Pseudotumor Cerebri: Clinical Signs, Treatment and Prognosis

PSÖDOTÜMÖR SEREBRİ: KLİNİK BULGULAR, TEDAVİ VE PROGNOZ

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SUMMARY

Idiopathic Intracranial Hypertension (IIH) is a syndrome characterized by symptoms and signs of increased intracranial pressure without localising neurological findings in an alert and oriented patient. There is not any deformity or obstruction of the ventricular system and neuroradiological studies are normal otherwise except for increased cerebrospinal fluid pressure.

Loss of visual function is the only major form of morbidity associted with IIH.

In this study, we investigated neurological and neuroradiological signs, treatment and prognosis of 15 patients with IIH.

Key Words: Idiopathic intracranial hypertension, Etiology, Treatment

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ÖZET

İdiopatik intrakraniyal hipertansiyon (psödotümör serebri) şuuru açık ve oriente hastalarda, artmış intrakraniyal basınç semptom ve bulgularının, lokalize nörolojik bulgu olmaksızın ortaya çıkması ile karakterize bir sendromdur. Ventriküler sistemde herhangi bir deformite veya obstrüksiyon yoktur, nöroradyolojik çalışmalar normaldir.

İdiopatik intrakraniyal hipertansiyonda morbiditeyi, görme kaybı oluşturmaktadır.

Bu çalışmada idiopatik intrakraniyal hipertansiyonu olan 15 hastanın nörolojik ve nöroradyolojik bulguları ile tedavi ve prognozları sunulmuştur.

Anahtar Kelimeler: İdiopatik intrakraniyal hipertansiyon, Etiyoloji, Tedavi

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Pseudotumor cerebri describes a heterogenous group of disorders that are characterized by increased intracranial pressure (ICP) when intracranial mass lesions, obstructive hydrocephalus, intracranial infection, and hypertensive encephalopathy have been excluded (1). It's also termed Benign Intracranial Hypertension or Idiopathic Intracranial Hypertension (IIH). It was first described by Quincke in 1897. The signs and symptoms of increased ICP are headache, with nausea and vomitting; visual findings are diplopia due to sixth cranial nerve paresis, transient visual obscurations, and papilledema with its associated loss of sensory visual function (2,3).

Several mechanisms have been considered as possible explanations for the pathophysiology of IIH. These include the following: 1-An increased rate of cerebrospinal fluid (CSF) pressure, 2-A sustained increase in intracranial venous pressure, 3-A decreased rate of CSF absorbtion by arachnoid villi apart from venous occlusive disease and, 4-An increase in brain volume because of an increase in cerebral blood volume or interstitial fluid volume (4,5).

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PATIENTS AND METHODS

The study group consisted of 15 patients who were hospitalized in SSK Ankara Hospital Neurology Clinic between 1992 and 1993. The patients were evaluated retrospectively from their medical files. A detailed medical history was taken and neurologic examination was performed. Neuroophthalmological examination included snellen visual acuity and visual fields by automated perimetry was first performed on admission and then repeated. All patients had blood and CSF testing to exclude other disorders known to cause raised ICP.

All patients had computed tomography (CT), and four of them had magnetic resonance imaging (MRI). All met Dandy criteria for IIH.

RESULTS

The mean age was 32.3 with a range of 21 to 43. Of the 15 patients in the study 10 were women. The female to male ratio was 3/2.

The most common symptoms were as follows; headache (%93.3) nausea and vomiting (%53.3), diplopia (40%) (Table 1).

Papilledema was the most common finding and was seen in 14 patients (93.3%). It was bilateral in 13 patients and unilateral in 1 patient. In two patients papilledema was associated with subhyaloid hemorrhage. One patient had temporal pallor.

Table 1. Symptoms of patients with IIH

Symptoms	Number of Patients	%
Headache	14	93.3
Nausea and vomitting	8	53.3
Diplopia	6	40
Visual loss-Blurred vision	4	26.6
Shoulder/Arm pain	1	6.6
Dyscoordination	1	6.6
Retrobulbar pain on eye movement	1	6.6

Visual acuity was 20/20 in 13 patients and 14/20 in one, 8/20 in another. Visual fields by automated perimetry were normal in 13 patients, bilateral constriction in one patient, inferior altitudinal loss in another. Bilateral N.abducens paresis was found in one patient.

CSF pressure values of first lumbar punctions on decubitus position, that were performed before treatment were between 150 to 700 mmH_2O. After the treatment the pressures were 40 to 290 mmH_2O and CSF pressure values declined 20 to 490 mmH_2O.

Computed tomographies were normal in 10 patients, 3 patients had slit like ventricules and 2 patients had brain oedema. Four patients had MRI and all were normal.

We investigated conditions and diseases that were assoated with IIH (Table 2).

Table 2. Frequency of conditions and diseases in patients

Conditions/Drugs	Number of Patients	%
Obesity*	5	33.3
Iron deficiency	5	33.3
Idiopathic	5	33.3
Sinus thrombosis	2	13.3
Corticosteroid therapy	2	13.3
Behçet's disease	1	6.6
Abortion	1	6.6
Tetracycline therapy	1	6.6
Hypothyroidism	1	6.6
Hyperthyroidism	1	6.6
Addison's disease	1	6.6
Intoxication with Vitamin A	1	6.6

*We defined obesity as weight >=120% of the ideal weight

Table 3. Results of visual acuity and perimetry at the initial and final examinations

	Visual Acuity		Visual Fields	
Patients	Initially	At 3rd Month	Initially	At 3rd Month
1 to 11	20/20	20/20	N	N
12	20/20	20/20	Visual field constriction	Minimal improvement
13	14/20	16/20	N	N
14	8/20	8/20	N	N
15	20/20	20/20	Inferior temporal loss	Same

Acetazolamide was administered to 5 patients, corticosteroid to 3 patients and acetazolamide+corticosteroid to 6 patients. In a patient, who previously was under corticosteroid therapy, the drug was discontinued. Specific treatments were given if treatable conditions or diseases were found. Repeated lumbar punctions were performed in 7 of the 15 patients. Symptoms were reduced during the hospitalized period in all patients.

Visual acuity and visual fields were examined at initial and after the treatment (3rd month) (Table 3).

DISCUSSION

IIH, also known as Pseudotumor cerebri is a condition of increased ICP of unknown cause (6,7). It is most commonly seen in young and obese women. Annual incidence in the general population was 0.9 per 100.000. While it became 19 per 100.000 for women 20 to 44 years of age who were 20% or more than ideal weight (3). In many previous reports female to male ratio was between 8/1 to 10/1 (2,3,7,8). In our study ratio is 1.5/1 and it is discordant with the other reports.

The symptoms most commonly reported by IIH patients were headache (94%), transient visual obscurations (68%), pulsatile intracranial noises (58%), photopsia (54%), diplopia (38%), visual loss (30%) and retrobulbar pain on eye movement (22%) (7). Headache is the most common symptom in our study (93.3%). Apart from diplopia and visual loss other symptoms are discordant with other reports (Table 1).

Papilledema is the most common sign (2,9). We found it in all but except for one patient. This patient had headache, nausea and vomiting. In ophthalmological examination his temporal sides of optic discs were pale, visual acuity was 20/20 and had visual field constriction on automated perimetry. In history he had described sinus trombosis. CT was normal and CSF pressure was 700 mm H_2O. Acetazolamide was given and minimal improvement was noted on automated perimetry. This phenomenon is important to show that papilledema might not be seen in some patients with IIH and its absence is not the rule to exclude IIH.

In previous reports 22% to 25% visual acuity loss and 60% to 92% visual field loss was reported (2,9). We found both of them in a ratio of 13.3% and it was significantly lower.

The diverse etiologic factors associated with IIH include a heterogenous group of endocrine and metabolic disorders, intracranial venous sinus thrombosis, drug and toxins, haematological and connective tissue disorders, high CSF protein content, "menengism" with systemic viral or bacterial infections, empty cella syndrome and idiopathic conditions (1,4,7).

Obesity was reported in high frequencies (69%, 90%, 94%) (3,7,2). In our group we found it in 5 patients (33.5%).

Iron deficiency is an important etiologic factor. We found it more frequently (33.5%) than the other studies

(0%, 10%, 16%) (3,10,2). This high rates might be in relation with socioeconomic factors. In undeveloped countries, phsycians should initially exclude iron deficiency in the cause of IIH.

Weight loss under guidance of a dietitian should be recommended for every obese patient. In morbidly obese patients with IIH, obesity surgery was recommended (11). At least a third of patients with Pseudotumor cerebri have recovered after repeated lumbar punctions (12). Prednisone or oral hyperosmotic agents such as glycerol or acetazolamide to reduce CSF formation all have their advocates (5,13). In patients with protracted high intracranial pressure and papilledema, especially with those a measurable impairment of vision that does not respond to the usual therapeutic measures, a lumboperitoneal shunt should be performed (5). But CSF diversion procedures have a significant failure rate as well as a high frequency of side effects (14). Progressive loss of visual function associated with Pseudotumor cerebri can be reversed or stabilized with optic nerve sheath decompression surgery (15,16).

In our patient group, 4 had visual loss. Two of them have showed minimal improvement and the other two were the same. None of our patients have needed surgical treatment.

CONCLUSION

IIH is a syndrome of obscure origin. It is particularly frequent in fat adolescent girls and young women. A variety of pathologic conditions are associated with IIH.

Striking feature of this study is the frequency of iron deficiency anemia with IIH. Loss of vision is the only major form of morbidity associated with IIH. In this study our patients had benign course. Surgical procedures should be performed if patients does not respond to medical treatment.

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