Chapter 29

A Young Woman with Sudden-onset Headache

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Case History

A 22-year-old college senior had been working on a new cheerleading routine involving high-impact aerobic activity and gymnastics, when she noted the onset of a sudden, severe headache. It was excruciating enough to bring her to the ground, although she never had a change in level of consciousness. She was able to describe the headache onset as if someone had taken a baseball bat and hit her in the back of her head. She became nauseated with blurred vision and photophobia and was taken to the emergency room because of the severe pain. She vomited once, and after approximately 1 hour, the headache began decreasing in severity, becoming a dull occipital ache associated with some neck tightness. She had no prior history of severe headache. She had a normal brief neurologic examination. A computed tomography (CT) scan of the brain was performed in the emergency room (ER) and read as negative by the on-call radiologist. She was given injections of meperidine and hydroxyzine that helped the pain, and she was sent home with the diagnosis of probable exercise-induced migraine.

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 emergency room physician's differential diagnosis?
- Because headache happened suddenly during maximal exercise, does this narrow the differential diagnosis?
- Does this differential diagnosis change with a normal CT of the head?
- Should any further testing be done?
- If she had a prior history of migraine, would that help you in your differential?
- Would you expect abnormal neurologic examination findings?

Case Discussion

Severe headache is a common cause for emergency room visits and, statistically, is likely to be a primary headache syndrome. However, the sudden onset of this "worst headache of her life," whether or not this patient had a prior migraine history, made imaging necessary. Her negative head CT, normal neurologic examination, and sudden onset of headache during exercise certainly put "benign exertional headache" high on the differential diagnosis list. The headache was brought on by exercise, and was improving by the time she came to the ER. Benign exertional headache can also have features of migraine such as photophobia and nausea. Blurry vision can often be an accompanying feature of migraine headache, and the majority of patients have associated neck and shoulder pain. Effort headaches usually rapidly decrease in intensity following cessation of the effort.

Other primary headache syndromes that can have a sudden onset include "crash migraine," benign thunderclap headache, and sexual and cough headache syndromes. This patient did not have a prior history of migraine, although her gender and age place her in a high prevalence group for migraine.

Appropriately, the ER physician ordered a head CT to rule out the most common secondary headache diagnosis in this age group. Subarachnoid headache (SAHA) is found in 1 to 4% of patients presenting to emergency departments with headache. The hallmark of SAHA is the triad of 1) sudden-onset "worst headache of your life," often

reaching peak intensity within a minute, 2) change in level of consciousness, and 3) stiff neck, but the spectrum of presenting symptoms will depend on the location of the aneurysm as well as the magnitude of the hemorrhage so that a smaller "sentinel bleed" or unruptured aneurysm may present with no neurologic deficits (Table 29-1). Neck stiffness, changes in level or content of consciousness, and nausea and vomiting are common. Other causes of sudden-onset, severe secondary headache include intracerebral hemorrhage or pituitary apoplexy.

A head CT is the neuroimaging study of choice for a SAHA, but it is not foolproof. Sensitivity of CT in detecting subarachnoid blood is approximately 92 to 93% within the first 24 hours, decreasing to 58% on day 5. The longer the time between the event and imaging, the more likely blood will be isodense on CT imaging and missed. In this case, results were false-negative based on small-volume bleeding. Other potential factors for misdiagnosis may be the expertise of the reading physician, which in some cases, particularly during night hours, may be the ER physician instead of a radiologist. Technical factors such as slice thickness may also decrease sensitivity of the CT scan, with < 3-mm cuts being preferable. Anemia, with a hematocrit less than 30%, may also show false-negative results for blood on a head CT.

In this case, there was high clinical suspicion for SAHA, given the lack of prior headache history, the sudden onset of severe pain, and the neck stiffness. A lumbar puncture should have been performed. Following a SAHA, red blood cells (RBCs) can be detected in cerebral spinal fluid (CSF) for 4 to 21 days. Similar RBC counts in serial tubes of CSF can differentiate SAHA from a "traumatic tap," in which a high RBC number can be obtained in the first tube with a decrease in RBC numbers in subsequent tubes. As RBCs break down in CSF, release of blood pigments causes the supernatant of centrifuged CSF to stain yellow (xanthochromia). Xanthochromia can be detected by spectrophotometry within 12 hours of hemorrhage and can last for 2 to 3 weeks following a SAHA.

Diagnosis of SAHA may also be missed following abrupt-onset headache by failure to recognize that the headache may improve spontaneously, or after treatment with analgesic medications or triptans.

Table 29-1. International Headache Society 2004 Classification for Headaches Attributed to Subarachnoid Hemorrhage

6.2.2 Headache attributed to subarachnoid hemorrhage
Diagnostic criteria:
A. Severe headache of sudden onset fulfilling criteria C and D
B. Neuroimaging (CT or T2-weighted MRI or flair) or CSF evidence of non-traumatic subarachnoid hemorrhage with or without other clinical signs
C. Headache develops simultaneously with hemorrhage
D. Headache resolves within 1 month
Comments:
Subarachnoid hemorrhage is by far the most common cause of intense and incapacitating headache of abrupt onset (thunderclap headache) and remains a serious condition (50% of patients die following subarachnoid hemorrhage, often before arriving at hospital, and 50% of survivors are left disabled).
Excluding trauma, 80% of cases result from ruptured saccular aneurysms.

Adapted from Headache Classification Committee of the International Headache Society, 2004.

CT = computed tomography; CSF = cerebral spinal fluid; MRI = magnetic resonance imaging.

Case Summary

The next day her neck stiffness persisted, and as she bent forward to wash her hair, she noted sudden-onset vertigo lasting several minutes, associated with severe nausea. She called her physician, who was able to obtain an immediate consultation with a neurologist. A magnetic resonance imaging (MRI) scan and MR angiography showed normal brain parenchyma, but a mass extending along the middle third of the basilar artery. Follow-up cerebral angiography showed a fusiform aneurysm approximately 11 mm in length, spindle shaped, and of 8 mm maximum width.

Neurosurgical consultation was obtained. Surgical intervention was deemed too dangerous because of the location of the aneurysm.

Within weeks, she developed postural headache with any movement of her head, and vertigo with any forward or backward head movement. Because of rapid symptom progression, aneurysm clipping was attempted using a suboccipital craniotomy approach, and there was old blood noted in the field. The area was immediately closed because the fusiform aneurysm was unable to be differentiated from the basilar for clipping. She was brought back to the neurology floor after awakening in the recovery room. While sitting up conversing with her family, she developed sudden severe head pain and lost consciousness. She went into immediate respiratory and cardiac arrest. Autopsy showed massive hemorrhage filling the cisterns and radiating into the neck and over the cerebral convexities.

Overview of Secondary Thunderclap Headache

Thunderclap headache describes a sudden-onset severe headache reaching peak intensity almost instantly. Because of the virtually instantaneous onset of the headache, it was compared to a clap of thunder and called thunderclap headache by Day and Raskin in 1986. The most common cause of secondary thunderclap headache is SAHA. However, all headaches secondary to SAHA may

not be instantaneous. In a prospective series of 102 patients with sudden severe headache, Linn and colleagues found that 42% were caused by SAHA, and only half had maximum intensity of their headache at onset. Duration of a SAHA-induced headache varies according to the severity of the hemorrhage, from 2 to 3 days with minor up to 7 to 8 days with major hemorrhages, although the excruciating pain at onset typically lasts an hour or two. Initial headache pain following SAHA is thought to be due to local distension and stretching of pain-sensitive cerebral vasculature and the adjacent arachnoid layer. Arteries are innervated by the fifth, ninth, and tenth cranial nerves and upper cervical nerves. Afferent nerve endings can be directly stimulated by blood in the subarachnoid space and release substance P or calcitonin gene-related peptide, similar to the neurogenic inflammation of migraine. Extravasated blood causes an aseptic inflammatory reaction, evident within 2 hours of the hemorrhage. Increased intracranial pressure, the development of hydrocephalus as a complication of SAHA, or cerebral vasospasm can all contribute to the generation of headache. Headache may also occur in the absence of rupture due to bleeding within the aneurysm wall, or due to rapid or slow progressive expansion of the aneurysm which exerts pressure on the meninges or sensory nerve fibers.

Approximately 80% of cases of SAHA result from ruptured saccular aneurysms, occurring in 30,000 patients annually in the United States. In population-based studies, the 30-day mortality of SAHA is nearly 50%, with 25% of the fatalities occurring within 24 hours, and the majority within a week of the hemorrhage. Leading causes of death are neurologic complications of the initial hemorrhage, recurrent aneurysmal hemorrhage, or vasospasm leading to ischemic stroke. Approximately 5% of the population have one or more saccular aneurysms in autopsy series, and over 50% are asymptomatic and undiscovered.

The presentation of patients with SAHA varies depending on the magnitude of hemorrhage due to rupture or other factors associated with unruptured aneurysms causing referred pain. Up to 50% of patients with documented SAHA have reported a thunderclap headache a few days or weeks prior to major rupture. Typical headache of SAHA is sudden and severe, usually bilateral but can be unilateral, accompanied by nausea and vomiting, and possible loss of consciousness. The neurologic examination can show nuchal rigidity, change in level of consciousness, or focal neurologic signs. In one patient series, two-thirds of patients with headache had associated signs and symptoms such as nausea or vomiting (20%), neck pain or stiffness (30%), visual changes such as blurry vision (15%), and sensory changes or motor deficits (15 to 20%). Electrocardiogram (EKG) changes are also common following SAHA, with more severe hemorrhages predicting more frequent EKG changes. Rhythm disturbances can be seen in almost all patients following a SAHA, and in about 20% of patients, arrythmias may be life-threatening. Elevated temperature, hypertension, and chest pain may also accompany SAHA.

Thunderclap headache as a warning sign of impending aneurysmal rupture is usually so severe and unusual to those who experience it that 40 to 75% of patients see a physician or get medical advice. However, once they see a physician, a missed diagnosis is all too common. In Hauerberg and colleagues' 5-year Danish study of 1,000 patients with SAHA, 15% had a history of sudden headache with neck pain, dizziness, vomiting, or drowsiness, and two-thirds were misdiagnosed by their physicians. Almost half died within 2 years. Thunderclap headache should always have SAHA or aneurysm at the top of the differential list because missing it can be catastrophic. Linn and colleagues' prospective study of 102 patients presenting with severe, sudden headache found that 64% (65/102) had SAHA. SAHA or a "sentinal bleed" should be suspected in any patient presenting with a severe sudden-onset headache, especially if nuchal rigidity or stiffness, change in level of consciousness, or vomiting is also present.

Selected Readings

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Editorial Comments

Thunderclap headache has to be considered to be an intercranial bleed until proven otherwise. The serious consequences of missing this diagnosis makes this a headache disorder that requires the utmost scrutiny and diligence by all physicians. In this case, the lack of any prior history of headache should raise the spectre of a major red flag. Appropriate imaging may well lead to an early diagnosis, or not, and the same applies for CSF examination. Modern neuroimaging with MRA or CT angiography are most helpful in this setting, and should be employed early and frequently if there is any doubt about the diagnosis. Digital cerebral angiography remains the definitive test in some cases. Even with accurate diagnosis, such as occurred in the present case, the nature of the aneurysm and/or its natural clinical history after rupture does not always lead to a favorable outcome. Nevertheless, Dr. Lucas shows us the right approach, and despite the outcome, this is a case worthy of further study.

Final diagnosis:

Thunderclap headache secondary to subarachnoid hemorrhage from fusiform basilar artery aneurysm