Case Report

A case report: Pediatric Idiopathic Intracranial Hypertension

Suntaree Thitiwichienlert, Kornkanok Saksritawee

Abstract

A 6-year-old girl presented with headache and binocular diplopia for one month. There were signs of papilledema and sixth nerve paresis. Magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) brain revealed no stenosis of cerebral venous sinuses. A lumbar puncture (LP) revealed elevated cerebrospinal fluid (CSF) opening pressure. The cytological analysis of the CSF was otherwise within normal limits. The patient was treated with a carbonic anhydrase inhibitor and her headaches have improved. One month later, papilledema was completely resolved.

Keywords: Idiopathic intracranial hypertension, Papilledema, Sixth nerve paresis, Headache

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Department of Ophthalmology, Faculty of Medicine, Thammasat University
Corresponding author: อาจารย์แพทย์หญิง สุนทรี ธิติวิเชียรเลิศ ภาควิชาจักษุวิทยา คณะแพทยศาสตร์ มหาวิทยาลัยธรรมศาสตร์
โทรศัพท์ 02-9269957 อีเมล punoipunoi@hotmail.com

Introduction

Idiopathic intracranial hypertension (IIH) is an uncommon neurologic condition in children. IIH can also occur in pediatric patients about 7% of cases, and there is no related significant risk factor demonstrated including gender predilection and obesity.[1] Diagnosis of IIH is made with the modified Dandy criteria; (1) signs and symptoms of increased intracranial pressure (2) no localizing neurological signs, except for unilateral or bilateral sixth nerve paresis (3) increased CSF opening pressure (opening CSF pressure at LP ≥ 25 cm H_2O in adults and ≥ 28 cm H_2O in children) but normal CSF composition (4) no evidence of hydrocephalus, mass, structural, or vascular lesion on neuroimaging (5) no other cause of intracranial hypertension identified.[2] We report a case of non-obese girl diagnosed with IIH presenting with a history of increased ICP. The study was approved by the Medical Ethics Committee of Thammasat University (MTU-EC-OP-1-042/62), Pathum thani, Thailand.

Case report

A 6-year-old non-obese girl complained of acute binocular horizontal diplopia with a history of slow progressive headache, and associated vomiting for about one month. She denied having visual loss, transient visual obscuration, and fever. She had no history of trauma, underlying diseases, obesity, and a history of drugs predisposing to IIH including antibiotics and vitamin A. The blood pressure was 109/66 mmHg. Her body weight was 22 kilogram (kg), while her height was 111 centimeters (cm). Her body mass index (BMI) was 17.85 kg/m^2 (weight 22 kg /height squared (1.11 m^2). Initial best corrected visual acuity (BCVA) was 20/20 in each eye. The intraocular pressure and anterior segment were normal both eyes. The pupils were 3 mm in size and reacted to light both eyes without afferent pupillary defect. Motility examination revealed limited abduction of both eyes. The fundus examinations revealed bilateral optic discs swelling (figure 1). The color vision was normal and the visual field test revealed enlarged blind spot. Neurological examinations revealed no focal deficits or other cranial nerves involvement.

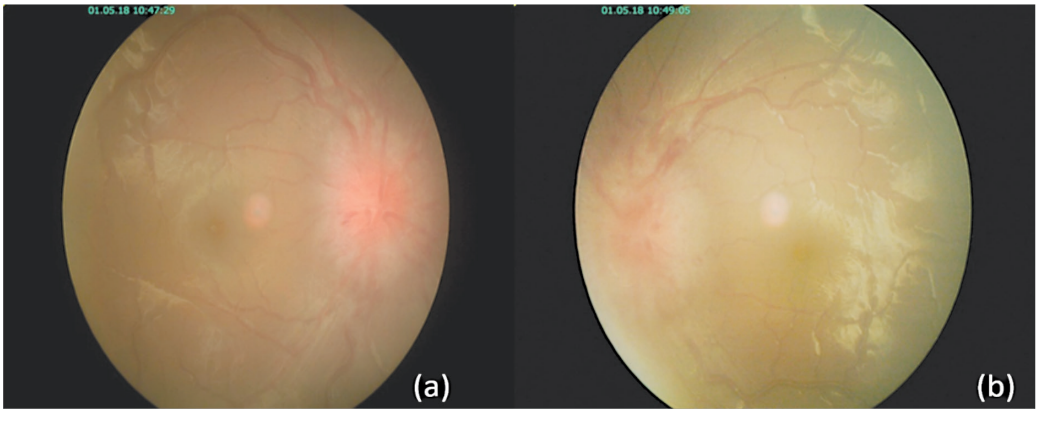

Figure 1 Bilateral optic disc swelling shown in fundus examination. Fundus photographs of the right eye (a) and left eye (b) showing diffuse hyperemic optic disc swelling.

Contrast-enhanced brain magnetic resonance imaging (MRI) was performed to exclude organic causes of papilledema. The findings revealed no space occupying lesion, focal area of brain edema, or leptomeningeal enhancement. As a result, lumbar puncture was performed. The CSF analysis revealed open pressure of 49 cm H_2O and close pressure of 38 cm H_2O, CSF protein of 47 mg/dL, CSF sugar of 71 mg/dL, white blood cell count of 0 cells per high-power field, and red blood cell of 0 cells per high-power field.

Because of elevated ICP, we stated her headache was possibly due to IIH. Subsequent contrast-enhanced brain MRI/magnetic resonance venography (MRV) was performed to rule out secondary causes of intracranial hypertension. The MRV findings revealed normal size of the venous sinuses, cortical veins and deep cerebral veins without filling defect or stretching. The straight sinus mostly drains into left transverse sinus which could be a normal variant. No definite evidence of venous thrombosis was observed (figure 2).

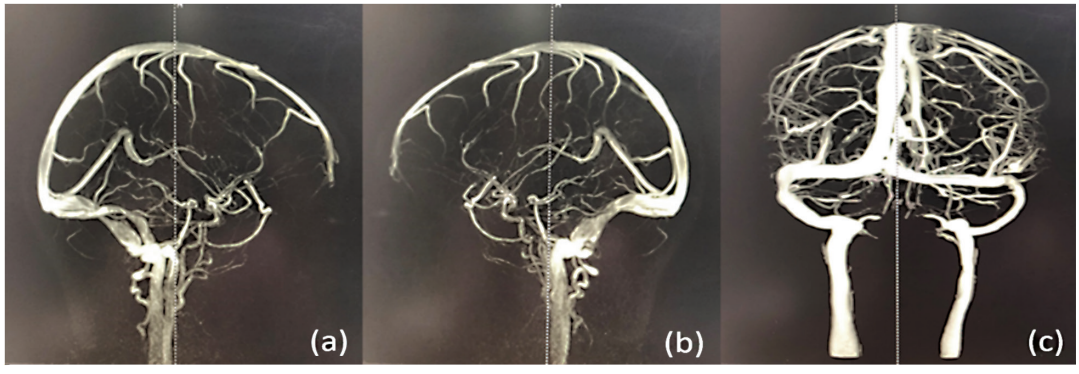

Figure 2 Contrast-enhanced brain MRV (a) right sided view (b) left sided view and (c) back view with no thrombosis or stenosis of cerebral venous sinuses.

The patient was diagnosed with IIH. Pediatric neurologist started carbonic anhydrase inhibitor therapy. The patient was received acetazolamide, a carbonic anhydrase inhibitor, tablet 250 mg (dose 25 MKD; BW 22 kg = 550 mg/day) 1 tab diluted to 5 ml per oral 3 ml (150 mg) every 6 hours (600 mg/day). Her headache was improved. On follow-up at one week at the outpatient clinic, the patient had BCVA of 20/20 OU and some improvement of lateral rectus palsy. Four weeks later, the fundus examination revealed completely resolved of papilledema.

Discussion

The incidence of IIH peaks in the third decade of life in which 90% of the cases are obese female.[3] In prepubertal children, there are no known risk factors nor gender predilection in patients with IIH. Several mechanisms are postulated to account for the disruption in CSF homeostasis, including increased CSF production and decreased CSF absorption. IIH is a diagnosis of exclusion that occurs primarily in obese female of childbearing age. However, many causes may be associated with secondary intracranial

hypertension include: (1) obstruction or impairment of cerebral venous drainage such as cerebral venous obstruction; (2) endocrinologic or metabolic dysfunction; (3) exposure to exogenous drugs and other substances; (4) withdrawal of certain drugs and (5) systemic conditions such as hypercoagulable abnormalities. Such causes especially cerebral venous sinus thrombosis (CVST) may resemble IIH, and therefore patients with suspected IIH should undergo neuro-imaging with contrast-enhanced brain MRI/MRV to exclude CVST. Lumbar puncture should be performed to confirm elevated ICP and to rule out infectious or inflammatory processes.[4]

The goals of treatment are to preserve vision and alleviate headache symptoms. Current practice patterns regarding the management of IIH are based largely on case series and clinical experience; there are no randomized controlled trials prospectively assessing the effectiveness of treatment. Medical treatment is the first-line treatment in the setting of good visual function. Enzyme carbonic anhydrase in the choroid plexus has a role in the production of CSF. Thus, carbonic anhydrase inhibitors (CAI) decrease production of CSF, resulting in decreased ICP.[5] In children, acetazolamide (Diamox) is started at a daily dose of 15 mg/kg/day in 2-3 divided doses.[6] Repeat lumbar punctures are sometimes useful to lower the ICP in selected cases with exacerbation of headache symptoms, but the role of procedure remains uncertain.[7] Surgical treatment is indicated in cases of intractable headache or progressive visual loss from full-developed papilledema despite medical treatment. Interventional optic nerve sheath fenestration (ONSF) often improve visual function by reduced papilledema especially in patients with marked papilledema, however, ONSF does not decrease ICP, so it may not be effective symptomatic treatment in intracranial hypertension.[8, 9]

The present case was non-obese girl complained of headache which is the presenting symptom in up to 91% of cases.[10] In patients with headache and suspected papilledema, neuroimaging should be performed to exclude structural brain lesions and other causes of increased ICP. In the present case, the contrast-enhanced brain MRI and CSF analysis were normal, but the ICP was increased, and we suspected the presumed diagnosis of IIH. Although no evidence of ventriculomegaly or mass effect in the contrast-enhanced brain MRI, further contrast-enhanced MRV or computerized venography (CTV) should be done to exclude the secondary causes attributing to the increased ICP. In the present case, the contrast-enhance MRV revealed no thrombosis or stenosis of cerebral venous sinuses and the patient was received acetazolamide therapy. The patient should be follow-up for the document visual function with stereo optic disc photography and visual field for full recovery within few months. Patients with IIH may require long-term acetazolamide therapy until headache, disc swelling, and visual field defect resolve typically in 3-9 months.

Conclusion

We reported a non-obese girl who developed papilledema from IIH. IIH is a diagnosis of exclusion, therefore, children with suspected IIH should undergo detailed physical and eye examinations, including BP measurement and BMI assessment. If initial brain CT/MRI unremarkable, then lumbar puncture should be done. Further investigations such as MRV to exclude CVST should be performed to rule out secondary causes of intracranial hypertension before IIH can be diagnosed.

Conflict of interest

None

Acknowledgement

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บทคัดย่อ

รายงานผู้ป่วย: ภาวะความดันในกะโหลกศีรษะสูงโดยไม่ทราบสาเหตุในเด็ก

สุนทรี ธิติวิเชียรเลิศ, กรกนก ศักดิ์ศรีทวี
ภาควิชาจักษุวิทยา คณะแพทยศาสตร์ มหาวิทยาลัยธรรมศาสตร์

เด็กหญิงไทย อายุ 6 ปี มาด้วยอาการปวดศีรษะและเห็นภาพซ้อนมา 1 เดือน ตรวจพบขั้วประสาทตาบวมและเส้นประสาทสมองคู่ที่ 6 อัมพาต ผลเอกซเรย์แม่เหล็กไฟฟ้าสมองและหลอดเลือดไม่พบภาวะอุดตันของหลอดเลือดดำในสมอง ผลตรวจพบความดันของน้ำหล่อเลี้ยงไขสันหลังสูง ผู้ป่วยได้รับการรักษาด้วยยากลุ่มคาร์บอนิกแอนไฮเดรสอินฮิบิเตอร์ ภายหลังการรักษาอาการปวดศีรษะดีขึ้น ตรวจติดตามที่ 1 เดือน หลังรักษาพบขั้วประสาทตายุบบวม

คำสำคัญ: ภาวะความดันในกะโหลกศีรษะสูงโดยไม่ทราบสาเหตุ, ภาวะขั้วประสาทตาบวมอันเนื่องมาจากความดันในกะโหลกศีรษะสูง, ภาวะเส้นประสาทสมองคู่ที่ 6 อัมพาต, ปวดศีรษะ