12 The Medical History: The Key to Correct Headache Diagnosis

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Abstract: Effective management of headache depends on correct diagnosis. The presence of warning symptoms should necessitate further investigation to rule out an underlying cause. Once pathology has been excluded, targeted questions and pattern recognition of typical presentations of headache enable more confident diagnosis. With primary headaches being more common in practice than secondary headaches, correct diagnosis is dependent on a good history. A good history is also important to prevent misdiagnosis when there are coexisting headache types.

Introduction

Headache accounts for around 4.4% of consultations in primary care and is the most common cause of referral to neurologists (Bone and Fuller 2002; Latinovic et al. 2006). And since different headaches have different treatments, it is important to arrive at the right diagnosis. (Lipton et al. 1998). History is crucial to effective diagnosis of primary headaches as the examination is essentially normal. Failure to recognize and manage coexisting headaches is a common cause of treatment failure. In a study of patients with a diagnosis of migraine who were referred to a specialist headache clinic, nearly one-third had at least two headache diagnoses (Blau and MacGregor 1995).

Taking a History

The first task is to exclude a condition requiring more urgent intervention by eliciting any warning features in the history (*Table 12.1*). This can enable identification of those who need further investigation to reach a diagnosis and treat specifically.

Changes in a patient with a long-standing history of primary headache are an alert to question further about the development of new, or unusual, symptoms. Age of the patient is important as the likelihood of pathology is greater with elderly patients. Seizure is a cardinal symptom of intracerebral space–occupying lesions; "thunderclap" headache should raise the suspicion of subarachnoid hemorrhage, carotid dissection, cerebral venous sinus thrombosis, and reversible cerebral vasoconstriction syndrome.

Once serious pathology has been excluded, it is appropriate to take a more detailed history. In an ideal world, patients should be allowed to tell their own story. Time constraints rarely make this feasible, and it is necessary to ask a few pertinent questions to structure the consultation. Unless the patient is particularly verbose or several headaches coexist, the diagnosis can usually be made in a few minutes.

Typical responses to a structured history are useful in diagnosing headaches (*Table 12.2*).

"*How many different headaches do you have?*" This can be a useful opening question, particularly for patients with long-standing primary headaches who develop a more insidious secondary headache (Laughey et al. 1993). Most patients can readily distinguish between different headaches as each type usually follows a typical pattern of onset, timing, and symptoms. If more than one pattern of headache is suspected, it is necessary to take a separate history for each.

"*Why are you consulting now?*" This may be because of the severity or frequency of headache, but it can also reveal patients' fears, or external pressure on them to "do something about their headaches" from family or work.

Table 12.1
Identifying secondary causes of headache: SNOOP4 (Dodick 2010)

	Ask about	Possible causes
*S*ystematic symptoms/signs	Fever	Giant cell arteritis
	Chills	Infection
	Night sweats	Malignancy
	Myalgias	
	Weight loss	
*S*ystematic disease	History of malignancy	Metastatic disease
	Immunocompromised state	Opportunistic CNS infection
	HIV	
*N*eurologic symptoms or signs	Focal or global neurological symptoms or signs, including behavioral or personality changes	Neoplasia
		Infection
	Diplopia, transient visual obscurations, pulsatile tinnitus (especially in obese patients)	Inflammation
		Vascular CNS disease
		Idiopathic intracranial hypertension
*O*nset sudden (thunderclap headache)	How quickly the pain went from 0/10 to 10/10	Vascular crises (stroke, subarachnoid hemorrhage, cerebral venous sinus thrombosis, reversible cerebral vasoconstriction syndrome, arterial dissection)
*O*nset after age 50 years		Neoplasia
		Infection
		Inflammation
		Giant cell arteritis
*P*attern change (if previous history)	*P*rogressive headache with loss of headache-free periods	
	*P*recipitated by Valsava	Chiari malformation
		Structural lesions which obstruct CSF flow
		CSF leak
	*P*ostural aggravation	Worse when standing or lying: intracranial hypotension from CSF leak; intracranial hypertension
		Worse with certain neck movements; cervicogenic headache
	*P*apilledema	Intracranial hypertension

"*When did the headache first start?*" Recent new-onset headaches are of greater concern than long-standing headaches. Headache as an isolated symptom for more than 10 weeks is rarely due to brain tumor (Vazquez-Barquero et al. 1994). Around half of migraineurs experience their first attack before age 25 and three-quarter before age 35 years

Table 12.2
An approach to the headache history (MacGregor et al. 2010)

1. How many different headaches types does the patient experience?	
Separate histories are necessary for each. It is reasonable to concentrate on the most bothersome to the patient but others should always attract some enquiry in case they are clinically important.	
2. Time questions	(a) Why consulting now?
	(b) How recent in onset?
	(c) How frequent, and what temporal pattern (especially distinguishing between episodic and daily or unremitting)?
	(d) How long lasting?
3. Character questions	(a) Intensity of pain?
	(b) Nature and quality of pain?
	(c) Site and spread of pain?
	(d) Associated symptoms?
4. Cause questions	(a) Predisposing and/or trigger factors?
	(b) Aggravating and/or relieving factors?
	(c) Family history of similar headache?
5. Response to headache questions	(a) What does the patient do during the headache?
	(b) How much is activity (function) limited or prevented?
	(c) What medication has been and is used, and in what manner?
6. State of health between attacks	(a) Completely well, or residual or persisting symptoms?
	(b) Concerns, anxieties, fears about recurrent attacks, and/or their cause?

(Stewart et al. 2008). Frequency of attacks fluctuates during a lifetime, and it is not uncommon for migraine to return after several years of respite (Bille 1997). Cluster headache is frequently misdiagnosed in primary care, with an average of 2–3 years until correct diagnosis (Bahra and Goadsby 2004; van Vliet et al. 2003).

"*How many days in a month do you NOT have a headache of any type for the entire day? How many headache days per month are severe/moderate/mild?*" Frequent headaches need detailed analysis of the pattern. Cluster headaches follow a stereotypical pattern of daily attacks over several weeks with periods of remission in-between. Daily, progressive headaches are a cause for concern. Daily headaches are not typically migraine but "chronic migraine" may manifest as near-daily headaches. Patients reporting daily should be asked how the pattern has changed over time. Although an underlying pathology needs exclusion, medication overuse is a more common contributory factor.

"*Is there any pattern to attacks?*" Patients may notice attacks occur more often at weekends or are linked to menstruation, which then needs to be diagnosed and managed accordingly. Cluster headache often wakes the patient a few hours after sleep. Headache associated with medication overuse and raised intracranial pressure are characteristically worse on waking.

"*How long does the headache typically last if you don't take treatment, or if treatment is not effective?*" Most migraines in adults will last part of a day, up to 3 days. Attacks are typically shorter in children – sometimes less than a couple of hours (Headache Classification

Subcommittee of the International Headache Society (IHS) 2004). Cluster attacks last between 20 and 180 min (usually 60 min); medication-overuse headache is mostly continuous. Tension-type headaches can vary from a few hours to daily symptoms.

"*How severe is the pain?*"; "*What does the pain feel like?*" The headache of migraine is usually described as a moderate-to-severe throbbing, pounding headache. The pain of cluster headache is extremely severe and prohibits any activity during the attack period.

"*Where do you get the pain?*" Patients will usually point to one side of the head, which may alternate between or during attacks. Although typically unilateral, bilateral headache does not discount migraine, occurring in about 30% of attacks. The headache can swap sides between and during attacks. Neck pain is also a common symptom before and during migraine, sometimes radiating to the shoulder. A more generalized and unremitting "pressure" headache is more typically associated with tension-type headache and medication-overuse headache. Cluster headache is strictly unilateral, centered on one eye, and while very uncommon, may shift sides.

"*What other symptoms have you experienced?*" The presence of associated symptoms can help secure a diagnosis. A positive response to the presence of the following symptoms can "PIN" the diagnosis of migraine (Dodick 2010):

- Photophobia: Does light bother you when you have a headache?
- Impairment: Do you experience headaches that impair your ability to function?
- Nausea: Do you feel nauseated or sick to your stomach when you experience a headache?

A positive answer to two or three of these three questions results in a 93% and 98%, respectively, positive predictive value for a diagnosis of migraine (Lipton et al. 2003).

Migraine may be preceded by premonitory symptoms, which occur hours to days before onset of headache and include symptoms such as unusual tiredness, difficulty concentrating, neck stiffness, yawning, and food cravings (Giffin et al. 2003). These generalized symptoms are often confused with the more specific symptoms of aura. Visual aura symptoms are usually symmetrical, affecting one hemifield of both eyes, although subjectively they may appear to affect only one eye – if there is any doubt, patients should be asked to assess their next attack. A migrainous scotoma is typically positive (bright), starting as a small spot gradually increasing in size to assume the shape of a letter C, developing scintillating edges that appear as zigzags (fortifications – a term coined in the late eighteenth century because the visual disturbances resembled a fortified town surrounded by bastions). The aura usually starts at or near the center of fixation, gradually spreading laterally, increasing in size over a period of 5–30 min. In contrast, ischemic events do not generally have the scintillating and spreading features of the visual aura of migraine and the visual loss is usually a monocular negative scotoma (black). Transient monocular blindness is not typical of migraine and prompts urgent investigation. Sensory aura symptoms are commonly positive, that is, a sensation of pins and needles rather than numbness. In an ischemic episode, a sense of numbness or "deadness" is described. Migraine symptoms have a characteristic unilateral distribution affecting one arm, often spreading over several minutes proximally from the hand to affect the mouth and tongue – "cheiro-oral distribution." This spread to involve the tongue is typical with migraine aura and is rarely seen in cerebrovascular ischemic episodes. Even when sensory symptoms occur, the majority of auras include visual symptoms. Hence, a useful screening question may be: "*Do you have visual disturbances that last up to one hour and resolve before or with the onset of headache?*"

Tension-type headache is often described as a "featureless" headache.

Associated symptoms of cluster headache and other trigeminal autonomic cephalalgias are most prominent on the side ipsilateral to the pain, and include lacrimation, conjunctival injection, nasal congestion, rhinorrhea, Horner's syndrome, facial swelling, erythema, or pallor.

"*Have you found any triggers for attacks?*" Most patients with migraine can list at least a couple of triggers spontaneously and will identify several more if prompted with a list. Within a cluster, alcohol, heat, high altitude, and sleep are common triggers.

"*What makes the headache better?*"; "*What makes the headache worse?*" This can elicit a response such as "lying still helps, while movement makes the headache worse" in the case of migraine. For cluster, patients rarely stay still, often crying and pacing restlessly during the pain (Blau 1993).

"*Who else in your family has similar headaches?*" Although a family history can confirm the diagnosis, absence of a family history does not prohibit migraine. Do not assume that the headache is the same as the patients – many family members of cluster headache patients have migraine.

"*What do you do when you have a headache?*" Patients should be encouraged to describe medication taken, as well as what they physically do – go to bed, lie still, sleep, etc. Those who continue working should be questioned on how well they function.

"*What do the headaches stop you from doing?*" Headaches can cause significant disability with time lost from work, household duties, and leisure, particularly if attacks are severe and/or frequent. This has important implications for treatment as disabling attacks require more aggressive therapy.

"*What medication do you treat the headaches with and what have you tried in the past? How many days in a month do you NOT take a medication of any type (prescription or OTC) for headache?*" Many treatments that appear to fail might succeed if taken in adequate doses, sufficiently early in attacks. Establish what has failed in the past, and why, before recommending alternatives. Patients with frequent headaches should be carefully questioned about frequency with which they take acute medications to exclude medication overuse.

"*How do you feel between attacks?*" The response for episodic headache is usually "*Fine.*" Patients with continued symptoms between attacks may have more than one type of headache.

"*What worries you about your headaches?*" Many patients have fears that they find hard to express. A simple question can prevent the patient leaving the consulting room still harboring fear of a brain tumor that they have been frightened to ask about. Isolated headache is rarely caused by an intracranial lesion, and in primary care, the risk of a brain tumor with headache presentation is only 0.09% (Hamilton and Kernick 2007). Directing this question has the advantage of enabling the doctor to know what the patient's ideas are so that they can be reinforced or refuted, and developed into management plans. This can help forge a better bond between doctor and patient. Patients often do not give an immediate answer to the question, but wait until they are being examined. This may be because they feel less vulnerable during a nonthreatening examination. Eye contact is lost and the break in tension may permit the patient to release information or ask questions that are important.

General Questions

Most of the following will already be known to the physician but confirm that there have been no recent changes to the patients' general health.

Systemic Review

Most patients presenting purely with migraine are otherwise fit and well. Symptoms suggestive of systemic disease require more detailed questioning. In particular, symptoms such as fever, night sweats, chills, and weight loss should be elicited.

Past Medical History

There is rarely a relevant medical history for migraine although some patients may time the onset subsequent to a head injury, illness, or emotional upset. In most cases, it is impossible to know if the event was truly the initiator of migraine. Travel sickness and recurrent abdominal pain in childhood have been linked to the development of migraine in later life, but this association is not diagnostic. Comorbid conditions should be considered, particularly depression, which may require specific management. Work difficulties, marital problems, alcoholism, etc., need consideration. Medical conditions relevant to therapy should be considered, for example, peptic ulcer or uncontrolled hypertension would contraindicate NSAIDs or triptans, respectively.

Medications

Headache is listed as a side effect of almost every available drug. However, some drugs have been particularly associated with increased headache. These include the combined oral contraceptive pill, although menstrual migraine sometimes resolves with continued use. Occasionally, increased frequency and severity of headache or migraine may necessitate adjustments to treatment or even withdrawal. Specific drugs, including dipyridamole, trazadone, nitroglycerin, among others, may worsen migraine. A careful drug history is necessary to ensure that there is no incompatibility between drugs used for migraine and those taken for other indications. Frequent use of acute headache treatments can lead to medication-overuse headache.

Social History

Alcohol sometimes triggers migraine and can worsen cluster headaches in the active phase. Several occupations can increase the likelihood of migraine. Stressful jobs create obvious triggers that can be specifically identified. Working at a computer screen for several hours can result in tension headache. Shift work can disrupt sleep and dietary routines. Unemployment and redundancy carry the risk of depression. Personal or family problems may be relevant.

The impact of headache on the patient's life should be discussed. It is not uncommon for patients to fear making arrangements in case they are disrupted by a migraine. This cycle of fear can be broken with effective management but may require additional psychological treatment.

Family History

A family history may be present but is not necessary to confirm the diagnosis. A family history of arterial disease may be relevant if vasoconstrictor drugs are considered.

Examination

The main purpose of the examination is to reassure patients. Patients and their family are often worried that there is a serious cause for the headaches such as brain tumor or stroke. Patients expect a physical examination and may be less likely to agree with the doctor's perspective and subsequent management recommendations if this is not done. The examination can be brief, but should be thorough. In patients seen in a specialist clinic, fewer than 1% have headaches secondary to intracranial disease, and all have signs of it. The mental state will have been assessed while taking the history. Pulse, blood pressure, and auscultation for cardiac abnormalities and bruits should be checked first and are particularly important if vasoconstrictor drugs such as ergotamine or the triptans are considered. Examining the jaw can identify temporomandibular joint dysfunction that can give rise to headache. Examination of the neck and cervical spine may reveal muscle contraction, cervical spondylosis, or even meningism.

While Taking the History

Speech, mood, and memory can be assessed by the patient's response to questions.

At the end of the history, request permission from the patient before the examination.

A Rapid Neurological Examination

It is unnecessary to check every aspect of neurological function, and a routine screen should take no more than 5 min (*Table 12.3*). Particular attention should be paid to examination of the cranial nerves, tendon reflexes, and optic disks. If the history suggests that there is a more sinister cause for the headache, a full neurological appraisal is necessary. It is of great comfort to patients when doctors explain that the findings are "normal." When time is short, a minimum examination should include blood pressure and examination of the optic fundi.

Investigations

In clinical practice, the initial concern is to differentiate primary headaches from sinister secondary headaches. Investigations do not contribute to the diagnosis of primary headaches and are not warranted in children or adults with a defined headache and normal neurological examination (Detsky et al. 2006; Weingarten et al. 1992). Investigations are indicated if secondary headache is suspected OR because of undefined headache, atypical symptoms, persistent neurological or psychopathological abnormalities, abnormal findings on neurologic examination, or recent trauma. A low threshold is indicated for new-onset headaches and if there is significant parental anxiety about a child with headache. Inappropriate investigations can increase morbidity, particularly in the presence of unrelated incidental findings and, with respect to computed tomography, unnecessary radiation exposure. Symptomatic brain abnormalities are identified in up to 14% of an asymptomatic population (Vernooij et al. 2007). Many patients request investigations for the reassurance that they do not have a brain tumor or other serious underlying pathology. This may be avoided if, on the basis of a sound history and examination, the doctor spends time with the patient directly discussing his or her concerns

Table 12.3

The neurological examination (MacGregor and Frith 2008)

While patient is standing	
Ask the patient to:	Tests:
Close your eyes and stand with your feet together (Romberg)	Midline cerebellar; dorsal column; proprioception
Open your eyes and walk heel to toe	Midline cerebellar; dorsal column; proprioception
Walk on your tip-toes	Power of dorsiflexion
Walk on your heels	Power of plantar flexion
Close your eyes and hold your hands out straight in front of you with your palms flat and facing upward	Hemisphere lesions (e.g., left hemisphere lesion, right hand will bend in and drift up)
	Neglect (e.g., left parietal lesion, right hand will drop down)
Keep your eyes closed. Touch your nose with the fingertip that I touch (person testing uses their own finger to touch a couple of the patient's fingertips in turn)	Light touch and finger-nose test (cerebellar or sensory ataxia and light-touch in fingertip)
Open your eyes and with your arms outstretched, pretend to play the piano	Fine finger movements
	Pyramidal and extrapyramidal function
Tap the back of one hand with your other hand. Change hands and repeat.	Ataxia
Screw your eyes up tight and then relax and open your eyes	Pupil dilation and constriction
	Horner's syndrome
	Lower motor neurone lesion
Bare your teeth/grin	Upper motor neurone facial weakness
Stick your tongue out and wiggle it	Bulbar and pseudobulbar palsy
Stare at my face at point at the fingers which move (person testing has arms out to the side with index finger pointing. Arms stop in an arc and index finger is wiggled on each side in turn or together)	Temporal field defects (important visual field defects always involve one or other temporal field)
	Inattention (parietal lobe lesion)
Keeping your head still, stare at my finger and follow my finger up and down with your eyes (person testing draws a wide "H" in the air)	Eye movements (cranial nerves III, IV, and VI)
	Nystagmus; saccadic (jerky) eye movements
While patient is lying down	
Examine:	Tests:
Limb reflexes	Upper motor neurone lesion (brisk)
	Peripheral nerve or nerve root lesion (absent)
Plantar response	Upper motor neurone lesion (Babinski/ extensor response)
Abdominal reflexes	Spinal cord disease

Table 12.3 (Continued)

While patient is lying down	
Funduscopy	Raised intracranial pressure (papilledema)
	Optic atrophy
Pulse and BP	Hypertension
If indicated, examine the chest, palpate breasts, and abdomen	Systemic disease, e.g., neoplasia

(Fitzpatrick 1996). Although it may be necessary for a few who will not be reassured in the absence of a "brain scan," any anxiolytic effects of a normal result may not be sustained beyond a few months (Howard et al. 2005).

Full blood count and erythrocyte sedimentation rate may detect the presence of infection, temporal arteritis, or malignancy.

Plain radiography of the skull is normal in most patients with headache but may be indicated if there is a history of head injury or if symptoms/examination are suggestive of a tumor, particularly of the pituitary gland. This has now been largely replaced by imaging studies.

Lumbar puncture confirms infection (meningitis or encephalitis). It should be used if subarachnoid hemorrhage (SAH) is suspected and CT is either unavailable or the results are inconclusive – CT may be normal in 10–15% of all subarachnoid hemorrhage, and its ability to detect SAH declines with time after the onset of symptoms. Lumbar puncture is also useful for the detection of elevated or low intracranial pressure.

Electroencephalography (*EEG*) is of little diagnostic value in headache but may be considered if a clinical diagnosis suggests features of epilepsy, such as loss of consciousness occurring in association with migraine.

Computerized tomography (*CT*) is of limited value for routine evaluation of headache as a number of secondary causes can easily be missed (*Table 12.4*). CT can demonstrate structural lesions including tumor, vascular malformations, hemorrhage, and hydrocephalus. If intracranial or subarachnoid hemorrhage is suspected, CT scan without contrast can detect recent bleeds – MRI may miss fresh blood. It may be necessary to give an intravenous injection of contrast material to highlight a suspected tumor or vascular lesion. Indications for CT are persistent focal neurological deficits, symptoms or signs suggestive of an arteriovenous malformation and hemorrhagic stroke.

Magnetic resonance imaging (*MRI*) produces better definition of soft tissue abnormalities than CT scanning. MRI with gadolinium is the investigation of choice for meningeal pathology. Although CT detects most tumors, MRI is superior to CT when imaging lesions in the region of the posterior fossa, axial tumors, the orbit and the paranasal sinuses, and demyelinating lesions.

Magnetic resonance or CT-angiography is indicated in patients with thunderclap headache when the lumbar puncture and unenhanced CT scan are unremarkable. Noninvasive angiography is reliable in detecting cervicocephalic arterial dissection, cerebral venous sinus thrombosis, dural arteriovenous fistula, reversible cerebral vasoconstriction syndrome, or intracranial aneurysm.

Cerebral angiography is rarely required as a primary investigation and its use is limited by its invasiveness. If CT or MRI confirms arteriovenous malformation, angiography is used to define the extent of the lesion and demonstrate feeding and draining vessels. Angiography is also still a gold standard for identifying the site, size, and morphology of intracranial aneurysms in the setting of subarachnoid hemorrhage.

Table 12.4
Secondary causes of headache that may be missed on computed tomography (Dodick 2010)

Vascular	Saccular aneurysms
	Subarachnoid hemorrhage
	Arteriovenous malformations (especially posterior fossa)
	Carotid or vertebral artery dissections
	Ischemic stroke
	Cerebral venous sinus thrombosis
	Vasculitis
	Reversible cerebral vasoconstriction syndrome
Neoplasia	Parenchymal and extra-axial neoplasms (especially in the posterior fossa)
	Meningeal carcinomatosis
	Pituitary tumor and hemorrhage
	Metastatic brain tumor
Cervicomedullary lesions	Chiari malformation
	Foramen magnum meningioma
	Acoustic schwanomma
Infections	Meningoencephalitis
	Cerebritis and brain abscess
Other	CSF leak (intracranial hypotension)
	Intracranial hypertension
	Idiopathic hypertrophic pachymeningitis

Isotope scanning and *doppler flow studies* are only of value for research in headache. Detection of carotid dissection may be possible with carotid Doppler studies when there is an index of suspicion and MR or CT-angiography is not available.

Conclusions: How to Get It Right

Making a diagnosis of headache based on the suggested approach may appear time consuming. However, primary care physicians often have the advantage of treating a patient for several years and so, for many of the questions, the answers will already be known. A brief but thorough neurological examination need not be time consuming, although blood pressure and fundoscopy are mandatory. The diagnosis should be reviewed at follow-up visits, particularly in cases of treatment failure. Be alert to coexisting headaches, which can confuse the picture.

Need to Know

- A new headache needs a new diagnosis.
- All patients presenting with sudden severe headache warrant further investigation.

- Increased frequency of headache should prompt suspicion of medication overuse.
- Unless directly questioned, patients may not reveal the true extent of medication use.
- Listen to symptoms that patients describe, not the diagnosis that they have been given.
- Pattern recognition from the history enables correct diagnosis.
- Different headaches need to be treated differently.

Case Histories

Case 1

PW is a 52-year-old white male. He presented to the emergency department 5 days after the abrupt onset of his "worse ever headache" associated with nausea and photophobia. At the time of review, his headache was less severe but he still felt unwell. He had a history of infrequent migraine without aura but described this recent headache as more severe and more prolonged than his usual attacks. Prescription drugs that usually helped his migraine had not been effective. He had no other relevant medical history and was not on any regular medication. Glasgow Coma Scale was 15, and neurological examination was unremarkable. A computed tomographic (CT) brain scan identified perimesencephalic subarachnoid hemorrhage (SAH), which resolved on serial CTs. Cerebral angiography showed no evidence of arteriovenous malformation or aneurysm. PW was discharged following full recovery.

Comment: Perimesenchephalic hemorrhage is a non-aneurysmal cause that affects around 10% of patients with SAH. The CT scan shows a characteristic pattern of bleeding confined to the midbrain cisterns. The risk of rebleeding is very low, and the long-term prognosis is good.

Case 2

AS is a 44-year-old Asian female. She visited her primary care physician because of weekly attacks of migraine that were not responding to her usual painkillers. The history revealed that her first attack of migraine when she was 12 but attacks were only once or twice a year. Since her 20s, she had experienced migraine once or twice a month. Over the last 4 years, the attacks had become more frequent and in the last 6 months, the attacks were occurring weekly. AS stated that she had a headache most days but could manage these; it was the weekly attacks that were troubling her. Direct questioning revealed that AS was taking painkillers most days, as this could get her through the day. AS was otherwise healthy and took no medication other than for her headaches. Physical and neurological assessments were unremarkable. The doctor considered that, given the frequency of medication use, the most likely diagnosis was medication-overuse headache. After appropriate management, AS stopped daily medication and reverted to a pattern of migraine once or twice a month. She felt well between attacks, which once again responded to their usual treatment.

Comment: Daily headache is associated with medication-overuse headache (MOH) in around 30% of the population and up to 60% of patients attending specialist clinics. It is most prevalent in those aged 40–50 years and affects three times more women than men. Any patient who has headache more than 10 days a month should be questioned about their use of medication. Specific questions should use of analgesics for reasons other than headache; use of over-the-counter as well as prescription drugs; acute medications becoming less effective;

and escalation to using more drugs. Assessment should also search for possible complications of regular drug intake (e.g., recurrent gastric ulcers, anemia).

Case 3

HM, a 35-year-old white male had been referred to the specialist because his migraine had returned and had failed to respond to treatment. The referral letter from the primary care physician stated that codeine-containing analgesia had controlled HM's pain and several standard migraine prophylactics had been prescribed without success. HM said that he had had migraine for the last 10 years. He usually only had them on-and-off for a couple of months but the present bout had started over 4 months ago and showed no signs of letting up. The history revealed attacks that woke HM a couple of hours after going to sleep, most nights. The pain was excruciating, centered on his right eye, which he felt was being pushed out of his skull. The eye looked red and watery, but the left eye was normal. The pain was so severe that he sat on the bed crying and rocking until the symptoms abated a couple of hours later. Normally a heavy social drinker, HM was avoiding alcohol as it almost instantly triggered an attack. No medication worked, and he felt he could not go on. Physical and neurological assessments were unremarkable. The specialist recognized the typical history of cluster headache and arranged appropriate management.

Comment: Cluster headache affects fewer than 1% of the population. It is frequently misdiagnosed as migraine and treated as such. Unlike migraine, it affects more men than women. The typical "clusters" of attacks, "clockwork" timing, severity of pain, and unilateral autonomic symptoms are distinctive symptoms that lead to the correct diagnosis.

Case 4

PN was a 41-year-old Hispanic male who presented with a 4-h history of a severe unremitting occipital headache that started abruptly while he was straining at stool. The headache was associated with nausea, photophobia, and recurrent vomiting. He preferred to lie still as movement worsened the pain. He had a history of episodic migraine with aura since the age of 14 with, usually, two or three attacks each year that were rarely troublesome. He described this new headache as very different from his migraine. There was no other relevant personal or family history. Examination was unremarkable, although PN was in obvious pain. There was no neck stiffness or focal neurological symptoms.

Complete blood count, serum chemistry, urinalysis, urine drug screen, and electrocardiogram revealed no abnormalities or remarkable findings. Unenhanced brain CT was normal. There was no evidence of subarachnoid hemorrhage, ischemic or intraparenchymal hemorrhagic stroke, or intracranial mass lesion. Lumbar puncture revealed an opening pressure of 12 cm water, CSF was clear and without red or white blood cells or xanthachromia, and serum-matched total protein and glucose were normal. Gram stain was negative.

The patient was discharged from the emergency department, only to return 2 days later with another abrupt onset, severe headache that occurred during sexual intercourse. Brain MRI, MR venography, and MR-angiography revealed multiple areas of vasoconstriction involving the anterior and posterior cerebral arteries. A diagnosis of reversible cerebral vasoconstriction syndrome (RCVS) was made. Nimodipine was initiated at a dose of 60 mg every 6 h for the first week, then 30 mg every 6 h for the next 3 weeks. MR-angiogram of the

head and neck was repeated 4 weeks after the first imaging study and demonstrated complete resolution of the cerebral vasoconstriction.

Comment: The differential diagnosis of sudden-onset severe headache (thunderclap headache: TCH) is important because of the morbidity and mortality associated with the conditions that can present with TCH. The diagnosis may be challenging when the headache occurs in isolation and in the absence of neurological symptoms or signs, thereby lowering the index of suspicion of a sinister secondary cause. Although subarachnoid hemorrhage and hemorrhagic and ischemic stroke are likely to be identified by brain CT and lumbar puncture, other causes such as cerebral venous sinus thrombosis, carotid or vertebral artery dissection, and RCVS require angiography. Hence, the clinical approach to the patient with TCH should be methodical, tailored to evaluate each of these causes in an appropriate and sequential fashion.

TCH associated with RCVS invariably occurs, and commonly recurs, within the first 7–10 days after the initial onset. However, all patients who present for the first time with TCH should be evaluated for RCVS, which is a much less benign condition than is suggested in the International Classification of Headache Disorders (ICHD-II), where it is listed as *6.7.3 Headache attributed to benign (or reversible) angiopathy of the central nervous system.* Rapid and accurate diagnosis is important since ischemic or hemorrhagic stroke occurs in up to one-third of patients in the ensuing weeks. It is important to note that up to 20% of patients eventually diagnosed with RCVS have normal initial CT- or MR-angiography. RCVS has been associated not only with serious disorders such as pheochromocytoma, severe hypertension, carcinoid, and porphyria but also with pregnancy, exposure to illicit (marijuana, cocaine) and pharmaceutical drugs (e.g., bromocriptine, SSRIs, intravenous immunoglobulin, and over-the-counter medications containing pseudoephedrine). In the absence of a demonstrable precipitating disease or drug, the headache is often triggered by a Valsalva maneuver. It is probably a commonly overlooked cause of TCH, as recent studies indicate that 40–60% of patients who present with TCH and a negative CT and LP have cerebral vasoconstriction on MR-angiography. Initial management includes control of blood pressure, hydration, analgesia, and avoidance of drugs with vasoconstrictor activity (e.g., triptans, ergots). Nimodipine is usually initiated at a dose of 30–60 mg every 6 h, to reverse the vasoconstriction and minimize the risk of stroke, and should be continued until reversal is complete and the patient has been without headache or other symptoms for at least 7 days. Reversal of vasoconstriction is demonstrated by repeated CT- or MR-angiography, and is usually complete by 2–4 weeks after the onset of headache but may take up to 2 months.

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