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Papilledema Associated with Puberty

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Case Report

A 9-year-old girl was brought by her mother to a hospital emergency room because of severe headache and vomiting. The symptoms began 2 weeks earlier while at summer camp. The child's examination was benign, and she was discharged with a diagnosis of migraine. The ocular fundi were not examined. The headaches continued and were sometimes associated with nausea, photophobia, and tinnitus. She was evaluated 2 weeks later in our pediatric ophthalmology clinic because of intermittent horizontal diplopia and blurred vision.

There was no relevant past medical history. The patient did not take tetracycline antibiotics or any other medications. The height was 142.5 cm and the weight was 58.0 kg, for a body mass index (BMI) of 28.6 kg/m² (obese). Physical development was at Tanner stage II.

The visual acuity was 20/20 in each eye without correction. Pupils were equal, round, and briskly responsive to light. There was no afferent pupillary defect. The extraocular eye movements were full. Ocular alignment was orthotropic by cover/uncover test. Stereopsis was intact to a hidden random dot butterfly pattern. Slit lamp examination was normal. Dilated fundus examination revealed acutely swollen optic nerves (Figure 1).

Clinical Course

Magnetic resonance (MR) imaging of the brain was normal, except for protrusion of the optic discs into the vitreous cavities, consistent with the findings on ophthalmoscopy (Figure 1). There was also a partially empty sella, a finding that sometimes occurs from raised intracranial pressure. An MR venogram showed no occlusion of the cerebral dural sinuses.

A lumbar puncture under sedation yielded an opening pressure of 370 mm H₂O in the lateral decubitus position. Cerebrospinal fluid analysis showed no bacteria, 2 white cells, 0 red cells, protein 20 mg/dL, and glucose 52 mg/dL. A panel of laboratory tests for hematologic, autoimmune, and inflammatory diseases was negative.

The patient was treated with acetazolamide 250 mg twice per day. Two weeks later she reported lessening of

headache. Fundus examination showed slight reduction of her papilledema. Over the next 6 months her symptoms gradually resolved and she stopped attending clinic appointments.

Two years after her original presentation, she returned to the eye clinic for a routine examination. On her own initiative, she had discontinued treatment with acetazolamide 18 months earlier. She denied headache or blurred vision. The height was 152.4 cm and the weight was 71.67 kg, for a BMI of 30.9 kg/m² (obese). She had regular menstrual periods. The visual acuity was 20/20 in each eye without correction. Pupils, eye movements, ocular alignment, visual fields, and slit lamp examination were normal. The papilledema had resolved (Figure 2).

Final Diagnosis

Idiopathic intracranial hypertension

Discussion

The term *pseudotumor cerebri* was coined by Max Nonne in 1904 to describe the curious occurrence of elevated intracranial pressure in healthy patients without brain tumor.¹ The term *benign intracranial hypertension* was later adopted to emphasize the absence of malignancy.² This name has been criticized because the disease course in some patients is hardly benign, inasmuch as irreversible vision loss may occur.³ For this reason, the name was revised again to *idiopathic intracranial hypertension*.⁴

In adults, the major risk factors for pseudotumor cerebri are female gender and obesity, suggesting that the underlying pathophysiology may involve estrogens and endocrinologically active adipose tissue.⁵ In adult

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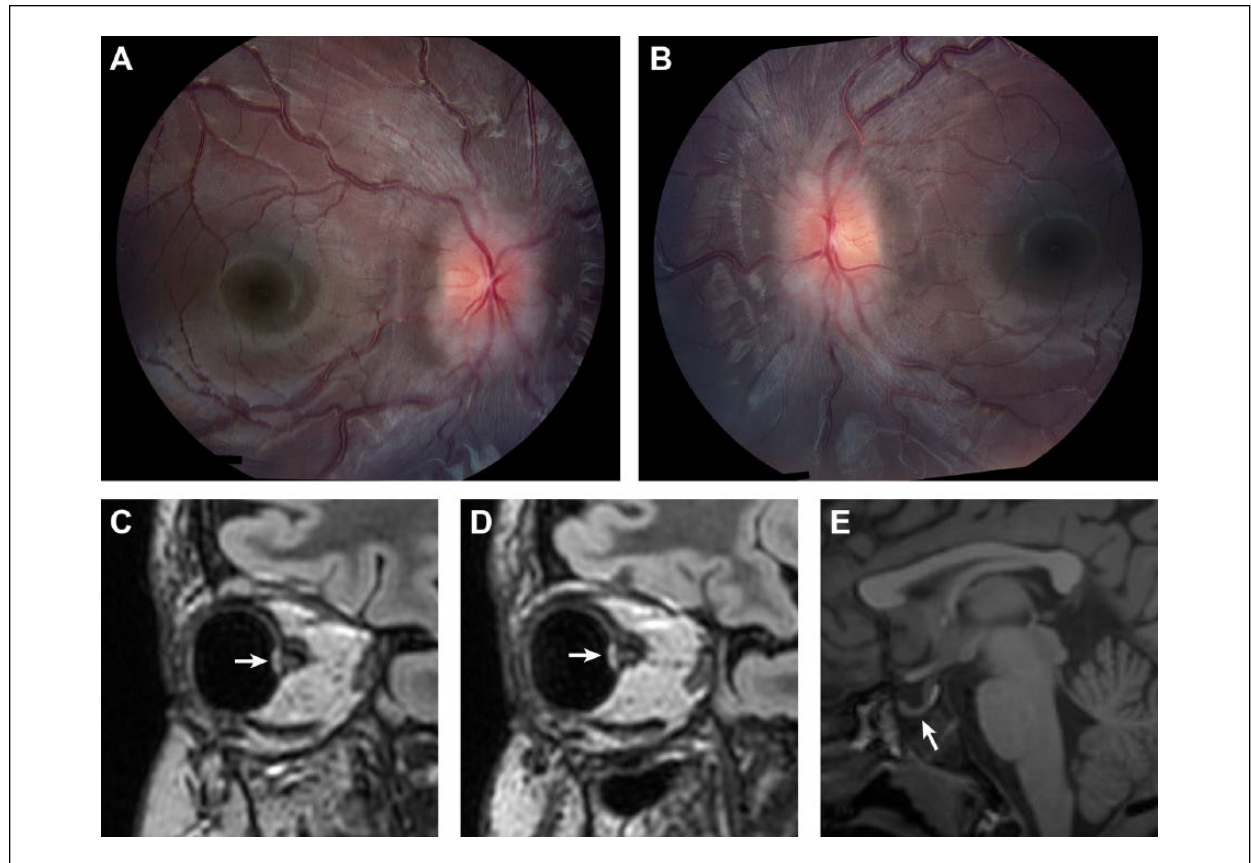


Figure 1. Right (A) and left (B) fundi showing acute papilledema. Right (C) and left (D) parasagittal T₁-weighted MR images showing elevated optic nerveheads (arrows). (E) Sagittal T₁ FLAIR MR images revealing a partially empty sella (arrow).

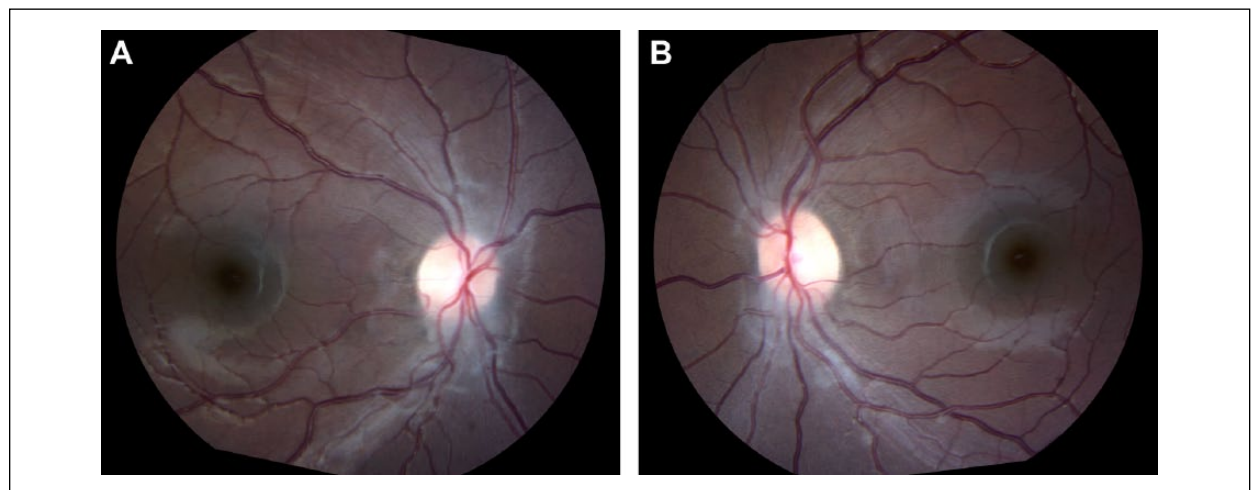


Figure 2. Right (A) and left (B) fundi showing resolution of the papilledema observed 2 years earlier.

females, the condition is often chronic, requiring careful long-term monitoring of visual function and

papilledema. Unfortunately, permanent and severe vision loss is not uncommon.³

In children with pseudotumor cerebri, the association with female gender and obesity is less pronounced. A recent population-based cohort study calculated odds ratios for 78 patients with pediatric pseudotumor cerebri. In prepubertal children, female sex conferred an odds ratio of 1.56, while extreme obesity (defined as BMI > 35 kg/m²) conferred an odds ratio of 3.44. In contrast, among postpubertal children, female sex conferred an odds ratio of 8.33, while extreme obesity conferred an odds ratio of 16.14.⁶ These odds approach those found in adult populations, in which the female-to-male ratio of pseudotumor cerebri is 8:1, and the overweight-to-normal weight ratio is 19:1.⁷

Greer was the first to recognize that pseudotumor cerebri often is associated with the onset of puberty in females. He noted that the condition was self-limited, with recovery in all 10 patients in his series by 3 weeks after diagnosis.⁸ Other studies have noted the relatively transient nature of pseudotumor cerebri in pre- or peripubertal children, with complete resolution of symptoms and papilledema.⁹

To our knowledge, this case report is the first to document photographically the resolution of papilledema in a pubescent girl with pseudotumor cerebri. Pictures taken at the child's initial clinic visit showed acute optic disc swelling. Pictures taken 2 years later, when the patient's symptoms had resolved, showed that the optic discs had returned to normal. It should be underscored that our patient's recovery was essentially spontaneous, although she was treated for a few months with a low dose of acetazolamide. This drug has been shown to reduce intracranial pressure and papilledema.¹⁰ No surgical intervention was necessary. Although weight loss was recommended, our patient's BMI at the end of her clinical course was in fact greater than her initial BMI. Her improvement, despite an increased BMI, is consistent with evidence showing that pseudotumor cerebri in children is less strongly linked to obesity than in adults.

Conclusion

The onset of puberty in girls can provoke increased intracranial pressure and papilledema. Although the presentation may be fulminant, the condition in children usually has a good prognosis.⁶ Therefore, in children the term *benign intracranial hypertension* usually provides an accurate description of the disease. MR imaging may reveal elevation of the optic discs, a partially empty sella, and other features of raised intracranial pressure.

However, the crux of the diagnosis is to examine the optic discs with an ophthalmoscope for evidence of papilledema. This child's optic discs were not examined when she was evaluated initially in the emergency room for headache and emesis. This omission led astray the physicians, who concluded that she was suffering from new onset of migraine headaches.

Declaration of Conflicting Interests

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