Chapter 1

Confusing one benign headache with another

Sometimes there is simply no mistaking which primary headache disorder is causing a patient's problem. In these lucky instances, a patient spontaneously provides a history so characteristic of a disorder that there can be no doubt about the diagnosis. In specialty headache practice, however, the patient who gives the doctor such a "silver platter" description of a headache problem is the exception and not the rule. Since the primary, nondangerous headache disorders are clinical diagnoses with, by definition, normal imaging and laboratory findings, the patient history is critical to making a diagnosis.

Unfortunately, as seasoned doctors know all too well, obtaining an accurate history can be difficult. This is particularly true for subjective symptoms such as headache, where patients are struggling to describe things like pain that cannot be directly observed or measured by the doctor. Then too, patients also are reporting symptoms that may come and go. Studies show that patient recall of headache frequency and features is low and deteriorates rapidly over time. A missed or delayed diagnosis of a primary headache disorder is unlikely to expose patients to life-threatening harm, but it does have consequences. For one thing, it may mean the patient does not have the benefit of new, highly effective treatments that work for some headache problems and not others.

In this chapter we deal with cases in which a patient with one of the "big three" common, nondangerous primary headaches – migraine, tension-type, or cluster headache – presented with ambiguous, overlapping features and was therefore mistakenly diagnosed. To minimize the chance of confusing these headaches, it is worth becoming familiar with the many ways in which the diagnostic features of common, nondangerous headaches can overlap or be missed. Far and away the most common pitfall is confusion about whether a patient has migraine or tension-type headache, so we'll start there.

Migraine or tension-type headache?

Case

A 34-year-old teacher was referred for consultation regarding recurrent headaches for the last five years. She was in good health and taking only occasional ibuprofen to treat the headaches. She estimated that headaches occurred on average twice a month, lasting a day or two at a time, and had not recently changed in character. They were bilateral, over her forehead, and sometimes accompanied by neck pain. She said that her job was stressful and wondered whether that might be causing the headaches. She missed a day of work every other month because of headaches, and reported she was seeking treatment because her missed work time had recently become "an issue" with her employer. Physical and neurologic examinations were normal except for mild tenderness on palpation over the posterior neck and upper trapezius muscles. She had previously been told that she probably had "tension headaches" and had been referred for physical therapy but did not find that treatment effective.

How can migraine be reliably distinguished from tension-type headache in this patient?

Careful evaluation of a full headache history is, in our experience, the most useful method of distinguishing between migraine and tension-type headache. In this case, the patient did not spontaneously report characteristic features of migraine such as nausea, vomiting, photo or phonophobia, or worsening with physical activity. On the other hand, we did not ask!

Table 1.1 lists migraine features that are contained in the diagnostic criteria for the disorder, along with examples of how these features can be missed or misinterpreted. The diagnostic criteria for migraine have not changed in the latest version (3-beta) of the International Classification of Headache Disorders (ICHD).

Table 1.1. Diagnostic features of migraine without aura: common pitfalls

"Silver platter" migraine features	"Not so obvious" migraine history
Duration of 4–72 hours	Duration uncertain because patient treats early or falls asleep. Shorter headaches often seen in children
Unilateral (often over the temple)*	Bilateral, posterior location of pain or prominent complaints of neck pain often lead to a diagnosis of tension-type headache, but neck pain occurs in almost three-quarters of migraine attacks
Throbbing*	Not all patients who otherwise have clear-cut migraine report throbbing pain; for many patients the throbbing quality of the pain is obvious only in fully developed, longer duration headaches, so patients who treat early or fall asleep may not experience this. Be alert for synonymous descriptions such as "pounding" or "with my heartbeat"
Moderate to severe pain intensity*	In migraine attacks that are treated or do not progress, pain may never reach severe intensity. Differences in pain reporting behaviors and pain perception among patients may affect patient ratings of pain intensity
Aggravated by or causing avoidance of physical activity*	Sedentary patients may not have noticed this feature of their headaches
Nausea and/or vomiting#	Vomiting is prominent in children with migraine, but often lessens as patients get older or headache frequency increases. Decreased appetite may be present instead
Photophobia and phonophobia#	Sensitivity to light or sound may become apparent only in headaches that have a chance to develop fully; these symptoms may not develop in milder *"forme fruste"* or treated attacks

* Only two of these four features are required for a diagnosis of migraine.
\# Only one of these two features is required for a diagnosis of migraine. At least five attacks meeting criteria are required before migraine without aura can be diagnosed.

Some diagnostic criteria for migraine are more useful than others when trying to decide between diagnoses of migraine and tension-type headache. A systematic review of diagnostic studies showed that the features most predictive of migraine as opposed to tension-type headache were nausea, photo and phonophobia. When present, a typical history consistent with migraine aura was, unsurprisingly, also highly predictive of a migraine diagnosis.

What benign headache disorder might account for this patient's headaches?

Taking the limited history at face value, this patient's presentation is compatible with a diagnosis of tension-type headache. It is tempting to think that the bilateral, posterior location of the headache and associated muscle tenderness clinch the matter. Migraine is, however, also in the differential. In fact, while tension-type headache is the most common type of headache in the general population, it is not the most common type of headache in patients whose headaches are troublesome enough to seek medical care. A wealth of good quality evidence suggests that once dangerous causes of headache have been ruled out, the likelihood is that patients consulting general physicians for troublesome headaches have migraine and not tension-type headache. This is true even in patients like the one in our case who present with features such as muscle tension and neck pain and attribute their headaches to stress. Muscle tension and neck pain are common in both migraine and tension-type headache, as is aggravation of headaches by emotional stress or tension. In fact, migraine patients who have these overlap characteristics (particularly neck pain) are most likely to receive an incorrect diagnosis of tension-type headache.

In this case, further questioning revealed that the patient did have some loss of appetite with her headaches and she became mildly sensitive to light, features which support a diagnosis of migraine instead of tension-type headache.

Discussion

Research in primary care settings shows that most patients who seek care for troublesome headaches receive a diagnosis of tension-type headache. This is particularly likely to occur when patients report features that are assumed to be highly characteristic of tension-type headache – as the patient in our case did. For example, many physicians (and patients, too) assume that muscle pain or tenderness in the neck or shoulders is synonymous with tension-type headache. They may also assume the same thing in patients who

Table 1.2. Headache features and diagnosis of migraine

Clinical feature	Sensitivity of diagnosis of migraine (% of patients)	Specificity of diagnosis for migraine vs. tension-type headache (% of patients)	Likelihood ratio for diagnosis of migraine (95% confidence interval)	
			Positive	Negative
Nausea	82	96	23.2 (17.7–30.4)	0.19 (0.18–0.20)
Photophobia	79	87	6.0 (5.2–6.8)	0.24 (0.23–0.26)
Phonophobia	67	87	5.2 (4.5–5.9)	0.38 (0.36–0.40)
Exacerbation by physical activity	81	78	3.7 (3.4–4.0)	0.24 (0.23–0.26)
Unilateral	66	78	3.1 (2.8–3.3)	0.43 (0.41–0.45)
Throbbing or pulsating	76	77	3.3 (3.1–3.6)	0.32 (0.30–0.33)
Duration 4–24 hours	57	67	1.7 (1.5–2.0)	0.64 (0.58–0.71)
Duration 24–72 hours	13	91	1.4 (1.0–2.0)	0.96 (0.92–1.0)
Duration less than 4 hours	26	51	0.52 (0.44–0.61)	1.5 (1.3–1.6)

report high levels of psychologic or emotional tension. Faced with a clinical situation like the one described above, many physicians might consider sending the patient for physical therapy or prescribing a muscle relaxant, both treatments for tension-type headache.

The assumption that these symptoms are indicative of tension-type headache probably stems from the fact that the term "tension-type headache" suggests that "tension" of some sort – perhaps psychologic or muscle – may be the cause of headache. This is a diagnostic pitfall, however, since evidence to support these views is lacking. Although patients with tension-type headache do, as a group, have more pericranial tenderness than patients with migraine, muscle pain, especially neck pain, is nonetheless very common in migraine patients.

Electromyography (EMG) is not useful in distinguishing between the two disorders. Over a third of migraine patients report neck pain with at least some of their attacks. The neck pain can come before, during, or even after attacks; this variability in its time course makes it unlikely that neck pain is the "cause" of headache. Both migraine and tension-type headache patients have lower thresholds for experiencing pain with pressure on muscles than do people without headache; interestingly, the upper trapezius is the most common site of tenderness.

Similarly, elevated levels of psychologic and emotional distress are common in patients who seek medical care for stubborn headaches, and may in part reflect the impact of poorly controlled headaches on their lives, rather than the other way around. It is certainly the case, however, that emotional stress is a commonly mentioned "trigger" of headache in both tension-type and migraine patients. In fact, as demonstrated in Table 1.2, there is considerable overlap of commonly reported triggers between migraine and tension-type headache. This underscores the surprisingly *low* diagnostic value of many triggers and other historical features commonly thought to be pathognomonic of one or the other disorder. Having patients keep a headache diary, such as the one illustrated in Table 1.3, should help in distinguishing between tension-type and migraine headaches when the diagnosis is uncertain.

In a large multinational study, over a thousand patients consulting physicians with a complaint of headache were asked to keep careful diaries of their headaches for up to six months. These records were then reviewed by headache experts, and the final diagnosis of headache type was compared with the diagnosis the treating physician had made at the patient's first visit. When physicians made a diagnosis of migraine, this diagnosis was correct in 98% of patients. When physicians diagnosed non-migraine headaches, such as tension-type headache, the diagnosis ultimately turned out to be wrong in 82% of patients. The predominant reason for misdiagnosis was having missed migraine. The authors of this study concluded that "These findings support the diagnostic approach of considering episodic, disabling primary headaches with an otherwise normal physical exam

Table 1.3. A headache diary that can help distinguish between migraine and tension-type headache

	Date	Date	Date	Date	Date
Just before the headache began, was there any disturbance of vision?	Yes OR No	Yes OR No	Yes OR No	Yes OR No	Yes OR No
Just before the headache began, did you have any weakness, numbness, or speech problems?	Yes OR No	Yes OR No	Yes OR No	Yes OR No	Yes OR No
What did the pain feel like?	Pounding, throbbing OR Steady, tightening, squeezing	Pounding, throbbing OR Steady, tightening, squeezing	Pounding, throbbing OR Steady, tightening, squeezing	Pounding, throbbing OR Steady, tightening, squeezing	Pounding, throbbing OR Steady, tightening, squeezing
Where was the headache located?	Right side of the head Left side of the head Both sides of the head	Right side of the head Left side of the head Both sides of the head	Right side of the head Left side of the head Both sides of the head	Right side of the head Left side of the head Both sides of the head	Right side of the head Left side of the head Both sides of the head
Did you experience nausea or vomiting with the headache?	Nausea OR Vomiting OR Both	Nausea OR Vomiting OR Both	Nausea OR Vomiting OR Both	Nausea OR Vomiting OR Both	Nausea OR Vomiting OR Both
Did the headache get worse with physical activity or keep you from being active?	Yes OR No	Yes OR No	Yes OR No	Yes OR No	Yes OR No
During the headache, were you bothered by light?	Yes OR No	Yes OR No	Yes OR No	Yes OR No	Yes OR No
During the headache, were you bothered by sound?	Yes OR No	Yes OR No	Yes OR No	Yes OR No	Yes OR No
At its worst, was your headache pain mild, moderate, or severe?	Mild OR Moderate OR Severe	Mild OR Moderate OR Severe	Mild OR Moderate OR Severe	Mild OR Moderate OR Severe	Mild OR Moderate OR Severe
How long did your headache last?	Less than 4 hours OR 4–72 hours OR Longer than 72 hours or constant	Less than 4 hours OR 4–72 hours OR Longer than 72 hours or constant	Less than 4 hours OR 4–72 hours OR Longer than 72 hours or constant	Less than 4 hours OR 4–72 hours OR Longer than 72 hours or constant	Less than 4 hours OR 4–72 hours OR Longer than 72 hours or constant

to be migraine in the absence of contradictory evidence."

In summary, once a diagnosis of a primary headache disorder has been made, it is appropriate for physicians to *think migraine*. Prospectively kept headache diaries are invaluable in making the diagnosis, as is a careful and probing history. Physicians should avoid placing too much emphasis on historical features such as pain location, muscle tension, psychologic stress, and headache triggers. In contrast, a history of nausea in conjunction with headaches is highly predictive of migraine.

Diagnosis

Migraine without aura.

Tip

Migraine often presents with features assumed to be highly characteristic of tension-type headache. Most patients consulting physicians for troublesome headaches have migraine and not tension-type headache.

Migraine with or without aura?

Case

An 18-year-old woman sought a second opinion because she had been told she should not use estrogen-containing contraceptives – "the pill" – due to her diagnosis of migraine with aura. Instead she was prescribed a progesterone-only pill but that had caused weight gain and irregular periods and she recently stopped it. She was not interested in an intrauterine device and did not think she would be able to use barrier methods reliably. Most of her friends were taking "regular birth control pills" continuously and thus did not have any withdrawal bleeding, and she wanted to do this too. The patient's headaches occurred only four or five times a year. She wondered if she might be able to use estrogen-containing oral contraceptives despite her headaches, since she did not want to get pregnant.

What important piece of information is still missing in this case?

It is not obvious that this patient's diagnosis of migraine with aura is correct. In fact, all we know is that she has headaches once every few months. In order to establish that she has migraine and/or aura, we need to know the details of her headaches and any accompanying features. In this case, the patient confirmed a diagnosis of migraine by describing unilateral, pounding headaches with nausea that last half a day when untreated and were severe enough to prevent her from usual activities, including her usual exercise routine. When asked if she has any warning signs, or other things that occur in association with her headaches, she mentioned that sometimes her vision was blurry prior to a headache. She added that occasionally "the pain is so bad I can't see." When questioned closely, she clarified this by saying that she actually could see just fine, but shut her eyes tightly because the pain was so bad. Even with further questioning she did not report additional associated symptoms.

Table 1.4. The Visual Aura Rating Scale (VARS)

Visual symptom characteristic	Risk score
Duration 5–60 minutes	3
Develops gradually over 5 or more minutes	2
Scotoma	2
Zig-zag line (fortification spectrum)	2
Unilateral (homonymous)	1
Maximum VARS score	10
Migraine with aura diagnosis	≥5

Adapted from: Eriksen *et al.* The Visual Aura Rating Scale (VARS) for migraine aura diagnosis. *Cephalalgia.* 2005;10:801–10, with permission.

Does this patient have migraine with aura?

Aura is a focal neurologic event, which means that it includes symptoms that can be attributed to dysfunction in a particular part of the brain. Aura symptoms can be visual, sensory, motor, or mixed. Visual aura is by far the most common form of aura. Most people who have any form of aura will, at least occasionally, *also* have visual aura. Common features of visual aura are the *scotoma*, an area of decreased visual acuity or visual loss (not seeing something that is there) or a positive visual phenomenon (seeing something that is not there) such as a zig-zag line. These areas of visual loss or distortion are surrounded by areas of normal vision. Figure 1.1 shows a typical scintillating scotoma drawn by a patient who has migraine with aura, who perceived that this scotoma was shimmering and pulsating. Often a scotoma will start as a small area in the center of the visual field, and then expand and move to the periphery of the visual field before fading away.

In migraine with aura, the aura typically precedes the headache; symptoms begin and fade away gradually and do not last longer than an hour. Symptoms also are unilateral (or, in the case of visual symptoms, homonymous – which means they occur in only half of the visual field). Once the aura begins to fade away, it is usually quickly followed by a headache. Sometimes aura can occur without a headache. Table 1.4 reproduces the Visual Aura Rating Scale, which is a method of diagnosing visual aura. This scale assigns points for the presence of certain aura symptoms; to diagnose

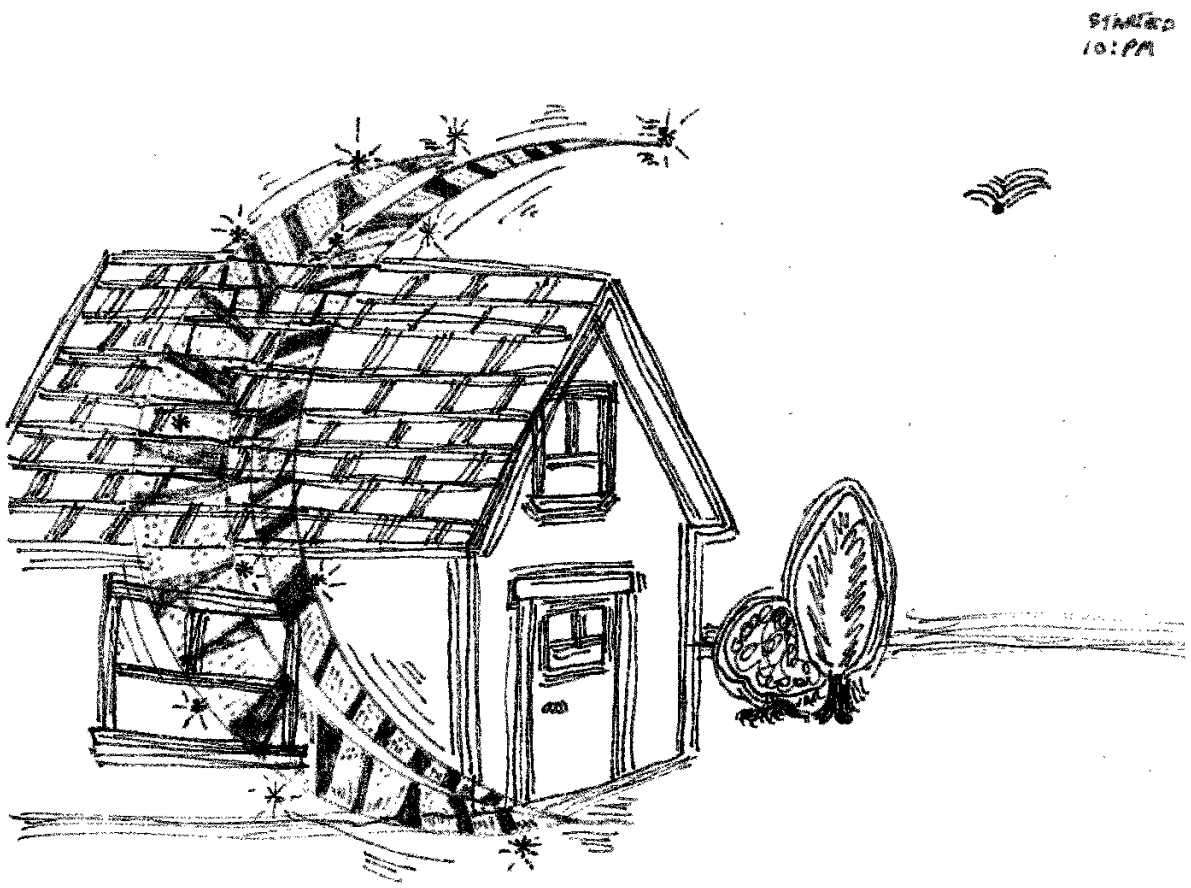

Figure 1.1 A typical scintillating migraine scotoma. Note the patient's attempt to convey the shimmering sensation of movement and vibration in the crescent-shaped, zig-zag visual phenomenon (fortification spectrum) that is drawn.

aura reliably requires a score of at least 5 of the possible 10 points.

Are estrogen-containing contraceptives contraindicated in women who have migraine with aura?

Migraine with aura is associated with an increased risk of ischemic stroke and so is the use of exogenous estrogens. The added risk of stroke with each of these things individually is quite small, but it is higher in the presence of additional risk factors, such as smoking or increasing age. Although it is difficult to place absolute numbers on these risks, authors of these studies note the increase in risk is likely to be multiplicative rather than additive. Because of these risks, guidelines from a number of authoritative groups, including the American College of Obstetrics and Gynecology, recommend against the use of estrogen-containing contraceptives in women who are over 35 and have any type of migraine, or women who have migraine with aura, regardless of age.

Discussion

This patient's headaches met criteria for migraine but the visual events she described were not consistent with a diagnosis of aura. Thus, she has migraine without aura and the use of estrogen-containing contraceptives is not contraindicated. General visual blurring and visual sensitivity, while commonly reported by migraine patients, are not aura. The blurred vision described by this patient is better thought of as part of her headache prodrome. Prodromal events occur before a headache but are not focal neurologic events. Changes in mood, appetite, or concentration are commonly reported migraine prodrome symptoms.

Distinguishing between migraine aura and migraine prodromal or associated symptoms is important because evidence that migraine with aura increases the risk of stroke continues to mount.

Diagnosis

Migraine without aura.

Tip

Focal neurologic symptoms such as positive or negative visual phenomena or sensory disturbance are required for a diagnosis of migraine aura. Since essentially all patients with any sort of aura *also* have visual aura, it is only necessary to establish a history of visual aura in order to make a diagnosis.

Severe unilateral headaches in a man

Case

A 30-year-old man presented for management of cluster headache diagnosed by another physician. He reported an average of three headaches a month that tended to occur towards the end of the month: "They come in bunches. As soon as one is over I might get another one a day or so later." Headaches were located over his forehead bilaterally and he described them as throbbing, with occasional mild nausea. He was not sure how long headaches would last without treatment because for many years he had been using subcutaneous sumatriptan as soon as headaches began.

Is this history consistent with a diagnosis of cluster headache?

Not all elements of the history in this case fit with cluster headache. The bilateral nature of the pain is not typical of cluster headache, which is a strictly unilateral headache, usually located behind an eye. The throbbing nature of the pain is more typical of migraine than of cluster headache, in which the pain is usually described as sharp. The patient does report that headaches come "in bunches," but the period of several days between attacks is not typical of true cluster headaches, in which short headaches usually occur daily or even several times a day. The patient's response to sumatriptan is not helpful in clarifying the diagnosis, since sumatriptan is effective for the treatment of individual attacks of both migraine and cluster headache.

Additional questioning about the features and duration of individual headaches, as well as the pattern of attacks over time, can distinguish migraine from cluster headache. In this case, when questioned, the patient reported that as a child he would occasionally vomit with his headaches, and that they could last up to a day. Both of these features are more consistent with a diagnosis of migraine than with cluster headache.

Why was the diagnosis of migraine missed in this patient?

The criteria for diagnosing migraine are the same for men and women. Unfortunately, they do not fully reflect differences between the sexes in the clinical profile and presentation of migraine. Evidence from the American Migraine Prevalence and Prevention Study shows that, *on average*, men with migraine have fewer clinical features of migraine than women with the disorder. That is, men with migraine are less likely than women with migraine to report nausea, vomiting, photo or phonophobia. That does not mean they do not have these symptoms, but rather that they have *fewer* of these symptoms than women. Because doctors rely on these characteristic historical features to make the clinical diagnosis of migraine, men are at a disadvantage in receiving a correct diagnosis.

In this case, it seems likely that the patient's physician correctly recognized that cluster headache is more common in men and migraine is more common in women. She failed, though, to realize that neither headache type occurs exclusively in one sex.

Discussion

In addition to the fact that men with migraine report fewer migraine-associated features than women, there are other ways in which migraine may differ in men compared with women. For example, men with migraine report that, on average, their attacks are shorter, less severe, and less disabling than migraine attacks in women. Males with migraine also are not exposed to the potent migraine trigger of monthly changes in sex hormones as are females with the disorder, so there is no increase in migraine prevalence at puberty in males. This is in contrast to the situation in females, where migraine prevalence increases

substantially at puberty and remains higher than the prevalence in men even into old age. This suggests that female hormones have an enduring effect on migraine susceptibility. In addition to this lifelong impact on disease risk, women with migraine also experience periodic increases in migraine attack frequency because of hormonal changes with the menstrual cycle.

Although the prevalence of migraine in men is lower than in women, migraine is an extremely common disorder in both sexes. By age 80, the cumulative incidence of migraine in men reaches almost 18% – meaning that almost one in five men will experience migraine during his lifetime. As high as this number is, though, it is certainly lower than the 44% lifetime cumulative incidence of migraine in women. Thus it is understandable that some physicians – and patients – view migraine as a "woman's disease." Unfortunately, the result is that when men seek care for their headaches, clinicians may be less likely to consider migraine as a diagnosis.

Diagnosis

Migraine without aura.

Tip

Migraine is more common in women than in men, but it is highly prevalent in both sexes. Diagnosis of migraine in men may be challenging because men with migraine have fewer typical migraine symptoms than do women.

Frequent, severe episodic headaches in a woman

Case

A 45-year-old woman had been treated for many years for a diagnosis of migraine. She treated individual headaches with 10 mg of oral rizatriptan. Headaches typically awakened her from sleep and were extremely severe, sometimes associated with nausea. They lasted two or three hours, so by the time the rizatriptan became effective the headache was almost over. For the last year her headaches had occurred nightly or every other night, although prior to that she would have headaches "only two or three months at a time and I could deal with that." With no break in her headaches she reported being sleep-deprived and anxious. She was taking 160 mg of long-acting propranolol and 200 mg of topiramate daily. At the last visit she reported that the medications were ineffective and that she was "tired and can't think straight. I am about to lose my job."

Has this patient developed chronic migraine?

Chronic migraine is unlikely in this patient. Although daily, her headaches are short, usually lasting three hours or less. Another clue that this patient may not have migraine is that the headaches have not responded to aggressive treatment with the typically used migraine preventive treatments of propranolol and topiramate. Of course, not all patients with migraine improve with appropriate preventive treatment, but failure to respond to migraine-specific therapy may also suggest an alternative diagnosis. Most features of this patient's headaches are consistent with a diagnosis of cluster headache and not migraine. Additional questioning about the features and duration of individual headaches, as well as the pattern of attacks over time, can distinguish the two disorders (see Table 1.5).

In this case, the patient gave additional history which further clarified the diagnosis. For ten years before the onset of her daily headache she had just one or two bouts of daily or near-daily headache each year. Those periods of frequent headache lasted on average two months and seemed to come in the fall and spring of each year. Individual headaches have always been located behind the left eye and associated with left-sided nasal congestion and tearing of the left eye. Although they last only two to three hours, headaches are extremely severe, "like someone is stabbing me in the eye with a hot poker." The patient is restless and paces the floor during attacks. Her story is more consistent with episodic cluster headache that has now become chronic than with chronic migraine.

Why was cluster headache misdiagnosed in this patient?

The presentation of cluster headache is highly characteristic but the disorder is uncommon. In contrast to migraine, it is more frequent in men than in women. It is caused by dysfunction of central nervous system pain control mechanisms and has distinctive circadian and circannual features. Most physicians have never

Table 1.5. Distinguishing migraine from cluster headache

Pain features	Migraine	Cluster headache
Location	Often unilateral over the temple or forehead area but may be bilateral	Strictly unilateral; typically highly localized to behind one eye
Duration of attack	4–72 hours (adults)	15 minutes to 3 hours*
Frequency of attacks	Sporadic. Can "cluster" in bunches but rarely follow the distinctive pattern of true cluster headache	Attacks can occur once every other day up to eight times a day (for more than half of the time during an active cluster bout – headache frequency may increase or taper slowly at the beginning or end of a bout)*
Associated features	Nausea, vomiting, photo and phonophobia	Agitation or restlessness OR one of the following seven symptoms or signs must occur on the side of the headache: (1) eye redness or tearing; (2) nasal congestion or runny nose; (3) edema of the eyelid; (4) sweating of the forehead and face; (5) flushing of the forehead and face; (6) a feeling of ear fullness; (7) decreased pupil size or ptosis*
Sex ratio	Females > males	Males > females
Behavior during attack	Quiet; prefer to lie quietly in a dark room	Agitated, restless
Temporal features	Attacks typically occur at random and are not easily predictable	Attacks commonly occur at specific times of the day or night. Named for the way they "cluster" together occurring daily or almost daily for 2- to 3-month bouts. In episodic cluster headache these bouts are separated by periods of remission lasting at least a month; in chronic cluster headache remissions do not occur or are shorter than a month

* According to ICHD-3 beta, all of these criteria must be met in at least five attacks in order to make a diagnosis of cluster headache.

seen or treated a patient with cluster headache and may be especially likely to miss it in women, perhaps because women with cluster headache are more likely than men to have migrainous symptoms such as nausea. Diagnostic delay is common in any case, however, with one study showing that the median time from onset to symptoms was three years, with a range of one week to 48 years.

This patient's partial response to a triptan medication may also have contributed to confusion about the diagnosis, since many doctors think of triptans as "migraine medications." In fact, triptans are useful for treating individual attacks of cluster headache as well as migraine.

Discussion

Cluster headache is correctly diagnosed after the initial evaluation only 21% of the time. The most common incorrect diagnoses made in these patients are migraine (34%) and sinusitis (21%). Migraine and cluster can be differentiated on the basis of headache duration, frequency, seasonality, triggering factors, and pain behavior during a headache. The presence of autonomic features is usually part of the presentation of cluster headache but is not strictly necessary for diagnosis if the patient is agitated or paces during an attack. Aura is very rarely seen in cluster headache but should not rule out the diagnosis.

Preventive treatment of cluster headache differs from that of migraine. Typical migraine preventive drugs such as topiramate and propranolol are unlikely to be helpful for cluster headache. In this case the delay in accurate diagnosis has delayed institution of appropriate preventive treatment aimed at reducing or eliminating attacks of cluster headache. The mainstays of prevention for cluster headache are verapamil or lithium. There are no US Food and Drug Administration (FDA)-approved preventive treatments for cluster headache, but clinical experience shows that one or the other of these drugs, or occasionally the combination, brings the disorder under control for most patients.

The slow onset of action of oral triptans makes them a poor treatment choice for most patients with cluster headache. Subcutaneous sumatriptan, which patients can self-administer via an auto-injector, has a more rapid onset of action. It is the only triptan formulation that is FDA approved for treatment of cluster headache.

Diagnosis

Chronic cluster headache.

Tip

Cluster headache is more common in men but can also occur in women. It is often missed in both sexes.

Headache with disturbing visual perceptual alterations

Case

A 43-year-old woman reported she had experienced bad headaches since childhood. In response to the open-ended question "Does anything else happen with your headaches?" she tearfully related symptoms that "might sound crazy." At age 11 she awoke one morning and, while still lying in bed, realized that her hands did not feel "like they belonged to me." When she held them in front of her they looked long and twig-like, not like normal hands. These perceptions disappeared in a few minutes, but she then developed a severe, unilateral headache with vomiting. Over the years she has had many similar episodes of visual abnormality, all followed by severe headaches that meet criteria for migraine. Once while driving she noticed that a fence and the trees behind it appeared weirdly distorted in size and shape. This abnormality was limited to the left side of her visual field but was still present when she covered her left eye. The patient has discussed her headaches with other physicians, but not her visual symptoms, because of her fear that they might be interpreted as psychiatric in nature.

What conditions may be causing these symptoms?

Bizarre visual illusions and distortions that affect the apparent size, volume, shape, or position in space of objects are described with the term "metamorphopsia." Metamorphopsia is part of *Alice in Wonderland* syndrome, thought to be a particular sort of migraine with aura, but can also reflect structural eye disease (usually retinal) and has been reported in patients with idiopathic intracranial hypertension. Metamorphopsia is a common symptom in age-related macular degeneration or other diseases that affect the macula. Typically, patients will complain of "waves" or "bending" in objects known to be straight, such as doorframes or roof lines. Careful examination of the retina, as well as ancillary testing when necessary, including Amsler grids or fluorescein angiography, can usually identify non-aura disorders that might be causing metamorphopsia.

In this case, the symptoms were associated with migraine and occur in consistent temporal relation to the headaches. The patient was otherwise healthy, and Alice in Wonderland syndrome is the most likely diagnosis.

How should this patient be treated?

As is the case for more typical forms of aura, there are no clinically available treatments that will specifically treat the aura of Alice in Wonderland syndrome. (Intravenous ketamine reportedly aborts aura in about half of sufferers, but is not a practical approach to outpatient therapy.) Rather, treatment is aimed at reducing the number of migraine episodes using typical migraine preventive therapy, and also focuses treating the pain of any headache that accompanies the aura. Triptans and other vasoconstrictive agents are *not* contraindicated in this disorder or in migraine with more typical forms of aura.

Discussion

This unusual form of aura is called "Alice in Wonderland syndrome" because of its similarity to the experiences of Lewis Carroll's fictional Alice in Wonderland. It was first described in 1955. The visual abnormalities in Alice in Wonderland syndrome are more peculiar than those of typical visual aura. The visual disturbance may also be associated with alterations in the perception of time, or feelings of depersonalization or derealization, which seem to be what the patient in this vignette experienced during her first childhood episode. Figures 1.2 and 1.3 depict an illustration of the metamorphopsic visual distortions of this illness.

Alice in Wonderland syndrome is said to be more common in children than adults. In our experience this apparent difference in prevalence may stem from the reluctance of adults to describe symptoms they fear will result in stigmatization. Children may be less worried about this. In their 2008 book *Headache in Children and Adolescents*, Winner *et al.* report that "'The children rarely seem frightened by these illusions and relate the experience in enthusiastic detail. Witnesses of the child's event will either remark that the child has an unusual, bemused look on the face or describe the child changing body positions so that they can 'get under a low ceiling.'"