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Case Report

Simultaneous Papilledema and Optic Disc Drusen in a Child

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ABSTRACT

Idiopathic intracranial hypertension is a headache syndrome characterized by elevated intracranial pressure with normal cerebrospinal fluid content, normal cranial imaging, and elevated appearance of the optic disc. We report on a 6.5-year-old boy with complaints of headache and right esotropia causing diplopia. A lumbar puncture indicated an opening cerebrospinal fluid pressure of 28 cm H_2O. The headache, diplopia, and esodeviation resolved after the lumbar puncture. However, at 2-week follow-up, the elevated appearance of the optic disc continued despite normal cerebrospinal fluid pressure. A second ophthalmologic consultation revealed optic disc drusen, as also demonstrated by ocular ultrasonography. To date, two such cases have been reported in the literature. To our knowledge, this patient is the youngest with coexisting optic disc drusen and idiopathic intracranial hypertension.

Introduction

Idiopathic intracranial hypertension is a headache syndrome characterized by elevated intracranial pressure with normal cerebrospinal fluid content, normal results of cranial imaging, and an elevated appearance of the optic disc [1,2]. On the other hand, optic disc drusen, containing hyaline bodies buried in the surface of the optic disc, constitutes the most common cause of an elevated appearance of the disc (pseudopapilledema) [3].

We report on a 6-year-old boy with idiopathic intracranial hypertension and optic disc drusen. Although two pediatric cases were previously reported in the literature, to our knowledge, this patient is the youngest with coexisting optic disc drusen and idiopathic intracranial hypertension.

Case Report

A previously healthy 6-year-old boy presented with complaints of headache for 2 weeks' duration and right esotropia, causing diplopia for the preceding 2 days. A complete ophthalmologic examination revealed bilateral optic disc swelling and right esotropia. His uncorrected visual acuity, color vision assessment, and papillary light reflexes were normal. The patient's ophthalmic and medical history was otherwise unremarkable. No similar family history was evident.

Upon physical examination, the patient's pupils were equal, round, and reactive to light. His extraocular muscles were intact, although he experienced difficulty abducting his right eye. Diplopia was reported on his right lateral gaze. A funduscopic examination revealed an elevation of the optic disc margin, which did not resolve after a lumbar puncture (Fig 1). Cranial magnetic resonance imaging revealed no intracranial pathology. The lumbar puncture indicated an opening cerebrospinal fluid pressure of 28 cm H_2O, which fell to 14 cm H_2O after his lumbar puncture. The headache, diplopia, and esodeviation resolved after the lumbar puncture. His laboratory tests on admission produced findings of hemoglobin, 13.6 g/dL; hematocrit, 38.5%; white blood cells, 16300/mm^3; platelets, 312,000/mm^3; iron, 22 μg/dL; serum transferrin, 425 μg/dL; and ferritin, 7 ng/mL. Levels of serum parathormone, vitamin A, and vitamin D were normal. A complete blood count demonstrated iron-deficiency anemia, and therefore iron treatment was initiated.

Three days after the first lumbar puncture, the opening pressure of the patient's cerebrospinal fluid was 17 cm H_2O. In view of the resolved signs and reduced cerebrospinal fluid opening pressure, medical treatment (acetazolamide and steroid) was not initiated. However, at 2-week follow-up, because the elevated appearance of the optic disc continued despite the reduced signs and cerebrospinal fluid opening pressure, a lumbar puncture was repeated, and the opening pressure was measured at 15 cm H_2O. A second ophthalmologic consultation revealed optic disc drusen, as also demonstrated by ocular ultrasonography (Fig 2). In consideration of the neurologic and ophthalmologic findings, the patient was diagnosed with mild idiopathic intracranial hypertension. He has been followed for 6 months and has been stable, with no subjective or objective findings, but his optic disc margin remains elevated (pseudopapilledema), secondary to optic disc drusen.

Discussion

The coexistence of idiopathic intracranial hypertension and optic disc drusen was first described by Rosenberg et al. in 1979 [4]. To date, the coexistence of idiopathic intracranial hypertension and optic disc drusen was reported in two pediatric patients [5,6]. The difficulty of performing a detailed ophthalmologic examination in children renders the discrimination between optic disc drusen and idiopathic intracranial hypertension more complicated. In the presence of optic disc edema, the existence of optic disc drusen can cause delays in the diagnosis and appropriate management of true

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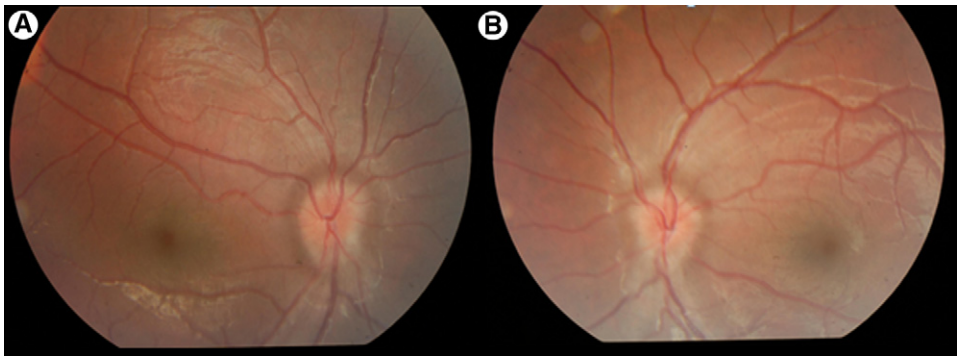

Figure 1. Fundus photography of the patient reveals bilateral, elevated optic discs after therapeutic lumbar puncture. (a) Right eye. (b) Left eye.

papilledema. Clinicians should bear in mind that optic disc drusen is rare in childhood [7].

The etiology of idiopathic intracranial hypertension may involve an identifiable underlying cause, or it may qualify as idiopathic. The most common clinical signs include optic nerve edema with or without venous pulsations, occasional abducens nerve palsy, headache, vomiting, blurred vision, diplopia, and photophobia. It does not commonly occur in childhood. Idiopathic intracranial hypertension characterizes a syndrome of increased intracranial pressure, normal cerebrospinal fluid content, and normal results of cranial imaging [1,7]. In our patient, the initial clinical signs pointed to a diagnosis of idiopathic intracranial hypertension, thereafter confirmed by an elevated cerebrospinal fluid opening pressure and the results of neurologic examinations, imaging, and laboratory tests.

Optic disc drusen is an uncommon congenital anomaly composed of hyaline material and calcified deposits that rarely begin in early childhood. Discrete hyaline bodies or papillary calcifications were first observed in one or both eyes at a mean age of 12.1 years, and became more prominent with age [3]. Optic disc drusen can be isolated, inherited in an autosomal dominant fashion, or associated with many neurologic disorders, including epilepsy, migraine, pseudotumor cerebri, mental retardation, tuberous sclerosis, and ataxia-telangiectasia [3,8]. Optic disc drusen occurs in 0.3-2.0% of the population, and is bilateral in 75% of cases. Impairment of visual acuity and of the visual field is rare. Patients with optic disc drusen are usually asymptomatic, and are only coincidentally diagnosed. Optic disc drusen has been mistaken for true papilledema by ophthalmologists as well as pediatricians [7]. Clinical findings, combined with B-scan ocular ultrasound and computed tomography, help in confirming optic disc drusen [3,5,6]. Imaging techniques can assist in establishing the diagnosis. Ocular ultrasound is very useful in the detection of optic disc drusen, including buried drusen. In our patient, a second ophthalmologic consultation revealed optic disc drusen, as also demonstrated by ocular ultrasonography.

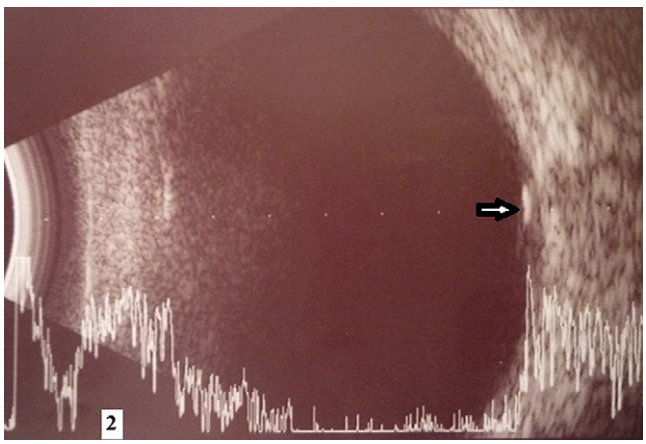

Figure 2. Ocular ultrasonography of the patient demonstrates optic disc drusen in the right eye (arrow), with the same condition in the left eye.

In conclusion, we report on an uncommon case of optic disc drusen and idiopathic intracranial hypertension appearing simultaneously. Optic disc drusen may coexist with optic disc edema. To differentiate optic disc drusen from papilledema is especially important in pediatric patients, to avoid overlooking important neurologic disorders such as idiopathic intracranial hypertension. To the best of our knowledge, this patient is the youngest so far described with coexisting optic disc drusen and idiopathic intracranial hypertension.

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