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CORE TRAINING FOR DOCTORS

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Diagnosis and management of headache

Introduction

Headache makes up 2% of emergency department presentations (Goldstein et al, 2006). While 98% of these are not associated with significant underlying pathology, the 2% that are can have significant morbidity and even mortality if missed (Goldstein et al, 2006). When the headache is the symptom of an underlying intracranial or systemic pathology it is classified as secondary headache. When there is no associated underlying abnormality it is known as a primary headache. Identification and treatment of the minority of patients with sinister secondary headache is imperative. However, diagnosis of primary headache disorders is also important, as while not life threatening, they may carry significant morbidity.

Primary headaches

According to the International Headache Society classification (Headache Classification Committee of the International Headache Society, 2013) primary headaches are divided into 'migraine', 'tension-type headache', 'cluster headache and other trigeminal autonomic cephalgias' and 'other primary headaches' (*Table 1*). Each group is then further subdivided on a hierarchical basis so that both generalists and specialists can use the classification system to differing levels of accuracy (http://ihs-classification.org/en/). Treatment of primary headache disorders involves symptomatic treatment as well as prophylactic treatments and varies according to the underlying condition (*Table 1*).

Migraine

Migraine is a significant problem: one in ten people have migraine (Davenport, 2008) and migraine is now ranked nineteenth by the World Health Organization among all diseases worldwide causing disability (Headache Classification Committee of the International Headache Society, 2013). Migraine is subdivided into migraine with and without aura.

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Migraine with aura

An aura is 'reversible focal neurological symptoms that develop gradually over at least 5 minutes and last for less than 60 minutes' (National Institute for Health and Care Excellence, 2012; Headache Classification Committee of the International Headache Society, 2013). Visual symptoms such as fortification spectra and scotoma are most common, but focal numbness, weakness, aphasia and dysarthria may also occur, as may vertigo, imbalance and presyncope or syncope (Gelb, 2011). This can make differentiation from stroke complex, especially on first presentation.

To complicate the matter further, patients known to have migraine with aura are at increased risk of stroke. Thus a difference from normal aura, a lack of recovery within the normal time frame or new negative symptoms should be taken seriously. Moreover, given the increased relative risk of thromboembolism in migraine with aura, women should be advised against use of the combined oral contraceptive pill and both men and women advised to stop smoking (Gelb, 2011).

Migraine without aura

This is the commonest subtype of migraine (Headache Classification Committee of the International Headache Society, 2013). Although classically unilateral, migraines are often bilateral, can occur in any part of the head or neck and can be associated with scalp or face tenderness. Similarly, the pulsating quality of the pain is important but not necessary for diagnosis. Photophobia and phonophobia are typical features, the headache is usually worse with movement and often associated with nausea.

Medication overuse headache

No matter what the headache type, a clear medication history is important: patients can develop a secondary medication over-

use headache in addition to their primary headache disorder. Ask about analgesic days or number of packets of analgesia bought per week (Davenport, 2008) and make sure medication is being taken appropriately, for example, triptans need to be taken at the start of the headache, and not the aura, to be effective (Davenport, 2008). Any form of analgesia on more than 3 days per week for more than 2 months puts patients at risk of medication-overuse headache. Until patients withdraw from excess analgesia, the chronic headache will usually not resolve and often worsens with the patient taking ever-increasing amounts of analgesia.

Trigeminal autonomic cephalalgias

Cluster headache

Cluster headache is the most common of the trigeminal autonomic cephalalgias. It is six times more common in men (Gelb, 2011). In contrast to migraine, those suffering from cluster headache are typically agitated and will pace about. The headache is characterized by clusters of severe unilateral pain that usually occur over a period of 4–8 weeks with subsequent spontaneous resolution and recurrence. Individual episodes can last up to 180 minutes but are usually between 30 and 60 minutes long. Headaches can occur at any time of the day or night, frequently occur at the same time and often wake the patient from sleep (Gelb, 2011).

Table 1. Features that aid diagnosis of primary headache syndromes

	Acute migraine	Tension-type headache	Cluster headache	Trigeminal neuralgia	Paroxysmal hemicrania
Age	Usually >12	Usually >12	Usually 20–50	Usually >50	Usually 20–50
Sex	F>M	F>M	M>F	F=M	F=M
Aura	Y	N	N	N	N
Photophobia	Y	May occur	N	N	N
Phonophobia	Y	May occur	N	N	N
Autonomic features	May have ptosis or watering	N	Y – ipsilateral to headache	N	Y – ipsilateral to headache
Pain quality	Throbbing or pulsating	Pressing or tightening	Severe	Electric-shock-like precipitated by shaving, eating, applying make-up	Severe
Pain location	Unilateral or bilateral	Usually bilateral	Unilateral	Unilateral*	Unilateral
Pain duration	4–72 hours	30 minutes – continuous	< 180 mins	5–30 seconds	2–30 minutes
Behaviour in attack	Sleep, rest	Able to continue activities of daily living	Agitated and pacing	Agitated	Agitated
Wake from sleep	May do	N	Y	N	N
Frequency	<15 days per month (if >15 days per month for 3 months then 'chronic migraine')	<15 days per month (if >15 days per month for 3 months then 'chronic tension headache')	1 every other day to 8 per day with remission >1 month (if <1 month remission then 'chronic cluster')	100s of episodes per day. Remission may occur and last months or years	>5 per day (may become chronic)
Acute treatment	900 mg aspirin orally or diclofenac rectally or paracetamol if non-steroidal anti-inflammatory drugs not tolerated. Antiemetic orally or intravenous. Triptan at onset of headache if no contraindication	Judicious use of analgesia <3 times/week	Subcutaneous or nasal triptan. High flow oxygen (organize domicillary oxygen if necessary)	Carbamazepine as first line. If medical therapy fails surgery may be option	Indomethacin >150 mg orally or rectally once daily
Preventative treatment	Avoid precipitants, consider stopping contraceptive pill and treat underlying mood and anxiety disturbances. Regular propranolol or topiramate (need to counsel for risk of fetal malformations and reduced efficacy of hormonal contraceptive with topiramate). If above ineffective consider acupuncture or gabapentin. Review the need for continuing prophylaxis after 6 months	Pain management programme, treat underlying mood and anxiety disturbances. Consider low dose tricyclics	Verapamil	As above	Daily indomethacin. If not tolerated consider lamotrigine or topiramate

*A small percentage of trigeminal neuralgia patients may have bilateral pain in which case a central cause should be excluded. Table adapted from Davenport (2002), Scottish Intercollegiate Guidelines Network (2008), Gelb (2011), Headache Classification Committee of the International Headache Society (2013) and National Institute for Health and Care Excellence (2013)

Other types of trigeminal autonomic cephalalgias

Paroxysmal hemicrania and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing are rarer types of trigeminal autonomic cephalalgias. Paroxysmal hemicrania (and hemicrania continua, its chronic counterpart) is exceptionally responsive to indometacin, so should be considered in any patient presenting with recurrent episodes of short lived unilateral headache.

Tension-type headache

There is some controversy over the existence of tension-type headache with many neurologists believing it a mild variant of migraine (Gelb, 2011; Headache Classification Committee of the International Headache Society, 2013). It may be associated with tenderness on palpation of frontal, temporal, masseter, pterygoid, sternocleidomastoid, splenius and trapezius muscles. Tension-type headache is often seen in patients with chronic daily headache who also have exacerbations of migrainous headache.

In contrast to migrainous headache and the trigeminal autonomic cephalalgias, tension-type headaches usually do not respond well to medication but may respond to improvements in sleep, mood and anxieties (Headache Classification Committee of the International Headache Society, 2013). Provision of a positive diagnosis alongside reassurance regarding the good prognosis and the absence of more sinister pathology may be therapeutic in itself.

Trigeminal neuralgia

Trigeminal neuralgia is not considered a primary headache syndrome. However, it may form part of the differential for those presenting with head, neck or facial pain as it is characterized by intense paroxysms of facial pain that are described as electric-shock-like. It can occur in patients with multiple sclerosis as a secondary process as well as in many other disorders affecting the pons. It is usually idiopathic although an ectatic artery in contact with the trigeminal nerve may be found. Magnetic resonance imaging may be indicated to exclude a structural cause of trigeminal neuralgia (Gelb, 2011).

Secondary headaches

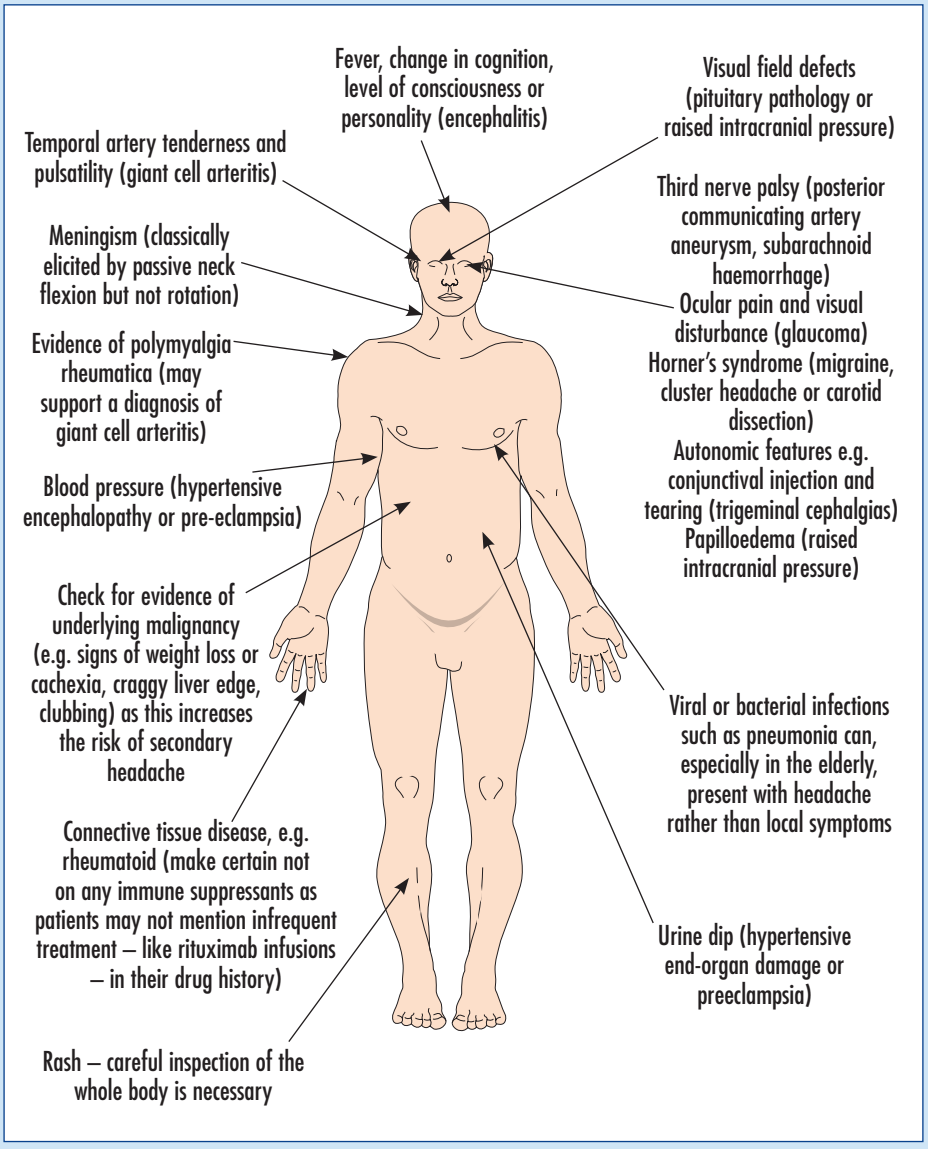
Secondary headaches can be the result of a wide range of pathology from relatively benign viral infection to life-threatening intracranial haemorrhage. Clinical features on history and examination may be associated with an increased risk of secondary headache (*Figure 1*). Management of secondary headache involves rapid identification and treatment of the underlying cause followed by appropriate symptomatic treatment.

Subarachnoid haemorrhage

Subarachnoid haemorrhage is an emergency: 50% of patients die following subarachnoid haemorrhage, often before reaching hospital, and 50% of survivors are left disabled (Headache Classification Committee of the International Headache Society, 2013). Subarachnoid haemorrhage commonly presents with thunderclap headache. This is defined as a headache reaching 'peak pain intensity within 1 minute' (Ducros and Bousser, 2013). However, not all patients with subarachnoid haemorrhage will meet this strict definition of thunderclap headache. In Bo et al's (2008) study only 70% of patients with subarachnoid haemorrhage reached peak pain intensity within 1 minute and a small minority (three of 71) took 30 minutes to reach the maximum pain intensity.

Other features associated with subarachnoid haemorrhage are transient loss of consciousness, vomiting, neck stiffness and onset with exertion such as exercise or

Figure 1. Features to look for on examination that may aid diagnosis.



sexual intercourse (Perry et al, 2010). However, these are not specific and may also occur with migraine. It is recurrence that is relatively reassuring: the patient who presents with his/her fourth thunderclap headache at the point of orgasm is far less likely to be having subarachnoid haemorrhage than if there is no such previous history.

Therefore, first presentation of thunderclap headache should be considered a subarachnoid haemorrhage until proven otherwise. All patients with new thunderclap headache should have neuroimaging performed. If a computed tomography scan is normal and a subarachnoid haemorrhage suspected then lumbar puncture should be done 12 hours after headache onset. This is sufficient time for red blood cells to be degraded to bilirubin. This degradation causes xanthochromia – the yellow discolouration of CSF – and allows a subarachnoid haemorrhage to be distinguished from a traumatic lumbar puncture.

A pyrexia may occur in subarachnoid haemorrhage and conversely bacterial meningitis may present with thunderclap headache (Dodick, 2002; Zarkou et al, 2010; Ducros and Bousser, 2013). Therefore, in patients with new headache and temperature, CSF should also be sent for culture, viral polymerase chain reaction, protein, paired serum and CSF glucose. It is often prudent to send an additional sample for storage as certain analyses may later be indicated as the clinical picture evolves.

Whether or not a normal computed tomography and CSF can fully exclude subarachnoid haemorrhage is controversial (Dodick, 2002). What is clear, as mentioned above, is that subarachnoid haemorrhage is not the only cause of thunderclap headache – indeed only 10–25% of patients presenting with thunderclap headache turn out to have subarachnoid haemorrhage (Ducros and Bousser, 2013). CSF opening pressure can be particularly helpful in differentiating non-subarachnoid haemorrhage causes of thunderclap headache (as discussed below) and should always be measured. Regardless of investigation results, if clinical suspicion of sinister pathology is high then further investigation or referral is indicated.

Management of subarachnoid haemorrhage can be radiological, neurosurgical or conservative. Patients are usually admitted under the care of the neurosurgeons. If a diagnosis of subarachnoid haemorrhage is considered then antithrombotic analgesia such as aspirin should be avoided.

Cerebral venous sinus thrombosis

Up to 10% of patients with cerebral venous sinus thrombosis present with an isolated thunderclap headache (Dodick, 2002; Zarkou et al, 2010). As mentioned above, computed tomography and CSF analysis can be normal. As CSF pressure is raised in 30–40% of patients with cerebral venous sinus thrombosis – an elevated CSF opening pressure on lumbar puncture may be the only pathological result on initial investigation. Additional features that can be associated with venous sinus thrombosis are focal neurological deficit, reduced consciousness, seizures and non-thunderclap headache (Zarkou et al, 2010). Risk factors for venous sinus thrombosis include a hypercoagulable disorder, dehydration and local infection. If there is clinical suspicion of venous sinus thrombosis, computed tomography or magnetic resonance venography should be performed. Treatment is with anticoagulation.

Arterial dissection

Carotid or vertebral artery dissection classically presents with unilateral head, neck or facial pain (Davenport, 2002). However, 13% of patients with carotid or vertebral artery dissection present with thunderclap headache (Dodick, 2002) and computed tomography and CSF analysis may be normal. Whiplash, trips to the hairdresser and osteopathic treatment are frequently cited as causative, but often there is no clear precipitant. Dedicated angiographic imaging is usually required to make the diagnosis and treatment may be conservative, radiological or neurosurgical.

Postural headaches: idiopathic intracranial hypertension and spontaneous intracranial hypotension

A headache that is worse on lying flat and improves with standing may indicate raised intracranial pressure. Idiopathic intracranial hypertension is a condition that classically occurs in obese young women who present with headaches secondary to raised intracranial pressure. The term 'benign intracranial hypertension' is no longer used because the condition may result in permanent visual loss if left untreated. It is important to enquire about visual obscurations, which are transient losses of vision associated with raised intracranial pressure. Papilloedema associated with an enlarged blind spot and peripheral visual field defects need to be excluded on examination. Secondary causes of raised intracranial pressure such as cerebral venous sinus thrombosis should also be excluded before a diagnosis of idiopathic intracranial hypertension is given.

A headache that is better on lying flat and worse on standing may indicate low intracranial pressure. This can occur spontaneously and again may present with a thunderclap headache (Dodick, 2002; Zarkou et al, 2010; Ducros and Bousser, 2013). Computed tomography and CSF analysis may again be normal. Low CSF pressure on lumbar puncture and characteristic changes on magnetic resonance imaging support the diagnosis.

Infective causes

When headache is associated with a fever, signs and symptoms of meningism (photophobia and neck stiffness on passive flexion but not rotation) or encephalitis (change in consciousness, cognition or personality) should be carefully sought on history and examination (*Figure 1*). If bacterial meningitis is suspected, blood cultures should be taken and antibiotic therapy initiated immediately. Antiviral treatment should also be administered if encephalitis is suspected. A full systemic examination should be performed looking for petechial rash from meningococcocal sepsis as well as alternative sources of infection.

Giant cell arteritis

Age over 50 years is an independent risk factor for secondary headache (Goldstein et al, 2006; Locker et al, 2006) and giant cell arteritis is a condition that is found over the age of 50 years. Patients may present with scalp tenderness, headache and/or general malaise. An erythrocyte

sedimentation rate and C-reactive protein level should be checked, a temporal artery biopsy performed and high-dose steroid treatment commenced as failure to diagnose or provide prompt treatment may result in irreversible visual loss.

Other causes of secondary headache

Although not within the scope of this article, it is important to remember that headache can originate from extra-cranial sources such as dental, ocular, ear, nose and throat. Glaucoma is an important example that warrants urgent ophthalmological opinion.

Special situations: the pregnant woman

Women of childbearing age are a key demographic in the headache population. When compared with the non-headache population presenting to emergency departments in the USA, patients presenting with headache were disproportionately female (71% *vs* 52%, $P<0.001$) and aged between 18 and 49 years (71% *vs* 47%, $P<0.001$) (Goldstein et al, 2006). The frequency of attacks of primary headache often reduces during pregnancy (Maggioni et al, 1997). However, a new headache presentation or new aura associated with known migraine should be assessed carefully to exclude a secondary cause (Macgregor, 2014).

Pathologies that are particularly important to consider in the pregnant patient include cerebral venous sinus thrombosis and pre-eclampsia. It is therefore critical that pregnant women presenting with headache have their blood pressure checked and urine dipped to assess for proteinuria. In the first 6 weeks post-partum women are also at increased risk of stroke and, particularly in the first week post-partum, of reversible cerebral vasoconstriction syndrome (Macgregor, 2014) which may present like stroke.

Examination

Physical examination is important for reassurance for the vast majority of patients who have benign headache (*Figure 1*). However, it also serves to identify uncommon features that might suggest a secondary headache. A full neurological exam that includes fundoscopy, level of consciousness and neck stiffness should be performed for all patients to exclude focal neurological deficits, signs of meningism or encephalitis. All patients should also have temperature and blood pressure checked.

Conclusions

Headache is a common problem with significant social morbidity. Obtaining a good history is critical in determining the underlying aetiology and making a positive diagnosis. This is important as appropriate treatment is critical for both primary and secondary headache disorders to reduce long-term morbidity, avoid mortality and improve patient outcome. BJHM

Conflict of interest: none.

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KEY POINTS

- About 98% of acute headache presentations are benign, but the 2% that are not can have potentially disastrous complications if the diagnosis is missed.
- History is the key to accurate diagnosis.
- Age >50 years, immunosuppression and known cancer diagnoses increase the likelihood of secondary headache.
- Subarachnoid haemorrhage is not the only cause of thunderclap headache and further investigation or referral is warranted in spite of normal computed tomography and lumbar puncture if clinical suspicion is high.
- Accurate diagnosis of primary headache syndromes is important to minimize morbidity and institute appropriate ongoing management.