THREE CASES OF "SIDE-LOCKED HEADACHE"

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Case History I

A 46-year-old female presented with a history of episodic tension-type headaches that were right-sided, frontal, and biparietal. Her headaches occurred rarely, lasting about 4 hours, and were usually treated successfully with two aspirin tablets. On March 11, 1984, at age 26 years, she had a mild headache, at which time she took two aspirin tablets and rested in bed. Shortly thereafter, the headache became significantly worse and then abruptly erupted into a "thunderclap headache." She suddenly lapsed consciousness, had major motor seizure activity, and was transferred by emergency ambulance to the hospital. Subsequently, she underwent a craniotomy, with evacuation of subarachnoid blood, evacuation of a subdural hematoma, and excision of an arteriovenous malformation. She was prescribed phenobarbital 30 mg tid as prophylaxis for a presumed seizure disorder, and was discharged home on March 22, 1984. She developed generalized pruritus in the arms, legs, and periorbital region on April 2, 1984, and was prescribed pyribenzamine.

On May 21, 1984, the patient underwent cerebral, right carotid, left carotid, and left vertebral arteriography scans. These arteriograms concluded that there was no evidence of residual arteriovenous malformation.

The patient is followed up on a yearly basis, and shows no signs of recurrence.

Headache frequency has been one tension-type headache every 2 months, and the patient has remained in relatively good health.

Questions on Case I

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

- What influence does preexisting headache have on the work-up of a new type of headache?
- · What constitutes an emergency in headache?
- What is the influence of the timeliness of a procedure of an acute subdural hematoma?

Case I Discussion

This case clearly identifies a de novo thunderclap-type headache in an otherwise healthy young woman. The dramatic change in her headache, the rapidity with which she was taken to the emergency room, and the skill and speed of an immediate operative intervention had a definite impact on her eventual outcome. She has had virtually no subsequent headache aside from mild tension-type headache attacks. The patient has been followed up yearly on an outpatient basis.

The change in this headache was a clear warning sign for impending disaster. Although headache practitioners often hear stories of changing headache, the immediate change to a severe headache is an important sign.

The more usual presentation of this type of headache is more prolonged. Differentiating the characteristics of migraine headache, preceded by visual aura, from a bleed secondary to a cerebral arteriovenous malformation (AVM) is often regarded as difficult. A study of 26 patients with occipital lobe AVM revealed two distinct syndromes in 18 patients—occipital epilepsy and occipital apoplexy. Occipital epilepsy is characterized either by elementary visual phenomena, such as brief flashes of light, or by dimming of a homonymous field. Occipital apoplexy results from hemorrhage and hematoma formation within the occipital lobe, and is characterized by sudden headache and homonymous visual field loss. The authors concluded that patients har-

boring occipital AVMs may, indeed, have visual phenomena and headache that should not be confused with migraine, because either a history of generalized seizure or bruits on examination will probably be present. Yet, in the above case study, there was only a seizure accompanying the severe headache and no other focal premonitory symptoms.

Reviews of the clinical presentations of AVMs describe frequent focal symptomatology, and many patients have preexisting migraine. The presence of premonitory symptoms as well as lateralized headache is also noted in the literature. The one thread throughout the reviews of intracranial vascular pathology including AVM and aneurysm is lateralized headache. Lateralized headache may initially respond to migraine therapy, which may mask the presence of an AVM or aneurysm. Therefore, it is even more important to watch for headache presenting with an explosive onset, and we emphasize the need for magnetic resonance imaging (MRI), and possibly magnetic resonance angiography, in a migraineur presenting with acuteonset lateralized headache. It is clear that this method of evaluation flies in the face of evidence-based guidelines issued by the American Academy of Neurology. If the patient lives in an outlying area without immediate access to emergency neurosurgery, then the importance of screening is even more crucial. Most survivors noted in the review by Guiotoku and colleagues had critically timeintensive care in an immediate fashion.

Case History II

A 30-year-old female arrived at the clinical office on December 11, 1998, for a second opinion regarding diagnosis of migraine headache. The patient and her husband were not satisfied with evaluations that had been done at her Health Maintenance Organization (HMO) facility. The patient and her husband both confirmed that they had requested a scan of the brain on at least two occasions at the HMO. The history indicated that the patient's headaches began at the age of 24 years, occurred at a frequency of four times monthly, and were accompanied by occasional scintillations in the left eye lasting a few minutes, followed by subsequent left-sided steady and throbbing pain, lasting 4 to 24 hours, and had on occasion lasted up to 5 days. Headaches were menstrual related. Neurologic examination including vital signs was entirely normal. The patient had previously treated her headaches with acetaminophen, acetaminophen/aspirin/caffeine, propranolol 10 mg bid, butalbital/aspirin/caffeine, and sumatriptan 25 mg. She stated these medications "were not particularly helpful."

The patient was concerned about a lateralized headache that had not remitted when she had taken a 25 mg sumatriptan tablet a "few days prior." After the normal examination, the patient self-injected sumatriptan 6 mg subcutaneously into her left thigh, with instruction by the physician and physician's assistant. Approximately 6 to 7 minutes after injection, the patient covered her left eye, stating she felt nausea. She then suffered a major motor seizure. The patient was transferred by emergency ambulance to the hospital.

Subsequently, she underwent a craniotomy for a subdural hematoma evacuation after it was discovered that she had a large middle cerebral artery aneurysm, which had bled. Postoperatively, she remained unconscious and intubated, until her death on December 15, 1998. This patient's vital signs at all times in the office and at the hospital were entirely normal.

Questions on Case II

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

- How does lateralized headache influence the decision to scan?
- Do triptans influence ongoing intracranial events?
- Do triptans cause intracranial events?

Case II Discussion

Once again, in Case II, the patient demonstrated by history clear-cut lateralized headache. The history was characteristic of migraine without aura associated with the menses. The patient had taken sumatriptan before she arrived at the office and had suffered no ill effects from the medication. Since an effect of the triptans is vasoconstriction and a slight increase in blood pressure, these two changes in the patient were considered as possible reasons for her deterioration. There was no change in the patient's blood pressure, and at postmortem examination, there was evidence of old organized clot from previous bleeding, with the new bleed. The official recommendations of the American Academy of Neurology is contradicted by the Frishberg article. The official evidence-based recommendation does not include scanning for consistently lateralized headache. We believe that consistently lateralized headache is an indication for scanning.

It is possible that the vasoconstrictive effect of the triptan could have helped control bleeding. This did not occur, probably due to the magnitude of the aneurysm and the fact that the vasculature involved in aneurysm was probably no longer responsive to vasoactive drugs.

Triptans can cause chest pressure or fullness and are contraindicated with Prinzmetal angina and cardiovascular disease. However, with over 10 years of experience with the class of medication and millions of doses prescribed, it can

be said with confidence that the triptans are one of the best studied and safest medications used in every day medical practice. Review of the very few cases of intracranial events after triptan use has not confirmed that the vasoactive effects of the triptans are responsible for those events. Evidence of serious central nervous system side effects is lacking for triptans, except with misuse, as noted by Nighnoghossian and Meschia and their colleagues.

Case History III

A third case is a 60-year-old woman with no history of headache prior to July 1970. In July of 1970, while under "a great deal of distress" over leaving her social worker position in East Africa, she began having a "bad headache" accompanied by diplopia, blurred vision, and neck pain, which subsequently resulted in failing consciousness for 18 days without any treatment. She was then sent to a neurosurgeon in England, where a ruptured intracranial aneurysm was repaired.

Two to 5 years later, the patient developed intermittent continually lateralized headaches, accompanied by nausea, occasional vomiting, photophobia, phonophobia, and occasional facial numbness, that would last 2 to 3 days, with a frequency of four per month. She treated these episodes of headache with sumatriptan and naproxen.

The patient currently has a diagnosis of migraine with and without aura, with response to triptan, analgesic, and antinauseant therapies.

The patient was followed up on a yearly basis, participated in clinical trials, and remained in relatively good health.

Questions on Case III

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

- What is the significance of migraine headache after a cataclysmic event?
- How predictive is an aneurysm to the development of migraine headache?
- How responsive is post-aneurysm migraine headache to triptan therapy?

Case III Discussion

Once again, we are faced with the patient who presents with lateralized headache, but this time, after an intracranial bleed and repair of an aneurysm. The incidence of persistent post-aneurysm surgery migraine headache is well defined in the literature. Headache frequency following radiosurgery techniques (coil) is somewhat reduced, but is definitely still a problem in postoperative manage-

ment. An aneurysm or AVM history is predictive of migraine headache.

Questions remain regarding the responsiveness of such post-aneurysm, post-AVM surgery headaches to triptan therapy, and of the safety for treating clipped aneurysms with vasoactive medications. The available clinical data do not permit conclusions on these two management strategies. However, it appears that this patient, in particular, is exquisitely responsive to triptan therapy, assuming that her migraine headaches are related to prior aneurysm surgery.

As indicated in the two prior cases, it is imperative that follow-up scanning be obtained to delineate the success of the surgery before attempting triptan therapy.

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Editorial Comments

Dr. Goldstein takes the views that "consistently lateralized headache" warrants an imaging study. The American Academy of Neurology Quality Standards Subcommittee, following a systematic review of the efficacy of imaging, concluded that the likelihood of routine CT or MRI scanning finding pathologic intracranial structural lesions is extremely low. The official recommendation is to not scan patients with conventional migraine, unless there is a history of seizures, a focal examination, or a recent change in headache pattern (Report of the Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter: the utility of neuroimaging in the evaluation of headache in patients with normal neurological examinations [summary statement]. Neurology 1994;44:1353–4). In

Case I, there was a dramatic change of pattern, with a sudden abrupt worsening, and all neurologists would recommend a scan urgently. In Case II, there may have been a subtle change in pattern, with new and more persistent lateralized headache unresponsive to low-dose sumatriptan. In this case, the need for a scan was less clear. In Case III, scanning was done after the initial ictus, but had to be repeated to ensure there was no neurosurgical change following surgery with the presentation of the postsurgical migraine. Thus, follow-up scanning in patients with change in headache postsurgery is also warranted. These cases illustrate that guidelines, no matter how carefully crafted, do not necessarily cover all clinical scenarios, and after all are only "guidelines" and do not replace good clinical judgment and individual consideration of each patient.

FINAL DIAGNOSES:

Case I, Arteriovenous malformation
Case II, Middle cerebral artery aneurysm
Case III, Post-aneurysm resection migraine without aura