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ORIGINAL RESEARCH

Behçet's Disease in Idiopathic Intracranial Hypertension Without Cerebral Venous Sinus Thrombosis: Retrospective Results of a Tertiary Headache Center

Fatma YILMAZ CAN[1], Mehmet Fevzi ÖZTEKİN[1], Hacı Asım KILIÇ[1], Aynur TURAN[2]

[1] Etlik City Hospital, Department of Neurology, Ankara, Türkiye.
[2] Etlik City Hospital, Department of Radiology, Ankara, Türkiye.

ABSTRACT

In our clinic, we frequently observed the comorbidity of Behçet's disease (BD) without cerebral vein thrombosis in patients with idiopathic intracranial hypertension (IIH). This study aimed to investigate the comorbidity of isolated BD in patients with IIH. The medical records of 98 patients diagnosed with IIH were retrospectively reviewed. IIH was diagnosed by an experienced neurologist. At the diagnosis stage, patients who responded to the question "Do you have any comorbidities?" with a statement of BD had their diagnoses confirmed by a rheumatologist. The radiological images of patients diagnosed with BD were reconfirmed by a neuroradiologist, and neuro-Behçet's (NBD) disease was excluded. Isolated BD was diagnosed in 4 of 98 IIH patients (4.1%). Ninety-two of the IIH patients were female (93.9%). The mean age of the patients was 36.7±12 (18-65) years, and their mean body mass index (BMI) was 32.39 (21-49). Four BD patients were female. There was no significant difference between those with BD and those without BD regarding gender (p= 1.0). The average age of those with BD was 43.50, and the average age of those without BD was 36.41, and there was no significant difference between them (p=0.167). The mean BMI of IIH patients with BD was 35.40, while the mean BMI of IIH patients without BD was 32.26. No significant difference was found between them (p=0.323). We found a 4.1% prevalence of BD in patients with IIH. We recommend questioning IIH patients for BD symptoms in regions where BD is endemic.

Key Words: Behçet's Disease. Comorbidity. Idiopathic intracranial hypertension. Inflammation.

İdiopatik Intracranial Hipertansiyonda Serebral Venöz Sinüz Trombozu Olmaksızın Behçet Hasatalığı: Üçüncü Basamak Bir Baş Ağrısı Merkezinin Retrospektif Sonuçları

ÖZET

Kliniğimizde idiyopatik intrakranial hipertansiyonlu (İİH) hastalarda sıklıkla serebral ven trombozu olmaksızın Behçet hastalığının (BH) eşlik ettiğini gözlemledik. Bu çalışmada, İİH'li hastalarda izole BH prevalansını araştırmayı amaçladık. İİH'li hastalara tanı aşamasında "Eşlik eden bir hastalığınız var mı?" sorusu soruldu. BH olduğunu beyan eden hastaların tanıları romatolog tarafından konfirme edildi, hastalık tanısı doğulandı. BH komorbiditesi olanlarin radyolojik görüntüleri tekrar tecrubeli bir nöroradyolog tarafından gözden geçirilerek nöro-Behçet hastalığı (NBH) ekaret edildi. Doksansekiz İİH hastasının 4'üne (%4,1) izole BH tanısı konuldu. Hastalardan 92'si kadın (%93,9), 6'sı erkekti (%6,1). Hastaların yaş ortalaması 36,7±12 (18-65) yıl, vücut kitle indeksi (VKİ) ortalamaları 32,39 (21-49) kg/m^{2}'di. Dört BH'li hasta da kadındı. BH'li hastalar ile BH'siz hastalar arasında cinsiyet açısından anlamlı bir fark yoktu (p=1,0). BH'li hastaların yaş ortalaması 43,50, BH'siz hastaların yaş ortalaması 36,41 yıldı ve aralarında anlamlı bir fark yoktu (p=0,167). BH'li İİH hastalarının ortalama VKİ'si 35,4 kg/m^2 iken, BH'siz IIH hastalarının ortalama VKİ'si 32,26 kg/m^{2}'ydi. Aralarında anlamlı bir fark bulunmadı (p=0,323). İİH hastalarında %4,1 oranında BH prevalansı saptadık. BH endemik olduğu bölgelerde İİH hastalarında BH semptomlarının sorgulanmasını öneriyoruz.

Anahtar Kelimeler: Behçet Hastalığı. İdiyopatik İntrakranial Hipertansiyon. İnflamasyon. Komorbidite.

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Dr. Fatma YILMAZ CAN
Etlik City Hospital, Department of Neurology, Ankara, Türkiye
E-mail: fatmayilmazcan33@hotmail.com

AUTHORS' ORCID INFORMATION
Fatma YILMAZ CAN: 0000-0003-0995-4911
Mehmet Fevzi ÖZTEKIN: 0000-0003-3582-2832
Hacı Asım KILIÇ: 0000-0002-5301-7039
Aynur TURAN: 0000-0001-6654-3129

Idiopathic intracranial hypertension (IIH) is the elevation of intracranial pressure with normal cerebrospinal fluid (CSF) without a known cause, excluding intracranial pathologies and cerebral venous thrombosis (CVT)[1]. The term "pseudotumor cerebri" is also used to define IIH. Its specific symptoms are headache, papilledema, temporary loss of vision, sixth cranial nerve palsy, diplopia, and pulsatile tinnitus[2]. Untreated IIH cases can lead to blindness. For this reason, diagnosis and treatment are important. Although the pathophysiological mechanism that

causes IIH is not exactly known, it is thought to stem from causes that disrupt the physiological balance such as an increase in CSF production, a decrease in CSF absorption, or an increase in cerebral venous pressure[2]. The syndrome affects mostly young obese women. Its prevalence in society is 0.5-2/100.000 [3]. The percentage of women is 82.4%[4]. It is suggested that obesity, toxins, steroids, oral contraceptives, tetracyclines, hypervitaminosis (vitamin A), collagen tissue diseases, endocrine and hematological disorders can have a role in its etiology[5]. It is assumed that chronic inflammation also plays a role in pathogenesis[6]. The results of a study investigating the role of inflammation in the pathogenesis of IIH using neutrophil-lymphocyte ratio and platelet-lymphocyte ratio were found to be significantly high for IIH[7]. One study, exposure to infectious and inflammatory diseases one year before the diagnosis of IIH was found higher in IIH patients compared to the control population[8].

Behçet's Disease (BD) is considered an idiopathic, chronic, recurrent, multisystemic vascular-inflammatory pathology[9]. Its prevalence displays regional differences. In epidemiological studies conducted, Türkiye is one of the high-prevalence regions with a rate of 17/10.000[10]. Central and peripheral nervous system involvement in BD is termed Neuro-Behçet's Disease (NBD)[11]. Central nervous system involvement in BD is classified as parenchymal and nonparenchymal (cerebral veins and arteries). CVT, post-capillary venules, and small parenchymal arteries are most commonly involved. Symptoms include cognitive changes, neurological deficits, and isolated headache syndrome[12]. Increased intracranial pressure occurs secondary to CVT in patients with BD. Fundus edema secondary to increased intracranial pressure, increased CSF opening pressure on lumbar puncture (LP), and normal CSF content may mimic IIH. In a single-center study investigating the etiology of 59 IIH patients, BD without SVT was found to coexist in 1.7% of cases. It is stated that small vessel vaculitis may play a role in the etiology[13]. Cases of IIH accompanying BD have been reported in the literature after excluding NBD and applying IIH criteria[14]. In a retrospective study of 105 BD patients, it was reported that two of the patients developed IIH without CVT [15]. The literature generally reports the presence of isolated IIH in patients with BD. We aimed to examine the situation from the opposite perspective. We planned to retrospectively investigate the comorbidity of BD in patients with IHH.

Materials and Methods

This study was approved by the institutional ethics committee of our hospital (approval No: AEŞH-EKI-2023-566; approval date: 22.11.2023); and informed consent was not required for this retrospective study.

The medical files of 98 patients diagnosed and followed up with IIH at our tertiary care hospital's Neurology Clinic Headache Center between 2022 and 2023 were retrospectively reviewed. The diagnosis had been made according to the Friedman Criteria[1]. Demographic characteristics, body mass index (BMI), CSF examinations and magnetic resonance imaging (MRI) features were recorded. The files of 4 patients who declared that they were diagnosed with BD in response to the question "Do you have any accompanying diseases?" in the medical files filled out by the physician during the diagnosis phase were examined separately. The presence of BD was confirmed a second time by a rheumatologist. The diagnosis of BD was consistent with the 1990 International Behçet's Disease Study Group Diagnostic Criteria[16]. Two of the BD patients were using colchicine, one was using methyl prednisolone and azathioprine along with colchicine. Of these three patients, two were diagnosed with IIH years after their BD diagnosis, and one was diagnosed with IIH months later. One patient was diagnosed with IIH and BD simultaneously. The newly diagnosed patient was not yet using medication.

Three patients who responded to the question "Do you have a family member with a genetic disease?" by indicating BD were re-evaluated for BD. The patients reported the presence of oral and genital ulcers. The three patients were evaluated by dermatologists and rheumatologists but did not fulfill the diagnostic criteria for BD. An experienced neuroradiologist reexamined the MRI scans with T1-weighted images, T2-weighted images, fluid-attenuated inversion recovery (FLAIR) images, and magnetic resonance venography (MRV) images. The examination was performed with a 1.5 Tesla MR unit (General Electric Medical System Milwaukee). Parenchymal lesions, brainstem atrophy, jugular vein thrombosis and CVT images that would exclude the diagnosis of IIH or suggest NBD were investigated, and NBD was excluded in all patients.

Statistical Analysis

The data were transferred to the IBM SPSS 29.0 software for statistical analysis. As a result of the analysis, categorical variables were presented as numbers and percentages, while continuous variables were expressed as minimum and maximum values, mean, standard deviation, and median values. In difference analyses, non-parametric test analysis was performed for the small sample of IIH+BD (n= 4), and the Mann-Whitney U test was used to evaluate differences. The statistical significance level was set at $p<0.05$ in the analyses.

Results

Out of 98 IIH patients, 92 were female (93.9%) and 6 were male (6.1%). The mean age of the patients was 36.7±12 (18-65) years, and their mean BMI was 32.39 (21-49). Four of the patients (4.1%) were diagnosed with BD, all females. There was no significant difference between those with BD and those without BD regarding gender (p=1.0). The average age of those with BD was 43.50 years, and the average age of those without BD was 36.41 years, and there was no significant difference between them (p=0.167). The mean BMI of IIH patients with BD was 35.40, while the mean BMI of IIH patients without BD was 32.26. No significant difference was found between them (p=0.323) (Table I).

Table I. Analysis findings of patient groups regarding age, BMI and gender

		Group	n	mean	p
Age		IIH	94	36.41	0.167
		IIH+BD	4	43.50	
BMI (kg%m^2)		IIH	94	32.26	0.323
		IIH+BD	4	35.40	
Gender	Female	IIH	88	93.6	1.000
		IIH+BD	4	100	
	Male	IIH	6	6.4	
		IIH+BD	0	0.0	

BD: Behçet's disease (BD); IIH: idiopathic intracranial hypertension; BMI: Body mass index

Papilledema was detected in all patients with both IIH and IIH+BD. All patients had CSF opening pressure >250. Laboratory test results showed CSF protein, glucose, and cell counts within normal limits.

The radiological images of these four BD patients determined no parenchymal or non-parenchymal lesions that would suggest NBD. In addition, the radiological images of three patients who stated in the survey That they had a family history of BD were normal.

Discussion and Conclusion

In the study, we investigated BD comorbidity in 98 IIH patients. In terms of demographic characteristics, mean age, BMI and gender are consistent with other studies in the literature[17]. Their mean BMI was 32.39 kg/m^2; in general, patients with IIH are defined as female, of childbearing age, and having a BMI >30 kg/m^2 [18]. We identified comorbid BD in 4 (4.1%) of the patients. There was no statistically significant difference between the IIH patients with BD and those without BD regarding age, gender, and BMI (p value respectively =0.167; 1.0; 0.323). Patients with a family history of BD and ulcers in the mouth and genital area did not fulfil the diagnostic criteria. All symptoms of BD patients usually do not appear simultaneously, and the symptoms are added on top of each other in different periods. While the most frequently seen symptom, oral ulcer, starts 7.5±10.2 years before the diagnosis, genital ulcer and eye and skin involvement may develop 1 or 2 years before the diagnosis[12]. In light of this information, it is possible that the three patients may be diagnosed with BD in the future. When signs of increased intracranial pressure are detected in patients with BD, the most suspected condition is CVT. CVT can cause increased intracranial pressure in the subacute and chronic stages and may show all the major criteria of IIH. Therefore, brain MRI and MRV images obtained at the time of diagnosis of IIH patients were re-examined to exclude CVT or parenchymal lesions that could cause increased intracranial pressure. Jugular vein thrombosis and CVT were clearly excluded. In acute NBH and chronic progressive NBH patients, CSF cell count and protein are significantly higher, and glucose levels are lower[19]. In our study, according to Friedman diagnostic criteria, CSF cell count, protein and glucose levels were within normal limits in both IIH and IIH+BD patients. Similar to our study, there are isolated IIH+BD case reports in the literature that included normal CSF protein, glucose, and cell count examinations, papilledema, and CSF opening pressure >250[14].

BD is frequently observed in Türkiye. The population prevalence is 17/10,000[10]. As a result of our study, we detected BD at a rate of 4.1% in IIH patients, and it was higher than the population prevalence. BD is an inflammatory disease, on the other hand, chronic inflammation is suggested to play a role as an etiological factor in the pathogenesis of IIH[6]. The pathophysiological mechanism is not fully known. Studies have found a significantly higher inflammatory status in IIH patients compared to age, gender and BMI matched controls[7]. There are studies and case reports in the literature that report the association of IIH with systemic and neuro-inflammatory diseases such as BD, Hashimoto's thyroiditis and systemic lupus erythematosus (SLE)[15, 20,21]. One study, exposure to infectious and inflammatory diseases one year before the diagnosis of IIH was found higher in IIH patients compared to the control population[8]. On the other hand, laboratory studies also emphasize the existence of an inflammatory condition in patients with IIH. Serum levels of leptin, a proinflammatory cytokine, have been shown to be significantly higher in IIH patients compared to both overweight and lean healthy control groups[22]. These findings suggest that inflammatory activation may be a possible factor in the development of IIH. A pathophysiological mechanism explaining

the association of IIH with inflammation has not yet been reported. One hypothesis has suggested that inflammatory products and cytokines may impair CSF drainage by causing fibrosis in the choroid plexus or arachnoid granulation tissue[23]. Decreased CSF absorption due to immune-mediated damage to arachnoid villi or vascular occlusion due to hypercoagulability are other proposed pathogenesis of this condition[24,25]. BD itself may have contributed to increased intracranial pressure through subclinical inflammation, microvascular dysfunction, or venous outflow disorders.

We found a higher prevalence of BD in IIH patients than in the general population. IIH can occur months to years after the diagnosis of BD. Patients with BD who present with headache should be evaluated for IIH. In areas with high prevalence, BD symptoms should be questioned when IIH is diagnosed.

Limitations

The retrospective design of the study, the small, single-center sample size, and the lack of a control group are significant methodological limitations. Additionally, the number of participants in the IIH+BD group was limited in our study. Although all CSF protein, glucose levels, and cell counts were within normal limits according to Friedman criteria, CSF laboratory data from the IIH and IIH+BD groups could not be compared due to the retrospective design of the study.

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