CHAPTER 51

THREE PATIENTS WITH "VASCULAR" HEADACHE

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Case History I

Since her early teens, a 28-year-old woman has had severe headaches nearly monthly. About half were preceded by spots and jagged lines that slowly traveled across her vision. The visual display varied and could begin on either side of vision, and usually lasted about 20 minutes. Headaches followed and were pounding and severe, and were usually followed by nausea. Headaches were common in her family. An aunt had died after a severe headache and the family was told that she had a "brain bleed."

While exercising in her home, after struggling to lift a particularly heavy weight, she very suddenly developed a severe generalized headache, more on the right side of the head. Her knees buckled and she felt dazed. She began to vomit. She called out to her husband who was having breakfast. He found her restless and a bit confused. He brought her to the local emergency room.

She explained to the doctor that this was the "worst migraine ever." The physician found her difficult to examine because of her restlessness and constant moving. She frequently interrupted the examination to vomit and once to defecate. Blood pressure was 115/70. Temperature and pulse were normal. There were no abnormalities on examination, except that her right eyelid was a bit drooped and the right pupil was larger than the left and did not react to light either directly or consensually.

Questions on Case I

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

- What is the differential diagnosis?
- Where is the lesion?
- How can migraine be separated from intracranial bleeding?
- How can intracerebral hemorrhage (ICH) be separated from subarachnoid hemorrhage (SAH)?
- How should the patient be evaluated?
- How should she be treated?

Case I Discussion

The main differential diagnosis is between migraine and an acute intracranial process. Although the patient had severe headache, she had no symptoms or signs that indicate brain dysfunction. The right ptosis and dilated nonreactive pupil suggest involvement of her right third cranial nerve. Does she have ophthalmoplegic migraine or a disorder that involves the meninges as well as the IIIrd nerve? The major relevant disorders that involve the meninges are infectious or chemical meningitis, and subarachnoid bleeding. Clearly, meningitis and SAH can be life threatening and are critical to diagnosis.

Bacterial and viral meningitis do not usually begin this abruptly, and she is afebrile. Chemical meningitis, such as that caused by leakage from a craniopharyngioma, can begin abruptly, but she has no signs of a previous pituitary region tumor. The disorder that does begin this suddenly is SAH from a ruptured aneurysm. Her history is typical for SAH. Bleeding from an aneurysm releases blood under arterial pressure into the space around the brain. Sudden-onset severe headache, transient loss of alertness or posture, and vomiting rapidly ensue, and are related to meningeal irritation and sudden increase in intracranial pressure. Reduced alertness or restless agitation is also typical. The vomiting and occasional frequent bowel movements can mimic gastroenteritis. Electrocardiographic abnormalities such as abnormal T waves can lead to misdiagnosis of a primary car-

diac disorder. Onset with exertion is also typical. Some patients with aneurysms have a history in the family of SAH as is likely in this woman. The right IIIrd nerve signs are typical for a posterior communicating artery aneurysm.

ICH indicates bleeding into brain parenchyma. Brain tissue has no pain-sensitive nerve endings. The earliest signs are related to the region into which the bleeding occurs; for example, bleeding into the left internal capsule causes right limb weakness. If bleeding continues and develops mass effect, then vomiting, headache, and decreased consciousness ensue. This patient's early-onset headache and vomiting without parenchymatous signs do not suggest ICH. Furthermore, she is normotensive and has no risk factors for ICH.

Ophthalmoplegic migraine is rare. Third nerve or other extraocular muscle palsy is usually found after the headache develops. Attacks usually begin early in life and are repeated. Her past history is that of migraine with aura. The headache onset in this case is not characteristic of migraine; the vomiting is earlier than in migraine, and leg buckling, confusion, and restlessness early in the course are typical for sudden increase in intracranial pressure, and are not consistent with migraine. Patients with SAH are invariably unable to function normally, while during migraine, patients seem to be able to carry out necessary obligations despite their pain.

Management Strategies for Case I

- Brain imaging should be done quickly. A computed tomography (CT) scan is probably more sensitive for SAH than magnetic resonance imaging (MRI), but MR with T2* (susceptibility) imaging can also pick up SAH.
- If the CT or MRI is nondiagnostic, then this woman needs a lumbar puncture, looking for blood or meningitis.
- Vascular imaging is essential once the diagnosis of SAH is made. CT and MR angiography (MRA) are good screening procedures, and can be performed at the same time as brain imaging. Usually, catheter angiography will be needed to better define the aneurysm and to decide on coiling versus neurosurgical clipping.

Case I Summary

- This is a typical case of SAH from a posterior communicating artery aneurysm.
- Very sudden-onset headache, often after exertion, with rapid vomiting and sudden interruption in behavior, are common features of SAH.
- Rapid brain and vascular imaging are essential. Lumbar puncture is needed when CT scan is normal in a patient with strong clinical suspicion of SAH.

Case History II

A 54-year-old white man had hypertension for a number of years, and his medical history included coronary bypass surgery and claudication of the legs when he walked. During the past 3 weeks, he developed headaches that had not been a problem in the past. The headaches were mostly right sided, came during the day when he was active, and lasted for hours. On one occasion, he had a momentary dimness of vision in his right eye that lasted about 30 seconds and completely cleared. The visual loss was not accompanied by headache. One morning, he awakened with a numb feeling in his left hand and face and came to his doctor's office. By the time he was examined, the numbness had gone.

Examination showed blood pressure of 150/85, pulse of 80 bpm and regular, no cardiac or neck murmurs or bruits, normal optic fundi except for vascular tortuosity, and normal neurologic findings.

Questions on Case II

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

- What is the differential diagnosis?
- What is the mechanism of the headache?
- How should he be evaluated?
- How should he be treated?

Case II Discussion

This patient has right internal carotid artery (ICA) occlusive disease and has had an attack of amaurosis fugax. Although late-in-life onset migraine enters into the differential diagnosis, the background history of severe risk factors for atherosclerosis (ie, hypertension and past coronary and peripheral vascular disease) make the diagnosis of ICA occlusive disease much more likely. The attack of right monocular visual loss is typical for ICA disease. The amaurosis is usually brief and consists of loss of vision. A gray or black shade or simply absence of vision is typical. The usual migraine visual aura consists of positive features: sparkles, lines, and jagged edges that are usually bright and move, and last longer than a minute. So-called retinal migraine, in which retinal arteries become vasoconstricted, can cause brief visual loss, but usually occurs in younger patients. The episode of transient numbness of the left hand and face was most likely related to ischemia in the right convexal paracentral sensory region of the cerebral cortex. Involvement of only the hand and face, which are adjacent on the homunculus of the sensory strip, would be very unusual in a deep internal capsular, thalamic, or brainstem

lesion. The combination of monocular visual loss and contralateral cerebral hemisphere ischemia is diagnostic of carotid artery disease. Atherosclerotic occlusive disease of neck arteries (ICAs and the origins of the vertebral arteries) is very common in white men. Women, blacks, and Asians have less extracranial and more intracranial occlusive disease. Atherosclerosis of coronary, peripheral vascular, and internal carotid arteries commonly coexist in white men with hypertension and hypercholesterolemia.

Headache is a common symptom in patients who have cervicocranial arterial occlusive disease. The headache is posited to be caused by the following:

1. Distention of arteries by thrombi; the pain-sensitive nerves on the outer wall of the arterial adventitia could be stretched
2. Dilatation of collateral arterial channels, also distending them and stimulating pain-sensitive fibers
3. Brain swelling with stimulation of pain-sensitive meningeal fibers; this explanation is only relevant in patients with brain infarcts

Headaches, unusual in the past for the patient, often precede strokes. Usually, there are accompanying transient ischemic attacks (TIAs), as in this patient. Anterior circulation occlusive disease is usually accompanied by headache in the forehead or temporal region, whereas vertebrobasilar occlusive disease is associated with headache that is in the occiput, posterior neck, or even the upper back.

Management Strategies for Case II

- This patient needs urgent evaluation. Begin with a brain image. Despite complete clearing of symptoms and a normal examination, some TIA patients do have brain infarcts on MRI or CT in the appropriate location for the transient symptoms. MRI is more sensitive for acute ischemia, especially if a diffusion-weighted imaging scan is included.
- Vascular imaging, either a CT angiography or an MRA, can be performed at the same time as brain imaging. The neck and intracranial arteries should be scanned.
- Ultrasound, including a duplex scan of the neck and transcranial Doppler ultrasound (TCD), is an alternative method of defining the presence and severity of ICA disease in the neck and of determining (using TCD) the intracranial impact of the neck disease and to screen for additional intracranial occlusive disease.
- MRI showed a small dot infarct in the right convexal parietal lobe and an additional dot in the right frontal lobe. MRA showed a gap in the signal in the neck ICA and normal intracranial arteries. Duplex ultrasound of the neck showed a very severe (90% luminal narrowing) of the right ICA.
- Carotid endarterectomy and endovascular ICA (angioplasty with or without a stent) are the two favored treatments. Choice of treatment depends on the experience and past performance of the various specialists available, and the decision of the patient after being given risk-benefit information.

Case II Summary

- The concurrence of transient amaurosis fugax in one eye and contralateral hemispheral ischemia is diagnostic of carotid artery disease.
- Some patients with carotid artery disease have prominent headache during days and weeks, often accompanied by retinal and/or cerebral ischemia.
- Patients with brain ischemia need urgent evaluation and treatment.

Case History III

A 65-year-old white man had been taking an angiotensin-converting enzyme inhibitor for hypertension, and a statin for elevated cholesterol, but had felt well. He had two brief attacks of dizziness while walking; these caused him to momentarily veer to one side. Four days after the last episode of dizziness, while working on his computer, he suddenly developed a severe pain in his right eye that was accompanied by a pounding headache above that eye. Headaches had not been a problem in the past. He noticed that he was having difficulty reading the text on the computer and could not find icons that he had placed on the left side of the desktop. He decided to visit his physician.

His blood pressure was 145/85, and his pulse was regular. The examination was normal, but the physician decided to send him to a neurologist, who saw him later that day. The neurologist's examination was normal except for a left homonymous hemianopia. The patient omitted words on the left of the page and often missed individuals on the left of pictures. There were no abnormalities of his motor or sensory examination and he walked normally.

Questions on Case III

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

- Where is his lesion?
- What are the likely causes?
- How would you evaluate this patient?
- How should he be treated?

Case III Discussion

The examination showed a left homonymous hemianopia. The lesion causing this must be in the right striate visual cortex, the lateral geniculate body, or the geniculocalcarine tract. The sudden onset, age, and vascular risk factors make stroke the most likely cause. The blood supply to the visual pathways comes from three different arteries: branches of the anterior choroidal artery supply the lateral geniculate body and proximal geniculocalcarine tract; the middle cerebral artery supplies the white matter that incorporates the geniculocalcarine tract as it courses deep near the temporal horn of the lateral ventricle; and the posterior cerebral artery (PCA) supplies branches to the lateral geniculate through posterior choroidal arteries, as well as supplies the striate and parastriate cortex in the occipital lobe.

The stroke involving the occipital lobe could be an infarct or a lobar-type brain hemorrhage. The first step is to look for clues that might differentiate ischemia from hemorrhage in this case. The patient was hypertensive, and hypertension does cause lobar hemorrhages, but the blood pressure was not high on admission. The occipital lobe is a site of predilection for amyloid angiopathy. Hemorrhage from amyloid angiopathy often occurs in the absence of hypertension, affects older individuals, and can mimic an ischemic stroke. The absence of a prior bleed argues only slightly against this possibility. He had two preceding spells characterized by dizziness. Could they have represented TIAs, making ischemic stroke likely? Dizziness is such a ubiquitous symptom, often related to benign peripheral vestibular disorders, such that it is hard to be certain initially that the dizziness was related to the stroke.

The dorsal surface of the tentorium cerebelli is innervated by the Vth cranial nerve. For this reason, patients with PCA territory infarction often have pain that localizes in the ipsilateral eye, and often the forehead above the eye, as in this patient.

Management Strategies for Case III

- First order an imaging study to separate hemorrhage from infarction. In this patient, an MRI showed an infarct involving the right occipital lobe. The infarct was clearly in the territory of the right PCA.
- The PCAs are the terminal branches of the basilar artery. Occasionally, one or both PCAs come from the ICA in a pattern common during fetal life.
- The vast majority of PCA territory infarcts are embolic. The donor source for embolism can be the heart, the aorta, or the extracranial (ECVAs) and intracranial vertebral arteries, or the basilar artery. These areas should be imaged to find the donor source.
- The preceding attacks of dizziness now assume more importance since the vestibulocerebellar regions that are usually involved in patients with vertiginous brain infarcts are supplied by the vertebral arteries.
- Duplex ultrasound with color-flow Doppler showed a severe stenosis at the origin of the right ECVA. The most frequent location for atherosclerotic disease of the ECVAs is at the vertebral artery origin (VAO) from the subclavian arteries. Atherosclerosis at this site shares epidemiologic features with ICA-origin atherosclerosis.
- MRA confirmed the severe VAO stenosis. Intracranial arteries were normal.
- Transesophageal echocardiography showed no important, potentially embolic lesion of the heart or aorta.
- Management of VAO disease is like that of ICA neck disease. Agents that affect platelet functions, standard anticoagulants, surgery, and angioplasty and stenting are all reasonable alternative treatments.
- Although surgery is less often performed for VAO disease than for ICA disease, it can be accomplished safely and effectively in experienced hands. No studies compare surgery with endovascular treatment or compare antiplatelet agents and anticoagulants in patients with VAO disease.
- Emboli from VAO occlusions are most likely red thrombi, so that if medical treatment is chosen, I favor standard anticoagulants.

Case III Summary

- Atherostenotic disease of the VAO caused several TIAs consisting of dizziness and veering.
- An embolus from the atherosclerotic VAO lesion went to the right PCA, causing a right occipital infarct.
- The occipital infarct caused a left hemianopia with left visual neglect.
- Occlusion of the PCA is often accompanied by pain in the ipsilateral eye and forehead above the eye.
- Multiple treatment options are available to treat vertebral artery disease in the neck, but no studies have been performed to test the superiority of any of these treatments over others.

Selected Readings for Case I

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Editorial Comments

In three succinct and clear cases, Dr. Caplan, once again, as in the first edition of this volume, brings the reader to the bedside of patients with cerebrovascular disease and headache. These cases are important to all physicians who see a large variety of headache patients, especially if there are many primary headache patients seen in practice or clinic. This is simply because, even in the presence of usually benign disorders, serious headaches lurk in the background. And of all the headache subtypes that can mimic migraine, those of cerebrovascular origin can be the most problematic, especially if the patient presents with one of the case scenarios noted above and has a primary headache disorder. Read these cases carefully, as they are full of classic neurology and new information as well.

Final diagnoses:

Case I, Subarachnoid hemorrhage
Case II, Transient ischemic attack in carotid territory
Case III, Transient ischemic attack in verterbrobasilar territory