



Idiopathic intracranial hypertension without headache: A case report and literature review

Baş ağrısız idiopatik intrakranial hipertansiyon: Olgu sunumu ve literatürün gözden geçirilmesi

Yasemin EREN,¹ Naciye KABATAŞ,² Neşe GÜNGÖR YAVAŞOĞLU,¹ Selim Selçuk ÇOMOĞLU¹

Summary

In this article, a 23-year-old man who has an idiopathic intracranial hypertension (IIH) case is presented with blurred vision and diplopia, without headaches is reported. Although headache is the most common symptom seen in IIH, sometimes it may not be observed clinically. This situation is defined more in males in young and children and thin patients. Crucial point here is that they are being presented with serious visual evidence, so they must be aggressively treated.

Keywords: Headache; idiopathic intracranial hypertension; men; papil edema.

Özet

Bu yazıda baş ağrısız olmaksızın, bulanık görme ve diplopi ile presente olan, 23 yaşında erkek İdiopatik intrakranial hipertansiyon (İİH) olgusu bildirilmiştir. İİH'de baş ağrısı en sık görülen semptom olmasına rağmen, bazen klinik olarak görülmeyebilir. Bu durum daha çok erkeklerde gençler özellikle çocuklarda ve zayıf hastalarda tanımlanmıştır. Buradaki önemli nokta bu hastalar ciddi görsel bulgularla presente olmaktadır, bu nedenle agresif tedavi edilmelidirler.

Anahtar sözcükler: Baş ağrısı; idiopatik intrakranial hipertansiyon; erkek; papil ödem.

Introduction

Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri (PTS) is characterized by an increase in cerebrospinal fluid (CSF) without occupying lesion in the brain or without ventricular dilatation. While its incidence is 1–2 at 100.000, it rises to 19 at 100,000 in fertile obese female population.^[1] Its etiopathogenesis is still unknown exactly. It may be due to primary (IIH), or secondary causes. Friedman and et al., recently introducing a new terminological approach, propose to assess the patients who have idiopathic and secondary caused intracranial pressure increase under the PTS syndrome (PTSS) umbrella.^[2] For IIH, most obvious risk factor is female gender and obesity. Secondary PTS may not be clinically distinguished from IIH; venous system abnormalities, medication toxicity, and a wide variety of systemic diseases such as risk

profile leads to increased intracranial pressure.^[3]

IIH diagnosis is made according to modified Dandy criteria. These are 1. Signs and symptoms due to intracranial increased pressure (headache, papilledema, visual signs and symptoms, tinnitus, nausea, vomiting); 2. Except the 6th nerve paralysis, lack of finding of lateralization in neurological examination result; 3. A reason to increase intracranial pressure in neuroimaging; 4. Having CSF opening pressure greater than 25 cm water and having CSF biochemical and cytology normal. 5. Unavailability of any other factor explaining the intracranial pressure increase.^[4] However with the diagnostics technology over the years, and revised new approaches in understanding the disease it has been revised.^[5] Outside the criteria defined in the literature, IIH cases not accompanied

¹Department of Neurology, Dışkapı Yıldırım Beyazıt Training and Research Hospital, Ankara, Turkey

²Department of Ophthalmology, Dışkapı Yıldırım Beyazıt Training and Research Hospital, Ankara, Turkey

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Correspondence (İletişim): Dr. Yasemin Eren. Dışkapı Yıldırım Beyazıt Eğitim ve Araştırma Hastanesi, Nöroloji Bölümü, İrfan Baştuğ Cad. Altındağ, Ankara, Turkey.

Phone (Tel): +90 - 312 - 596 20 00 **e-mail (e-posta):** yeren.72@hotmail.com

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Figure 1. In the Fundus examination, in the right eye grade 4 papilledema, on the left eye grade 4 papilledema and macular edema.

by headache or papilledema have been reported. In this paper, an IIH case who applied with blurred vision and diplopia, without accompanying headache will be presented.

Case Report

24-year-old male, with blurred vision began one week ago and diplopia has applied. Headache, nausea, tinnitus did not exist, but he described neck pain that lasted about ten days about 1 month ago. His history did not have systemic diseases, drug use, weight change. In his neurological examination both eyes were limited to outward vision and outward glance was prognosing horizontal diplopia. During application, best corrected visual acuity was 20/20 in

the right eye and 20/50 on the left eye. Microscopic findings were natural and intraocular pressure was within normal limits. In the Fundus examination, in the right eye grade 4 papilledema, on the left eye grade 4 papilledema and macular edema is revealed. In the optical coherence tomography (OCT) done, optical disc bulging in both eyes and subretinal edema in the macula in the left eye were observed. Expansion in the blind spot of vision area and central vision loss were observed. In lumbar puncture CSF opening pressure was 330 mm /water, and closing pressure was measured as 250 mm /water. In cranial MRI and MR venography pathology was not detected. Routine blood tests were in the normal range. 1500 mg/day of acetazolamide, topiramate 50 mg/day is started to patient. At the end of the first month, in the controls visual acuity reached the 20/20 level. In the fundus examination, papilledema level dropped to grade 3 level in both eyes and in the left eye macular edema was lost and in OCT examination macula was seen attached.

Discussion

Headache is the most common symptom in IIH, seen in 90% of patients.^[3] However, less has been reported in men than in women.^[6] It does not have a specific

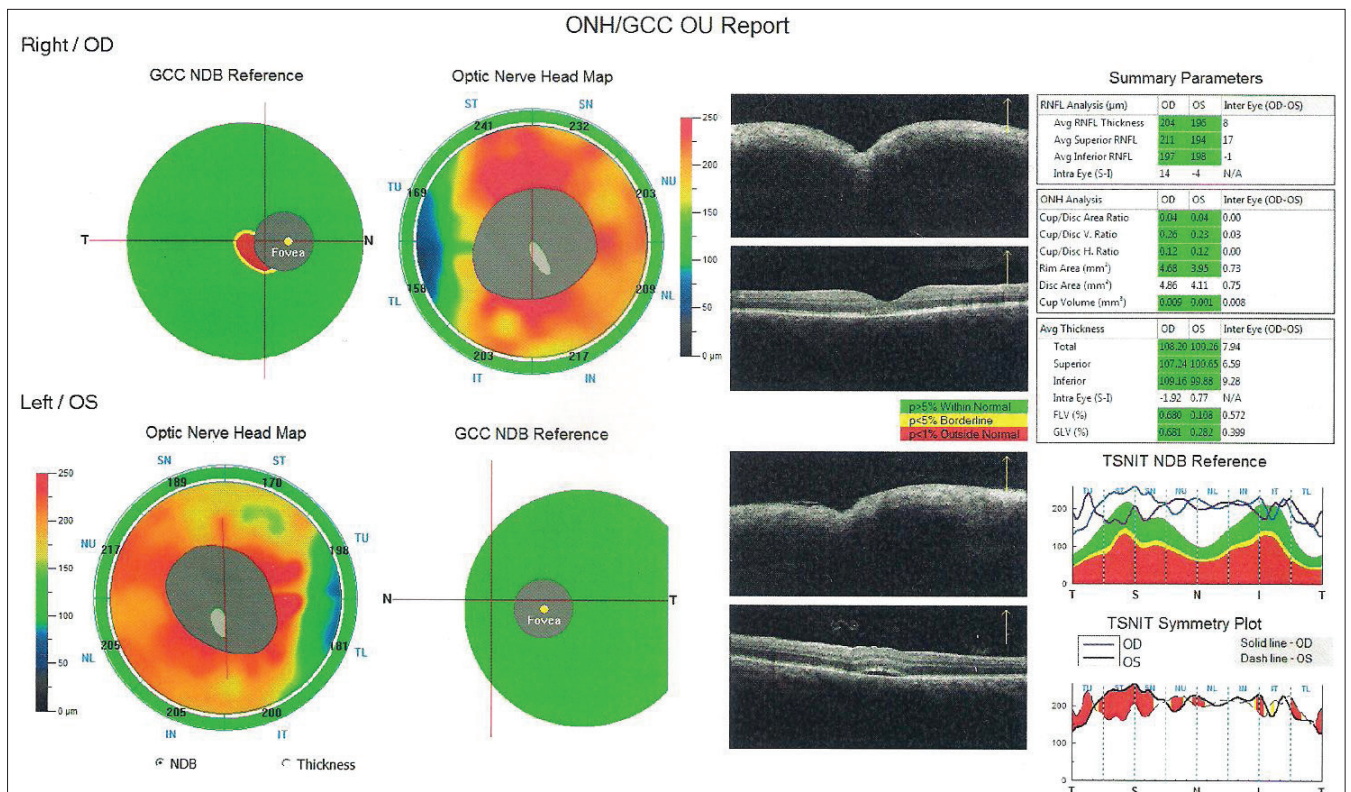


Figure 2. In the optical coherence tomography (OCT), optical disc bulging in both eyes and subretinal edema in the macula in the left eye.

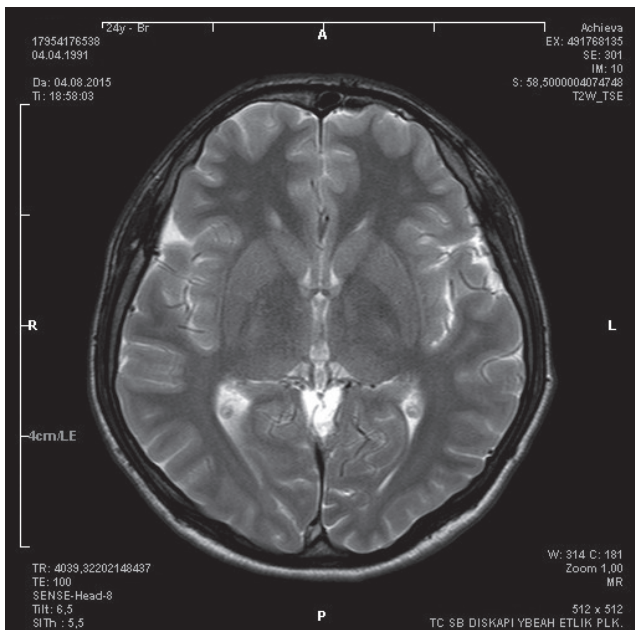


Figure 3. Normal MRI T2 axial image.

characteristic; it can be bilateral, frontal or retroocular. Its severity varies from mild to severe. It worsens on waking up and increases with movement. Usually it is characterized as throbbing or pulsating. Pain accompanied by migraine like nausea and vomiting may be observed. Primary headaches of migraine or tension headache may accompany the PTS. Further, PTS is included in the new daily persistent headache differential diagnosis.^[4] Cervical or back pain may occur in patients. Neck stiffness depends on the stress due to CSF pressure increase in the spinal nerve sheath.^[5] In our patient although he did not have headache, he had severe neck pain.

Bruce et al., in a study conducted with 66 men and 655 women participated, found that there is less headache in men as the initial symptom (55% vs. 75%), more often visual symptoms (35% vs. 20%). They reported that visual symptoms are more serious compared to women.^[6] IIH cases without headaches are rather identified in children. In the publications, PTS cases without headaches has been suggested in the rate of 9–38% Lim and et al., have reported higher, around 29%, PTS patients without headaches compared to other publications. 7 of these patients were female (58.3%), and 5 of them were men (41.7%). When groups of with headache and without headache are compared; patients without headache were being presented with younger, more neurological symptoms, severe vision loss and visual field defects.^[7] In a retrospective study on 152 children suffering

from PTS, in 22 of the patients (14.5%), headache was not detected. The ratio of girls and boys in these patients are (13; 9) (59.1%: (48.1%). It has been reported that these patients have younger and lower body mass index (BMI). In addition, CSF opening pressure is not any different from patients with headache. Our patient was also a male and in the group of young adults. BMI=19.80 and he was not obese. Similar to the literature, visual symptoms were noisy and serious. Moreover, CSF opening pressure in LP was 330 mm and similar with patients who had headache.^[8]

Besch and et al., reported PTS accompanied by vision loss in two female prepubertal patients taking growth hormone therapy. These patients did not describe nausea and vomiting. One patient had back pain and intermittent eye blackened. These patients were not obese.^[9] Barnett and et al., reported a child with PTS having severe visual loss with nephrotic syndrome presented without headaches.^[10]

Various opinions have been proposed to explain the clinical differences in IIH between patient groups with or without headaches. It has been reported that it could be associated with ventricular compliance and increased pressure process.^[7] This variability in symptoms is reported to be more similar to differences in headache threshold of women and men. Reasons such as having migraine and TTH more common in women, having greater total time of painful mechanical stimulation in women compared to men associate women to have lower threshold to pain than men. In chronic stimulus IIH such as increased intracranial pressure, also sexual differences may play a role in the headache.^[6] Moreover, in the studies mentioned earlier rate of not having headaches in IIH in childhood, while the ratio in women is higher the ratio in men is observed less. We think that this is in IIH in women beyond hormonal factors both etiological factors and it's a reason that changes the prevalence of headache frequency.

As a result, besides classic symptoms in IIH, where headache or papilledema may not be observed, it may present with different symptoms in women and men. Although headache is not observed frequently clinically in men, a closer and aggressive monitoring and treatment must be made as visual symptoms are serious.

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