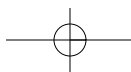
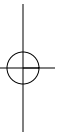
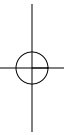


PART II

SECONDARY AND RARE HEADACHE DISORDERS



CHAPTER 25

THE PATIENT WITH THE CHANGING HEADACHE

JOHN S. WARNER, MD

Case History

A 31-year-old lady came to the headache clinic with a history of migraine that had started early in childhood. During the previous 10 years these headaches occurred three times each month and sometimes lasted for 24 hours. The headache might start at anytime during the day or night, and she stated that she might see spots for 2 days prior to these headaches. No other prodrome had been noted and she denied an aura. The pain started in the right temple and spread to the right half of the head and neck, sparing the face, and had a throbbing sensation when at its maximum. Nausea, vomiting, lightheadedness, phonophobia, and photophobia accompanied most headaches. The pain was accentuated by cough, strain, or movement. Prior to her hysterectomy, there had been no relationship of these headaches with menses. "Migraine" never occurred on the left side. A magnetic resonance imaging scan 2 years earlier was reported to be normal. Ergotamine/bellafoline/phenobarbital tablets and metoprolol failed to prevent these headaches.

A different headache began 2 months prior to her initial visit to the clinic. These headaches were daily and nocturnal during the first 2 weeks. Thereafter they occurred as often as four times each day or night without a true circadian pattern. She noted the sudden onset of intense pain in the right temple and face spreading rapidly into the neck. She described this as "like a knife that is continuously being pushed deeper into the head," more intense than the previous migraine, and without the prodrome or throbbing sensation. This pain would last for 10 to 15 minutes, rapidly subside to become a minimal dull discomfort for the next 2 to 3 minutes,

then suddenly return as the intense pain. These episodes of repeated intense 10 to 15 minute pains, interrupted by 2 to 3 minutes of discomfort, might last for 45 minutes or continue as long as 7 hours. Vomiting often accompanied these headaches and the patient noted unilateral photophobia and scalp tenderness on the right. Movement of the eyes increased the pain, and pressure on the right orbit would provide partial relief. The pain was unrelated to position and did not change with cough or strain. During each of these headaches, she experienced ipsilateral conjunctival injection, increased lacrimation, nasal obstruction, and swelling of the face. Frequently, she experienced the sensation of her heart having skipped a beat.

The past medical history was otherwise noncontributory. Two days earlier she had been started on verapamil 80 mg t.i.d. and prochlorperazine 25 mg p.r.n. which provided partial relief. The neurologic examination was performed when she was headache free and it revealed no abnormalities.

Questions about This Case

- Were her earlier headaches migraine?
- How do you classify the more recent headaches?
- What neurodiagnostic studies would you request?
- What therapy would you prescribe?

Case Discussion

This case study addresses the problem of changing patterns of headache, and occurrence which frequently precedes a patient's initial visit to a primary care physician

or referral to a headache specialist. Most often, the change in the headache presents no immediate or urgent risk to the patient's future health. Exceptions include the warning or sentinel headache of a bleeding intracranial aneurysm or the initial headache of bacterial meningitis, conditions that often escape appropriate or timely evaluation.

Changing pattern of headache occurs in three forms. The first consists of changes of features of one headache type over a period of time. These changes are common in migraine and are revealed by taking a careful history from the patient or by comparing the notes from recent office visits to the notes of previous visits. A migraine might originally begin in the temple and then in later years start with pain in the upper neck and, in both instances, spread to become a unilateral headache. It is not unusual for a person to have migraine without aura prior to menopause and to have auras without headache after menopause. These changes in pattern of a single headache usually do not disturb the patient nor present a problem to the physician.

The other two forms of changing pattern of headache involve the development of an entirely different headache type. During their lifetime, most people will experience different types of headaches. Rare is the person with an alcoholic hangover who has not had at least one previous tension-type headache. That individual almost always recognizes the cause of and method of avoiding the intense early morning headache and vomiting which follows the prior evening's overindulgence. Although they may experience repeated hangover headaches, they almost never seek medical attention for this combination of headache.

Changing pattern of headache might consist of changing from one type of primary headache to another primary headache disorder as occurred in the patient whose history was cited at the start of this case history. Many patients with migraine following puberty will have noted tension-type headache prior to puberty. At times they have difficulty recognizing the onset of each of these primary headaches, and some authors question whether tension-type and migraine represent two extremes of only one condition.

On the other hand, changing pattern of headache might occur in a patient having an initial primary headache disorder and later developing a "secondary headache" caused by toxic, metabolic, infectious, vascular, neoplastic, and other conditions. Further into this discussion, it will be shown that the patient whose history was presented went on to develop two different secondary headaches and later had two additional pain-in-the-head and face problems, a total of six distinct conditions.

At the end of the case history, four questions were raised and this is an appropriate place to discuss them. The patient's prior headaches were migraine, fulfilling the International Headache Society's rigid definition of migraine without aura. She had noted more than five headaches of greater than 4 hours' duration. The pain was pulsating, unilateral, and accentuated by movement. Nausea, vomiting, photophobia, and phonophobia accompanied these migraine attacks. There is no indication for neurodiagnostic studies for migraine attacks of this duration. In the past, the patient had seen numerous physicians who had given various medications to abort the migraine attacks, but she had never received satisfactory treatment to prevent the episodes.

Her more recent headaches were episodic, unilateral, and accompanied by ipsilateral changes of the autonomic nervous system. Three headache syndromes present with these features. The first is true cluster headache, a condition that usually occurs in males. The pain of cluster headaches is periorbital, excruciating, lasting from 15 to 120 minutes, and rarely occurring more than four times a day. This patient's headache duration was shorter, lasting only 10 to 15 minutes. The attacks occurred more frequently, and were more widespread. There was no circadian pattern, a feature that is often noted with cluster. Thus, the more recent headaches which this patient was experiencing are not cluster headaches.

A second unilateral headache with ipsilateral autonomic features is the short-lasting neuralgiform headache with conjunctival injection and tearing (SUNCT) syndrome, an extremely uncommon condition which thus far has been described in less than 30 Europeans, mostly male, Scandinavian, and over the age of 47 years at the time of their initial attack. The pain in SUNCT syndrome lasts less than 120 seconds and is reported to occur as often as 30 times an hour. Interestingly, the SUNCT syndrome has never been recognized in North Americans of pure Norwegian or Swedish descent.

The third cause of paroxysmal headache with ipsilateral changes of autonomic nervous system function is the primary headache disorder initially described as chronic paroxysmal hemicrania (CPH) but later recognized as occurring in an episodic pattern. This condition occurs at less than 10% of the incidence of cluster headache and less than 1% of the incidence of migraine. The typical patient with CPH is a female with unilateral headache of 20 minute duration that occurs at least 11 times each day. The intensity and location of the maximum discomfort of this unilateral pain varies from one patient to the next. This is in contrast to the more prolonged, less frequent, excruciating periorbital pain of a cluster headache, the latter sparing the parietal, occipital, and vertex area. Characteristically the headache of CPH responds to indomethacin.

To date, there is no satisfactory explanation for the neurophysiologic changes of CPH nor any understanding of the neurochemical changes that probably are occurring. Isolated reports of somewhat similar headaches secondary to organic disease have been published in recent years. Therefore, it would be appropriate to order magnetic resonance imaging and a platelet count for patients suspected of having CPH.

This particular patient's headache pattern was unusual for CPH in that she had flurries of intense headaches each day, each 10 to 15 minute pain separated by brief episodes of minimal pain at the same site. These flurries occurred daily and would last for as long as 7 hours.

Her headaches were classified as CPH presenting in the episodic pattern. She was started on indomethacin 75 mg t.i.d. at the first visit and experienced almost immediate relief. She was pain-free the following 2 days. At noon on day 4, she noted the gradual onset of a generalized throbbing headache with subsequent vomiting, the pain accentuated as she flexed her neck. The headache and vomiting continued throughout the night and she returned to the clinic on the morning of day 5. The presumptive diagnosis at that time was a generalized headache secondary to indomethacin therapy. A lumbar puncture was performed to exclude meningeal infection. Acellular fluid with normal protein and sugar was obtained, the opening pressure being 12 cm. The indomethacin was stopped and the generalized headache ceased, only to be followed the next day by typical post-spinal headache which required a blood patch on day eight. These latter two iatrogenic headaches appeared during a one week interval and represented an example of primary headache(s) changing to secondary headache(s). Fortunately, the etiology of each of these secondary headaches was immediately recognized and appropriately treated without prolonged suffering by the patient.

Her episodes of paroxysmal hemicrania returned 1 week after the indomethacin was terminated. Verapamil 120 mg t.i.d. provided relief, and after 1 month, was discontinued. During the following 18 months she had migraine headaches about once every 3 weeks lasting 2 to 3 days. She was not on prophylactic therapy and was using only simple analgesics. After having been quiescent for 18 months, the right sided headache of CPH returned. The location of the pain enlarged; the pain spread into the tongue and throat. Initially, the episodes were daily but over the following 4 weeks these increased to three per day. The usual episode lasted 1 or 2 hours during which time she noted intense pain for 10 to 15 minutes followed by minimal pain for 10 or 15 minutes, then a return to the intense pain. Ipsilateral autonomic changes accompanied each intense pain. These attacks ceased when she restarted verapamil 80 mg t.i.d., a drug that she continued for only 3 weeks.

Eleven months later, she was injured in a motor vehicle accident. During the following 3 months, she had constant daily, dull, generalized headache and more intense generalized migraine-like attacks 3 days each week. She was taking various over-the-counter analgesics at least four times each day. When she returned to the clinic with this history of 3 months of constant post-traumatic headache, she was told that these were a typical pattern of analgesic-induced or analgesic-rebound daily headache following trauma. She was instructed to stop all analgesics and her daily continuous headaches ceased approximately 1 week later.

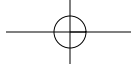
Four months later, she was seen at another medical center for right-sided jaw, ear, and temple pain of 6 months' duration, a complaint that she had not voiced at previous visits. Surgery was performed on the right temporomandibular joint following which she noted some relief of the right-sided facial pain.

When last seen at the headache clinic, she reported the resumption of the pattern of one to three migraine headaches each month. In addition, she was noting frequent episodes of paroxysmal hemicrania with autonomic changes lasting for 3 to 4 hours each. Interestingly, she insisted that these latest attacks had shifted to the left side. Verapamil was again prescribed and she again failed to return for follow-up visits.

Since her last visit, she has not returned to the headache clinic. Attempts to contact the patient by phone and mail have been unsuccessful. In reviewing her records from the prior 3½ years, it is noted that she had experienced every form of changing headache pattern. Her migraine had changed from right to left. A second primary headache problem, namely chronic paroxysmal hemicrania in its episodic presentation, had appeared and later shifted to the other side. She had experienced iatrogenic indomethacin induced headache and lumbar puncture headache. Following a motor vehicle accident, she developed analgesic induced headache. Lastly, she developed what another center diagnosed and treated as temporomandibular joint pain. There is no record of simple tension-type headaches.

The lesson to be learned from this case is that over time, there are usually changes in headache pattern in a given individual. The physician encountering the headache patient must always carefully inquire as to the different types of pain that the patient might be experiencing. This has to be followed by specific questioning regarding the date of initial onset, frequency, duration, character of pain, and the accompanying features of each headache type. The proper therapeutic approach to each headache type often differs.

Portions of this patient's history were included in Case Histories from The Vanderbilt Headache Clinic #15. Headache Q 1994;5:159-60. It is reprinted with permission of the editor.



Editorial Comments

All physicians caring for patients with headache will fully appreciate this case from Dr. Warner. Most patients with headache have more than one type, and it is well known that migraine presents with multiple subtypes in many patients. Variability is common in headache as are changing patterns.

It may seem that this patient has a remarkable array of headaches, and she does, but with careful historical inquiry and appropriate therapy for each subtype or type of headache, she can be effectively managed, as outlined by Dr. Warner.

We should all be vigilant to detect the changing headache as it could represent a new organic problem, even an iatrogenic one.

