Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(2):95-98 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2015.v03i02.008

Changes of Papilledema in a Case of Idiopathic Intracranial Hypertension

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Abstract: We present a case of idiopathic intracranial hypertension in an 18-year-old boy. The ophthalmoscopic examination revealed marked papilledema in the right eye and mild papilledema in the left. Magnetic resonance imaging demonstrated a torturous right optic nerve, flattened right posterior sclera, and bilateral enlargement of the optic nerve sheath. The bilateral papilledema worsened during 2 months after the initial visit. On lumbar puncture, the cerebrospinal fluid pressure was 270 mm H_2O. He was prescribed acetazolamide, and the bilateral papilledema gradually improved. Six months later, the papilledema was completely resolved.
Keywords: Idiopathic intracranial hypertension, Papilledema, Magnetic resonance imaging, Acetazolamide

INTRODUCTION

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is characterized by increased cerebrospinal fluid (CSF) pressure and papilledema without focal neurologic symptoms, except for the occasional abducens nerve palsy [1]. It is a diagnosis of exclusion, and radiologic examinations are traditionally performed to help exclude other causes of intracranial hypertension, such as obstructive hydrocephalus, tumor, chronic meningitis, arteriovenous fistula, internal jugular vein stenosis, and dural sinus thrombosis [1].

The revised diagnostic criteria for IIH were established as follows [2]: (A) papilledema; (B) normal neurologic examination, except for cranial nerve abnormalities; (C) normal brain parenchyma on neuroimaging without evidence of hydrocephalus, mass, or structural lesion, and no abnormal meningeal enhancement on plain and gadolinium-enhanced magnetic resonance imaging (MRI) in typical patients (female and obese), and on magnetic resonance venography for others; if MRI is unavailable or contraindicated, then contrast-enhanced CT may be used; (D) normal CSF composition; and (E) elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF in awake and normal weight children]) in a correctly performed lumbar puncture. A diagnosis of IIH is definite if the patient fulfills criteria A–E. The diagnosis is considered probable if criteria A–D are met but the CSF pressure is lower than specified for a definite diagnosis.

In this report, we describe the progression of papilledema in a patient with IIH.

CASE REPORT

An 18-year-old boy was referred to Jichi Medical University Hospital because of transient diplopia. The eye position was orthotropic, and ocular motility was mildly restricted in the right lateral direction. In both eyes, his visual acuity was 1.2, and the anterior segments and ocular pressures were normal. Ophthalmoscopic examination revealed marked papilledema in the right eye (Fig. 1A) and blurring of the upper and lower margins of the left optic disc (Fig. 1B).

The axial T2-weighted MRI revealed tortuosity in the right optic nerve (Fig. 2A; arrow) and a flattened right posterior sclera (Fig. 2A; arrowheads). A hypointense lesion was observed protruding mildly into the posterior vitreous cavity, which corresponded to the position of the swollen right optic disc (Fig. 2B; arrowhead).

The coronal T2-weighted MR image showed bilateral widening of the optic nerve sheaths (Fig. 3A arrows). The axial T2-weighted MR image revealed right perioptic CSF distension (Fig. 3B arrows).

In addition, an arachnoid cyst was detected in the right middle fossa (Fig. 4A and 4B; arrows). Based on these collective findings, the patient was diagnosed with IIH.

Although his visual acuity did not deteriorate, the papilledema gradually worsened during 2 months after the initial visit (Fig. 5A and 5B).

A lumber puncture was performed and showed a CSF pressure of 270 mm H_2O and normal CSF composition. The diagnosis of IIH was confirmed, and the patient was prescribed oral acetazolamide 250 mg three times daily.

After beginning acetazolamide treatment, the bilateral papilledema gradually improved (Fig. 6 A–F). Six months later, the papilledema was completely resolved (Fig. 6G and 6H), and visual acuity was maintained at 1.2 in both eyes.

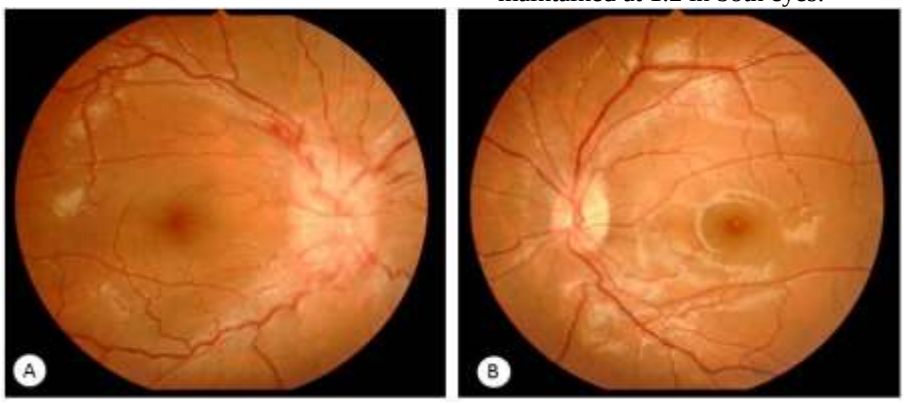

Fig. 1: Photographs of the right (A) and left (B) fundus on initial examination. The right eye exhibited marked papilledema (A). The upper and lower margins of the left optic disc were blurred as well (B).

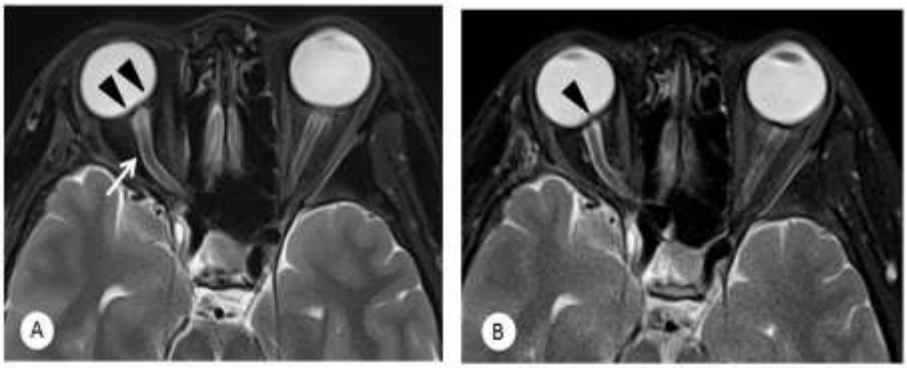

Fig. 2: Initial axial T2-weighted MRI. The right optic nerve appeared torturous (A; arrow), the right posterior sclera was flattened (A; arrowheads), and a hypointense lesion was observed mildly protruding into the right globe (B; arrowhead).

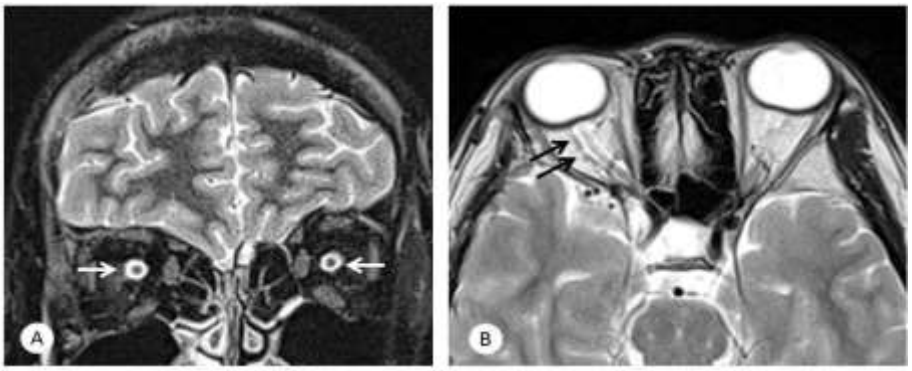

Fig. 3: Initial coronal (A) and axial (B) T2-weighted MRI. Bilateral widening of the optic nerve sheath (A; arrows) and right perioptic CSF distension (B; arrows) were observed.

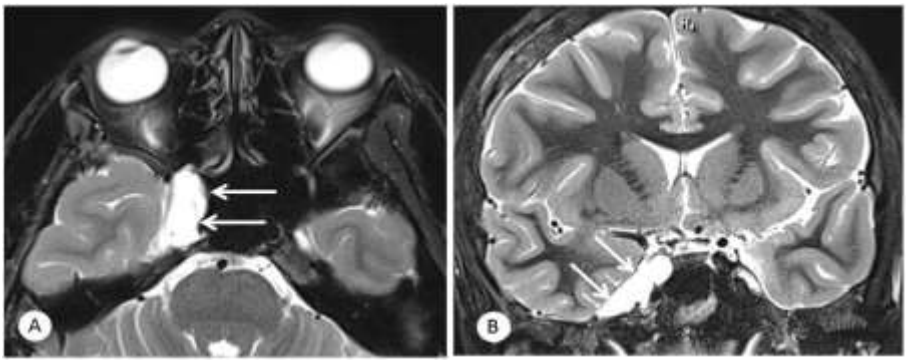

Fig. 4: Initial axial (A) and coronal (B) T2-weighted MRI. An arachnoid cyst (arrows) was detected in the right middle fossa.

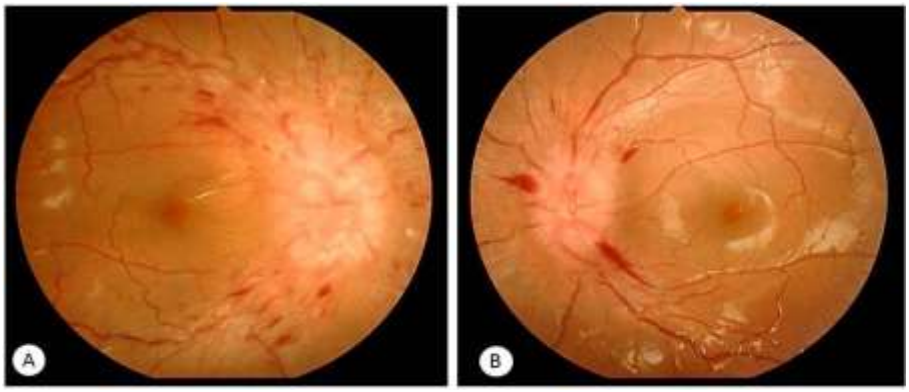

Fig. 5: Photographs of the right (A) and left (B) fundus 2 months after initial examination. The papilledema was worsened in both eyes.

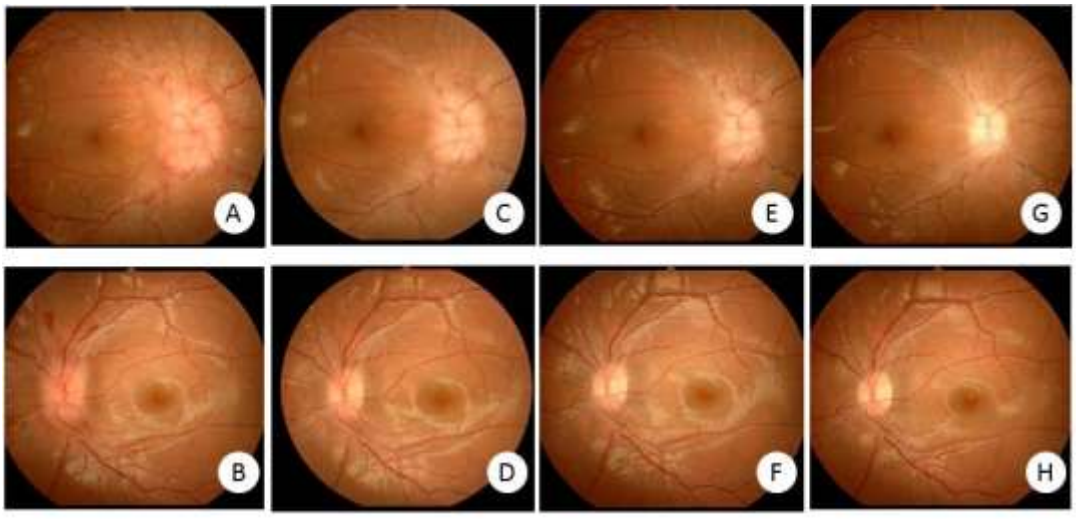

Fig. 6: Photographs of the right (top) and left (bottom) fundus during 6 months of acetazolamide treatment. The bilateral papilledema gradually improved in both eyes during 6 months of oral acetazolamide treatment. A, B: 1 month after beginning treatment; C, D: 2 months later; E, F: 3 months later; G, H: 6 months later

DISCUSSION

The most commonly reported macroscopic findings on MRI of patients diagnosed with papilledema are as follows: (1) enlarged optic nerve sheath, (2) flattened posterior sclera, (3) optic papilla protrusion into the globe, and (4) torturous optic nerve [3-6]. The present case fulfilled the diagnostic criteria of IIH [2] and showed these MRI findings.

Current theories suggest that IIH is the result of abnormal CSF hydrodynamics; therefore, treatment generally aims to modify CSF production and flow [7]. Medical therapy for IIH typically includes oral

acetazolamide, which is a carbonic anhydrase inhibitor with diuretic properties that decreases the CSF [7, 8]. Acetazolamide treatment was successful in this case with no deterioration of the visual acuity noted.

CONCLUSION

Finally, this patient also had an arachnoid cyst in the right middle fossa. Although the presence of IIH and the arachnoid cyst in this patient is most likely is coincidental, we speculate that the differing severity of papilledema between the eyes at the initial visit may reflect influence from the arachnoid cyst.

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