Delirium, Headache, And The Type I Arnold-Chiari Malformation

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Acute delirium can occur in a variety of states including migraine,[1] head trauma,[2] nonconvulsive status epilepticus,[3] and cerebral infarction.[4] Acute delirium accompanied by migrainous headache is much more common in children than adults and is termed acute confusional migraine.[5]

In 1883 John Cleland, a British poet and anatomist, first described the abnormality of the brainstem and cerebellum now called the Arnold-Chiari malformation (ACM).[6] Chiari in 1891and Arnold in 1894 further detailed this anomaly in children. Students of Arnold coined the term Arnold-Chiari malformation in 1907. In type I ACM, the cerebellum extends downward through the foramen magnum. Associated abnormalities can include hydrocephalus and syringomyelia or cavitation within the spinal cord. Type II ACM is characterized by all the features of type I plus elongation of the fourth ventricle into the spinal canal, displacement of the medulla, and crowding of the cervical cord roots into an upward course. Severe hydrocephalus and myelomeningocele are usually apparent. The focus of this review will be the type I malformation.

Misdiagnosis of ACM is common because of the vague nature of the symptoms and the variable course of the disease. The vague and bizarre symptoms might initially suggest a psychiatric disorder.[7] The incidence, although not precisely known, is considered to be between 1 and 2 per 1000 births.[8]

We present a case report of a woman with acute delirium, migrainous-like headache, and the type I Arnold-Chiari malformation.

Submitted, revised, 28 February 1994.

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Case Report

The patient was a 32-year-old white woman who had recently moved to Vermont from Boston. She was married and had 2 daughters, aged 2 and 3 1/2 years. She had worked for the past 10 years as a manager for a large manufacturing company.

She was well until 6 days before her first admission to the Medical Center Hospital of Vermont, when she experienced the rapid onset of numbness and weakness first in her right leg, then her left leg, then her right arm, then her left arm, then her lower face (including an inability to speak words that she knew in her mind). This progression was followed by a severe headache. She went to the emergency department and was treated as having migraine, given meperidine and promethazine with only some relief, and discharged. The headache returned to a manageable level until 3 days before her admission, when it recurred in the same manner. This time she was treated with sumatriptan with very good results. She returned on the day of admission with another severe headache, this time without weakness or numbness but with unsteadiness on her feet and confusion. Now she could speak well but could not think clearly of her words. Ketorolac injection was tried with minimal results. She was admitted to the hospital and treated with nifedipine and naproxyn. A lumbar puncture revealed normal cerebrospinal fluid. Findings on a computerized tomogram of the brain were normal. An electroencephalogram showed diffuse slowing in the left hemisphere. The headache slowly improved, and the patient was discharged on the 3rd day. She returned the next day, again confused with a severe headache, dizziness, and unsteadiness. Again there were no arm or leg symptoms.

Her headache was primarily occipital with accompanying neck discomfort. She described it as alternately constant or throbbing, sometimes stabbing. The neck ache was constant even when the headache resolved. Sitting or standing immediately exacerbated the headache, sometimes severely. A psychiatric consultation was requested

because the neurology resident believed that the condition might represent a schizophrenia or other psychosis.

Mental status examination failed to reveal any affective disorder or anxiety disorder. The patient lacked any psychiatric diagnosis. Cognitive testing, however, revealed rather specific and marked impairments in visual construction, visual memory, and word recall. Her ability to interpret proverbs was entirely concrete. She was able to grasp similarities. Her ability to calculate was not impaired. Her attention was intact. The Minnesota Multiphasic Personality Inventory was administered, and the results were entirely normal. Neuropsychological testing confirmed the above general findings, which suggested a specific frontoparietal defect. Findings from a neurological examination were unremarkable except for some signs of cerebellar dysfunction. The Romberg test was positive with dizziness. The finger-to-nose maneuver resulted in a gross miss of the nose by 8 inches. Her gait was unsteady. Two-point tactile discrimination was intact at 2 mm on the pad of the index finger but was abnormal on the skin of the forearm and the thigh, and she was unable to discriminate between points less than 8 cm apart. Patting was normal, but she could not coordinate rapid alternation between pronation and supination and could not maintain rapid synchronous patting activity. She also had an air of indifference to the obvious defects on examination.

A magnetic resonance scan of the brain revealed a type I Arnold-Chiari malformation with extension of the cerebellar tonsils 5 mm below the foramen magnum.

The patient was discharged with prescriptions for nifedipine and naproxyn, which had improved her headache by at least 50 percent, she said. She remained mildly delirious. The discharge plan called for conservative treatment for 1 year unless symptoms worsened, at which time prompt neurosurgical consultation would be obtained.

Discussion

A host of symptoms and types of headaches have been reported with the type I Arnold-Chiari malformation, including diplopia;[6] severe fatigue;[6] rotatory nystagmus;[6,9] depression;[6] tinnitus;[9] weakness;[6,7] visual changes;[6,7,9,10-12] nausea and vomiting;[7,9] vertigo and dizziness;[7,11] cerebellar dysfunction;[7,11,13] peripheral pain;[7,11] generalized headache;[11,13-16] neck pain;[7,11,14] frontal headaches sometimes worsened by position changes;[7,9] occipital headaches also sometimes worsened by position change or by laughing, coughing, or exercising;[7,9-14, 17-19] no headache;[7,11,13,15,16] migrainous headache;[9,14,19,20] tension headache;[14,19] headache with frontotemporal radiation;[14] trigeminal neuralgia;[10,14] shoulder pain;[11,14] speech difficulties;[9] photophobia;[9] sonophobia;[9] hyperreflexia;[9] and loss of consciousness.[11,12]

Several facts serve to differentiate our patient's symptoms from those of a more typical migrainous type headache. Her pain was occipital, frontal, severe, and accentuated by activity, especially by sitting and standing. She had accompanying neck pain. The peripheral symptoms consisted of numbness and weakness in her arms and legs. Cerebellar findings are also atypical of migraines. Our patient also experienced dizziness and an unsteady gait.

Migraine patients usually have a history of headaches and a family history of headaches.[21] This type of history is often absent from patients with type I ACM, as it was from our patient. The most common headache of type I ACM is an occipital and frontal headache with neck pain. The average age for symptom onset varies from 40 years[8] to 46 years[11] in adults with type I ACM with a range from 21 to 64 years. The most common presenting complaint is pain, which most often takes the form of occipital and frontal headache with neck pain.

Syncope has also been reported with type I ACM.[12] Our patient did not have syncope, but developed a waxing and waning confusional state. For 1 patient who had syncope in the Stovner[11] series, her headache always followed loss of consciousness, and both headache and syncope disappeared after surgical therapy.

Our patient had gait disturbance, which Levy, et al. reported in 43 percent of their type I ACM patients. She had upper extremity weakness, which was also found in 33 percent of type I ACM patients.[19] She manifested cerebellar abnormalities, which Paul, et al.[22] found in 27 percent of their type I ACM patients, a finding consistent with data presented by Susman, et al.[6] and Pascual, et al.[14]

That this case was not migraine was supported by the absence of structural abnormalities associated with migraine.[9] Stovner[11] compared symptomatic type I ACM patients (with headache)

with a group of common migraine and cervicogenic headache patients. Cervicogenic headache patients were much more likely to have had previous head trauma. Type I ACM headache patients were more likely to have neck pain and pain in the ipsilateral shoulder and arm. They (along with the cervicogenic headache group) were more likely to have restriction of neck movement (never found in the common migraine group).

Cervicogenic headache patients had a later onset of symptoms than either common migraine or type I ACM patients. Migraine patients tended to have an earlier onset of symptoms than type I ACM headache patients. Migraine patients reported dizziness much less often and experienced occipital pain less often than did cervicogenic headache patients or type I ACM headache patients. Neither type I ACM headache nor migraine patients in this series experienced headache precipitated by abdominal straining. Type I ACM headache patients never had pain related to the menstrual cycle, which was very common for migraine patients and less common for cervicogenic headache patients. Psychological tension was found more often in migraine patients.[11]

According to Stovner's[11] description, our patient fell into the type I ACM headache category. Stovner reported a discrimination function to classify patients. In his series of patients, his discrimination function was 95 percent correct in classifying patients' headaches into type I ACM headache, migraine, or cervicogenic headache. Using his classifying variables, our patient's headache was classified as resulting from type I ACM.

Migraine sufferers also can generally be distinguished from patients with other types of headache using the MMPI. Elevations of MMPI scales 1, 2, and 3 are often found in headache patients, with increasing elevations in the order of cluster headaches, migraines, combination headaches, tension headaches, posttraumatic headaches, and conversion headaches. Our patient had no elevations on any of these scales.

The cause of the type I ACM headache has been related to a valve-like blockage at the foramen magnum preventing egress of cerebrospinal fluid from cranial to spinal subarachnoid space. This craniospinal pressure dissociation causes pain by distorting sensitive structures in the arachnoid and in the blood vessels.[23] Blood vessel involvement could explain the similarity of type I ACM headaches to migraine headaches. Lower brain stem and upper cervical cord regions are both implicated in migraine and ACM. It is known that stimulation of the trigeminal nerve, locus coeruleus, and dorsal raphe nuclei can induce cerebral and extracranial vascular changes of the same order as those in migraine.

Khurana[9] found a characteristic of ACM is that the patient has symptoms and history suggestive of organic disease which cannot be substantiated by cranial computed tomogram or analysis of cerebral spinal fluid. Clues to the diagnosis include the prolonged, persistent, and progressive nature of the patient's complaints and the subtle but important findings on neurological examination.[6]

From all the above, we conclude that our patient was suffering from delirium and other symptoms secondary to symptomatic type I ACM. The alternative explanation would be acute confusional migraine in an adult. Acute confusional migraine was first described by Gascon and Barlow.[1] The majority of the reported cases have occurred during adolescence.[3,5] Seventy-five percent of patients had a family history of migraines, and 83 percent had a personal history of migraines, neither of which were true in our patient.

Episodes of acute confusional migraine can occur as the initial manifestation of migraine.[5] The attacks can last several hours and are associated with agitated confusion.[3] A headache can precede, occur simultaneously with, or follow the period of confusion, and visual difficulties rather than a headache can be the primary symptoms of an acute attack. Focal neurologic deficits might be noted during the attacks, but they tend to resolve within 24 hours. Results of cerebrospinal fluid studies and computerized tomography are normal. Electroencephalographic recordings during the attacks show diffuse or focal slowing of background activity, as well as frontal intermittent rhythmic delta activity.[3] The patient might have partial or total amnesia of the episode.

Treatment

The symptoms of type I ACM can resolve spontaneously or require treatment, which is usually surgical.[10,19,24,25] The operation of choice is posterior fossa decompression, which usually not only resolves the headache immediately but also treats the syringomyelia in a number of cases.[10] When syringomyelia is not improved with posterior fossa

decompression, a syringoperitoneal shunt can be performed. Success has been reported with a calcium channel blocker. Our patient improved somewhat on nifedipine and naproxen but remained symptomatic. Her cognitive functions have not entirely returned 6 months later.

We suggest that surgical intervention be considered for patients who are clearly worsening despite medical management; who are incapacitated as a result of their symptoms, such as being unable to work or to engage in their normal activities of daily living; or who desire to assume the risks of surgery because their perception of their suffering is sufficiently great.

To our knowledge this report is the first in the literature of type I ACM presenting as acute delirium. The type I ACM condition is important for primary care physicians to remember, as it is encountered in 2 of every 1000 patients and could be symptomatic. The differentiation of ACM from migraine and cervicogenic headache is not always easy, but the guidelines outlined above can assist in doing so.

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