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Clinical Challenges

A baby with a lot of nerve

Hema L. Ramkumar, MD[a], *Rohan Verma, MD*[b], *Janet Crow, MD*[c], *Shira Robbins, MD*[d], *David B. Granet, MD*[d,*], *Claire A. Sheldon, MD, PhD*[e], *Fred M. Henretig, MD*[f,g], *Grant T. Liu, MD*[h,i,j]

[a] *Department of Ophthalmology, University of California, San Diego, San Diego, California, USA*
[b] *School of Medicine, University of California, San Diego, San Diego, California, USA*
[c] *Department of Pediatrics, University of California, San Diego, California, USA*
[d] *Pediatric Ophthalmology and Adult Re-Alignment Services, Ratner Children's Eye Center & Shiley Eye Institute, University of California, San Diego, San Diego, California, USA*
[e] *Division of Neuro-Ophthalmology, Department of Ophthalmology & Visual Sciences, University of British Columbia, Vancouver, British Columbia, Canada*
[f] *Section of Clinical Toxicology, Division of Emergency Medicine, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA*
[g] *Department of Pediatrics, Perelman School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania, USA*
[h] *Neuro-ophthalmology Service, Division of Ophthalmology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA*
[i] *Department of Neurology, Perelman School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania, USA*
[j] *Department of Ophthalmology, Perelman School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania, USA*

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1. Case report

A 10-month-old girl was referred by her pediatrician for new onset esotropia. Her mother reported a 5-day history of intermittent inward turning of the left eye. One month before presentation, the patient had 2 episodes of vomiting. A pediatric physical examination at the time was normal. The infant was playful and had no history of fever, seizures, other gastrointestinal symptoms, or altered consciousness.

* Corresponding author: David B. Granet, MD, Anne F. Ratner Professor of Ophthalmology & Pediatrics, Ratner & Shiley Eye Institute, University of California San Diego, 9415 Campus Point Drive, La Jolla, California 92037, USA.
E-mail address: dgranet@ucsd.edu (D.B. Granet).
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The child was born after an unremarkable pregnancy at 40 weeks of gestation by uncomplicated spontaneous vaginal delivery at 3,841 grams. The mother's medications during the pregnancy included lexapro for well-controlled depression as well as lysine, sunflower lecithin, calcium, fish oil, and a prenatal vitamin that contained 4,000 IU vitamin A (retinal palmitate). She denied using alcohol or other drugs and continued to take the vitamin postnatally. The child was otherwise healthy, immunized, and met all developmental milestones. She was breast-fed and was taking vitamin D supplementation prescribed by the pediatrician. There was no family history of strabismus or neurologic disease.

Her refractive error was $+2.00 + 0.50 \times 180$ OU. She had a 30-prism diopter esotropia at near with a right eye preference that increased on left gaze and decreased on right gaze. Her ductions and versions revealed full range of motion OD with a −2.5 abduction deficit OS without nystagmus. Dilated fundus examination revealed mild hyperemia of both optic disks with blurred disk margins. The infant's anterior fontanelle was open and soft. Because her ophthalmic examination was consistent with a left cranial nerve VI palsy in the setting of bilateral disk edema, she was transferred to the local children's hospital emergency room for further evaluation.

What are the diagnostic considerations?

What would you do next?

2. Comments

2.1. *Comments by Claire A. Sheldon, MD, PhD and Grant T. Liu, MD*

The presenters describe the case of a 10-month-old child with vomiting, acute esotropia, and papilledema. Together, these clinical features are concerning for raised intracranial pressure (ICP). The differential diagnosis remains broad at this point and includes an intracranial mass lesion, hydrocephalus, intracranial hemorrhage, pseudotumor cerebri (PTC), venous thrombosis or obstruction, and meningitis.

In light of the differential diagnosis, she requires urgent neuro-imaging. Given the child's young age and the potential radiation exposure[28] of computed tomography, magnetic resonance imaging (MRI) of the brain (with and without gadolinium) should be performed to rule out an intracranial mass lesion, hydrocephalus, or intracranial hemorrhage. Given her age, we would also recommend completing MR venography to exclude a venous thrombosis.

If imaging is normal, a lumbar puncture is necessary to document elevated ICP and cerebrospinal fluid (CSF) composition. CSF fluid analysis should include, at a minimum, cell count, cytology, and concentrations of glucose and protein. Meningitis should be excluded.

3. Case report (Continued)

MRI of the brain with and without gadolinium demonstrated no intracranial abnormalities, but protrusion of the optic nerve into the globes, consistent with the bilateral optic disk edema (Fig. 1). Major vascular flow voids at the skull base suggested that there was no venous sinus thrombosis. An MR venography was not performed because postcontrast images were deemed sufficient to rule out a dural venous sinus thrombosis. A lumbar puncture performed in the left lateral decubitus position showed an elevated opening pressure of 50 cm H_2O with normal CSF composition. A serum vitamin A level was elevated to 52 mcg/dL. The normal range of vitamin A in children age 1–6 is 20–43 mcg/dL, but there is no accepted normal range for infants.

What is the relevance of this CSF opening pressure?

4. Comments (Continued)

4.1. *Comments by Claire Sheldon, MD, PhD; Grant T. Liu, MD*

In the past, the diagnosis of PTC was based on the modified Dandy criteria; however, subsequent modifications have been made based on increasing recognition of typical signs and symptoms of this disorder. The most recent updated diagnostic criteria were published in 2013 and incorporated timely insights into common neuro-imaging characteristics of raised ICP and reference ranges of normal CSF opening pressure.[13] In the current case, many criteria are fulfilled. First, the contrast-enhanced MRI revealed normal brain parenchyma and, with attention paid to the cerebral veins, there was no evidence of venous thrombosis or obstruction. The MRI did reveal at least 2 radiographic features of raised ICP, namely globe flattening and perioptic distention (Fig. 1). Second, an opening pressure of 50-cm H_2O documents raised ICP. Opening pressure greater than 28-cm H_2O in children is considered elevated; however, greater than 25-cm H_2O is considered elevated in those not sedated during the lumbar puncture and nonobese children.[2] Third, there was normal CSF composition. In summary, as illustrated by this case, key diagnostic requirements for PTC include 1) papilledema, 2) normal brain parenchyma on neuro-imaging with contrast-enhanced MRI or computed tomography, 3) normal venous imaging, in select cases, with MRI- or computed tomography–venogram, and 4) normal CSF

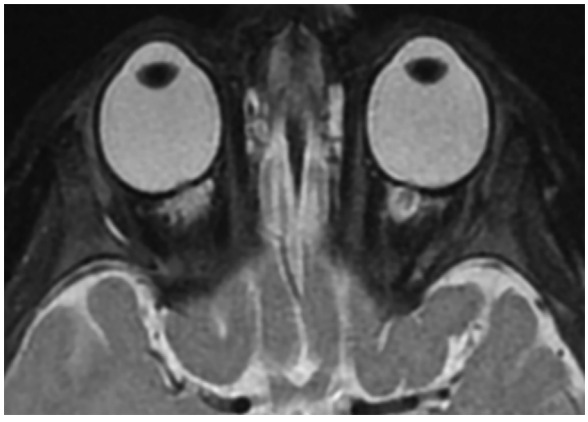

Fig. 1 – Magnetic resonance imaging (MRI) T2 axial section demonstrating edema of the optic nerve sheath and bilateral protrusion of the optic nerves into the globe consistent with clinical optic disk edema.