

Is this just a Headache?



By Werner J. Becker, MD, FRCPC

Headache is one of the most common reasons for a patient to see a doctor. While most patients have a benign headache type, the headache can be the initial presentation of a serious underlying illness. In fact, many patients harbour the fear they may have a brain tumour. It is important, therefore, that the physician understand the possible etiologies of headache, as well as the clinical features suggestive of a serious underlying cause for headache.

Headache diagnosis may also be difficult because some headache types are quite uncommon (*i.e.*, cluster headache), while others may appear superficially very similar to each other (*i.e.*, those conditions that result in chronic daily headache). These factors can also lead to uncertainty in headache diagnosis by the family physician.

In this article, the author will review headache classification, some of the features which suggest a serious underlying cause for headache, and the diagnostic features of some of the less common headache syndromes.

Classifying Headaches

It has been said that a good classification makes order out of chaos. This is especially true for headache, and if a physician is unable to classify a patient's headache disorder, this leads to uncertainty with regard to diagnosis and to treatment.

Case

A 25-year-old woman presents to your office complaining of headache. These headaches were one day in duration, occurred about once a month and were described as severe, throbbing, bilateral, and primarily occipital. There was associated nausea, phonophobia and, at times, vomiting. At this stage, her headaches were diagnosed as migraine. She had a normal neurologic examination, and her headaches met diagnostic criteria for migraine without aura. They were purely occipital, which is unusual, but can occur in a small percentage of patients with migraine. A variety of symptomatic and prophylactic migraine medications were eventually tried.

Eight months later, the patient had daily headaches, which were bilateral, occipital, and frontal. She also complained of problems with nausea, even with no headache. Her headaches would occasionally awaken her at night, were worse in the mornings and when standing up. She complained of ringing in her right ear. Her neurologic examination remained normal, including her optic fundi.

The International Headache Society (IHS) has worked hard to establish a comprehensive headache classification, which tries to group all headache types into 13 overarching categories.¹ Each of these categories has many sub-categories,

Headache

Table 1

Diagnostic Criteria For Migraine Without Aura

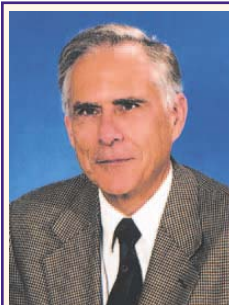
The patient must have had at least five attacks meeting the diagnostic criteria below:

- For headaches that are untreated or unsuccessfully treated, headache duration should be between four and 72 hours.
- The headache should have at least two of the following characteristics:
 - Unilateral location;
 - Pulsating quality;
 - Moderate or severe intensity; and
 - Aggravation by walking stairs or similar physical activity.
- During headache there should be at least one of the following:
 - Nausea and/or vomiting; and
 - Photophobia and phonophobia.
- History, physical and neurologic examinations do not suggest another cause for the patient's headache.

Modified from Headache Classification Committee of the International Headache Society: Classification and diagnostic criteria for headache disorders, cranial neuralgias, and facial pain. *Cephalalgia* 1988; 8(Suppl. 7):1-96.

and detailed discussion of many of these is beyond the scope of this article.

Migraine is the headache type that brings most patients to physicians, and it comes in many guises. Particularly problematic are patients who have



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migraine aura without headache, as consideration must be given to whether the patient has a transient ischemic attack or perhaps a focal seizure. Migraine with prolonged aura, where a migraine aura symptom goes beyond the usual limit of 60 minutes, also causes concern with regard to cerebrovascular disease.

All the headache types in the IHS headache classification have diagnostic criteria. It is unrealistic to expect the family practitioner to know all of these, but the diagnostic criteria for migraine without aura are shown in Table 1. These diagnostic criteria make the point that the diagnosis of many headache syndromes, including migraine, is basically a clinical diagnosis. As for many other benign headache types, however, the final diagnostic criterion for migraine is that no other cause must be apparent. For the most part, a clinical examination, including a careful neurologic examination, is sufficient to meet this diagnostic criterion. The point is that patients with headache deserve a careful clinical examination.

When Should I Suspect A Serious Underlying Cause?

This is a difficult area, where a lot of clinical judgment and experience comes into play. One can list six clinical features that should cause some concern about a possible serious underlying cause for the patient's headache. Again, none of these is diagnostic, and clinical discretion must be used as to how significant any one of these is in any particular patient.

Each of the clinical features shown in Table 2 requires clinical interpretation and judgment. For example, although most patients who develop migraine will have done so by the age of 30, some patients develop migraine later in life. Also, patients with migraine do have their ups and downs, so their headache pattern may change even though no serious underlying cause for their headaches is present.

Table 2

Clinical Signs To Consider A Possible Serious Underlying Cause

- Recent onset of a significant headache condition, especially after middle age.
- A recent change in an established headache pattern.
- The presence of other neurologic symptoms besides headache (*i.e.*, seizures).
- Abnormal neurologic signs on examination (*i.e.*, focal neurologic signs that cannot be explained by another known neurologic condition).
- A rapidly progressive headache syndrome.
- The inability to classify a patient's headaches into one of the benign headache syndromes (*i.e.*, the patient's headaches do not meet diagnostic criteria for one of migraine, tension-type or cluster headache syndromes).

The presence of other neurologic symptoms, that might suggest a structural lesion (*i.e.*, seizures) should always be taken seriously. Symptoms may include, for example, the presence of nausea between headache attacks at times when the patient is pain-free. Abnormal focal neurologic signs on examination should always be taken seriously.

A rapidly progressive headache syndrome, where the patient's headaches are increasing rapidly in severity and/or frequency, should also be taken seriously. This development may suggest an enlarging intracranial mass lesion or the development of increased intracranial pressure.

Finally, there are patients who have unusual headaches, which simply do not meet diagnostic criteria for migraine, tension-type, or cluster headache. Although such patients may well have negative investigations, structural or metabolic

causes may need to be excluded. Once again, clinical judgment must be exercised.

The fear of many patients is that they may be harbouring a brain tumour. Patients may transmit this concern to their physicians and, for the physician to be able to reassure the patient, it is important the physician have confidence in his or her clinical diagnosis. Such confidence is best gained by a careful history and physical, and a knowledge of the clinical features of the benign headache syndromes. If neuroimaging is necessary, a brain computed tomography (CT) scan without contrast will usually suffice. It is not appropriate, however, to image every patient with recurrent migraine or tension-type headache.

With regard to the issue of brain tumour and headache, the study by Vasquez-Barquero is instructive.² These authors reviewed the presentation of a large series of patients with brain tumours and found

that, at the time of diagnosis, virtually all patients with brain tumour had at least one other neurologic symptom besides headache. Furthermore, although headache could be the only symptom of the patient presenting with brain tumour, in their series the longest duration of headache as the only symptom was 77 days. By this time, other symptoms were also present. It is unlikely, therefore, that cases of recurrent or chronic

headache lasting for many months or years are caused by a brain tumour.



**For a good move
see page 109**

What the CT shows

As can be seen from the patient history and the clinical features listed in Table 2, this patient certainly merits neuroimaging from several standpoints. She clearly has a rapidly progressive headache syndrome

Headache

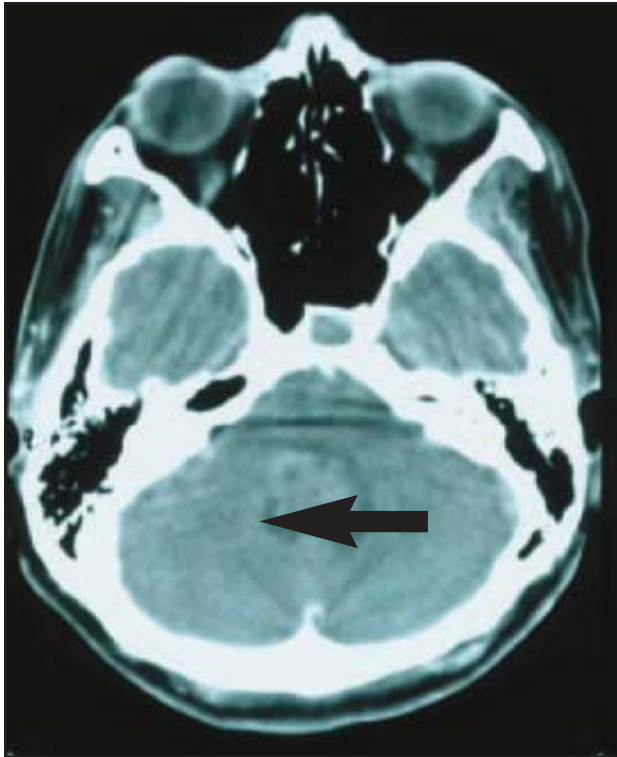


Figure 1. Computed tomography scan showing an ependymoma of the fourth ventricle.

and has other neurologic symptoms (*i.e.*, nausea between headaches and tinnitus). It is true she still has a normal neurologic examination, which is usually a fairly solid guarantee that no structural cause is present for the patient's headache. However, no rule is absolute, and the patient's clinical features at this point certainly merit neuroimaging. Her brain CT scan is shown in Figure 1.

As can be seen, this patient had an ependymoma of the fourth ventricle, which was no doubt causing symptoms both through the displacement of local structures and by some increased intracranial pressure. The latter had not yet resulted in papilledema, but likely would have done so eventually. A non-contrast CT scan was sufficient to show this lesion, as well as to determine the need for further investigation and treatment.

Examination of the optic fundi is an important part of the neurologic examination in the patient with headache. This is never more true than in the patient with benign intracranial hypertension (*pseudotumor cerebri*). In these patients, intracranial pressure is increased, but neuroimaging shows no mass lesion. The cause for the increased pressure is not well understood, but must relate to the dynamics of cerebral spinal fluid formation and absorption. These patients typically present with generalised progressive headache, often with nausea. Diagnosis is important because the papilledema can eventually lead to visual loss. Although this syndrome is more common in obese young females, it can occur in other individuals, including men.

Uncommon Headache Types

The headache type that causes the most difficulty in this area is cluster headache. Even though it is a very characteristic headache, which is usually easily diagnosed clinically, cluster headache is uncommon. In fact, it occurs in approximately 1:1,000 males and in 1:10,000 females. It may be, therefore, that a family physician will only encounter one of these patients every few years. Nevertheless, this headache type causes severe pain and is relatively treatable.

As can be seen from the diagnostic criteria in Table 3, this headache type causes relatively short headache attacks, lasting anywhere from 15 minutes to three hours. The headaches may occur many times per day, and often pop up at night. They also are unilateral, usually centred around the eye and can be very severe. Unlike patients with migraine, patients with cluster headache will generally move around and not lie down during the headache. In fact, if a patient tells you that they get up at night with a headache and go outside in the winter to help them cope with it, this is a strong sign of cluster headache. The short duration

Table 3

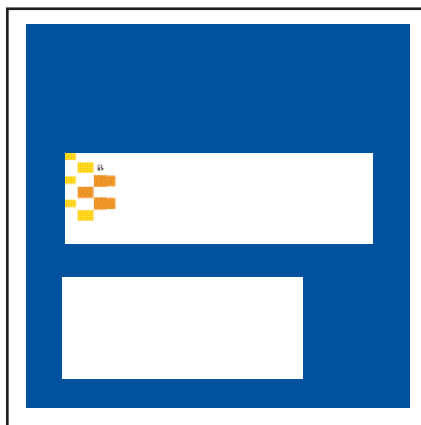
Diagnostic Criteria For Cluster Headache

- Severe unilateral, orbital, supraorbital and/or temporal pain lasting 15 to 180 minutes untreated.
- Headache has one of the following:
 - Conjunctival injection;
 - Lacrimation;
 - Nasal congestion;
 - Rhinorrhea;
 - Forehead and facial sweating;
 - Miosis;
 - Ptosis; or
 - Eyelid edema.
- Frequency of attacks varies from one every other day to eight per day.
- No other cause apparent on history and physical.

Modified from Headache Classification Committee of the International Headache Society: Classification and diagnostic criteria for headache disorders, cranial neuralgias, and facial pain. *Cephalalgia* 1988; 8(Suppl. 7):1-96.

of these headache attacks and the accompanying autonomic features, which usually involve redness and tearing of the eye, with plugging of the nostril on that side, should lead to an easy diagnosis.

Cluster headaches have become of great interest because positron emission tomography studies appear to indicate they are triggered by a small area in the hypothalamus.³ This likely explains some of the unusual features of cluster headache (*i.e.*, occurrence in some patients at the same time each night and, in some patients with episodic cluster, the seasonal occurrence of the cluster headaches at the same time each year). Keep in mind, however, there are patients with chronic cluster headache, who have the typically brief



severe cluster headache attacks throughout the year without prolonged periods of remission.

There are other causes of short-lasting unilateral headaches, but these are much more rare than cluster headache. Such patients are best referred to a neurologist for diagnosis and treatment.

Chronic Daily Headache

This descriptive term is applied to patients who have headache on 15 days or more per month. Many of these patients have exactly what the name implies, namely headache every day. In Western populations, it would appear that between 4% and 5% of the population have chronic daily headache, with over one-half of these patients meeting diagnostic criteria for chronic tension-type headache, and with more than one-third of patients meeting diagnostic criteria for transformed migraine.⁴

The patients with chronic tension-type headache usually have headaches that are bilateral, of a dull pressing and tightening quality, and are usually of only mild or moderate severity. An important feature that helps distinguish chronic tension-type headache from migraine is the fact that the headache is not generally aggravated by walking stairs. In addition, nausea and photophobia usually are not prominent in patients with chronic tension-type headache.

Many patients with chronic daily headache, however, do not meet the above diagnostic criteria.

They may have unilateral headaches, they may have very significant nausea at times and, in fact, their headaches may become quite severe and appear very much like a migraine attack. For some time, such patients were considered to have both migraine and

Headache

tension-type headaches. However, the concept of transformed migraine may better explain what happens to these patients.

Basically, it appears that a significant minority of patients with migraine eventually develop daily or near-daily headache (known as transformed migraine). To make this diagnosis, one must attempt to characterise the nature of the patient's headache attacks prior to the development of chronic daily headache. Many patients will give a clear-cut history of intermittent migraine headaches prior to the development of chronic daily headache. Also helpful in diagnosis is the clinical characterisation of the patient's current headache exacerbations, if such are present. If these headache exacerbations satisfy migraine diagnostic criteria, except for the fact that the attacks may be too long, this is symptomatic of transformed migraine (Table 1).

The point is that, in patients with transformed migraine, the basic headache problem is migraine. The headache exacerbations in such patients may respond to migraine medication, such as the triptans. In addition, there is always the question of why the

Take-home message

Patients with headache may present a diagnostic dilemma. It is important that the physician have a good working knowledge of the common benign headache syndromes, and that he or she know when to suspect a serious underlying cause for the patient's headache. This article has focused on headache secondary to brain tumour, cluster headache and the chronic daily headache syndromes. There are, however, many other important diagnostic entities and diagnostic considerations. Headache diagnosis is indeed challenging, both to the family physician and specialist alike.

Table 4

Diagnostic Criteria For Transformed Migraine

- Headaches occurring more than 15 days per month for longer than one month.
- Average headache duration of longer than four hours per day (if untreated).
- At least one of the following:
 - History of episodic migraine; or
 - Current headache exacerbations meet diagnostic criteria for migraine (except they may be prolonged).
- No other cause apparent.

Modified from Silberstein, Stephen D, Lipton RB, et al: Classification of daily and near-daily headaches: Field trial of revised IHS criteria. *Neurology* 1996; 47:871-5.

patient's migraine headaches transformed in the first place. It is not fully understood why such a headache transformation occurs, but important factors appear to be overuse of symptomatic headache medications like analgesics, ergotamine and the triptans, and also medical and metabolic factors, such as oral contraceptives and hypertension. These should all be considered in the patient with transformed migraine (Table 4).⁵ [CME](#)

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