CHAPTER 48

THE WOMAN WITH OCCIPITAL HEADACHE AND ACUTE NEUROLOGIC DEFICITS

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Case History

This 29-year-old woman initially developed migraine at the age of 19, when she experienced one episode of left-side pulsatile headache associated with transient tingling paresthesia in the right arm and leg. Her headaches were typically unilateral and pulsatile in nature, and were associated with photophobia and nausea. They were easily treated with ibuprofen, and would occur just a few times a year.

The patient works as a nurse and also has what she calls "typical tension" headaches from time to time at the end of a hard day of work. She is otherwise healthy.

She was in her usual state of good health until one day she developed a strong occipital headache, which persisted for 2 days. On the third day at 7:30 AM the pain in her occiput acutely worsened, and she experienced numbness, which traveled from the right leg to the occiput to the right arm, followed by a claw-like contraction of the right hand. During this episode, which lasted 30 minutes, she also had right facial weakness and difficulty speaking. She was admitted to a local emergency room. At that point her deficits had completely resolved and she was released.

The next day, she had a second episode, which began with a severe bilateral occipital headache followed shortly thereafter by numbness, which traveled up both legs, beginning in the feet, progressing to the thighs, then to both arms and then to the mouth. At this point she was completely unable to speak. Her parents took her to another emergency room where she was treated with oxycodone and ibuprofen. After approximately 40 minutes, as these deficits resolved she experienced severe photophobia and saw white spots. A computed tomography (CT) scan was obtained and was negative. She was released on pain medication.

One day later, she saw a neurologist who diagnosed complicated migraine and prescribed a sumatriptan injection. That evening, while driving home from the appointment, she experienced a third episode, which again began with a severe occipital headache, followed by numbness in the right leg, which traveled to the right arm. When she got home the symptoms persisted and she took a dose of sumatriptan. Her symptoms resolved immediately.

The next day her neurologist ordered a magnetic resonance imaging (MRI) and magnetic resonance angiography of the brain, which was completely normal.

Finally, in the evening three days later, while at home, she experienced a fourth episode of severe occipital headache. At the onset of this attack she noticed difficulty holding the phone in her left hand. She then rapidly developed an acute agitated confusional state. She was taken to another local emergency room where her temperature was 104°F. At that time she had a left hemiplegia. It was also noticed that as she tried to dress to go to the ER she had not put her clothes on the left side of her body, and a neurologic exam confirmed the presence of left hemispatial neglect.

A spinal tap was performed which showed a lymphocytic pleocytosis and elevated protein (Table 49–1). She was transferred to a tertiary care hospital for further management. On arrival at the intensive care unit she was agitated. A mental status exam showed her to be oriented but mildly encephalopathic, with delayed verbal responses, verbal perseveration, and emotional lability. Over the next few days her mental status completely

TABLE 48–1. Lumbar Puncture Results

Date	WBC (cells/mm^3)	RBC (cells/mm^3)	Protein (mg/dL)	Glucose (mg/dL)	Differential Lymphocytes/Monocytes
Baseline	280	300	142	67	99/1
4 days later	135	15	65	70	95/5
9 days later	215	20	36	51	97/3
21 days later	50	0	47	72	97/3

WBC = white blood cell count; RBC = red blood cell count.

cleared and her left hemiparesis resolved. Two days after admission her condition was normal except for bilateral knee clonus, and a mildly unsteady tandem gait. A second lumbar puncture (LP) showed slight improvement in the lymphocytic pleocytosis (Table 48–1). Her workup at this time included an electroencephalogram, which showed mild bilateral slowing, another CT scan which was normal, and a brain single photon emission computed tomography scan which was normal. She was treated with acyclovir, antibiotics, and phenytoin and experienced no further episodes. She was discharged after 10 days with a normal exam and was off all medications.

Negative laboratory tests included cerebrospinal fluid (CSF) herpes simplex virus–polymerase chain reaction (HSV-PCR); CSF Lyme antibody; antinuclear antibodies (ANA); Coxsackie virus titers; echovirus titers; arboviral titers; viral rectal and throat cultures; CSF bacterial, fungal and viral cultures; CSF immunoglobulin G (IgG) index; CSF cryptococcal antigen; CSF Venereal Disease Research Laboratory (VDRL); and CSF angiotensin converting enzyme (ACE) activity. Oligoclonal bands were positive in the CSF.

Five days after discharge from the hospital, she experienced a fifth stereotyped episode of occipital headache followed by numbness which ascended from the right leg to the occiput to the right arm followed by speech difficulty. She called her doctor and had evidence of anomic aphasia with semantic paraphasic errors on the telephone. This episode again lasted 30 minutes. She was told to begin taking valproic acid, but did not fill the prescription.

When driving home from her doctor's appointment, she experienced another severe attack, which began with occipital headache and numbness spreading from the right leg to the occiput to the right arm, followed rapidly by a confusional state and aphasia. She was again taken to the emergency room where she was in an agitated delirium with aphasic speech and a right hemivisual deficit. Another lumbar puncture was performed which again showed lymphocytic pleocytosis (Table 48–1). She improved markedly over the next 24 hours and by the next day had a normal mental status with mild right hyperreflexia and difficulty with tandem gait. At this point, all serologic tests for viral meningitis were negative. She was treated with dexamethasone and loaded with valproic acid. On hospital day 1 she had a trans-cranial doppler examination which showed evidence of accelerated flow consistent with spasm of the distal basilar artery (Table 48–2). This was repeated 2 days later and showed normalization of basilar flow, confirming the presumed diagnosis of complicated basilar artery migraine with secondary pleocytosis. An MRI of the brain was normal.

TABLE 48–2. Basilar Artery Transcranial Doppler Examination Results

Date	Depth (mm)	Peak Velocity (cm/sec)	Mean Velocity (cm/sec)	Pulsatility Index
Baseline	104	105	70	0.82
2 days later	100	63	46	0.62

Thereafter, the dexamethasone was tapered off over a period of 10 days and she continued to take valproic acid with levels ranging from 75 to 120 mg per dL. She experienced no further episodes of complicated migraine. After 3 months the valproic acid was tapered off and the patient has done well ever since with no further episodes of basilar migraine.

Questions about This Case

- What are your diagnostic considerations in this case?
- Do you think that sumatriptan was an appropriate treatment after the second attack of occipital headache and neurologic deficit?
- How would you have managed this patient's clinical course?

Points for discussion:

- This patient clinically had a history compatible with basilar migraine. The differential diagnosis would have included partial seizures or viral meningitis.
- Sumatriptan and other vasoconstricting agents are generally considered to be contraindicated in this condition but happen to be effective in this case.

- Pleocytosis has been described in association with complicated migraine, and supports the hypothesis of a neurogenically-mediated inflammatory response (trigeminovascular hypothesis) in the etiology of complicated migraine.
- The presence of distal basal artery constriction documented by transcranial doppler is strongly confirmatory for the presumed diagnosis of basilar artery migraine.
- Valproic acid was associated with a dramatic cessation of symptoms in this case and may be effective for the treatment of complicated migraine. Other treatment possibilities might include calcium channel blockers.

Case Discussion

Diagnostic considerations in the case include a recurring encephalitis, a cerebral vasculitis such as extrinsic compression from rheumatoid inflammatory tissue, basilar artery dissection or the syndrome of mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS). A lactic acid level was not obtained and she might also have undergone a muscle biopsy looking for ragged red fibers which are characteristic of a mitochondrial cytopathy, albeit nonspecific. Infarctions in MELAS do have a predilection for the posterior cerebral region, but this patient has exhibited a benign course, unlikely to be the case in MELAS.

Sumatriptan is contraindicated in the treatment of basilar migraine but appeared to be effective in this case. It is contraindicated because of fear that it might further reduce cerebral perfusion in a situation felt likely to be associated with cerebral ischemia. Practically, it is contraindicated because the studies on this agent which led to FDA approval excluded any patients with basilar migraine. Therefore it is untested in such patients. Sumatriptan is clearly a potent constrictor of human dural arteries but less so of cerebral and temporal arteries. Sumatriptan, as well as ergotamine, does appear to affect flow velocity on transcranial Doppler (TCD) studies in some migraineurs, particularly involving the middle cerebral and basilar arteries, although the majority of patients studied do not show any alteration in blood flow velocities. A theory still exists that arteriovenous anastomoses open in migraine and that sumatriptan and dihydroergotamine constrict these anastomoses. Should that be the case, these drugs could prove useful by improving perfusion.

Several convincing cases of migraine with CSF pleocytosis and elevations in CSF protein have been reported. Pleocytosis supports the trigeminovascular hypothesis that is described in detail elsewhere in this book. Most cases of migraine have not been associated with any abnormalities of the CSF, but several cases of "hemiplegic" migraine have had similar CSF profiles to that of our patient. It is recognized that lumbar punctures are not routinely performed in patients with migraine with aura and therefore the actual incidence of these abnormalities is not known. Cerebrospinal fluid viral cultures and CSF Lyme titers in this patient were normal, but that does not entirely exclude an infectious cause of the attacks. We also know that vasodilator polypeptides accumulate in the CSF of migraineurs and that there is an increase in capillary permeability. It is not difficult to imagine that a migraine headache could therefore simply reflect the neurogenic inflammation of a severe migraine attack.

The presence of distal basilar artery constriction may confirm the presumed hypothesis of basilar migraine. We know that this disorder can be associated with cerebral infarction, which is thought to be secondary to vasospasm, particularly since morphologic changes within these patient's vessels have not been identified. Cerebral angiography performed during attacks of basilar migraine is generally normal, but at the height of an attack, constriction of the basilar artery has been seen. Unfortunately studies of TCD features have not yielded consistent results. It appears that the intracranial mean flow velocities may be high in all cerebral arteries. It is also possible in this case that an unidentified cause of meningoencephalitis served as a nonspecific trigger for attacks of basilar migraine. Others (Barleson) have concluded that a syndrome of migrainous attacks with a mononuclear CSF pleocytosis suggests a benign syndrome that might not justify an invasive evaluation with cerebral angiography nor any therapeutic intervention. Furthermore, it has been suggested from the anecdotal experiences of several authors that migrainous patients may have an excessive complication rate from cerebral angiography.

The final point deals with how to manage this patient. Valproate appeared to have helped resolve these attacks. In similar cases, attacks are often infrequent making a conclusion about the efficacy of any preventive agent difficult. Most patients with basilar migraine have their attacks widely spaced and are nonprogressive. Therefore prophylaxis is often not suggested. In fact, all treatment recommendations for basilar migraine are based on anecdotal experiences and no controlled studies really exist to guide us. Another reasonable treatment is with calcium channel blockers, noting that these agents reduce vasospasm since calcium regulates vascular smooth-muscle contraction. A few case reports suggest that propranolol may have precipitated attacks and some believe these agents are contraindicated. Other headache experts have recommended these agents for the prophylaxis of basilar migraine.

Management Strategies

- Rule out infectious causes of the encephalopathy and treat for a meningoencephalitis if the diagnosis is uncertain. It is preferable to overtreat for an infectious etiology than to undertreat for migraine.
- Avoid treatment with serotonin-1 agonists if the differential diagnosis includes hemiplegic or basilar migraine.
- Remember that a response or lack of response to an abortive antimigraine cannot be used as evidence for the diagnosis of migraine.
- A neurology consultation is always appropriate in such a gravely ill patient.
- A transcranial doppler can be a valuable study if available in a hospital setting if an attack might be observed.
- Should the diagnosis of basilar migraine be made, a decision about whether to use prophylactic agents will be dependent upon the frequency and severity of such attacks, but recognizing that there is no proven effective therapy.

Selected Readings

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Editorial Comments

Unusual variations of migraine are not infrequent in clinical practice. However, this case by Drs. Green and Mayer raises numerous questions about diagnoses and appropriate management, as well as reminding clinicians that mononuclear pleocytosis in the CSF is seen in many headache disorders, including migraine. The authors conclude that this patient has basilar migraine possibly triggered by meningoencephalitis. Readers should draw their own conclusions.