Woman With Severe Headache and Neurologic Deficit

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THE CASE:
A 32-year-old woman complains of severe throbbing pain at the top of her head; numbness and weakness on the left side of her face and in her left arm and leg; and nausea, vomiting, and light and noise sensitivity. She rates the severity of the pain as 8 on a 10-point scale. The headache started about 36 hours earlier, with pain in the left temporal area. As the pain increased, it radiated to the opposite side and eventually involved the entire head.

When the pain started, the patient felt light tingling in her lips (more on the left side) that slowly spread down to her left arm and leg. The tingling increased, and the affected areas eventually became numb and weak.

Her blood pressure is 130/90 mm Hg; heart rate, 84 beats per minute; and respiration rate, 16 breaths per minute. A neurologic examination reveals decreased pain sensitivity on the left side of the face, neck, and ipsilateral extremities. Muscle strength is rated as 5 of 5 in her right arm and leg and 3 of 5 on the left side. Deep tendon reflexes are somewhat elevated on the left side. No other neurologic abnormalities are noted.

The patient initially experienced headaches during adolescence. These occurred about once or twice a month, were usually located at the top of her head bilaterally, and occasionally radiated and localized to one side. At that time, she experienced an aura (flashing lights or dark spots in both visual fields) 20 to 40 minutes before the headache began. Over the next few years, the headaches occurred more regularly and episodes became more prolonged, lasting up to 5 or 6 days. Three years ago, an MRI scan of the brain showed no abnormalities.

During the past 3 years, some migraine episodes (usually the prolonged attacks) became associated with numbness and weakness in her face and extremities on one side or the other that lasted from a few hours up to 2 to 3 days, with gradual but complete resolution. Most of her headaches now are typical migraines (with or without aura), but about every 1 to 2 months, the neurologic symptoms accompany the headache.

The patient is otherwise healthy and does not smoke. Her mother, sister, and an aunt (her mother's sister) have a history of migraine. The patient said that her mother has migraine attacks with visual aura, but no family member has migraine episodes associated with neurologic deficits.

What is the differential diagnosis of severe headache with accompanying neurologic deficit?
What tests and clinical signs are most helpful in pinpointing the diagnosis?
What therapeutic strategy will relieve both the migraine pain and the neurologic symptoms?

THE DIALOGUE:

Primary care doctor: This patient seems to have several types of headache; the most evident is migraine with and without visual aura. What is the significance of the neurologic symptoms that accompany her headaches, and how can I best determine their cause?

Headache specialist: The symptoms are most likely associated with her migraines; however, the diagnosis can be established only after other causes of the neurologic deficits are excluded. I would first consider possible stroke and transient ischemic attack. When a patient complains of the worst headache of his or her life, it is important to rule out subarachnoid hemorrhage. In this case, the severity of the pain and its gradual onset facilitate ruling out the latter diagnosis.

A neurologic deficit caused by a transient ischemic attack should last less than 24 hours. This woman presented with signs of paresis that had persisted for 36 hours, thus ruling out transient ischemic attack.
Primary care doctor: Which imaging studies would you order to exclude an ischemic event?

Headache specialist: The procedure of choice is a CT or MRI scan of the brain. CT is more valuable when subarachnoid hemorrhage is suspected. In the case of suspected ischemia, brain MRI is preferred because it may take as long as 48 hours for a CT scan to demonstrate ischemic changes. When an aneurysm is suspected, consider magnetic resonance angiography (MRA) of the brain.

Primary care doctor: What were the results of these neuroradiologic studies in this patient?

Headache specialist: We decided to repeat brain MRI because her symptoms had persisted and worsened over the past few years. An MRA scan of the brain was also performed. Although the office neurologic examination showed hemiparesis, the MRI scan did not demonstrate any ischemic changes of the brain and MRA revealed no blood vessel abnormalities. Thus, we were able to rule out stroke and cerebral blood vessel malformations.

Primary care doctor: What diagnosis did you consider after you excluded cerebrovascular disease?

Headache specialist: The diagnosis was hemiplegic migraine. This is a variant of migraine with aura, although it represents a separate clinical entity. Hemiplegic migraine consists of typical migraine features, such as unilateral location, throbbing quality, and duration of pain, and migraine-associated symptoms (nausea, vomiting, photophobia, and phonophobia). In addition, this type of migraine is associated with reversible weakness of one or both extremities on one side of the body.\(^1\)

Two major types of hemiplegic migraine have been identified: sporadic and familial. They have similar clinical presentations. The familial form is a genetic disorder in which mutations in \textit{CACNA1A} (for hemiplegic migraine type 1) and \textit{ATP1A2} (for hemiplegic migraine type 2) genes have been identified.\(^2\) For familial hemiplegic migraine to be diagnosed, the patient must have at least one first- or second-degree relative with the same disorder. This patient denies any family history of similar symptoms; thus, her hemiplegic migraine is the sporadic type.

Primary care doctor: How long do the neurologic symptoms typically persist?

Headache specialist: According to the classification of the International Headache Society, neurologic symptoms should continue for more than 5 minutes but less than 24 hours. However, patients with hemiplegic migraine frequently have neurologic deficits that persist much longer. In some patients, symptoms do not completely resolve for several weeks. In such patients, the nature of the symptoms must be thoroughly investigated to prevent misdiagnosis of a catastrophic vascular event. Consider the results of CT, MRI, and MRA, as well as whether there is a history of similar episodes that completely resolved. In this patient, her relatively young age, lack of risk factors, and insignificant family history of major vascular diseases also help rule out a cerebrovascular event.

Primary care doctor: What is the prevalence of hemiplegic migraine?

Headache specialist: A survey of the Danish population (5.2 million) showed that the prevalence of hemiplegic migraine was 0.01%.\(^3\) It occurs more frequently in women than in men (ratio, 3:1), and there is equal distribution of familial and sporadic forms.

Primary care doctor: How would you treat a patient with hemiplegic migraine?

Headache specialist: The list of safe, effective medications for hemiplegic migraine includes analgesics, corticosteroids, and NSAIDs. Some reports have described the effectiveness of the calcium channel blocker verapamil (either oral or parenteral) in the treatment of both familial and sporadic forms of hemiplegic migraine. As with basilar artery migraine, triptans and ergotamine-containing medications are not recommended in the management of hemiplegic migraine.

In another patient who had the sporadic form of hemiplegic migraine, both analgesics and corticosteroids proved ineffective. However, the patient obtained rapid relief from the migraine episode and the neurologic symptoms after intravenous administration of 1 g of valproic acid. In this patient, the headache diminished within 30 minutes after the injection. The headache and neurologic symptoms completely resolved within 2 hours.\(^4\)

The problem of hemiplegic migraine is not yet well understood. We continue to wait for results of newer clinical studies and newer methods of treatment.

References: REFERENCES:


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