subarachnoid haemorrhage (SAH) patients die from the initial bleed. Another third (half of the survivors) will have another bleed within 6 weeks, and 50% of the re-bleeds will be fatal. The likelihood of further bleeds in the remaining patients gradually falls over a year to a baseline level of 1% per year. It is therefore vital not to miss the diagnosis, as these patients remain at risk of sudden death.

The risk of re-bleeding in AVMs is much less at 6% in the first year and 3% for each subsequent year, so treatment can be delayed until the patient has settled.

Clinical features

- ‘Thunder clap’ headache—a sudden severe occipital headache radiating over the head and down the neck.
- Impaired conscious level—ranging from transient loss of consciousness to comatose, dependent on site and size of bleed.
- Meningism and neck stiffness.
- Photophobia.
- Vomiting.
- Fitting.
- Hemiparesis and/or dysphasia (in poor grade cases)—focal neurology will only occur if bleeding has also occurred into the brain substance.

Some patients report a sudden onset headache, that eased within a few hours, several weeks before a major SAH—a ‘herald bleed’. **All patients with a sudden-onset headache should therefore be investigated for SAH, even if the headache eased within a few hours.**

Management

- **Resuscitation**, IV fluids, clotting studies.
- **Analgesics, anti-emetics**.
- **CT scan**. This should be performed as soon as possible after the bleed, when it will be diagnostic in 90% of cases. A delay in performing the scan reduces its diagnostic rate as the blood lyses. **A normal CT scan does not rule out a SAH.**
- **Lumbar puncture (LP)**. This is required in all suspected cases if the CT scan is normal, but if performed too soon, the CSF can be normal, as the blood has not reached the lumbar region. The LP should not therefore be performed within 6h from the bleed. The diagnostic

finding is xanthochromia, but all cases with bloodstained or equivocal CSF should be discussed with the neurosurgical unit.

- **Oral nimodipine.** Improves outcome by reducing the risk of ischaemic complications.
- In the neurosurgical unit, **cerebral angiography** will be performed to determine the cause.
- Aneurysms are secured by either **surgical clipping or endovascular embolization**. AVMs can be excised, embolized, or treated with extremely high-dose, finely localized radiation (**stereotactic radiosurgery**), which leads to gradual obliteration over a 2-year period. If no structural cause is found, the patient can be reassured that they are not at risk of further bleeds.

! Intra-cerebral haemorrhage

Intra-cerebral haemorrhage, a form of stroke, is most commonly due to **hypertension**. Bleeding disorders, AVMs, aneurysms, tumours, and venous hypertension due to central venous thrombosis can also be responsible.

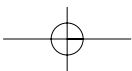
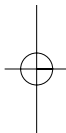
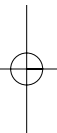
Clinical features

These include the following, but not all need be present:

- headache
- loss of consciousness
- focal neurological deficit.

Management

- **Resuscitation**, IV fluids, clotting studies.
- **CT scan**. This should be performed as soon as possible after the bleed, especially if the patient is unconscious or an aneurysmal SAH is a possibility. MRI scans are best avoided, as the appearance of a haematoma on them is variable. An LP is unnecessary and potentially dangerous.
- **Angiograms** might be performed, especially if the clot is close to the Circle of Willis or Sylvian Fissure (possible aneurysmal cause) in younger non-hypertensive patients (possible AVM), or if surgical evacuation is being considered.
- **Surgical evacuation**. The role of surgery is controversial, as evacuating a cerebral haematoma has not been proven to improve the outcome. Most neurosurgeons would evacuate the haematoma in a semi-conscious patient, or a patient who starts in good condition but deteriorates later due to cerebral oedema.
- **Stroke rehabilitation**. This will be necessary in the majority of patients.



! Cavernous sinus thrombosis

Often fatal in the pre-antibiotic era, cavernous sinus thrombosis is essentially a septic thrombosis within the cavernous sinus. **It usually arises from an infection in the face**, most commonly the peri-orbital region, but can also arise from paranasal sinus infection. Propagation of an infected thrombus to the cavernous sinus occurs against venous flow, because of the absence of valves in the facial, angular, ophthalmic, and pterygoid plexus of veins. Thrombosis might spread to other venous sinuses and the infection may spread to cause subdural empyema or meningitis. Infective endocarditis and thrombosis of the internal carotid artery can also occur.

Clinical features

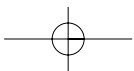
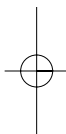
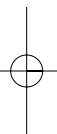
- Systemic upset: swinging pyrexia/tachycardia/rigors/sweats.
- Facial or peri-orbital pain.
- Venous obstruction—eyelid oedema/dilated facial veins.
- ‘Pulsating exophthalmus’ a transmitted carotid pulse with periorbital oedema.
- Blindness with papilloedema and retinal haemorrhages.
- Ophthalmoplegia: classically abducens (VIth) first followed by IIIrd and IVth.
- Obvious site of infection: usually unilateral initially; most commonly a peri-orbital cellulitis.
- Central signs—developing evidence of meningeal irritation.
- Bilateral signs develop with contra lateral extension of thrombus.

Investigations

- CT or MRI scans: usually show brain swelling and possible local infection. An occluded sinus might be visible on MRI scans.
- Coagulation studies.
- Cerebral angiogram, with venous phase (if diagnosis uncertain).
- Investigations into the cause of the infection.

Management

- Antibiotics and drainage of any collection of pus.
- Neurosurgical opinion.
- Anticoagulation.
- Thrombolytic therapy might be given if the patient is deteriorating.



! Saggital sinus thrombosis

Saggital sinus thrombosis can affect any age and either sex, but most commonly affects young and middle-aged females. It can be caused by trauma with depressed fractures overlying the sinus, tumours invading the sinus, and post-neurosurgery.

Clinical features

- Headaches, especially of a morning.
- Visual disturbance.
- Papilloedema.

Investigations

CT or MRI scans might show brain swelling. The 'delta' sign is a triangular filling defect in the sinus on a contrasted CT scan. An occluded sinus is usually visible on MRI scans. Infarction or haemorrhage due to venous hypertension might also be visible.

Management

- Anticoagulation.
- Thrombolytic therapy might be given if the patient is deteriorating.
- CSF diversion: might be necessary later if benign intra-cranial hypertension results.