



Dural Ectasia of the Optic Nerve and Unilateral Proptosis-Two Rare Comorbidities Associated with Idiopathic Intracranial Hypertension

İdiopatik İntrakranial Hipertansiyonda Nadir Görülen İki Ayrı Komorbidite Optik Sinir Dura Ektazisi ve Unilateral Proptozis

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Summary

A 24-year-old woman with a recent history of visual abnormality (obscurations) had bilateral optic disc swellings, dilatation of the optic nerve sheaths, monocular proptosis, and increased cerebrospinal fluid pressures. She was diagnosed as idiopathic intracranial hypertension (IIH). IIH can present with both papilledema and dural ectasia although the latter is usually seen as an isolated entity. Various conditions also have been implicated in IIH. However, apart from female sex, recent weight gain and obesity, there are no proven associations. In this case report, we present a case of IIH associated with two different pathologies: dural ectasia of the optic nerve sheaths and monocular proptosis. (Turk J Ophthalmol 2013; 43: 297-300)

Key Words: Idiopathic intracranial hypertension, dural ectasia of the optic nerve sheaths, proptosis

Özet

Görme şikayetleri nedeniyle başvuran 24 yaşında bir bayan hastanın yapılan muayenesinde bilateral optik disk ödemi, optik sinir kılıflarında genişleme, monoküler proptozis ve beyin-omurilik sıvı basıncında artış saptanmıştır. Hastaya idiyopatik intrakranial hipertansiyon (IHH) tanısı konulmuştur. IHH, papilödem ve dural ektazi ile birlikte görülebilmekle beraber, optik sinir kılıfında dural ektaziye izole bir bulgu olarak rastlanabilmektedir. IHH ile birlikte çeşitli patolojilere rastlanılabilmektedir. Ancak bayan cinsiyeti, son dönemlerde kilo alma ve obezite dışında, bu hastalık ile ilintiler henüz ispatlanamamıştır. Bu olgu sunumunda IHH'ye eşlik eden iki farklı patoloji; optik sinir kılıfında dural ektazi ve monoküler proptozis rapor edilmektedir. (Turk J Ophthalmol 2013; 43: 297-300)

Anahtar Kelimeler: İdiyopatik intrakranial hipertansiyon, optik sinir kılıfı dural ektazisi, proptozis

Introduction

Idiopathic intracranial hypertension (IHH), also called "pseudotumor cerebri", presents with signs and symptoms of elevated intracranial pressure (ICP). Headache, nausea, and vomiting are common symptoms. Transient visual obscurations due to papilledema, diplopia secondary to abducens nerve paresis, visual field loss, pulsatile tinnitus, and dizziness may be additional complaints.¹⁻³ Most patients with IHH have papilledema.

Dural ectasia of the optic nerve is saccular dilatation of the nerve sheath. It is characterized by expansion of the cerebrospinal fluid (CSF) around the optic nerve without orbital

or cerebral neoplasm and inflammation.^{4,5} Although the term dural ectasia is preferred to describe the optic nerve sheath dilation, optic hydrops, primary cyst of the optic nerve sheath, patulous subarachnoid space, cystic hygroma, arachnoid cyst, and perioptic subdural hygroma terms were all used to describe this entity since its first description in 1918.^{4,6,7} Magnetic resonance imaging (MRI) of the patients with dural ectasia reveals tube-like enlargement of the optic nerve.^{4,7}

Proptosis is an abnormal protrusion of the globe which may be caused by retrobulbar lesion or, less frequently, a shallow orbit. Space occupying lesions within the muscle cone cause axial proptosis, whereas extraconal lesions usually give rise to eccentric

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proptosis. Facial asymmetry, severe ipsilateral enlargement of the globe, ipsilateral lid retraction or contralateral enophthalmus can cause a false impression of proptosis.⁸

Various conditions have been implicated in IHH. Female sex, recent weight gain, and obesity are proven associations with IHH. There are some other conditions reported which are not proven associations.

In this case report, we present a patient with visual disturbances related with IHH associated with dural ectasia and monocular proptosis.

Case Report

A 24-year-old woman presents with symptoms of transient visual “dimming” over the past 1 month. The episodes are bilateral, occur several times per week, and last for approximately 10 seconds. This was the only complaint of the patient and she neither had headache or other neuro-ophthalmic complaints. She denied to take any kind of medicines including oral contraceptives.

On our examination, the left eye had a proptotic appearance. Hertel measurements were found as 19 mm in the right eye and 23 mm in the left eye. The proptosis was axial, non-pulsatile, and there was no bruit. Her visual acuity (VA) was 20/20 OU. Both pupils reacted briskly to light and constricted to near stimulation. The ocular alignment was normal and the eye movements in all cardinal positions were within the normal range. There was no history of diplopia. The anterior segment structures were normal in slit-lamp biomicroscopy. Funduscopy revealed a bilateral swelled, hyperemic, enlarged optic discs with blurred margins (Figure 1). She could identify all of the Ishihara color plates OU. Visual fields showed bilateral blind spot enlargement with no additional defect (Figure 2).

In our patient, blood tests included complete blood counts, routine chemistry, coagulation panel, erythrocyte sedimentation rate, CRP, RF, ANA, and thyroid function tests which were all normal.

In this patient with suspected papilledema, neuroimaging has been performed immediately, prior to lumbar puncture (LP), to exclude disorders with a risk for herniation during the procedure and to search for any secondary cause of increased ICP.

In MRI study, there was no evidence of a space-occupying lesion, but the neuroimaging revealed a dilated optic nerve sheath surrounding the optic nerves (Figure 3 a-b). T2-weighted and gadolinium-enhanced MRIs exhibited flattening of the posterior sclera, perioptic dilation of the CSF space, tortuosity of the orbital optic nerve and protrusion into the vitreous, and enhancement of the optic discs. Dural ectasia of the optic nerve is considered in diagnosis.

LP was performed with an opening pressure of 22 cm of H₂O. CSF analysis showed protein level of 51.3 mg/dL and no white blood cells.

The patient received diuretics therapy (Acetazolamide 250 mg, 4x1, per os, with potassium supplement, and her visual disturbances improved over the following several days even

though the papilledema persisted. Repeat Goldmann perimetry also documented a decrease in the enlarged blind spot (Figure 2 b). The change in optic disc was prominent after treatment (Figure 1b). Informed consent was obtained from the patient.

Discussion

Bilateral optic disc edema in the presence of normal optic nerve function frequently indicates an increased ICP. Diagnosis of IHH is based on symptoms and signs attributable to increased

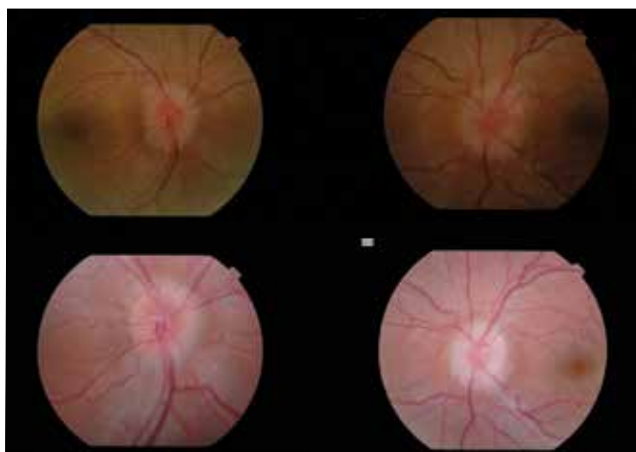


Figure 1. Fundus photography showing bilateral hyperemic, enlarged optic discs with blurred margins. b; disc changes after treatment

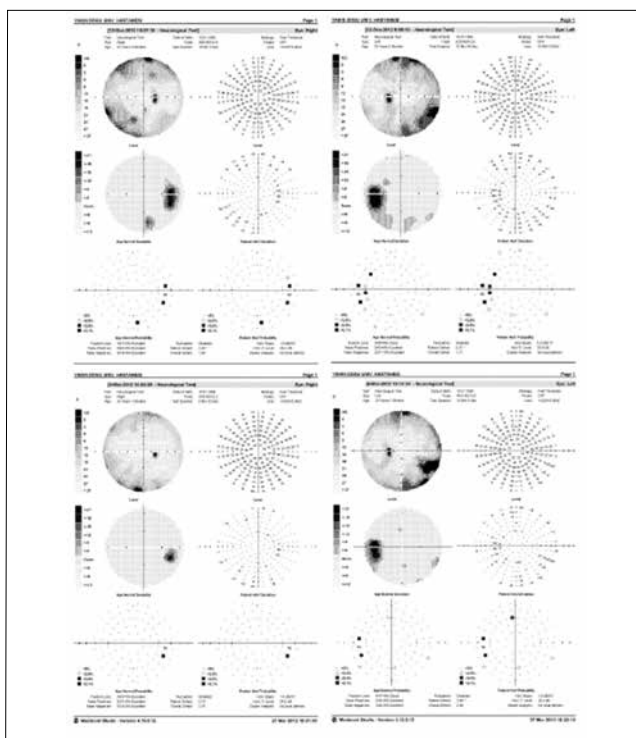


Figure 2 a-b. Standard automated perimetry showing enlarged blind spots in both eyes.(before, after treatment)

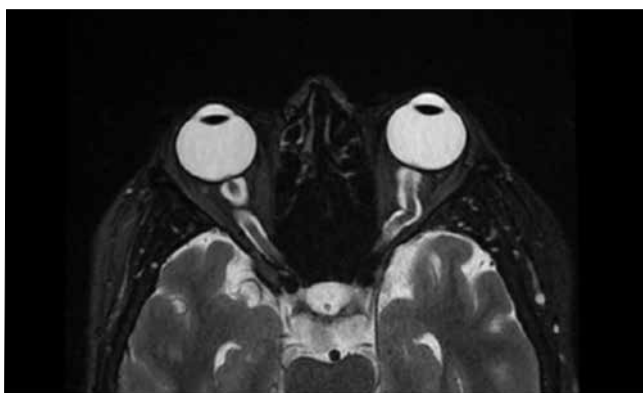


Figure 3 a. MRI revealing the dural ectasia of the optic nerve.(a; axial MRI scan)

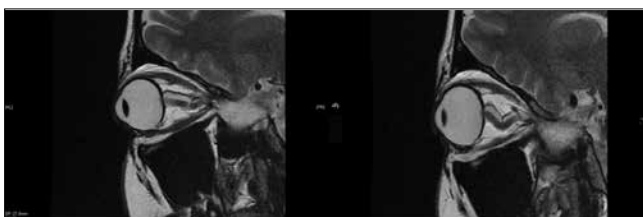


Figure 3 b. MRI revealing the dural ectasia of the optic nerve.(b; sagittal MRI scan)

ICP, elevated CSF pressure, normal CSF profiles in the absence of ventriculomegaly, mass, or vascular lesions on neuroimaging, and no other etiology of intracranial hypertension identified.⁹ However, various other probable causes of increased ICP should be considered including benign or malignant mass lesions of the brain, venous sinus thrombosis, infectious, inflammatory and malignant disorders involving the meninges.

In this instance, a previously healthy young woman developed visual obscurations. Given the appearance of bilateral papilledema, increased ICP with or without a space-occupying lesion was a prime suspicion. In view of the visual symptoms and fundus appearance without other focal neurologic signs in a young woman, IHH should be the top differential.

IHH is not a rare disease. Population-based studies have documented annual incidence rates of 1-2 per 100000 people.¹⁰ The pathophysiology of IHH is still unknown.² There are various proposed hypotheses for the IHH which are mostly concerning deranged CSF homeostasis, and include diffuse brain edema, excessive CSF production, reduced CSF absorption, and increased cerebral venous pressure, which all require further elucidation.^{2,4} Various conditions also have been implicated in IHH.² Case reports have associated IHH with iron deficiency anemia, hypothyroidism, polycystic ovary syndrome, and use of steroids, vitamin A, and oral contraceptives, however, apart from female sex, recent weight gain, and obesity, there are no proven associations.²⁻⁴

The report concerning diagnostic criteria for IHH has proposed that the lumbar opening pressure should be greater than 250 mm of water measured with the patient in the lateral decubitus

position.¹¹ The values between 200 and 250 mm of water are accepted as nondiagnostic.¹² However, LP may provide an inaccurate measure of ICP, given its known moment-to-moment fluctuations and the effects on ICP of straining, positioning, and pharmacologic sedation.¹³ The CSF pressure naturally fluctuates just like intraocular pressure. Although we recommended a repeat LP which is usually necessary if the opening pressure is low in the appropriate clinical settings to prevent obtaining the false negative test results because of the fluctuation phenomena, the patient refused the repeat LP measurements with insistence. In this manner, we had to diagnose this patient empirically. The clinical picture of the optic nerve heads, symptoms of patient and the prompt response of patient to typical IHH treatment were all strong indicators that this patient could be accepted as IHH. Furthermore, the generally accepted normal range in adults is 80–200 mm of water.¹⁴ In this case, we could not classify this patient's CSF pressure as within normal limits.

There are some questionnaires to identify the symptoms and coexisting medical conditions associated with IHH.¹⁵ Ninety percent of the IHH patients were women; the mean age was 33. Obesity and recent weight gain were much more common among patients than controls. Symptoms most commonly reported by IHH patients were headache (94%), transient visual obscurations (TVO) (68%), and intracranial noises (ICN) (58%). Other common symptoms observed in IHH included photopsia (54%), diplopia (38%), and visual loss (30%).¹⁵

Daily occurrence of these symptoms was much more common among patients than controls. Controls also reported these and other IHH symptoms, but at lower frequencies. Several conditions previously associated with IHH were no more common in patients than in controls including iron deficiency anemia, thyroid disease, pregnancy, antibiotic intake, and use of oral contraceptives. We conclude that previous studies of IHH, mostly uncontrolled and retrospective, have underestimated the frequency of symptoms in IHH patients and reported chance and spurious associations with common medical conditions and medications.

Radhakrishnan et al. presented a variety of additionally described risk factors of varying strength, including the coexistence of endocrinopathies, menstrual irregularities, autoimmune disorders, chronic renal failure, and concurrent medication use - such as steroids, estrogens, vitamin A, thyroid supplements and antibiotics - among patients with IHH.¹⁶ These authors have suggested that patients developing IHH may possess an underlying abnormality or predisposition which, when acting in concert with an aggravating agent such as recent weight gain or antibiotic use, produces the disease.

Papilledema and optic nerve sheath enlargement can be seen with IHH.^{17,18} There are some common features of both diseases; however, there is no significant evidence suggesting the role of raised intracranial pressure in the etiology of the dural ectasia of the optic nerve.^{4,19} Although optic nerve sheath dilation is one of the radiological features of IHH, the enlargement of the optic nerve sheath to the degree we found in our patient is unusual. A rare case of dural ectasia of the optic nerve sheath have also been

reported. In this case, a 36-year-old woman with a 3-month history of progressive visual loss had papilledema, dilatation of optic nerve sheath, and normal CSF pressure; she had been treated with acetazolamide 250 mg qid for 8 weeks elsewhere. LP was performed with an opening pressure of 22 cm of water and the CSF pressure at repeat LP was significantly low which could not be recorded.²⁰ MRI is mostly performed to exclude other intracranial pathologies which may increase ICP. However, MRI may exhibit radiographic evidence of increased ICP, which includes slit ventricle, empty sella, flattening of the posterior sclera, distension of the perioptic subarachnoid space, tortuous optic nerve, protrusion and enhancement of the optic disc.

Endocrine dysfunction is a rare but known cause of IIH in adults. A rare case of IIH in the pediatric age group associated with autoimmune hyperthyroidism have also been reported.²¹ In this case, a 12-year-old girl presented with a 3-month history of headaches. Ophthalmic examination revealed bilateral papilledema. The ocular findings were otherwise normal, with no exophthalmos. Cranial and orbital MRI was unremarkable. Lumbar CSF opening pressure in recumbent and relaxed position was elevated (31 cm water). Thyroid hormones (fT3 and fT4) were elevated, while TSH was completely suppressed. As TSH receptor stimulating antibodies (TSHR-Ab) were elevated, Graves' disease was diagnosed. Thyroid suppressive therapy with carbimazole was initiated and supplemented by propranolol. As hyperthyroidism improved over two weeks, the headaches subsided, and the papilledema slowly resolved over the next 2 months. This case illustrates that hyperthyroidism should be considered as a cause of IIH in children.

In summary, our case is an IIH associated with unilateral proptosis and bilateral prominent dural ectasia. To our knowledge, this is the first case reported with these associated pathologies together. Although the proptosis may be the only sign of the euthyroid Graves' disease and thus patient may have an endocrine dysfunction, we could not detect this clinically and serologically. In this condition, we thought that this case with these isolated pathologies should be reported for providing an additional information for the IIH literature.

References

1. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. *Brain*. 1991;114:155-80.
2. Friedman DI, Jacobson DM. Idiopathic intracranial hypertension. *J Neuro-Ophthalmol*. 2004;24:138-45.
3. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*. 2002;59:1492-5.
4. Garrity JA, Trautmann JC, Bartley GB, et al. Optic nerve sheath meningoceles. Clinical and radiographic features in 13 cases with a review of the literature. *Ophthalmology*. 1990;97:1519-31.
5. Jungkim S, Khurshid SG, Fenton S. Dural ectasia of the optic nerve sheath. *Acta Ophthalmol Scand*. 2005;83:266-7.
6. Lövblad KO, Remonda L, Ozdoba C, Huber P, Scroth G. Dural ectasia of the optic nerve sheath in neurofibromatosis type 1. CT and MR features. *J Comput Assist Tomogr*. 1994;18:728-30.
7. Lunardi P, Farah JO, Ruggeri A, Nardacci B, Ferranti L, Puzzilli F. Surgically verified case of optic sheath nerve meningocele: case report with review of the literature. *Neurosurg Rev*. 1997;20:201-5.
8. Kanski JJ. *Clinical Ophthalmology*. 5th edition. Butterworth-Heinemann International Edition. Orbit, 559.
9. Friedman DI. pseudotumor cerebri. *Neurol Clin*. 2004;22:99-131.
10. Durcan FJ, Corbett JJ, wall M. The incidence of pseudotumor cerebri: population studies in Iowa and Louisiana. *Arch Neurol*. 1988;45:875-7.
11. Corbett JJ, Mehta MP. Cerebrospinal fluid pressure in normal obese subjects and patients with pseudotumor cerebri. *Neurology*. 1983;33:1386-8.
12. Digre KB, Corbett JJ. Idiopathic intracranial hypertension (pseudotumor cerebri): a reappraisal. *Neurologist*. 2001;7:2-67.
13. Warden KF, Alizai AM, Trobe JD, Hoff JT. Short-Term Continuous Intracranial Pressure Monitoring in Presumed Idiopathic Intracranial Hypertension; *J Neuro-Ophthalmol*. 2011;31:202-5.
14. Friedman DI. Idiopathic Intracranial Hypertension With Dan and Beyond: The 2010 Jacobson Lecture, *MPH Journal of Neuro-Ophthalmology*. 2010;30:380-5.
15. Giuseffi V, Wall M, Siegel PZ, Rojas PB. Symptoms and disease associations in idiopathic intracranial hypertension (pseudotumor cerebri): A case-control study. *Neurology*. 1991;41:239-44.
16. Radhakrishnan K, Ahlskog JE, Garrity JA, Kurland LT. Idiopathic intracranial hypertension. *Mayo Clin Proc*. 1994;69:169-80.
17. Helmke K, Hansen HC. Fundamentals of transorbital sonographic evaluation of optic nerve sheath expansion under intracranial hypertension. II. Patient study. *Pediatr Radiol*. 1996;26:706-10.
18. Agid R, Farb RI, Willinsky RA, Mikulis DJ, Tomlinson G. Idiopathic intracranial hypertension: the validity of cross-sectional neuroimaging signs. *Neuroradiology*. 2006;48:521-7.
19. Shanmuganathan V, Leatherbarrow B, Ansons A, Laitt R. Bilateral idiopathic optic nerve sheath meningocele associated with unilateral transient cystoid macular edema. *Eye*. 2002;16:800-2.
20. Berker B, Hava D, Tülay K, Hayyam K. Dural ectasia of the optic nerve sheath: is it always benign? *Eye and Brain*. 2009;1:5-7.
21. Merckenschlager A, Ehrh O, Müller-Felber W, Schmidt H, Bernhard MK. Reversible benign intracranial hypertension in a child with hyperthyroidism. *J Pediatr Endocrinol Metab*. 2008;21:1099-101.