Chapter 38

The Glassblower Who Couldn't Blow

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Case History

A 24-year-old student and avid glassblower complained of daily headache exacerbated by pursuing her glassblowing hobby. She first developed headache at around the age of 17 years, following removal of her wisdom teeth. These headaches consisted of infrequent nonthrobbing generalized headache lasting about 24 hours, with no associated nausea, photophobia, or phonophobia. These headaches increased in frequency to once a week, and in her early twenties, were described as a steady occipital and vertex pain, which involved both temporal regions and which radiated down her neck. At this time, her headaches were exacerbated by coughing and straining. For some years, it had been difficult for her to attend the hairdresser, because the neck extension that she would undergo at these times caused headache exacerbations. Approximately 6 months prior to her presentation to the headache specialist, her headaches had become continuous and daily. She gradually developed intractable nausea, and became almost bedridden. She had to give up her university studies as well as her glassblowing. In addition to her headache, she described some unusual symptoms, which included intermittent hearing loss, bilateral arm numbness, and at times, some difficulty with walking downstairs.

She was taking ketorolac and amitriptyline daily. In the past, numerous migraine prophylactic medications had proved unhelpful, and all symptomatic medications had been stopped for over a month because of concerns about rebound headache, but this had not produced any improvement. Triptans had not been of significant benefit to her.

Physical examination showed no definite focal neurologic signs. Extraocular movements were normal, and coordination was normal except for some very questionable reduction in rapid alternating movements in her nondominant left hand. Examination of her neck showed some limitation in neck rotation. Neck muscles were diffusely tender, but there was no radiation of this pain on palpation.

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 most likely diagnosis, and what would be an appropriate differential diagnosis?
- Are her headache precipitants (coughing, straining, neck extension) of diagnostic significance?
- Do her associated symptoms of intractable nausea, intermittent hearing loss, bilateral arm numbness, and some difficulty with walking on stairs assist in differential diagnosis?
- What investigations should be done?
- What are the treatment options in this case?

Case Discussion

Diagnosis

This patient presented with a very disabling headache syndrome, which had basically put her in bed for much of the time over the several months prior to consultation with the headache specialist. Up to that point, many clinicians who had seen this patient felt the most likely diagnosis was transformed migraine with medication overuse. The prominent nausea in her case would indeed suggest a diagnosis of migraine, and at the time of presentation, she was taking ketorolac on a daily basis. However, she did not seem to have headaches consistent with intermittent

migraine prior to the onset of chronic daily headache. This is not a prerequisite for a diagnosis of transformed migraine, but is often present. Although this patient did have intermittent headaches prior to the development of chronic daily headache, these headaches did not have sufficient associated symptoms at that time to be diagnosed as migraine. A trial of stopping her analgesics had been done in the past, and this had provided no relief, although one could argue that a month is not long enough to definitively exclude medication-overuse headache. Her headaches had however proved especially intractable to treatment, and had pursued a progressive course.

In a patient with intractable headache, who obtains some relief from lying down, a diagnosis of intracranial hypotension secondary to a cerebrospinal fluid (CSF) leak should also be considered. This diagnosis was not considered seriously in this patient because of her other clinical features and diagnostic imaging findings. The response of her headaches to standing up or lying down was not dramatic, and she clearly had intractable headache even while supine. A gadolinium-enhanced magnetic resonance imaging (MRI) scan to exclude dural enhancement, and a lumbar puncture to measure CSF pressure could have been helpful in excluding this diagnosis, although they were not done in this patient.

This patient did have features to suggest that her headaches might be the result of a Chiari type-I malformation (CM-I). She clearly described coughing, straining, and certain neck movements as major headache precipitants. These are very consistent with headache secondary to a CM-I. Unfortunately, she did not have typical short "hindbrain hernia" headaches in response to these precipitants. Had she experienced such headaches in the past, this likely would have been a clear diagnostic indicator for the physicians who had been taking care of her. The location of her headache is also important, in that it had a strong occipital component, although it did involve the vertex and the temporal regions as well. Headaches related to Chiari malformations can involve any part of the head, but a major occipital component is usually present. Radiation of the headache down her neck is also consistent with Chiari malformation. Her unusual associated symptoms are also of significance. Headache-associated symptoms related to hearing abnormalities and numbness in the face or arms have been described in patients with headaches secondary to CM-I.

In summary, there were several features in this patient's history, which suggested that a CM-I should be looked for, and that she did not suffer from a primary headache disorder. These included the following:

- The prominent occipital component to her headache, and its progressive nature
- The unusual associated hearing-related and sensory symptoms
- The marked exacerbation of her headaches by coughing, straining, and neck extension

The lack of typical short hindbrain hernia headaches lasting a minute or two in her history in response to coughing or straining was somewhat unusual, as was her presentation of chronic daily headache. These factors no doubt contributed to the delay in definitive management of her case.

Investigations

In a patient with a marked degree of disability and whose headaches do not fit well into one of the primary headache categories, a brain MRI scan should certainly be considered. It is also the primary investigation for ruling out a CM-I as a contributing cause to the patient's symptoms. This patient had in fact had a cervical spine MRI scan done over a year previously, primarily because her physicians were considering a cervicogenic cause for her headaches. This MRI scan had shown a CM-I, with the inferior margin of the cerebellar tonsils reaching to the inferior aspect of the first cervical vertebrae. However there was controversy as to whether the CM-I was indeed a major cause of this patient's headaches. At that time, facet joint injections were booked for her, to help exclude a cervical spine pain generator as a cause for her headaches. These were never carried out. A brain MRI scan done over a year after the first cervical spine MRI scan showed herniation of the cerebellar tonsils 1 cm into the spinal canal (Figure 38-1). However, the radiologist commented that adequate CSF flow was present both anteriorly and posteriorly on

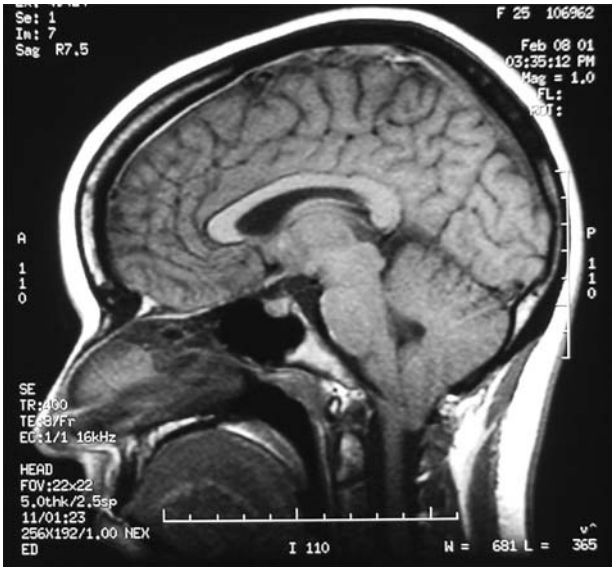

Figure 38-1. Brain magnetic resonance image from our patient showing a Chiari type-I malformation, with at least 10 mm of tonsillar herniation below the foramen magnum.

cine MRI. At that point, her physicians were still uncertain as to whether or not the CM-I was the cause of this patient's headaches. However, this patient's later clinical course would suggest that the reluctance of her physicians to accept the CM-I as a major contributor to her headache syndrome, given her overall clinical picture, was unwarranted.

Management Strategies

This patient had intractable disabling headache, a well-documented CM-I, and a number of symptoms which suggested that the CM-I was significant in the generation of her headaches. The main issue is at what stage surgical management of the CM-I should be pursued. While the neurosurgical consultation was being pursued, she was tried on indomethacin, as there is some evidence that this nonsteroidal anti-inflammatory drug may be particularly helpful in patients with a CM-I. In addition, she was placed on flunarizine, a migraine prophylactic she had not tried previously. When these were not helpful, a long-acting oral morphine preparation was tried. None of these medical approaches were successful and she underwent a posterior fossa decompression and reconstruction. This involved a posterior fossa craniotomy, removal of the arch of C1, and a duraplasty, with a cadaver dura graft. The occipital bone flaps were then replaced in a more vertical position and were fixed with two bioplates.

Following the surgery, she had considerable headaches for 2 months postoperatively, although she felt these were different from her previous headaches. Her headaches then improved dramatically, and by 3 months postoperatively, she felt she was some 90% better. On final follow-up 18 months after her surgery, she was having approximately one headache day a month without nausea or photophobia, which responded well to a few acetaminophen tablets. She was doing glassblowing again, was back at University full time, and considered herself normal. Coughing, straining, and in particular glassblowing, no longer precipitated headache.

In retrospect, this patient's CM-I should have been seriously considered as a cause for her headache much sooner and definitive therapy offered.

On the other hand, there are many patients with CM-I and headache whose headache syndromes are not severe enough to warrant surgery, and who can be managed satisfactorily medically. However, the headaches that these patients experience do change over time, and for these patients, surgery is always an option should their headache syndrome progress.

Case Summary

Our patient was a young woman who gradually developed intractable headache with severe nausea, which caused severe disability. Although she had a history of intermittent headaches for some years prior to the onset of chronic daily headache, these headache attacks did not meet migraine diagnostic criteria. Although her CM-I was discovered relatively early once her headaches became progressive, its significance with regard to her headache syndrome was not appreciated, and a number of other differential diagnoses were pursued. She illustrates that although a CM-I may be asymptomatic in some patients, it should be taken seriously in the appropriate clinical setting in a patient with a difficult headache problem. Medical treatment options can be helpful for some of these patients, but when these fail, surgical options should not be excessively delayed.

Overview of Chiari Type-I Malformations and Headache

Chiari Type-I Malformations

A CM-I is present when the cerebellar tonsils herniate 5 mm or more through the foramen magnum, although some authors consider 3 mm to be significant. Approximately 0.5 to 0.7% of patients undergoing MRI in neurologic centers are found to have CM-I. The degree of tonsillar herniation present in any individual patient with CM-I likely should not be overemphasized. It has been suggested, for example, based on successful surgery for syringomyelia, that disturbances of CSF flow can occur at the foramen magnum secondary to tonsillar crowding without evident tonsillar herniation. In fact, the tonsillar herniation typically found in a CM-I is likely secondary to a congenitally small posterior fossa, which cannot accommodate the cerebellum, brainstem, and the usual volume of the CSF. It must be kept in mind however that some degree of tonsillar herniation may occur in patients with CSF leak and low-pressure headaches. This can imitate a true CM-I, although such patients will also usually show sagging of the brainstem against the clivus, and enhancement of the dura on gadolinium-enhanced MRI studies.

Spectrum of Headache Syndromes Associated with Chiari Type-I Malformations

Headache occurring in association with CM-I is well recognized. Typically, these take the form of the classical hindbrain hernia headache, also called cough headache, with a brief severe occipital or global head pain lasting seconds or minutes after a provoking factor, usually a Valsalva such as straining or coughing. In addition, a spectrum of headache types has been described in association with CM-I. Given the high prevalence of primary headache types in the general population, such as migraine and tension-type headache, determining

whether a patient's CM-I is causally related to the patient's headache can be problematic. Chronic daily headache has been described in association with CM-I. In one patient series, 8 of 20 patients with CM-I and headache had continuous head pain, although this did fluctuate in intensity from time to time. Three of these patients with continuous headaches and CM-I had posterior fossa decompressive surgery, and two had a good result. There are some potential clues that may indicate whether a CM-I is contributing to the patient's headache symptoms. One of these is the presence of typical "hindbrain hernia" precipitants for headache exacerbations. These include sneezing, coughing, bending over, laughing, head turning, straining, and exertion. As a result, one could give headache syndromes in patients with CM-I a variety of interesting names. For example, our patient described here had what might be best described as "hairdresser headaches." Neck extension at the hairdresser had for many years triggered problematic headaches for her. Another of our patients regularly developed headaches when she would watch her children play hockey. Shouting for prolonged periods while her children scored goals would bring on a severe headache, which might best be termed "hockey headache" or perhaps "yelling headache" as this patient with CM-I described them. Many patients with CM-I will also describe exertion as the prominent trigger for their headaches. A third patient of ours found that she could cross-country ski without developing symptoms, but if she put on a burst of speed to pass someone on the ski track, this would regularly bring on a headache, which might be described as "cross-country ski headache." The same patient with a CM-I found she had to stop playing tennis, because bending over to pick up the tennis ball would regularly bring on a troublesome "tennis headache."

Another potential clue to the presence of a CM-I in patients with headache, is the presence of unusual associated features, such as visual symptoms suggesting oscillopsia, blackouts, trouble walking downstairs, imbalance, numbness in the face, palate or arms, or intermittent hearing loss or tinnitus. Finally, some patients with CM-I and headache experience prominent radiation of their headache into the neck, shoulders, and upper back and interscapular area. It must be remembered that although the pain usually involves a prominent occipital component, the maximal head pain may be elsewhere, even frontally.

Nausea can also be prominent in patients with CM-I. However, as can be seen, given the frequency of nausea in migraine, and also the fact that many patients with migraine experience pain on head shaking or other head movements, there is considerable overlap of symptoms between patients with CM-I and headache and patients with primary headache syndromes.

Pathophysiology of Headache Associated with Chiari Type-I Malformations

How a CM-I results in headache in some patients is not clear. The pathogenesis of these headaches likely involves interference with normal CSF dynamics. With the CM-I, there are complex alterations in the CSF flow at the foramen magnum during the cardiac cycle, and these have been reported to normalize with surgery. Normally, CSF flows caudally through the foramen magnum during systole as the stroke volume from the heart reaches the brain, and this flow reverses during diastole. The foramen magnum is also an active region of CSF flow during straining. As intrathoracic pressure rises, this is transmitted through the valveless venous system to the epidural veins and eventually to the spinal CSF. Cerebrospinal fluid is thus rapidly displaced from the spinal canal to the head. As intrathoracic pressure drops following straining, the flow of CSF is reversed and flows from the head to the spinal canal. In the presence of a CM-I, the cerebellar tonsils become impacted as the CSF pressure in the spinal canal drops following straining, and this return of CSF from the head to the spine is therefore partially blocked. The relevance of these transient increases in intracranial CSF pressure to headache production is not clear. More likely perhaps, the head pain is generated by local factors in the region of the foramen magnum, perhaps related to pain-sensitive structures in the arachnoid and blood vessels around the cerebellar tonsils and the upper spinal canal. Interference with cervical nerve roots has also been implicated. Perhaps combinations of these mechanisms are active to produce headache in patients with CM-I. This might help to explain the wide spectrum of headaches that have been observed clinically in association with this disorder.

Recommendations

Much more research is needed to elucidate the relationship between CM-I and headache. However, the following seems reasonable, given our current state of knowledge:

- In patients with problematic headaches that do not fit well into the well-defined primary headache categories, consider other etiologies such as a CM-I.
- A strong history of headache-exacerbating factors in a patient consistent with some of those observed in headache related to CM-I, such as sneezing, coughing, bending over, head turning, laughing, exertion, and straining, should raise suspicion of a possible CM-I.
- The presence of unusual headache-associated symptoms such as oscillopsia, blackouts, trouble walking downstairs, numbness of the face or arms, and intermittent alterations in hearing should alert the clinician to the possibility of a CM-I.
- A prominent occipital component to the patient's head pain, and with significant radiation of the pain

into the neck, shoulders, and upper back, may be a diagnostic clue.

- In patients with disabling headache disorders, including chronic daily headache, the presence of a CM-I should be taken seriously, and if other therapeutic modalities are not successful, then the patient should be considered for possible surgery. Most patients with problematic chronic daily headache syndromes do not have CM-I. It is therefore important that patients with chronic daily headache and CM-I be carefully evaluated by physicians expert in headache; and if invasive treatments are undertaken, the results of these should be carefully documented, as much more needs to be learned about this area.

Selected Readings

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Editorial Comments

When it comes to headache and CM-1, several problems arise for the clinician. First, it can be a very difficult diagnosis to make, especially if the patient also has a primary headache disorder or chronic daily headache, and if a clear history of classic features or exacerbating triggers is not forthcoming, or if there are no telltale signs. Cough headache should always be worked up with an MRI for Chiari malformation before assuming that it is primary. Secondly, once the diagnosis is sorted out from other CM-1 mimics, such as low-pressure headache, it can be difficult to decide whether surgical intervention is warranted and when to intervene. On balance, this is a clinical scenario where the combination of a good neurologic diagnostician along with an experienced neurosurgeon and neuroradiologist is essential for a favorable outcome. Read this case carefully—it teaches us a lot!

Final diagnosis:

Headache secondary to Chiari type-I malformation