Chapter 11

The Woman with Prolonged Visual Symptoms

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Case History

DT is a 44-year-old female nurse, with headaches since she was 34 years old. Her headaches are usually right-sided, throbbing, moderate to severe, and aggravated by physical activities. She has no nausea or vomiting, but describes phonophobia and photophobia. The pain occurs frequently on arising in the morning, and lasts for 2 to 3 days. The frequency of the attacks is 3 to 4 times a month. The pain is partially aborted by 1 or 2 tablets of acetaminophen 750 mg. In virtually all attacks, the headache is accompanied by visual symptoms: in some, the pain is preceded by the visual phenomena; in others, they occur simultaneously.

She describes the visual auras as multicolored, bright, zigzag lines, some with "blind spots." These two types of visual phenomena are scattered across both visual fields. The flickering lights usually last for 60 to 90 minutes, and the scotomata persist for up to 24 hours. She denies any sensory, aphasic, or motor symptoms.

The patient has no family history of migraine. She has a history of asthma, and denies any other medical problems. She is married, has four children, and is not on the birth control pill. She does not smoke, does not drink alcohol, and does not use illegal drugs.

Her clinical and neurologic examinations were completely normal. The blood pressure was 120/80, and the pulse was 76 bpm.

Questions on the Case

Please read the questions, try to answer them, and reflect on your answers before reading the author's discussion.

- What is the most likely diagnosis?
- Would you be concerned about an underlying disease?
- Would you recommend some testing? Which ones?
- How would you treat this patient?

Case Discussion

The most likely diagnosis for this patient is migraine with prolonged aura (code 1.2.2, using the 1988 classification of the International Headache Society [IHS], and now code 1.6.2, probable migraine with aura, in the 2004 IHS classification). The pain meets the criteria for migraine, and the aura lasts for about 24 hours. The typical migraine aura develops over 5 to 15 minutes and lasts less than 60 minutes (mean = 20 minutes). We consider an aura "prolonged" when it lasts from 60 minutes to 7 days. A "persistent" aura lasts for more than 1 week. Migraine with prolonged aura represents 3 to 11% of the migraine subtypes.

This patient had only visual auras. These are the most common (79 to 99%) of the migraine auras, followed by sensory (31%), aphasic (18%), and motor (6%) symptoms. Only visual aura occurs in isolation. The visual symptoms present with a wide variety of manifestations. The typical fortification spectra (teichopsia) are present in only 20% of the patients. Most migraineurs (60 to 90%) have "atypical" visual auras, such as small bright dots ("stars"), flashes of lights (photopsias), black dots, hemianopsia, "tunnel vision," "foggy vision," or visual changes "like a mosaic." This patient described both positive (bright zigzag lines) and negative (blind spots) visual phenomena.

She describes a visual aura in 100% of her attacks. This is not common, and it is mentioned by only 20% of the patients with migraine with aura. Most migraineurs have attacks both with and without aura. She also says that in all attacks the aura symptoms last for more than 60 minutes. This may occur in a few attacks in a typical migraine with aura patient; they usually have some attacks with prolonged aura.

The patient says that in most of her attacks the aura does not precede the pain, but the headache and the visual symptoms come together. This may be due to the fact that she already wakes up with headache in many of her attacks. Migraine auras precede the pain in 90% of the cases.

The typical visual aura starts in the center of the visual field and gradually progresses toward the periphery. Nonetheless, many patients (50 to 56%) describe their auras starting in the periphery of one or both visual fields, and only 9% have visual auras diffuse in both visual fields, as described in this case.

Prolonged migrainous aura may reflect ongoing spreading depression, triggered by any lesion of the brain. Although we must rule out pathologies of the central nervous system, particularly stroke, prolonged auras are not usually indicative of an underlying organic disease. There is no significant association between migraine with prolonged aura and cardiac and hematologic abnormalities, such as mitral valve prolapse, arrhythmias, platelet aggregation, or activation dysfunctions.

The IHS criteria for migraine with prolonged aura require that the neuroimaging be normal. If neuroimaging is abnormal, showing ischemic infarction, then the diagnosis becomes migrainous infarction, 1988 IHS code 1.6.2, or 2004 IHS code 1.5.4. True migrainous infarcts are rare. The cranial magnetic resonance image (MRI) of this patient was normal, as well as her electrocardiogram (ECG) and the routine laboratory examinations. In migraineurs with prolonged aura and white matter abnormalities on brain MRI, cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) should be suspected. In cases with prolonged motor auras, we must think of familial hemiplegic migraine (1988 IHS code 1.2.3, 2004 IHS code 1.2.4), which is a rare autosomal dominantly inherited subtype of migraine with aura. These patients often have prolonged visual and sensory symptoms as well.

The treatment of migraine with prolonged aura is based on anecdote. As abortive management, there is some concern about the use of ergotamines and triptans, because of the potentiality for cerebral vasoconstriction, which would increase the risk of brain infarction. However, some headache specialists have used triptans for this kind of migraineurs without any complications. Some think we should try simple analgesics and nonsteroidal anti-inflammatory drugs (NSAIDs) before trying vasoconstrictive agents.

As preventive treatment, many authorities recommend caution with the use of beta-blockers, because they may limit any compensatory vasodilator capacitance. This is based on an old report of 7 patients in whom there was an association with stroke and the use of propranolol in migraineurs. In a survey of some headache specialists about the use of preventives for treating persistent migraine auras, 55% preferred verapamil and 18% favored divalproex sodium. None would prescribe beta-blockers as a first choice. For patients with migraine with aura, and particularly with prolonged aura, my drugs of first choice are verapamil, flunarizine (not available in the United States), and divalproex sodium. For this patient, I prescribed verapamil 80 mg tid, which was very effective. The frequency of her attacks was then much lower, and the aura, when present, much shorter. To abort the attacks, she only needed NSAIDs.

Case Summary

- This is a case of migraine with prolonged aura.
- The patient has only visual auras, which last for about 24 hours.
- She has prolonged visual symptoms in every attack, with positive and negative phenomena, diffuse in both visual fields.
- The visual auras either precede or come together with the headache.
- She has no other underlying disease causing the prolonged aura. The laboratory tests, ECG, and brain MRI were normal.
- As abortive treatment, simple analgesics and NSAIDs should be tried first. Some specialists use ergotamines and triptans with caution.
- As preventive treatment, the drugs of first choice are verapamil, flunarizine, and divalproex sodium. Beta-blockers should be avoided.

Selected Readings

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Editorial Comments

Dr. Queiroz is an expert on the presentations of aura, having prospectively followed and reported a very large number in a classic study (Headache 1997;37:137–41). As noted, the genesis of persistent aura without infarction is likely to be associated with cortical spreading depression, similar to typical aura. This really should be called spreading activation, since the initial event is neuronal activation and hyperemia, followed by an almost postictal neuronal depression of activity, with oligemia, but not ischemia, due to the decrease in need for blood flow. The oligemia reaches 40%, and the debate rages as to whether vasoconstrictive agents are appropriate during the oligemia, despite the lack of ischemia. The case against using ergots and triptans is made as follows: 1) During the oligemia, with additional drug-induced vasoconstriction, sludging, thrombosis, and infarction could occur; 2) No adequately powered trial has been performed to establish the safety of the use of these medications, and anecdotal reports do not provide enough statistical power to make an evidence-based recommendation. For this reason, vasoconstrictive medications such as triptans or ergots are not used to abort migraine with prolonged or persistent aura.

Final diagnosis:

Probable migraine with (prolonged) aura