Surgical treatment of pseudotumor cerebri

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AIM: *To share our experience with idiopathic intracranial hypertension.*
MATERIAL AND METHODS: *All patients believed to have pseudotumor cerebri underwent a fundus oculi examination to confirm the existence of papillary stasis and lumbar puncture (LP) to measure cerebrospinal fluid (CSF) pressure. Patients who did not respond to medical treatment underwent fundus oculi examinations at 3-week intervals. Patients with CFS pressures exceeding 240 mm H_2O underwent at least three LPs at 3-day intervals. Patients with higher CFS pressures were treated surgically.*
RESULTS: *The mean patient age was 40.8 (range 31-58) years and the mean body mass index (BMI) was 30.9 (range 28.8-36.4) kg/m^2. Papillary stasis was observed in 15 (46.8%) cases. The mean initial CSF pressure was 455.6 (range 360-560) mmHg, and after a mean of 4.3 (range 3-6) repeat measurements, this decreased to 213.4 (range 160-320) mmHg (Table I). Complications in our series included a lumbar pouch in three patients, and an abdominal pouch, meningitis, and abdominal migration in one patient each.*
DISCUSSION: *Surgical treatment of idiopathic intracranial hypertension is necessary when the intracranial pressure does not decrease despite medical treatment and repeat LP.*
CONCLUSIONS: *Idiopathic intracranial hypertension is a clinical syndrome of unclear pathogenesis that is closely related to obesity.*

KEY WORDS: Cerebrospinal fluid, Idiopathic intracranial hypertension, Pseudotumor cerebri, Obesity

Introduction

Idiopathic intracranial hypertension (IIH) is a clinical syndrome of unknown origin [1]. The cerebrospinal fluid (CSF) and neurological imaging findings are mostly not significant and typically no cause is identified. Although it can affect patients of all ages and both genders, it most frequently affects women of childbearing age and the obese [2]. It may develop following the use of lithium, tetracycline, non-steroidal anti-inflammatory drugs, and vitamin A overdose, after rapidly discontinuing the use of steroids, as a secondary retraction syndrome, or with uremia [3,4]. It can be treated successfully if diagnosed in a timely manner, but may cause blindness when the diagnosis is delayed [5,6]. It is twice as frequent in the Middle East compared with Western countries [1]. Although the exact prevalence here in Turkey is unknown, it is similar to that in other Middle Eastern countries.

Here, we share our experiences with cases followed in our clinic.

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Material and Methods

Between August 2013 and September 2015, we treated 32 patients (1 male, 31 females; age range 31–59 years) diagnosed with pseudotumor cerebri based on their symptoms in our Neurology and Neurosurgery Department. Each patient underwent a fundus oculi examination for papillary stasis. A lumbar puncture (LP) was performed and CSF pressure measured. Fundus oculi examinations were repeated at 3-week intervals in patients who did not respond to medical treatment with acetazolamide (500 mg tid). LP was repeated at 3-day intervals in patients with CFS pressures exceeding 240 mmH_2O. Patients with higher CFS pressures were treated surgically if surgical treatment was not contraindicated. None of the patients had communicating hydrocephalus requiring lumboperitoneal shunt placement or a CFS fistula requiring surgical treatment.

Results

Table I summarizes the demographics of the 32 patients in our series, the existence of pupil stasis (PS), number of CFS measurements by LP, CFS pressure at LP, and postoperative CFS pressure. PS was present in 15 patients (46.8%). All but one of our patients were female (96%). The mean age of the patients was 40.8 (range 31-58)

TABLE I - *Demographics, clinical data, number of CFS pressure measurements with LP, CFS pressure measured with LP and postoperative CFS pressure values of 32 patients*

No. of case	Papillary stasis	Weight (kg)	Height (m)	VKI (kg/m^2)	Age (year)	Count of CSFP measure-ment	Initial pressure (mmHg)	Last pressure (mmHg)
1	+	86	1.68		58	3	470	220
2	+	72	1.62		52	4	520	200
3[1]		81	1.65		51	5	390	210
4	+	76	1.61		42	4	420	230
5		74	1.56		41	5	560	240
6	+	79	1.62		44	5	410	190
7[2]	+	81	1.56		32	4	400	180
8		82	1.59		38	5	460	260
9		84	1.62		31	5	500	310
10		85	1.59		32	5	510	210
11		71	1.56		42	5	410	210
12	+	79	1.63		34	4	420	200
13		76	1.61		42	6	380	190
14[1]		84	1.62		44	3	470	160
15	+	76	1.66		41	3	420	200
16		79	1.64		38	5	400	230
17		79	1.59		42	4	520	210
18[1]	+	71	1.57		42	5	390	300
19		81	1.58		36	3	420	260
20	+	84	1.62		39	3	480	190
21		79	1.58		37	3	510	210
22		76	1.57		40	5	490	200
23	+	80	1.61		41	5	460	210
24		82	1.68		38	4	430	230
25	+	78	1.62		41	4	470	220
26[3]	+	75	1.59		42	4	490	230
27		92	1.59		46	5	390	190
28	+	81	1.58		42	5	560	180
29	+	84	1.61		41	4	480	160
30[4]		81	1.59		42	5	510	190
31		79	1.60		41	5	360	200
32	+	92	1.62		35	3	480	210

CSFP; cerebro-spinal fluid pressure, BMI; body mass index
Complications: 1-lumbar pouch; 2-abdominal migration; 3-abdominal pouch; 4-meningitis

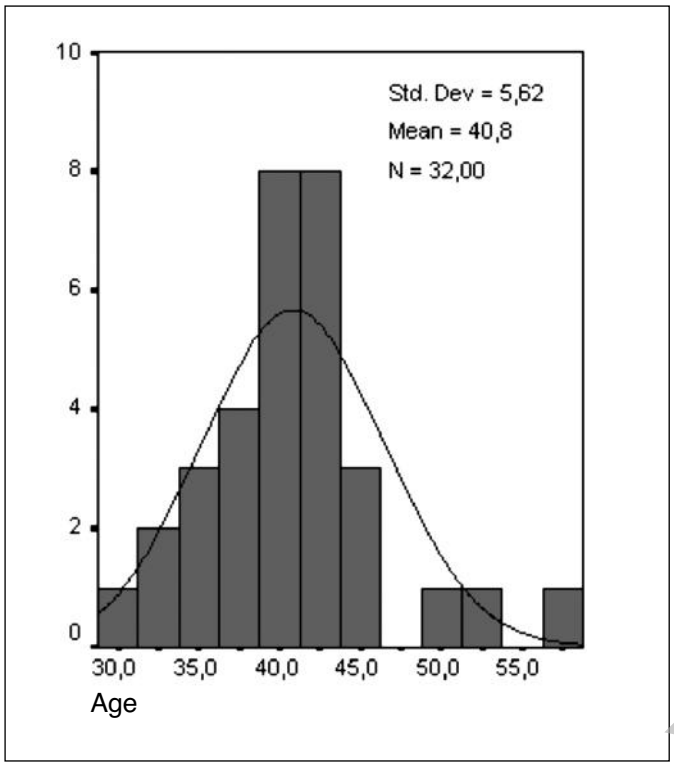

Fig. 1: Age distribution and mean of the patients involved in the study.

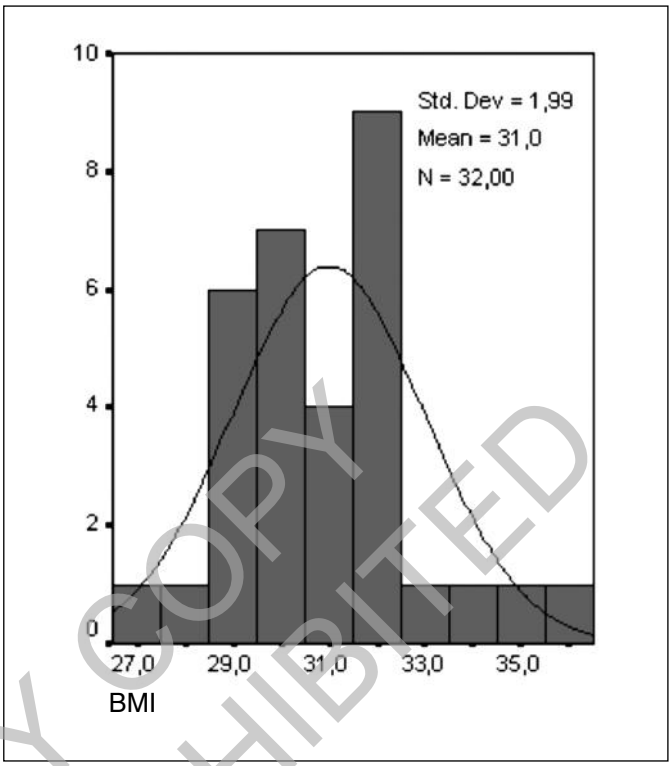

Fig. 2: Body mass index distribution and mean of the patients involved in the study.

years (Fig. 1). The mean BMI was 30.9 (range 27.4–36.4) (Fig. 2), with 37.5% of the patients classified as overweight and 62.5% as obese. All of our cases were treated surgically using a lumboperitoneal shunt. During follow-up, a lumbar pouch developed in three patients, and meningitis, migration to the abdomen, and pouch and pseudocyst were observed in one patient each (Table I).

Discussion

Idiopathic intracranial hypertension is usually seen in females of childbearing age [1]. Its incidence is about 1/100,000 in Western countries and 2/100,000 in the Middle East [1]. The incidence is higher in women than in men. Being of childbearing age and overweight markedly increase the incidence of IIH. In America, the incidence in overweight females of childbearing age is 13/100,000, while it is about 19.3/100,000 in obese females of childbearing age. Marked differences are seen in these rates among countries [1]. The incidence in our country, Turkey, is not known. In a series studied in western Turkey, 76% of 62 patients were female and 24% were male, and the mean age was 32.7 (range 18–56) years [7]. The mean age of our patient population was 40.8 (range 31-58) years, and all but one of our patients were female (96%). The increased number of females may be due to the number of women of childbearing age in our province, excessive obesity in women of childbearing age, and low access to healthcare services. The high mean age might be based on disregarding headache, which was the most frequent symptom of our patients, and delays in seeking healthcare.

Obesity is closely related to IIH [8]. In some studies, 94% of new patients were obese [9]. Obesity is an important problem in our country, especially in our region. In Turkey, every other person is obese or overweight [10]. In our series, the mean BMI was 30.9 (range 27.4 - 36.4) kg/m^2, and 62.5% of our cases were obese, 37.5% were overweight, and none of the patients had a normal weight. This demonstrates a health risk of being overweight in our country.

Patients suspected of having IIH underwent comprehensive eye and neurological examinations. Computed tomography (CT) and magnetic resonance imaging (MRI) are important for the diagnosis of IIH and for identifying additional pathology. MRI is especially helpful for showing venous sinus occlusions [11]. An empty sella, flattened posterior globe, widened optic nerve sheath, cerebral tonsillar herniation, and meningocele are important MRI findings [12]. In IIH, stenosis and occlu-

sion at the junction of the distal sigmoid sinus and transverse sinus can be incidental or causative [11,12]. All of our patients underwent preoperative brain CT; no organic pathology was seen in any patient.

Even if the imaging is negative, a lumbar puncture (LP) is necessary for both determining the real intracranial pressure and identifying any inflammatory, infectious, or neoplastic causes [11,12]. All of our patients presented complaining of a headache or visual impairment. In our series, 47% of the patients had papilledema compared with 22% in previous reports by Wall and George [13] and 27% in Çelebisoy and Seçil [14]. The high rate in our series might have resulted from delayed surgical treatment. None of our patients had a neurological deficit. Each patient underwent LP at least three times. The mean initial CFS pressure at LP was 456 mmHg and the mean postoperative CFS pressure was 213 mmHg, a decrease of about 50%. The patients are still being followed with regular fundus oculi and visual space examinations.

Weight loss is important in the treatment of IIH, as it is effective at eliminating most patient complaints [15,16]. If symptoms are mild, medical treatment with acetazolamide is the first treatment of choice. Surgical treatment is preferred in patients with marked visual impairment and severe symptoms who do not respond to medical treatment and weight loss [1]. The surgical treatments include optic nerve sheath fenestration and CFS drainage procedures such as a lumboperitoneal or ventriculoperitoneal shunt. All of our patients were treated surgically with a lumboperitoneal shunt. During follow-up, a lumbar pouch developed in three patients, and meningitis, migration to the abdomen, and pouch and pseudocyst developed in one patient each.

There is a close relationship between IIH and obesity, although the pathogenesis of IIH has not been completely elucidated. It frequently causes headache and visual impairment. The diagnosis is based on increased intracranial pressure in the absence of a mass with normal CFS findings. Surgical treatment is necessary when the intracranial pressure does not decrease with medical treatment. Clinicians must consider IIH as a rare complication of obesity in patients who present with a headache or visual impairment.

Riassunto

SCOPO: Condividere la nostra esperienza di ipertensione intracranica idiotapica osservata e trattata nella nostra clinica

MATERIALE E METODO: Ogni paziente che per la sintomatologia presentata si riteneva fosse sofferente di pseudotumor cerebri, è stato sottoposto allo studio del fondo oculare per la conferma dell'esistenza di una papilla da stasi, aggiungendo la puntura lombare (LP) per misurare la pressione del liquor cerebrospinale (CSP). Lo studio del fondo oculare è stato ripetuto ad intervalli di tre settimane presso il dipartimento oculistico nei pazienti refrattari al trattamento medico. Sono state eseguite almeno tre puntura lombari reiterative nei pazienti con pressione cerebro-spinale superiori ai 240 mm H_2O con intervallo di tre giorni. I pazienti con pressione cerebrospimale più elevata sono stati trattati chirurgicamente.

RISULTATI: L'età media della nostra casistica è stata di 40,8 anni (intervallo 31.58) e in media l'indice di massa corporea (BMI) è stato di 30,9 kg/m^2 (intervallo 28.8-36.4). la stasi papillare è stata osservata in 15 pazienti della casistica (46.8% del totale). La pressione cerebrospinale inziale era in media 455.6 mmHg (intervallo 360-560), ed è stata misurata almeno tre volte (in media 4,3 – intervallo 3-6), con decremento a 213.4 mmHg (intervallo 160-320) (vedi Tab. I). Le complicazioni osservate sono state il sacchetto lombare in tre pazienti, la tasca addominale in un paziente, la meningite in un paziente e la dislocazione in addome in un paziente.

DISCUSSIONE: Il trattamento chirurgico dell'ipertensione intracranica idiopatica è necessario nel caso che non ceda al trattamento medico ed alle reiterative puncture lombari.

CONCLUSIONI: L'ipertensione intracranica idiopatica (IIH) è una sindrome clinica di cui è noto lo stretto rapporto con l'obesità, e la cui patogenesi non è stata ancora chiarita completamente.

References

1. Almarzouqi SJ, Morgan ML, Lee AG: *Idiopathic intracranial hypertension in the Middle East: A growing concern*. Saudi J Ophthalmol, 2015; 29(1):26-31.

2. Dhungana S, Sharrack B, Woodroofe N.: *Idiopathic intracranial hypertension*. Acta Neurol Scand, 2010; 121(2):71-82.

3. Mrejen S, Vignal C, Bruce BB, Gineys R, Audren F, Preechawat P, at Gaudric A, Gout O, Newman NJ, Vighetto A, Bousser MG, Biousse V: *Idiopathic intracranial hypertension: a comparison between French and North-American white patients*. Rev Neurol (Paris). 2009; 165(6-7):542-48.

4. Kaya B, Altun E, Paydas S, Balal M*: Pseudotumor cerebri associated with uremia and NSAIDs*. Saudi J Kidney Dis Transpl, 2015; 26(3):602-03.

5. Alkali NH, Daif A, Dorasanamma M, Almoallem MA: *Prognosis of idiopathic intracranial hypertension in Saudi Arabia.* Ann Afr Med, 2011; 10(4):314-15.

6. Laemmer R, Heckmann JG, Mardin CY, Schwab S, Laemmer AB: *Detection of nevre fiber atrophy in apparently effectively treated papilledema in idiopathic intracranial hypertension.* Graefes Arch Clin Exp Ophthalmol, 2010; 248(12):1787-93.

7. Celebisoy N, Seçil Y, Akyürekli O: *Pseudotumor cerebri: Etiological factors,presenting features and prognosis in the western part of Turkey.* Acta Neurol Scand, 2002; 106(6):367-70.

8. Daniels AB, Liu GT, Volpe NJ, Galetta SL, Moster ML, Newman NJ, Biousse V, Lee AG, Wall M, Kardon R, Acierno

MD, Corbett JJ, Maguire MG, Balcer LJ: *Profiles of obesity, weight gain, and quality of life in idiopathic intracranial hypertension (pseudotumor cerebri)*. Am J Ophthalmol, 2007; 143(4):635-41.

9. Hannerz J, Ericson K: *The relationship between idiopathic intracranial hypertension and obesity*. Headache, 2009; 49(2):178-84.

10. *http://www.tuik.gov.tr/PreHaberBultenleri.do?id=13490*

11. Friedman Deborah I, Jacobson Daniel M: *Idiopathic intracranial hypertension.* J Neuroophthalmol, 2004; 24(2):138-45.

12. Bidot S, Saindane AM, Peragallo JH, Bruce BB, Newman NJ, Biousse V: *Brain Imaging in Idiopathic Intracranial Hypertension.* J Neuroophthalmol, 2015.

13. Wall M, George D: *Idiopathic intracranial hypertension*. Brain, 1991; 114:155-80.

14.Celebisoy N, Secil Y, Akyurekli O: *Pseudotumor cerebri: Etiological factors, presenting features and prognosis in the western part of Turkey*. Acta Neurol Scand, 2002; 106:367-70.

15. Handley JD, Baruah BP, Williams DM, Horner M, Barry J, Stephens JW: *Bariatric surgery as a treatment for idiopathic intracranial hypertension: A systematic review*. Surg Obes Relat Dis. 2015 12. pii: S1550-7289(15)00804-7.

16. Levin AA1, Hess D, Hohler AD: *Treatment of idiopathic intracranial hypertension with gastric bypass surgery*. Int J Neurosci. 2015; 125(1):78-80.