Imaging Miscues in Pediatric Headache

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Much has been written regarding the indications for imaging in pediatric headache to facilitate the detection of potentially catastrophic structural lesions (i.e., tumors and hydrocephalus). Detection of such lesions is typically straightforward with CT or MRI. Imaging correlates for certain well-described causes of pediatric headache, however, may be more subtle and, therefore, potentially overlooked. Furthermore, the overlap of imaging patterns may result in the potential for misinterpretation. The purpose of this article is to highlight some of the entities in which imaging findings are subtle and/or overlooked in this population of children with headache. In particular, we discuss: (1) inferior cerebellar tonsillar ectopia as a manifestation of Chiari I malformation, and its mimics; (2) subtle imaging findings of vascular etiologies for headache, including circle of Willis vasculopathy and dural sinus thrombosis; and (3) typical imaging features of migraine variants presenting in the pediatric population, including hemiplegic and ophthalmoplegic migraine.

Headache With Chiari I Malformation and Its Mimics

Chiari I malformation conventionally is defined as cerebellar tonsillar descent below the level of the foramen magnum by at least 5 mm, in the absence of intracranial mass lesions, hydrocephalus, and cerebral or cerebellar edema. As so defined, Chiari I malformation is noted in up to 3.6% of children referred for head MRI.1 Symptomatic patients with Chiari I malformation often present with headaches that are short in duration, occipital or nuchal in location, and exacerbated by the Valsalva maneuver. Management of symptomatic patients with Chiari I malformation is surgical, consisting of posterior decompression with inferior occipital craniectomy, C1 laminectomy, and possibly duraplasty.

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A subgroup of patients with Chiari I malformation, recently described as “complex Chiari malformation,” may require additional posterior occipitocervical fusion and/or ventral decompression for optimal therapy. In addition to cerebellar tonsillar descent, these children with complex Chiari malformation are identified by brainstem descent, with an obex position below the level of the foramen magnum and a dorsal cervicomedullary hump (Figure 1). This constellation of findings has been referred to as “Chiari 1.5.” Osseous anomalies of the cranio cervical junction commonly are demonstrated in patients with complex Chiari malformation, including retroflexion of the dens, reduced cranio cervical angle, basilar invagination, platybasia, and atlanto-occipital assimilation.\(^2\)

Peg-like cerebellar tonsils meeting these measurement criteria are not specific for Chiari I malformation. They also may occur in the setting of intracranial hypotension. The headache in patients with intracranial hypotension is characteristically orthostatic, increased with sitting or standing, and improved in the supine position. Intracranial hypotension results from low cerebrospinal fluid (CSF) pressure/volume of any cause, including from a persistent CSF leak in the spine. Although most commonly seen after violation of the spinal dura (e.g., after lumbar puncture), leaks may be spontaneous and are thought to result from tearing at a site of congenital or acquired weakness of the dura, such as a spinal diverticulum or dural ectasia. As such, patients with connective tissue disorders may be at increased risk for spontaneous intracranial hypotension. In our practice, overshunting is probably the most common cause of cerebellar tonsillar descent from intracranial hypotension. Regardless of the cause, treatment of intracranial hypotension with surgical posterior fossa decompression is inappropriate. Therapeutic options to treat the cause of the spinal leak include epidural blood patch, fibrin glue injection, or surgery.\(^3\) Although potentially similar in appearance to Chiari I malformation on MRI, often there are additional MR signs that suggest the diagnosis of intracranial hypotension. These additional MR signs include diffuse, smooth pachymeningeal enhancement; venous sinus distension, enlargement of the pituitary gland; and descent of the brain (including reduction in size or effacement of the prepontine or interpeduncular cisterns, inferior displacement of the optic chiasm, and descent of the iter, which is the entrance of the cerebral aqueduct) (Figure 2).

**Figure 1.** Complex Chiari malformation in a 4-year-old boy with recurrent headaches and vomiting for 3 months. Sagittal T1-weighted MR image demonstrates pointed cerebellar tonsils extending to the posterior arch of C1, an obex below the foramen magnum, a dorsal cervicomedullary hump (long arrow), a short clivus (asterisk), and basilar invagination (arrowhead). The obex is the point on the midline of the dorsal surface of the medulla oblongata that marks the caudal angle of the fourth ventricle. Note syrinx in the cervical cord.

**Patients with connective tissue disorders may be at increased risk for spontaneous intracranial hypotension.**

Idiopathic intracranial hypertension also can be associated with inferior cerebellar tonsillar ectopia, with the tonsils extending 5 mm below the foramen magnum in as many as 21% of individuals in a recent adult series.\(^4\) Headache is the most common presentation and tends to be variable in character. The diagnosis usually is made by the constellation of headache, papilledema, elevated opening CSF pressure, and absence of an intracranial mass lesion or hydrocephalus. Its
etiology is unclear but may be secondary to altered CSF resorption or altered venous outflow. Although idiopathic intracranial hypertension most often occurs in adolescent obese girls, there is no definitive sex dominance or relation to weight before puberty. Idiopathic intracranial hypertension may be difficult to distinguish from Chiari I malformation on MRI, but ancillary MR findings such as papilledema, tortuous or dilated optic nerve sheaths, and a partially empty sella turcica should raise the possibility of idiopathic intracranial hypertension (Figure 3).

In adolescents, idiopathic intracranial hypertension most often occurs in obese girls.

Headache With Vasculopathy and Stroke

Headache often accompanies arterial ischemic stroke in children. In these individuals, clinical findings and/or the presence of cerebral infarction on diffusion-weighted MR images leads to appropriate vascular imaging. However, headache occasionally can occur in the setting of pediatric vasculopathy in the absence of ischemia; in such cases, abnormal findings on conventional MR images may be overlooked.

The most well known of the cerebral vasculopathies presenting in the pediatric age group is moyamoya disease. The moniker comes from the Japanese term for “puff of smoke,” which is in reference to the lenticulostriate collateral arteries observed on conventional cerebral angiography. Moyamoya...
Moyamoya disease consists of stenosis or occlusion of the terminal internal carotid artery and/or its proximal branches, local arterial collateral networks, and bilaterality.

Cerebral Sinovenous Thrombosis

Headache is one of the most common, and occasionally the only symptom of cerebral sinovenous thrombosis involving school-age children and adolescents; other nonspecific signs and symptoms include seizures, motor deficits, altered mental status, nausea, vomiting, and papilledema. A variety of risk factors associated with cerebral sinovenous thrombosis may prompt clinical consideration of dedicated CT or MR venous imaging in the setting of headache, including dehydration, central nervous system infection, prothrombotic states, chronic systemic disease, use of oral contraceptives, iron-deficiency anemia, pseudotumor cerebri, or malignancy in general. However, cerebral sinovenous thrombosis often is not a clinical consideration at the time of routine imaging. In addition, signs suggesting sinus thrombosis may be overlooked easily on an unenhanced head CT scan, which typically is obtained in the urgent or emergent setting. Direct evidence of cerebral sinovenous thrombus, as manifested by increased density in expanded cerebral veins or dural sinuses, may or may not be present. Indirect signs of cerebral sinovenous thrombosis, including parenchymal edema or hemorrhage, also are variably present (Figure 5). Classically, parasagittal distribution of parenchymal edema/hemorrhage raises the possibility of superior sagittal sinus thrombosis. A posterior temporal location suggests lateral sinus/vein of Labbe thrombosis. A thalamic distribution potentially indicates deep venous system thrombosis. In infants and young children, or in those with polycythemia, the dural sinuses are often dense on a physiologic basis, confounding assessment. From a practical

Figure 4. Moyamoya disease in an 11-year-old girl with seizure disorder and headaches. A: Axial, T2-weighted MR image demonstrates substantially decreased caliber of the right proximal middle cerebral artery (lateral arrowhead) and smaller supraclinoid internal carotid artery (medial arrowhead). B: Lateral projection cerebral angiogram from a right internal carotid artery injection shows severe narrowing of the distal right internal carotid artery (arrow), with collateral vessels contributing to the anterior cerebral artery and middle cerebral artery territories. Notice that the right posterior cerebral artery territory has already promptly opacified (via the posterior communicating artery) during this early arterial phase.
Segmental increases in dural sinus attenuation, increased deep venous attenuation, or typical patterns of parenchymal edema/hemorrhage suggest cerebral sinovenous thrombosis.

Hemiplegic Migraine

Motor weakness during migraine aura qualifies as hemiplegic migraine, with a peak onset noted in childhood or adolescence. Hemiplegic migraine may be sporadic or familial and has been linked to several mutations in ion transport channels. Although imaging in hemiplegic migraine is usually normal, severe cases characterized by prolonged confusion, coma, and/or hemiparesis often have evidence of unilateral cerebral edema, with or without accompanying diffusion-weighted MR signal changes (Figure 6). Generally, these cross-sectional MRI abnormalities are reversible. Alterations of hemispheric perfusion with advanced imaging and vasospasm or dilation on angiographic imaging also have been reported. Raising the diagnosis of hemiplegic migraine as a differential diagnostic consideration in the appropriate clinical setting is important to facilitate genetic testing in these patients. In practice, as severely affected patients may have fever and/or seizures accompanying their clinical presentation, encephalitis and postictal edema usually are proffered differential diagnostic considerations until the diagnosis is established.

Figure 6. Hemiplegic migraine in an 11-year-old girl with a family history of hemiplegic migraine. She presented with acute onset of headache; right face, arm, and leg weakness; and confusion. Axial FLAIR MR image demonstrates diffuse left hemispheric cortical edema. Diffusion-weighted images (not shown) revealed no restriction. A genetic mutation in the sodium-potassium pump (ATP1A2) was confirmed.

Figure 5. Cerebral sinovenous thrombosis in a 17-year-old girl with persistent headaches and vomiting. A: Axial, unenhanced CT scan with subtle left thalamic edema (arrow). B: Sagittal, unenhanced CT reconstruction demonstrates expanded and dense vein of Galen (black arrow) and straight sinus (white arrows), particularly with respect to the superior sagittal sinus (arrowheads).

standpoint, segmental increases in dural sinus attenuation, increased deep venous attenuation, or typical patterns of parenchymal edema/hemorrhage suggest the diagnosis and should lead to appropriate head CT or MR venographic imaging.
Ophthalmoplegic Migraine

Ophthalmoplegic migraine is characterized by at least two episodes of headache with paresis involving the third, fourth, or sixth cranial nerves in the absence of an orbital apex, parasellar, or brainstem lesion. Peak incidence is in childhood. On MR images, ophthalmoplegic migraine typically manifests with third cranial nerve enhancement and thickening, most pronounced near its root entry zone about the interpeduncular cistern (Figure 7). Importantly, on follow-up MRI after corticosteroid therapy, there should be a significant reduction or resolution in enhancement about the involved nerve. The pathophysiology is unknown, with possibilities including recurrent demyelination or ischemia. As such, some authorities argue that the term “ophthalmoplegic migraine” is a misnomer and that the syndrome might better be called “recurrent ophthalmoplegic cranial neuropathy.” The differential diagnosis includes other infectious, inflammatory, or neoplastic processes involving the third cranial nerve, but the diagnosis should be considered with the appropriate clinical history and evidence of reduced third nerve enhancement on follow-up studies after treatment.6

On MRI, ophthalmoplegic migraine manifests with third cranial nerve enhancement and thickening, most pronounced near its root entry zone about the interpeduncular cistern.

Conclusion

This CME activity highlights the imaging appearance of some important etiologies of headache presenting in the pediatric population, with a focus on findings that may be easily overlooked and/or misconstrued. It is our belief that knowledge of such findings will allow for accurate diagnosis in these cases and, therefore, will facilitate optimal patient management.

References

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1. In which one of the following individuals in the pediatric population is idiopathic intracranial hypertension most likely to develop?
   A. An obese adolescent girl
   B. A normal-weight adolescent boy
   C. A newborn
   D. An underweight 5-year-old boy
   E. A normal-weight 5-year-old girl

   See Reference No. 4 for further study

2. All of the following may be associated with inferior cerebellar tonsillar ectopia, except
   A. Chiari I malformation
   B. intracranial hypotension
   C. cerebellitis with edema
   D. ophthalmoplegic migraine
   E. idiopathic intracranial hypertension

   See Reference No. 1 for further study

3. All of the following are MR features of complex Chiari malformation, except
   A. a dorsal cervicomedullary hump
   B. a cerebellar mass lesion
   C. an obex position below the foramen magnum
   D. cerebellar tonsillar descent
   E. brainstem descent

   See Reference No. 2 for further study

4. Figure 8 is an axial FLAIR head MR image of an 8-year-old boy who presented with migrainous headaches. Which one of the following MR findings suggests the diagnosis of moyamoya disease?
   A. Cortical vein thrombosis
   B. “Ivy sign”
   C. Subacute cortical infarction
   D. Intracranial hypotension
   E. Intracranial hypertension

   See Reference No. 5 for further study

Figure 8.
5. Which one of the following is expected on follow-up head MR images of a 6-year-old girl recently treated with corticosteroids for recurrent ophthalmoplegic cranial neuropathy with enhancement of the third cranial nerve (CN III) on a prior head MR examination?
   A. No change in CN III enhancement
   B. New CN IV and CN VI enhancement
   C. Worsening in CN III enhancement
   D. A new parasellar lesion
   E. Improvement in CN III enhancement

See Reference No. 8 for further study

6. On head MR images, parenchymal thalamic edema/hemorrhage suggests venous thrombosis involving which one of the following structures?
   A. Superior sagittal sinus
   B. Transverse sinus
   C. Vein of Labbe
   D. Deep venous system
   E. Cortical veins

See Reference No. 6 for further study

7. All of the following are appropriate treatments for intracranial hypotension resulting from a spinal CSF leak, except
   A. inferior occipital craniectomy
   B. epidural blood patch
   C. fibrin glue injection
   D. surgical CSF leak repair

See Reference No. 3 for further study

8. All of the following conditions in the pediatric population are associated with moyamoya syndrome, except
   A. sickle cell disease
   B. neurofibromatosis type I
   C. Down syndrome
   D. prior radiation therapy including the brain
   E. infantile hypothyroidism (cretinism)

See Reference No. 5 for further study

9. In addition to descended, pointed cerebellar tonsils, all of the following are MR features of intracranial hypotension, except
   A. smooth dural enhancement
   B. pituitary enlargement
   C. papilledema
   D. venous sinus distension
   E. inferior displacement of the optic chiasm

See Reference No. 3 for further study

10. All of the following are MR features of Chiari I malformation, except
    A. cerebellar tonsillar descent below the level of the foramen magnum by at least 5 mm
    B. absence of an intracranial mass lesion
    C. presence of hydrocephalus
    D. absence of cerebral edema
    E. absence of cerebellar edema

See Reference No. 1 for further study