CHAPTER 2

THE PATIENT WITH VISUAL SYMPTOMS AND HEADACHE

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Case History

This 25-year-old woman has suffered from headaches since the age of 17 years. They originally occurred quite sporadically, approximately once every 6 to 8 weeks. Over the past 3 years, she has been averaging about three headaches per month.

The pain is unilateral and throbbing and often begins in the back of the neck. It ultimately involves the entire left side of her head, being most pronounced around the left eye. It is almost always accompanied by nausea and only rarely by vomiting. There is always some degree of irritation of the gastrointestinal tract. During a headache she is usually quite sensitive to light, to the degree that on the few occasions that she came to the office during a headache, she was wearing sunglasses. During a headache attack, she is often quite sensitive to noise.

Occasionally, the headaches are associated with her menstrual period but obviously some occur where there is no hormonal relationship. One other precipitating factor for her headaches is stress. She was likely to have more frequent headaches during exam times in college, and that has carried over into her business life, with headaches often being associated with stressful times at work.

Her typical headache will last 2 days. Occasionally she is bedridden with the headache but there are days when she can complete the work day and then return home and go to bed. In the early days of her headaches, they lasted less than a day but for the past 2 to 3 years her current pattern has been in place.

Approximately once a month, or one out of every three headaches, she has an unusual set of visual experiences. Approximately an hour before the headache begins, she has a sudden development of wavy lines before her eyes. This visual disturbance lasts about 15 minutes. There is usually a period of 30 to 45 minutes when she is symptom free. At that point her headache begins and develops as described.

Her past medical history is relatively unremarkable. She is not on the birth control pill. She does not smoke and drinks only socially. She does not suffer from allergies.

During the first few years of her headaches she merely used over-the-counter medications for her headaches, without any significant relief. At the age of 20 years she had her first visual aura. At that time she consulted her family physician and he did an evaluation which consisted of a computed tomography (CT) scan with contrast as well as an electroencephalogram.

These tests were all normal. She was seen by an ophthalmologist who did a complete eye exam which was normal. She was treated initially with analgesic-barbiturates for over a year and these provided very little relief. They never totally eradicated a headache, more often merely dulling the pain, only to have it worsen when the medication wore off. She was then prescribed isometheptene mucate which would occasionally relieve her headache, but only if she caught it early. She has had to resort to going to the emergency room for the relief of her headaches on average four times a year.

When we saw this woman, her physical exam was normal. Her blood pressure was 126/76, and her pulse was 70 and regular. Her neurologic exam was completely normal. At that time she had no headache.

Questions about This Case

- What is the diagnosis in this case?
- How would you manage this case?
- What long-term management strategies would you suggest for the patient?
- What agent would you use for abortive therapy?
- Would you place her on a prophylactic agent?

Case Discussion

It is apparent that this patient suffers from two different headache patterns. Some of her headaches are preceded by a visual aura whereas the majority of her headaches begin without the aura. The first type of headache would be classified as migraine with aura, denoting the syndrome of headache associated with characteristic premonitory sensory, motor, or visual symptoms. This has been referred to in the past as "classic migraine." The second type of headache described would be migraine without aura, which has been previously described as "common migraine." It is important to recognize, first of all, that this is a straightforward example of a patient who suffers from both migraine with and without aura. Unfortunately, not all cases of migraine are as straightforward as this. For example, while the majority of all migraines are unilateral, the pain can also be bilateral. This patient is typical in that she tries to function with her headaches but often has to give in to it and go to bed. When I hear patients tell me that they are disabled by their headaches, the first diagnosis I consider is migraine. This patient's headaches are a perfect example of how the pattern of migraines may change throughout life. It is not unusual for migraines to start off occurring infrequently and to increase in frequency as time passes.

It is a well-known fact that approximately 75% of all patients who have migraines are women. Many women experience their headaches only during menstruation while others have headaches during menstruation and ovulation. It has been studied and determined that this is caused by the sudden changes in estrogen at those times of the month. It is also well known that migraines can stop occurring during pregnancy, only to recur once the pregnancy has terminated. Fortunately for many women, their migraines stop entirely after they have completed menopause; however, many women are currently being placed on estrogen replacement therapy to prevent osteoporosis, and this often causes the migraines to continue after menopause. If a women needs to be placed on estrogen following menopause, then the lowest dose of estrogen possible is the best choice. We find that the estrogen patch is the least likely form of replacement therapy to exacerbate a woman's migraines. Unfortunately it is often not strong enough to overcome the postmenopausal symptoms of the patient.

In this patient, the initial concern is the visual aura that accompanies her headaches. It is important to determine the etiology of the aura, and to rule out any organic pathology that could be precipitating the neurologic symptoms. If this patient had not already had a CT scan or magnetic resonance imaging (MRI) with contrast of the head, these tests would have to be performed. If the scan was done more than 2 years previously, one would have to consider repeating the test. This would be especially important if the characteristics of the headaches had changed or more importantly, if the visual aura had changed in any way. In rare situations, a cerebral angiography may be warranted.

It is interesting that the patients who suffer from migraine with aura describe their visual disturbance in a variety of ways, some describe it as flashing lights while others describe it as wavy lines. There are numerous descriptions in literature of patients' experiences.

The typical aura precedes the headache and lasts anywhere from a few minutes to half an hour. This is usually followed by a symptom-free period of up to an hour, which is followed by the migraine. If the visual aura lasts more than an hour, I would be very concerned about ruling out any pathologic cause for the patient's headaches.

Investigations in humans as to the pathophysiology of this aura phenomenon have been done but we are far from being able to draw clear conclusions. Most investigations have been limited to the measurement of blood flow, facilitated in migraine patients by noninvasive imaging techniques. Cerebral blood flow falls, but only to oligemic values in posterior regions of the cortex, in some patients during attacks of migraine with aura. It has also been noted that the regional hypoperfusion develops before and outlasts the focal symptoms. For the most part these cerebral blood flow changes have been demonstrated in patients with migraine with aura and not in those with migraine without aura.

A recent positron emission tomography study measured by means of O-15 labeled water done on a patient who, by chance, had a migraine while in the machine, supports the theory of decreased neuronal function. It demonstrated that there is bilateral hypoperfusion, starting in the occipital lobes and spreading anteriorly during a migraine headache.

The most common symptoms reported in the aura phase are visual, arising from the dysfunction of occipital lobe neurons according to some authors. Scotomas may appear in the central portion of the visual field. A characteristic pattern develops in about 10% of patients; the aura begins as a small paracentral scotoma which slowly expands into a "C" shape. Luminous angles appear at the enlarging outer edge, becoming colored as the scotoma expands toward the periphery of the involved half of the visual field. It eventually disappears over the horizon of peripheral vision, with the entire process lasting less than half an hour. This often occurs during the period of time before the headache and infrequently during the headache. This set of circumstances is not usually associated with a cerebral structural anomaly.

Management Strategies

The first and most important thing to do for a patient with headaches is to make an accurate diagnosis. After all, it is difficult to render the appropriate care if we have not made the proper diagnosis. In this case, the diagnosis is straightforward. This patient has two types of headache, but both are migraines. She has migraine without aura as her most frequent type of headache but she also has migraine with aura a smaller percent of the time. If this patient was presenting to me for the first time, I would probably order an MRI if she had not had one done in the last few years. While I am quite comfortable with the diagnosis according to her history, when neurologic symptoms other than headache become involved, some physicians might feel more comfortable with an imaging scan.

This patient is looking for abortive therapy, in other words, something to get rid of the pain. It is important to keep in mind that while we are dealing with migraine headaches, we are dealing with two different types of migraine. It is my strong opinion that the best types of abortive therapy for migraines of these types are the ergotamines and the triptans. It is very important to keep one fact in mind when dealing with migraine with aura, and that is that the triptans may not work when the headache is in the aura stage, while the ergotamines will sometimes abort the entire process, occasionally preventing the onset of the headache itself.

For this patient we have two options. We could prescribe an ergotamine for both types of headache and instruct her to take it immediately at the onset of her symptoms, whether they are the headache itself, or the visual aura. The second option would be to prescribe a triptan and instruct her to take the medication immediately upon recognition of the headache. Keep in mind that with the use of the triptans we have different formulations available and we need to individualize the prescription for each patient. Some patients may even require two different formulations for different headaches. Some patients awaken with a severe pain and at that point they will benefit more from a subcutaneous form of therapy or perhaps an intranasal spray. I prefer, for my own patients, the use of a sumatriptan nasal spray as the initial form of therapy in patients who do not require a subcutaneous injection. The reason for this is the drug's rapid onset of action and its relatively high degree of efficacy.

The other consideration for this patient is whether or not to place her on some form of preventive therapy. There is no specific therapy in the prophylactic area for migraine with aura. The same drugs that work in migraine without aura could be used in this case. I would consider the use of a prophylactic agent for the headaches that are hormone related. The drugs most commonly used for this purpose are the nonsteroidal anti-inflammatory agents (naproxen sodium), diuretics, and acetazolamide. These drugs are best used if the woman has a pattern with her headaches coming at a specific time during her cycle. The drug can be given a few days before the headache is likely to begin and taken for the next several days. Sometimes there is no specific pattern; therefore, these drugs need to be used for the entire month.

For the patient we have described in this case, the use of a prophylactic drug is questionable. If she averages three headaches per month and we can find her an abortive agent that can rid her of her headache within an hour, it is likely that she would not accept the idea of using a drug on a daily basis, especially when we have finished discussing the possible side effects of any of the prophylactic agents, and have also answered her questions about the likelihood of success with these drugs.

Selected Readings

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Editorial Comments

Migraine varies between patients and within each patient's history and lifetime. This case by Dr. Ryan illustrates these diagnostic variations and the importance of

accurate diagnosis. His approach is reasonable, yet some would defer investigations, while others would select other abortive agents or formulations. Some might include metoclopramide and ASA during the aura. Preventive strategies may include one 8-mg dose of ASA per day and verapamil for migraine with aura.

Since the time of this writing, several triptans have become available including zolmitriptan, naratriptan, and rizatriptan.

Nevertheless, Dr. Ryan clearly illustrates what the "thinking" is behind the decisions of the skilled clinician caring for the headache patient with variable migraine.