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Original Article

Novel Magnetic Resonance Imaging Findings in Children With Intracranial Hypertension

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ABSTRACT

BACKGROUND: Increased intracranial hypertension is defined as elevated intracranial pressure with absence of hydrocephalus, vascular or structural abnormalities, and normal cerebrospinal fluid content. Magnetic resonance imaging abnormalities of the optic nerve and sheath that have been described in adults include increased nerve tortuosity, flattening in posterior aspect of globe, intraocular protrusion of the optic nerve, and enlarged optic nerve sheath. **PURPOSE:** We evaluated accuracy of those proposed markers on magnetic resonance imaging in children with increased intracranial hypertension that are described in adults. **MATERIALS AND METHODS:** Eleven patients between 3 and 15 years of age with intracranial hypertension were selected for re-evaluation of magnetic resonance imaging that had been previously described as normal to determine the presence of: (1) increased tortuosity and elongation of the optic nerve; (2) enlargement of the optic nerve sheath on axial and coronal T2 so called by us "target sign" and postcontrast T1 sequences; (3) flattening in posterior aspect of the globe; and (4) intraocular protrusion of the optic nerve head. **RESULTS:** Of the 11 patients, tortuosity of the optic nerve (10/11, 90.9%) and enlarged optic nerve sheath—target sign (7/11, 63.6%)—were the most common findings. Flattening in the posterior aspect of globe (5/11, 45.5%) and intraocular protrusion (3/11, 27.3%) were also detected as a novel magnetic resonance imaging findings. **CONCLUSION:** Magnetic resonance imaging findings of the optic nerve and sheath include valuable signs of intracranial hypertension not only in adults but also in children. This is the first detailed analysis of the magnetic resonance imaging findings in children with increased intracranial hypertension.

Keywords: intracranial hypertension, MRI findings, children, increased intracranial pressure

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Introduction

Idiopathic intracranial hypertension (IIH) is defined by increased intracranial pressure with no clear etiology and the absence of hydrocephalus, vascular, or other structural lesion and normal cerebrospinal fluid (CSF) content.[1-12] Common complaints are headache, blurred vision, and vomiting.[1-16] The most important complication is permanent visual disturbance in approximately 10% of the patients.[1-16] Although magnetic resonance imaging (MRI) may be unremarkable in terms of gross pathology, some studies point to subtle radiological markers including compression of the optic globe, particularly in the posterior aspect, intraocular protrusion and enlargement of optic nerve sheath, and increased tortuosity in optic nerve that may be important observation in the early diagnosis of IIH.[14-28] These findings have been studied in adult populations, but there are few studies in children with IIH.[7,20,26-31]

Our aim was to investigate these MRI markers and their possible contribution to the diagnosis of IIH in the pediatric age group.

Material and Methods

Eleven patients aged between 3 and 15 years old (mean age 10.45 years) with IIH were selected retrospectively from Department of

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Pediatric Neurology, Gazi University School of Medicine, between January 2012 and December 2013. The diagnosis of IIH was based on the Modified Dandy Criteria: (1) signs and symptoms of increased intracranial pressure; (2) absence of localizing findings on neurological examination; (3) increased CSF opening pressure with normal CSF ingredients; (4) no evidence of hydrocephalus, mass, structural, or vascular lesions on imaging; and (5) no other causes of increased intracerebral pressure identified.[1,9] To confirm the diagnostic criteria, medical records of the patients were evaluated in terms of detailed history, physical and neurological examination, laboratory screening, and MRI findings as follows.

1. History and clinical features: Recent history of infection and medication comprising antibiotics, vitamins, hormones including oral contraceptives, and the presence of headache or additional complaints such as visual disturbances, nausea, and vomiting were investigated. Physical and neurological examination including abducens/facial nerve palsy and funduscopic examination for papilledema, if available, Snellen visual acuity, and Goldman visual field perimetry was evaluated. Body mass index was also noted for each patient. A higher percentile of 95% for each age was defined as obesity.
2. Laboratory results: Cerebrospinal fluid and blood testing was performed to exclude secondary reasons for IIH: Lumbar puncture was performed to evaluate the CSF profile and to measure opening pressure that was accepted as increased levels at >180 mm H_2O for children younger than 8 years of age and >250 mm H_2O for those older than 8 years.[4-6,8] All patients underwent some additional laboratory testing including complete blood count, liver and renal functions, electrolytes, sedimentation rate, serum ferritin, iron and iron-binding levels, thyroid function, parathyroid hormone, adrenocorticotropic hormone, cortisol, anti-nuclear antibody, anti—double-stranded DNA, and vitamins A and D levels.
3. MRI slices that were previously reported as normal in terms of congenital malformations, hydrocephalus, and mass, structural lesions were re-evaluated by T.H.; K.A., who is most experienced and educated physician in MRI interpretation in the group; and A.S. independently in terms of proposed MRI findings. The axial and coronal T2 and postcontrast T1 sequences were investigated for the presence of proposed subtle signs in optic nerve and sheath for each patient: (1) flattening at posterior aspect of the globe are described as disturbed normal convexity and also straightening of the normal outwards of the globe in the area of attachment to optic nerve, (2) intraocular protrusion of optic nerve: beyond the flattening as described previously, there is a concave protrusion towards to the globe; and (3) enlargement of optic nerve sheath on T2 axial serial—we newly proposed a “target sign” on coronal T2 sequences. The measurements of optic nerve sheath were performed from 5 mm posterior to the globe.[18,20,32-34] If the mean optic nerve sheath diameter was greater than 5 mm in diameter, it was considered as abnormal and meaningful for IIH,[19,20,22,25,30,31] (4) increased tortuosity in optic nerve can also be described as vertical tortuosity of the optic nerve which are observed as “redundant” or “S-shaped” appearance and (5) a narrowed ventricle so called “slit-like ventricles,” empty sella, and transverse sinus abnormalities were evaluated.[17-31]

Approval for this study was granted by Gazi University School of Medicine Ethical Committee (N:2014-534). Statistical Package for Social Science, version 16.0, SPSS Inc, Chicago, IL, was used for statistical analysis. Descriptive analyses were performed and data were presented as mean ± standard deviation. Correlation coefficient values were calculated for comparing CSF opening pressure and MRI findings.

Results

Eleven patients were found to fit the criteria for re-evaluation for subtle MRI findings of IIH. Five of the patients were female (45.5%) and 6 (55.5%) were male. Symptom duration was highly variable, ranging from 2 days to 1 year before admission to our department.

The most common complaints were headache (100%) and ophthalmologic symptoms (81.8%). Nausea and vomiting occurred in five (45.5%) patients. Four (36.4%) patients had diplopia and one had an abducens palsy. Eight (72.8%) patients had a problem in visual acuity, and one of these had a significant visual field defect. Bilateral papilledema was detected in all patients (11/11, 100%).

A total of eight patients (72.7%) was older than 8 years and the remaining three (27.3%) were younger than 8 years of age. The CSF pressure was measured as a mean of 190 mm H_2O (ranged 180-200 mm H_2O) in those younger than 8 years old and 465 mm H_2O (ranged 250-790 mm H_2O) in those older than 8 years old.

For etiology, basic hematologic, and biochemical results, hormone levels including thyroid functions, parathyroid hormone, adrenocorticotropic hormone, cortisol, and vitamins were within normal limits. The most important factor was obesity with increased body mass index (5/11, 45.5%). As to secondary IIH, one patient had Behcet disease with congenital hypoplasia of the transverse and sigmoid sinus and jugular vein; one patient had thrombosis in superior sagittal sinus with a factor V Leiden mutation. Detailed demographics and clinical signs are presented in Table 1.

MRI scans of all patients were within normal limits in terms of congenital, vascular, mass, structural lesions, and hydrocephalus in idiopathic IIH. One patient with Behcet disease had hypoplasia in transverse and sigmoid sinus; another one with factor V Leiden mutation had thrombosis in the anterior third of the superior sagittal sinus in the secondary IIH group (Fig. 1).

TABLE 1.
Demographics and Clinical and Laboratory Findings of the Patients

	Number (Range or %)
Age	10.45 (3-15)
<8 years old	3 (27.3%)
>8 years old	8 (72.8%)
Sex	
Female	5 (45.5%)
Male	6 (55.5%)
Symptom duration before admission	2 days to 1 year
Headache	11 (100%)
Nausea ± vomiting	5 (45.5%)
Ophthalmologic symptoms: findings	9 (81.8%)
Diplopia	4 (36.4%)
Disturbed visual acuity	8 (72.8%)
Abducens nerve palsy	1 (9%)
Papilledema on funduscopic examination	11 (100%)
CSF pressure	
<8 years old	190 mm H_2O (180-200)
>8 years old	465 mm H_2O (250-790)
Etiology	
Obesity	5 (45.5%)
Venous sinus thrombosis (factor V Leiden mutation)	1 (9%)
Hypoplasia of venous sinus (Behcet disease)	1 (9%)
Treatment	
Acetazolamide	11 (100%)
Acetazolamide and topiramate	1 (9%)
Ventriculoperitoneal shunt	1 (9%)