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CASE REPORT

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

Başağrısız idiopatik intrakranial hipertansiyon: Olgu sunumu ve literatürün gözden geçirilmesi

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Summary

This study reports the case of a 23-year-old man with idiopathic intracranial hypertension (IIH) who presented with blurred vision and diplopia, without accompanying headache. Although headache is the most common symptom associated with IIH, occasionally, it may not be observed clinically. This situation is more frequently observed in males, young adults, children, and in patients with low body mass index. This case highlights a crucial aspect; patients who present with serious visual symptoms without headache must be treated aggressively because vision loss will develop rapidly.

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

Özet

Bu yazıda başağrısı olmaksızın, bulanık görme ve diplopi ile presente olan, 23 yaşında erkek İdiopatik intrakranial hipertansiyon (İİH) olgusu bildirilmiştir. İHH'de başağrısı 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 olmaktadırlar, bu nedenle agresif tedavi edilmelidirler.

Anahtar sözcükler: Başağrısı; erkek; idiopatik intrakranial hipertansiyon; 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. The incidence of IIH is 1-2 per 100.000 in the general population, but it increases to 19 per 100,000 in the fertile obese female population.[1] Its exact etiopathogenesis remains unknown. It may be due to primary (IIH) or secondary causes. Friedman et al.[2] recently introduced a new terminology and proposed to assess patients with increase in intracranial pressure due to idiopathic and secondary causes under the umbrella term PTS syndrome (PTSS). For IIH, the most obvious risk factor is female sex 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 is diagnosed according to the following modified Dandy criteria: 1) signs and symptoms due to intracranial increased pressure (headache, papilledema, visual signs and symptoms, tinnitus, nausea, vomiting), 2) excepting a 6th nerve paralysis, lack of finding of lateralization in the neurological examination result, 3) a reason for increased intracranial pressure in neuroimaging, 4) having a CSF opening pressure greater than 25 cm of water as well as normal CSF biochemical and cytological results, and 5) unavailability of any other factor explaining the increase in intracranial pressure.[4] However, these criteria have been revised owing to advances in diagnostic tech-

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nology over the years and new approaches in understanding the disease.[5] Outside the criteria defined in the literature, IIH cases not accompanied by headache or papilledema have been reported. Herein, we report a case of a patient with IIH presenting with blurred vision and diplopia, without accompanying headache.

Case Report

A 24-year-old male presented with blurred vision, which began 1 week ago, as well as diplopia. He did not exhibit headache, nausea, and tinnitus but described neck pain that persisted for approximately 10 days approximately 1 month ago. He had no history of systemic disease, drug use, or weight change. Neurological examination revealed that both eyes were limited to outward vision and outward glance was prognosing horizontal diplopia. During application, best corrected visual acuity was 20/20 for the right eye and 20/50 for the left eye biomicroscopic findings were normal, and intraocular pressure was within normal limits. Fundus examination revealed grade 4 papilledema in the right eye and grade 4 papilledema and macular edema in the left eye (Fig. 1). Optical coherence tomography (OCT) showed a bulging optic disc in both eyes and subretinal edema in the macula of the left eye (Fig. 2). Expansion in the blind spot and central vision loss were observed in visual field examination. In a lumbar puncture, the CSF opening pressure was 33 cm H_2O and the closing pressure was 25 cm H_2O. Cranial magnetic resonance imagining (MRI) and MRI venography were normal (Fig. 3). Routine blood tests were in the normal range. The patient was started on 1500 mg/day of acetazolamide and topiramate 50 mg/day. In the follow-up at the end of the first month, his visual acuity was 20/20. A fundus examination revealed that the papilledema had improved to grade 3 in both eyes and the macular edema in the left eye had disappeared; OCT showed that the macula was attached.

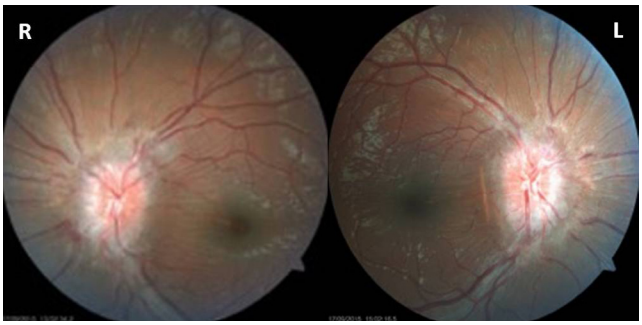

Figure 1. Fundus examination reveals grade 4 papilledema in the right eye and grade 4 papilledema and macular edema in the left eye.

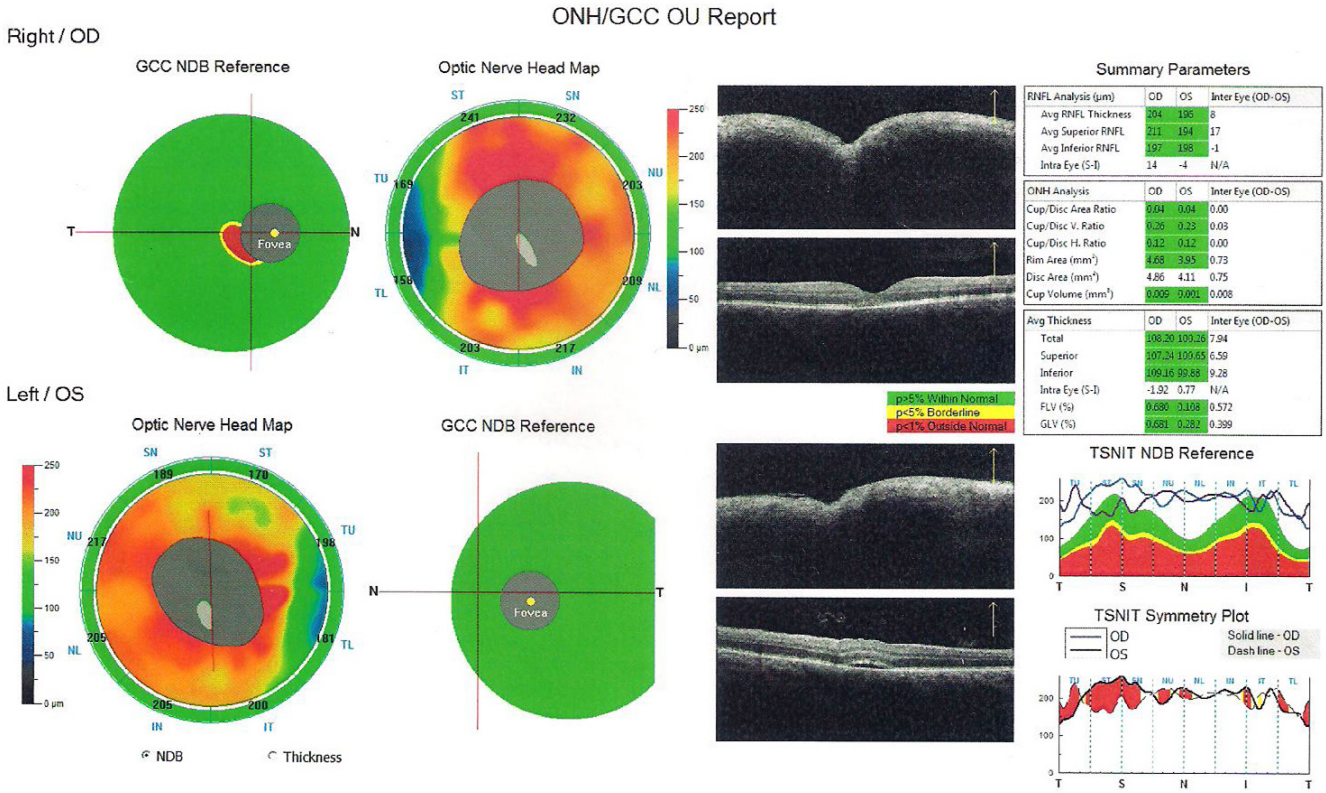

Figure 2. Optical coherence tomography (OCT) reveals a bulging optic disc in both eyes and subretinal edema in the macula of the left eye.

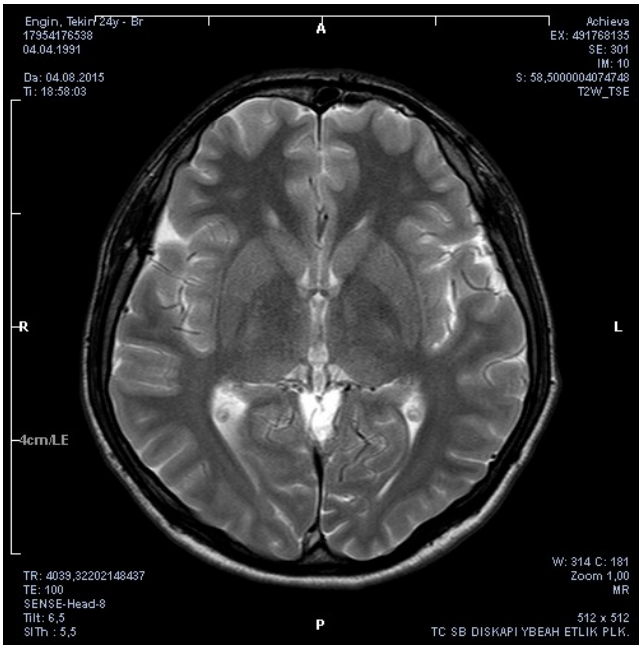

Figure 3. Normal T2 axial Magnetic Resonance Imaging (MRI).

Discussion

Headache is the most common symptom in IIH, occurring in 90% of patients.[3] However, it has been reported to occur less frequently in men than in women.[6] Headache has no specific characteristics; it can be bilateral, frontal, or retroocular. Its severity can vary from mild to severe and can worsen on waking up and increases with movement. Usually, it is characterized as throbbing or pulsating. Pain may be accompanied by migraine, nausea, and vomiting. Primary headaches of migraine or tension may accompany the PTS. Furthermore, PTS is included in the differential diagnosis of new daily persistent headache.[4] Cervical or back pain may occur in patients. Neck stiffness depends on the stress due to increased CSF pressure in the spinal nerve sheath.[5]Although our patient did not have headache, he had severe neck pain.

In a study conducted with 66 male and 655 female participants, Bruce et al. found that headache as the initial symptom occurred less frequently in men than in women (55% vs. 75%) and visual symptoms were more frequent (35% vs. 20%). They reported that visual symptoms were more serious in men than in women.[6] IIH cases without headaches are rather identified in children. Research has suggested that the prevalence rate of PTS cases without headaches is 9%–38%. Lim et al. have reported a higher rate, approximately 29%, of patients with PTS without headaches [7 females (58.3%), 5 males (41.7%)] in comparison to other studies. When groups of patients with headache and without headache are compared, patients without headache were younger and presented with more neurological symptoms, severe vision loss, and visual field defects.[7] In a retrospective study of 152 children with PTS, in 22 of the patients (14.5%), headache was not reported. The female-to-male ratio in these patients was 13:9 (59.1%:48.1%). It has been reported that these patients were younger and had a lower body mass index (BMI). In addition, the CSF opening pressure was not different from that in patients with headache. Our patient was also a male who belonged to the group of young adults. He had a BMI of 19.80 and was not obese. Similar to the literature, visual symptoms were noisy and serious. Moreover, the CSF opening pressure at LP was 33 cm H_2O, which was similar to that reported in patients who had headache.[8]

Besch et al.[9] 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. Barnet et al.[10] reported a child with PTS having severe vision loss with nephrotic syndrome who presented without headache.

Various opinions have been proposed to explain the clinical differences in IIH between patient groups with or without headaches. It has been reported that these differences could be associated with ventricular compliance and duration of increased intracranial pressure.[7] This variability in symptoms is reported to be more similar to differences in the headache threshold of women and men. Migraine and TTH are more common in women, with greater total time of painful mechanical stimulation in women compared with men, associate women to have lower threshold to pain than men. The more frequent occurrence of headache in women in IHH may be related to continuous stimulus such as increased intracranial pressure.[6] We think that this situation in IIH in women is beyond both hormonal and etiologic factors, and it is a reason that changes the prevalence of headache frequency.

Therefore, apart from classic symptoms in IIH, wherein headache or papilledema may not be observed,

IIH may present with different symptoms in women and men. Although headache is not frequently observed clinically in men, a closer and aggressive monitoring and treatment must be established as the visual symptoms are serious.

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