TEACHING CASE: CHIARI TYPE I/CEREBELLAR ECTOPIA HEADACHES: COMPLETE RESOLUTION FOLLOWING POSTERIOR FOSSA DECOMPRESSIVE SURGERY

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CASE PRESENTATION

A 21-year-old woman presented to our neurology outpatient clinic with a 9-month history of headaches. No preceding event was identified before the onset of the headache such as trauma or infection, though she had visited a dentist for a dental procedure as well as had visited a chiropractor a few times for the symptoms she was presenting to us with. The exact day of onset of headaches was not clear. Her headaches, over the period of time, had been progressively getting worse. She described a primary headache that was daily, would be present when she woke up and gradually get worse as the day progressed, fluctuating from a mild to a severe headache by the end of the day. It was described as a dull headache that was primarily present bilaterally in the frontal as well as the occipital regions. She could not identify any particular triggers that would make the headache worse. She denied having any pain-free days. She denied its relationship with exercise, food, amount of sleep, menstrual cycles or stress. She could not identify anything that made it better including any over-the-counter medications though ibuprofen sometimes partially helped. She denied any associated symptoms of lacrimation, conjunctival injection, rhinorrhea, photophobia, phonophobia, nausea, vomiting, visual changes or vertigo. No auras had ever been reported. She denied other symptoms such as weight loss, appetite changes, fevers, chills, hearing changes, incontinence, syncopal episodes, paresthesias, episodic paresis or plegia or visual symptoms.

Associated with these headaches was another type of headache which was usually unilateral, sudden onset, sharp and stabbing lasting usually no more than 5 min. These were associated with some nausea, photophobia, and phonophobia and were severe enough to interrupt whatever the patient was doing at that time. These usually had been infrequent but could happen up to 3 times a day.

The patient had a medical history significant of scoliosis since she was teenager though she never had any surgery nor did she feel that her symptoms of backache had worsened in the last few months. She was currently taking oral contraceptives but noted that her headaches had preceded the starting of these pills and she had not noticed any change in her symptoms in relationship with the medication. She denied any other prescribed or alternative medication. She had no history of any drug abuse. She did not consume alcohol or tobacco in any form. Her caffeine intake was minimal.

On exam, her blood pressures were normal with a normal rhythmic pulse. Her temporomandibular joint examination showed asymptomatic clicking and popping on the left. Percussion on sinuses was negative. She did have some occipital tenderness. Her pupils were normal and her fundi were benign. The cranial nerve exam, motor exam, sensory exam, cerebellar exam, reflexes, and gait were unremarkable and normal.

Considering a dual diagnosis of chronic daily headache and idiopathic stabbing headache, she was started on indomethacin and nortriptyline. Unfortunately, due to side effects, she did not tolerate nortriptyline and that was discontinued. After a few weeks, indomethacin was also discontinued. After no benefit from these medications and no change in her symptoms, she was started on propranolol and brain imaging was planned with a magnetic resonance imaging (MRI). Laboratory data were also obtained. Her MRI showed low lying cerebellar tonsils of about 3 mm, though not meeting the criteria of Chiari type I malformation (CM1). Incidentally, the cervical MRI also showed a syrinx at C6-C7 level.

A lumbar puncture was performed and her opening pressure was normal. Her laboratory data revealed a normal white count, no anemia, normal platelets, normal prothrombin and partial prothrombin time, normal sodium, chloride, potassium, bicarbonate, blood urea nitrogen and creatinine, glucose, calcium, protein, normal liver function tests, normal sedimentation rate, angiotensin enzyme level, normal thyroid function tests, cyanocobalamin level and normal antinuclear antibody (ANA) and lyme titers. Her
Cerebrospinal fluid (CSF) analysis showed normal protein, normal glucose, 2 white blood cells with no red blood cells and negative for xanthochromia, myelin basic protein and oligoclonal bands.

Over the course of the next 5 months, the patient was tried on multiple medical therapies. Propanolol did not help either and she was switched to topiramate with triptan use as rescue for the stabbing headaches. After failing to find any relief, she underwent greater occipital nerve blocks, which failed to provide any relief either. She was tried on tizanidine and the extended release preparation of indomethacin, which seemed to decrease the number of unilateral stabbing-pain episodes. Her chronic daily headaches continued to increase in severity. She also started to experience new episodes of pre-syncpoe where she would start to experience lightheadedness, feeling of passing out and would have put her head down for 15 min with resolution of her symptoms. These episodes, although infrequent, remained persistent despite the therapy.

At this point, she was thought to have Chiari type I of headaches, despite not meeting the radiological criteria of the syndrome. Being refractory to medical management, she was referred to neurosurgery for evaluation of posterior fossa decompressive surgery. She underwent the surgery and 9 months out has remained headache and symptom-free and is off all medication (Figs. 1-3).

**EXPERT COMMENTARY**

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There are many important clinical issues in this case that deserve discussion. First is the initial diagnosis. In evaluating patients who present with headache, it is imperative that the clinician excludes a secondary headache disorder before
assigning a primary diagnosis. A useful approach then is to look for warning signs or “red flags” that suggest the presence of secondary conditions. These ominous signals may be recalled by using the mnemonic SNOOP, which lists: Systemic symptoms such as fever, myalgias, weight loss; Secondary risk factors such as cancer or HIV; Neurological signs or symptoms such as mental status changes, or focal deficits; Onset of headache (sudden, severe, thunderclap, or those triggered by coughing, straining, exertion or sexual activity); Older age of headache onset; and Prior headache history because a new onset or change in an established headache pattern should be a cause for concern. It is also important to investigate patients in whom the features of the headache are atypical for that of a primary headache disorder.

This patient was initially diagnosed with 2 distinct primary headache disorders (although neither is an actual International Classification of Headache Disorders, 2nd edition [ICHD-II] diagnosis), chronic daily headache and idiopathic stabbing headache, but should she have been? We are told that this patient has no prior headache history (she is young so that in itself is not necessarily worrisome), but also a relatively new onset of headache, and headaches that have been progressively worsening. These headaches are bilateral, of varying intensity and without any associated features. The patient is unaware of any precipitating factors. Although she could not recall the exact day of onset, she tells us that she has never been pain-free since its onset. She also describes a unilateral headache of short duration that is associated with nausea, photo-, and phonophobia that would recur up to 3 times daily. Two important issues should now be evident. First, this patient’s history should have raised several “red flags” and second, the headache characteristics are quite atypical for either of the diagnoses given and therefore a more thorough investigation should have been ordered.

If we focus on the diagnosis of “idiopathic stabbing headaches,” there are clearly atypical features. The ICHD-II criteria for primary stabbing headaches mandate that these headaches last up to a few seconds, have no accompanying symptoms, and are not attributable to another disorder. This patient’s headaches are longer-lasting, have associated features and she had not been previously neuro-imaged. The diagnosis chronic daily headache is not an ICHD-II term, although it is used to designate any of the primary headache disorders that occur with a frequency of greater than 15 days monthly. Here, too, further investigations were warranted.

Another cause for concern was initiating treatment before establishing a diagnosis, especially when using indomethacin. Because indomethacin may lower intracerebral pressure, a clinical response could sway the physician to falsely assume that the diagnosis of a primary headache disorder and miss the secondary headache disorder responsible for the headaches.

Lastly, when the physicians finally ordered an appropriate neuro-imaging procedure, they discounted the significance of their findings. The Chiari malformations are congenital deformities that are thought to arise from intrauterine underdevelopment of the posterior cranial fossa. The resultant crowding of the posterior fossa causes a downward displacement of the cerebellar tonsils through the foramen magnum and into the upper cervical spinal canal. In the CM1, the tonsils extend at least 3 mm below the level of the foramen magnum. In the Chiari type II malformation (CM2), there is descent of the cerebellar tonsils, cerebellar inferior vermis, and portions of the cerebellar hemispheres into the spinal canal along with displacement of the brainstem and fourth ventricle. CM2 is almost always associated with spina bifida and hydrocephalus. Its prevalence is around 0.02% of live births. Although the exact prevalence of CM1 is unknown, it is believed to be a rare disorder, affecting less than 1% of the population, with a slight female predominance.

Many patients with CM1 are asymptomatic, and the deformity is discovered incidentally. Headache is the most common symptom of CM1 although other symptoms can include dizziness, diplopia, dysphagia, nausea, weakness, and ataxia. Pain localization is often occipital-nuchal but may be generalized. Approximately 30% of patients with CM1 report headaches that are precipitated by Valsalva maneuvers such as sneezing, laughing, straining, lifting, or bending over, and approximately 20% of CM1 patients experience cough headache. These cough-induced headaches are characterized by a sudden onset, short-lasting (seconds to minutes), sharp or stabbing pains of moderate to severe intensity, without associated features. CM1 can also be associated with headaches that last up to several days and rarely may cause a continuous headache of fluctuating intensity. The headache of CM1 is by definition a secondary headache and there is no evidence suggesting that this anomaly produces a primary headache disorder.

This patient had headaches that are well described with the CM1 and this case highlights the need for clinicians to be vigilant in their search for secondary headache disorders, especially when the patient presents with atypical features.

The next question that arises in a case like this, in which the hindbrain descent is minimal, is whether surgery is necessary. It is easy for me to be an armchair quarterback having read the resident’s report of complete improvement following the surgical intervention. In truth, however, I am not certain what course I would have chosen in this case. The diagnostic criteria for CM1 are somewhat arbitrary. The most widely accepted definition is a 3- to 5-mm descent of the tonsils below the plane of the foramen magnum. The clinical features and neurological findings seen in CM1 are believed to be the result of mechanical compression of the brainstem at the level of the foramen magnum or through disturbance of CSF dynamics, which over time produces syringomyelia. In a patient such as the one described here, the minimal tonsillar descent (3 mm) would seem unlikely to produce this patient’s symptoms through a compressive mechanism. Since the patient was also noted to have a syrinx in the cervical cord, a more likely etiology of the symptoms may have been as a result of altered CSF dynamics. CSF flow varies with the cardiac and respiratory cycles. Increased flow of CSF into the central subarachnoid space occurs with systole and during inspiration. During diastole and with expiration, CSF displacement occurs in the reverse direction. In patients with CM1, the displaced tonsils crowd the foramen magnum resulting in reduced CSF flow at the craniovertebral junction, and a compensatory pulsatile descent of the cerebellar tonsils is observed during systole. It is this cyclic alteration
in CSF dynamics that may lead to headaches upstream and syrinx development downstream.\textsuperscript{5}

Perhaps a prudent approach in a symptomatic patient who is discovered to have a CM1 would be to look at CSF dynamics via a cine cardiac-gated MRI. If CSF flow is altered, even in the setting of minimal tonsillar herniation, then posterior fossa decompression should be considered. Surgical intervention is associated with complications that include recurrent infection, CSF leaks, and the formation of fluid accumulations and cysts.

REFERENCES


QUESTIONS FOR DISCUSSION

1. What are the differences between Chiari type I and II malformations?
2. What is the proposed etiology of syrinx formation in Chiari type I malformation?
3. What are the typical features of headaches related to the Chari I malformations?