

## IIH with normal CSF pressures?

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Idiopathic intracranial hypertension (IIH) is a condition of raised intracranial pressure (ICP) in the absence of space occupying lesions. ICP is usually measured by lumbar puncture and a cerebrospinal fluid (CSF) pressure above 250 mm H<sub>2</sub>O is one of the diagnostic criteria of IIH. Recently, we have encountered two patients who complained of headaches and exhibited disc swelling without an increased ICP. We prescribed acetazolamide and followed both patients frequently; because of the definite disc swelling with IIH related symptoms. Symptoms and signs resolved in both patients after they started taking acetazolamide. It is generally known that an elevated ICP, as measured by lumbar puncture, is the most important diagnostic sign of IIH. However, these cases caution even when CSF pressure is within the normal range, that suspicion should be raised when a patient has papilledema with related symptoms, since untreated papilledema may cause progressive and irreversible visual loss.

**Key words:** Acetazolamide, idiopathic intracranial hypertension, normal pressure pseudotumor cerebri, papilledema

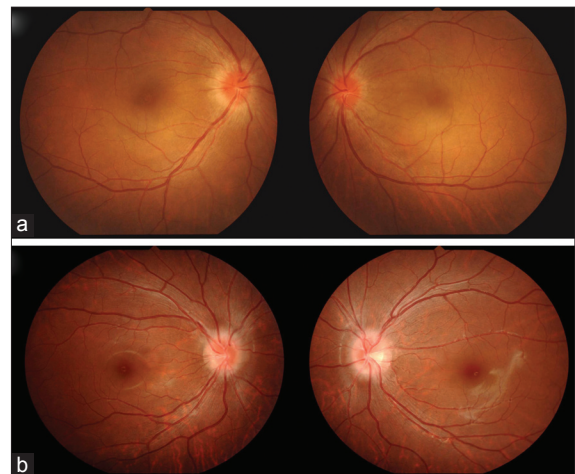
Idiopathic intracranial hypertension (IIH) is a condition of raised intracranial pressure (ICP) in the absence of a space-occupying lesion.<sup>[1]</sup> IIH patients usually present with typical symptoms and signs of increased ICP, such as, a headache, vomiting, blurred vision, and papilledema.<sup>[1]</sup> ICP is usually measured by lumbar puncture and a cerebrospinal fluid (CSF) pressure of above 250 mm H<sub>2</sub>O is one of the diagnostic criteria of IIH.<sup>[1]</sup> However, some patients do not fulfill diagnostic criteria, as demonstrated by some reported variants of IIH.<sup>[2,3]</sup> Recently, we encountered two Korean patients with a headache and other IIH related symptoms with bilateral disc swelling but without ICP elevation. Here, we introduce these two cases of IIH patients whose symptoms were relieved by ICP-lowering medication.

### Case Report

#### Case 1

A 52-year-old woman presented with blurred vision and headache. She said her headache started abruptly a week

previously and that this co-occurred with impaired color perception and blurred vision. When she visited our office, she still complained of blurred vision, especially in her left eye, but her vision was 20/20 and color vision was normal in both eyes. Both of her optic discs were swollen and Goldmann perimetry showed enlarged blind spots bilaterally [Figs. 1a and 2a]. Lumbar puncture was performed and the opening pressure was checked at 130 mm H<sub>2</sub>O; the CSF composition was normal. Brain magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) neuroimaging results were also normal; and no empty sella, globe flattening, or venous narrowing was detected by brain imaging. She was on no medication regimen, and had no other significant past medical history. We recommended another lumbar puncture, but she refused; and thus, because of the definite bilateral disc swelling and headache, which are typical features of IIH, we prescribed acetazolamide 500 mg twice daily. A week later, she told us that her headache and blurred vision had subsided, and thus, we gradually tapered the dosage during follow-up. At last visit, 4 months after the onset of her symptoms, both optic discs were normal without any swelling or temporal pallor and she no longer complained



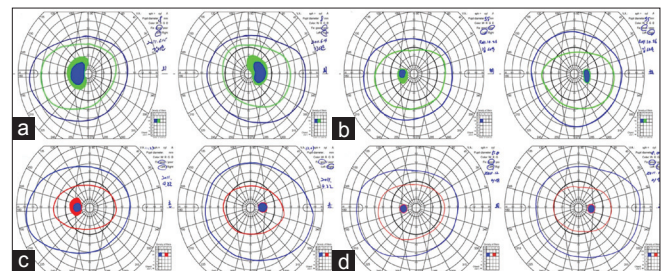
**Figure 1:** Initial fundus photographs of the patients. Bilateral optic disc edema with surrounding retinal nerve fiber layer swelling in a 52-year-old woman; who presented with blurred vision, impaired color perception, and headache. (a) Bilateral disc swelling with a blurred disc margin in a 15-year-old girl who presented with headache, pulsatile tinnitus, and transient visual obscuration (b)

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**Figure 2:** Visual field tests of the patients before and after treatment with acetazolamide. Initial visual fields showing the presence of enlarged blind spots in both eyes (a and c) Follow-up visual fields demonstrating diminished enlarged blind spot sizes after treatment (b and d)

of a headache or visual symptoms. The sizes of enlarged blind spots were prominently reduced on follow-up Goldmann perimetry [Fig. 2b].

### Case 2

A 15-year-old girl presented with a headache, pulsatile tinnitus, and transient visual obscuration of 2 months duration. She had bilateral disc swelling with a blurred disc margin and Goldmann perimetry showed enlarged blind spots in both eyes [Figs. 1b and 2c]. The lumbar CSF opening pressure was 200 mm H<sub>2</sub>O and CSF composition was normal. Brain MRI/MRV was also normal with no empty sella, globe flattening, or venous narrowing. A few days after the initial lumbar puncture, the puncture was repeated, and the rechecked CSF opening pressure was 205 mm H<sub>2</sub>O. Even though her ICP was still within the normal range, IIH was strongly suspected since she had several typical symptoms, and she told us her headache improved after the lumbar punctures. We prescribed acetazolamide 250 mg twice daily and made frequent follow-ups. Her symptoms improved after taking acetazolamide and Goldmann perimetry returned to normal [Fig. 2d]. Three months after her first visit, she had minimal swelling of both optic discs, but did not have headache or tinnitus. Currently (at 7 months after her initial visit), she is on acetazolamide 250 mg twice daily. We intend tapering the drug dosage when the disc swelling and symptoms resolve.

## Discussion

Johnston *et al.*, reported a series of atypical IIH patients, one of which was a 13-year-old boy whose disc edema rapidly resolved after lumboperitoneal shunt insertion even though his initial CSF pressure was normal.<sup>[3]</sup> Subsequently, Green *et al.*, reported the case of an 18-year-old woman IIH patient with a normal ICP, and introduced the term “normal pressure IIH” to describe this variant of IIH.<sup>[2]</sup> The mechanism responsible for this IIH variant is not known, but it has been proposed that patients may have different susceptibilities to ICP levels.<sup>[2]</sup>

Repeat lumbar puncture or 24-h ICP monitoring is occasionally suggested in patients who are suspected of having IIH but with a normal ICP by single lumbar puncture.<sup>[1]</sup> We recommended a second lumbar puncture to both of our patients, but our adult patient refused, and repeat lumbar puncture in the 15-year-old girl confirmed a normal ICP. We had considered continuous ICP monitoring in both cases, but we decided to start treatment first because they complained of a headache and showed definite

bilateral disc swelling with related visual field defects.

Papilledema is a warning sign of an elevated ICP and ophthalmologists should be alert when a patient has papilledema without any symptoms. It is known that papilledema is usually reversible with appropriate treatment in IIH patients, but untreated papilledema can result in progressive and irreversible visual loss.<sup>[4]</sup> IIH patients with persistent signs and symptoms can be treated using medical or surgical approaches, and carbonic anhydrase inhibitors, such as, acetazolamide, are perceived as the main medical treatment of IIH.<sup>[5]</sup> Surgical interventions, such as ventriculoperitoneal/lumboperitoneal shunting or optic nerve sheath fenestration, are currently the mainstay treatments when medical therapy is insufficient.<sup>[5]</sup> As both of our patients responded well to acetazolamide, surgical intervention was not considered in our patients.

Prompt diagnosis and treatment are essential in IIH patients to preserve vision. Although increased ICP, defined as an opening pressure of over 250 mm H<sub>2</sub>O, is one of the diagnostic criteria of IIH, it should be kept in mind that some patients may have discs that are more susceptible to lower ICP than others. Even when a patient's ICP is within the normal range, the possibility of IIH should always be considered in a patient with typical clinical features of IIH, such as, papilledema, a headache, pulsatile tinnitus, and blind spot enlargement by visual field testing.

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