An 8-Year-Old Girl with Headache and Syncope

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An 8-year-old black female with hypertension and insulin resistance presents to the emergency department with a severe headache and fainting episode. She reports that she has had headaches over the past few days that have been treated with ibuprofen, but had sudden onset of a severe headache while standing up giving an oral presentation at school today. Her teacher allowed the patient to sit down at her desk, and she does not recall anything after sitting down. Her teacher reported that as soon as the patient sat down, her head fell to her desk. Then she “awoke” a few seconds later, complaining of a severe headache.

On further questioning, the patient characterizes the headache as pain in the front of her head that worsens with both noise and bright lights. She reports blurred vision when she lifted her head from her desk.

Other than mild headaches over the past few days, her parents report that she does not get headaches very often. She denies fever, dizziness other than this episode, runny nose, cough, sore throat, vomiting, chest pain, weight loss, weakness, difficulty walking, altered behavior, or shortness of breath. Her recent headaches have not awakened her from sleep, and she has not had recent head trauma.

Past medical history includes insulin resistance and hypertension with good blood pressure control on a single medication. She also has obstructive sleep apnea and is supposed to be using continuous positive airway pressure (CPAP), but uses it only intermittently.

The patient is obese and has been treated in a pediatric obesity clinic for the past 2 years. She also has gastroesophageal reflux disease, dyslipidemia, and reactive airway disease. Her current medications are enalapril and metformin. She has no known drug allergies. Relevant family history is positive for diabetes, obesity, and hypertension, and social history is unremarkable.

On exam, her weight is 78 kg. She had both height and weight measured about 1 week before, and at that time, her body mass in-
dex was 39 kg/m², which is more than 99% for her age and sex.

She is afebrile with a blood pressure of 126/60 mm Hg; heart rate of 95 beats/minute; respiratory rate of 18 breaths/minute; and oxygen saturation of 99% on room air. Her pain was 4/10 at entry to the emergency department.

A complete exam (including neurologic exam) is within normal limits with the exception of obesity and acanthosis in skin folds.

Her neurologic exam demonstrates normal mental status, cranial nerves II-XII, fundi, motor tone and strength, sensation, deep tendon reflexes, and coordination.

Laboratory studies, including electrolytes and a complete blood count, are normal. An electrocardiogram is performed because of her syncope and is normal. Her non-fasting blood glucose is 102 mg/dL. Non-contrast computed tomography (CT) of the head is performed, which is normal, with no evidence of space-occupying lesions or ventriculomegaly.

Next, a lumbar puncture is performed. Opening pressure is elevated at 29 cm H₂O, which decreases to 17 cm H₂O with removal of 7 mL of cerebrospinal fluid (CSF). CSF cell counts, glucose, protein, cytology, and bacterial culture are sent and are all normal. Ophthalmology is consulted and finds no evidence of papilledema.

Given her presumptive diagnosis, she is started on acetazolamide 250 mg orally twice daily and instructed to follow up with her physician. She is scheduled for magnetic resonance imaging (MRI) of her brain as an outpatient.

On follow-up with her primary care physician 6 days later, she reports complete resolution of her headaches. Her physician discusses the importance of weight loss, given her new diagnosis.

She follows up with the pediatric obesity clinic in 3 months. She continues to be headache-free on acetazolamide. Her brain MRI is normal. Through lifestyle modification (including consumption of fruits and vegetables, daily breakfast, limits on screen time, and limited sugar-sweetened beverages), she has been able to reduce her weight; her body mass index is 0.5 kg/m² lower, now at 38.5 kg/m² (although she remains severely obese).
Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure without hydrocephalus or an intracranial mass and normal CSF. The etiology of IIH remains unclear; hypotheses include increased brain water, increased CSF production, decreased CSF absorption, and elevated cerebral venous pressure.

**Epidemiology and Risk Factors**

The incidence of IIH in children has been estimated to be 0.9 per 100,000 children. Adolescents are more likely to experience IIH than younger children. One study found that approximately 60% of children with IIH are older than 10 years. There is no difference by sex in younger children; whereas IIH predominantly affects females if the diagnosis is in older children. For example, Balcer and colleagues found that 50% of 3- to 11-year-old children with IIH were female; 88% of children 12 to 14 years old; and 100% of children 15 to 17 years old.

Obesity is also a well-known risk factor for IIH, although more prominently in older children. One study found that 43% of patients with IIH 3 to 11 years old were obese; 81% of those 12 to 14 years old; and 90% of those 15 to 17 years old.

**Symptoms and Signs**

Children presenting with IIH commonly complain of a headache; headache frequency and severity are variable. The headache may be worsened with Valsalva maneuvers and changes in posture. Children also may describe the headache as associated with photophobia, nausea, and vomiting.

Occasionally, IIH may present without headache, and these children are more likely to present with vision loss and have a poorer prognosis. Patients may also complain of tinnitus, visual changes, ataxia, paresthesias, stiff neck, or myalgias. Visual changes may include restriction of visual fields and loss of visual acuity. Papilledema is also a well-described sign; however, it is not present in all children. If present, it may be unilateral or bilateral.

In addition, cranial nerve palsies, most commonly sixth nerve palsy, leading to double vision may also occur. Unlike this patient, syncope or loss of consciousness is not a typical manifestation of IIH.

**Diagnosis and Evaluation**

The history and exam should focus on the symptoms and signs discussed thus far. Families should be asked about medications associated with idiopathic intracranial hypertension (eg, growth hormone, chronic steroids, tetracycline).

A detailed ophthalmic exam is necessary, including evaluation of visual acuity, pupils, ocular motility, visual fields, and dilation. Neuroimaging studies are necessary for diagnosis and need to be performed before a lumbar puncture. MRI can be considered (specifically to evaluate for venous sinus thrombosis), but at a minimum CT should be performed.

Assuming a normal CT exam, lumbar puncture is performed. Opening pressure should be measured and fluid sent for CSF studies. There is debate over what constitutes an elevated opening pressure. Adult studies typically consider an opening pressure of at least 250 mm H2O to be consistent with IIH. Guidelines for opening pressure in the pediatric population are less clear.

**Treatment**

Multiple treatment options are available for IIH. It is important to prevent progression of visual symptoms and the potential for permanent visual loss. Weight loss has been reported to yield improvement in IIH.

In addition, repeated lumbar puncture is occasionally considered but typically only yields short-term resolution of symptoms. In general, repeat lumbar puncture is not recommended in the pediatric population. Occasionally, symptoms may resolve with a single lumbar puncture.

**Oral Pharmacologic Options**

The mainstay of pharmacologic therapy remains acetazolamide. Acetazolamide is a carbonic anhydrase inhibitor and reduces CSF production. Side effects include poor appetite, gastrointestinal symptoms, paresthesias, and metabolic acidosis. Furosemide is an alternative to acetazolamide or can be used as adjunct therapy.

Topiramate is promising as headache treatment if neither acetazolamide or furosemide are tolerable.
and effective. Topiramate has some carbonic anhydrase activity and has a typical side effect of weight loss, which is beneficial in obese patients.\textsuperscript{15}

Although these medications with carbonic anhydrase activity often resolve headaches, occasionally more typical headache medication, such as beta-blockers, are needed as adjunct therapy.\textsuperscript{16}

Additional Treatment Options

In severe cases of acute visual loss, intravenous steroids in conjunction with acetazolamide are useful to prevent progression.\textsuperscript{17}

Surgical options are also available for severe cases or refractory cases. Optic nerve sheath fenestration is an option if visual symptoms are the primary concern and can provide stabilization or improvement in visual symptoms in children. An incision is made along the optic nerve sheath, which removes pressure off the optic nerve.\textsuperscript{18}

CSF shunting (lumboperitoneal or ventriculoperitoneal) can also be effective in reducing headaches and visual damage when previous treatments have failed. However, the risk for the well-known complications of shunt procedures, such as shunt revision and infections, should be considered.\textsuperscript{19}

PROGNOSIS

The most concerning symptom of IIH is the potential for permanent visual damage. Most children have resolution of papilledema in 4 to 5 months.\textsuperscript{20} Most patients regain normal visual acuity, but permanent loss of acuity can occur.\textsuperscript{9} Up to 17% of patients may also experience permanent visual field loss.\textsuperscript{6}

Weight gain in the year before diagnosis appears to predict visual deterioration despite appropriate treatment for IIH.\textsuperscript{21} Recurrences are described in up to about 20% of children.\textsuperscript{22}

REFERENCES