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A CASE OF WERNICKE ENCEPHALOPATHY PRESENTING WITH PAPILLEDEMA AS A RAREINITIAL FINDING

NADİR BİR SUNUM OLARAK PAPİLÖDEM İLE BAŞVURAN BİR WERNİCKE ENSEFALOPATİ VAKASI

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ABSTRACT

Wernicke encephalopathy is an acute encephalopathy, characterized by the triad of mental confusion, ophthalmoplegia, and gait ataxia. It may develop due to many reasons that may be associated with insufficient intake or malabsorption of thiamine. Here, we report a rare case of Wernicke's encephalopathy presenting with papilledema as the initial finding. In the patient who developed tetraparesis and ophthalmoplegia after gastric by-pass surgery, there was also a change in consciousness at follow-up. Wernicke's encephalopathy was considered as diagnosis. Her clinic improved progressively under high dose intravenous thiamine treatment. The diagnosis was delayed because the characteristic findings were obscure at the initial stage, the patient was obese, and had papilledema. This case was thought to be important as it would increase awareness of the uncommon findings of Wernicke's encephalopathy.

Keywords: bariatric surgery, papilledema, thiamine, Wernicke encephalopathy

ÖZET

Wernicke ensefalopatisi konfüzyon, oftalmopleji ve ataksi üçlüsü ile karakterize bir akut ensefalopati tablosudur. Etiyolojide tiaminin yetersiz alımı veya emilim bozukluğu ile ilişkili olabilecek birçok neden bildirilmiştir. Burada, nadir bir sunum olarak papilödem ile başvuran Wernicke ensefalopatisi olgusunu sunuyoruz. Gastrik by-pass cerrahisi sonrası gelişen bu vakada takip sırasında oftalmopleji ve tetraparezi gelişti ve tabloya bilinç değişikliği de eklendi. Ayırıcı tanıda Wernicke ensefalopatisi düşünülür düşünülmez yüksek doz intravenöz tiamin tedavisi başlandı ve hastanın kliniği progresif olarak düzeldi. Bu vakada papil ödemi, obezite ve tablonun karakteristik bulgularının başlangıçta silik olması tanıda gecikmeye yol açmıştır. Bu vakanın Wernicke ensefalopatisinin sık karşılaşılmayan bulguları hakkında farkındalığı artıracağı için önemli olduğu düşünülmüştür.

Anahtar Kelimeler: bariatrik cerrahi, papil ödemi, tiamin, Wernicke ensefalopatisi

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INTRODUCTION

Wernicke encephalopathy (WE) is a well-known neurological complication of thiamine deficiency, requiring immediate attention to avoid mortality and neurological morbidity. The total thiamine store in the body is 30-50 mg, and the daily requirement is 1-2 mg/day (1). Because the plasma half-life of thiamine is quite short (1-12 hours), its stores in the body can be depleted in 1-3 weeks (2). WE is an acute encephalopathy, characterized by the triad of mental confusion, ophthalmoplegia, and gait ataxia. Chronic alcoholism, anorexia nervosa- diets, starvation, gastrointestinal surgery, etc. may be the reason (2). Here we present a case of having WE after bariatric surgery with papilledema, delaying diagnosis.

CASE

A 19-year-old female patient was admitted to our hospital for headaches, blurred vision, and gait disturbance. She had a body mass index (BMI) of 49.2 and got bariatric surgery two months ago. Objective neurological examination was normal except fundus findings. On fundus examination grade 4 papilledema was detected, not any sign of retinal hemorrhage. Cerebrospinal fluid (CSF) pressure was measured to be 35 cm H_2O on lumbar puncture. CSF findings including biochemistry and microscopy were normal. Her initial brain magnetic resonance imaging (MRI) study was normal. Considering the diagnosis of idiopathic intracranial hypertension (IIH), acetazolamide treatment was started. Her clinical situation got worse and she became comatose in approximately six days. She had near-total ophthalmoplegia that started with limited outward gaze and worsened rapidly. Also, she had a flask tetraparesis dominantly on the lower extremities (deep tendon reflexes and plantar responses were absent). In the lower extremity, distal muscle strength was Medical Research Council (MRC) 2/5 while proximal muscle strength was MRC 3/5. In the upper extremity, distal muscle strength was Medical Research Council (MRC) 3/5 while proximal muscle strength was MRC 4/5. Her paresis was clinically ascending to mimic Guillain-Barré Syndrome. Electromyographic findings were not diagnostic because of technical troubles. Cardiac examination and electrocardiography were normal. As soon as, WE was thought of in differential diagnosis, after sampling blood for thiamine level, intravenous thiamine replacement treatment had been established. On analysis, the thiamine level was found to be 17 mg/L (Normal reference 25-75 mg/L). Because of the change in the patient's clinic, a control brain MRI was performed one week later. The control MRI study supported the diagnosis of WE **(figure 1)**. She showed clinical improvement on high dose thiamine replacement, two days of 3x500 mg followed by five days of 1x500 mg intravenous (IV) thiamine, and long-term oral 100 mg thiamine supplementation. Both optic disk findings and CSF pressure on control lumbar puncture were normal. The patient's other findings, except tetraparesis, had gradually improved at the time of discharge. Although we didn't have electromyographic evidence, we thought she had peripheric neuropathy and she went on physical treatment. In the first year after evaluation, her BMI was 23.4 and her other findings, except mild tetraparesis, had improved. Informed concept was obtained from the patient.

DISCUSSION

Thiamine is a cofactor of many enzymes such transketolase, α-ketoglutarate dehydrogenase and, pyruvate dehydrogenase (3). Thiamine requirement depends on metabolic state and increases in high carbohydrate intake (4). Wernicke encephalopathy (WE) is a well-known neurological complication of thiamine deficiency, characterized by the triad of mental confusion, ophthalmoplegia, and gait ataxia. Although the main diagnosis is based on clinical condition, the clinical triad is seen only in about 1/3 of the patients and this sometimes leads to misdiagnosis. Also in neurological examination, there might be signs of peripheric neuropathy and dysarthria and some rare cases papilledema, psychosis, hearing loss, seizures, or coma (2).

Here we present a rare case of Wernicke encephalopathy presenting with papilledema. It is very rare in the literature, especially when the first presentation is with papilledema. In a review with 84 patients with Wernicke's encephalopathy after bariatric surgery, seven cases had been reported with ophthalmological signs; four with papilledema, and 3 with peripapillary or retinal hemorrhages (5). Other reported neuro-ophthalmological findings were more horizontal and less vertical nystagmus, ophthalmoplegia, reduced pupillary response to light and light -near dissociation (6). The reason for optic neuritis might be similar to the nutritional neuropathies. Bohnsack BL and Patel SS.

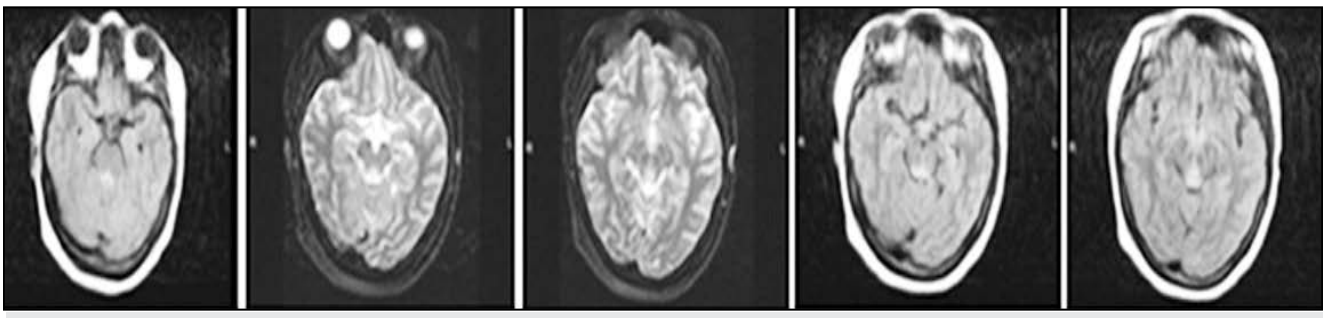

Figure1. Bilateral symmetric hyperintensity alterations on T2-weighted, and FLAIR MR images in the periphery of the third ventricle, periaqueductal area, and mammillary bodies.

reported that peripapillary nerve fiber layer thickening, telangiectasia, and retinal hemorrhages may be related to impaired mitochondrial function in retinal ganglion cells and capillaries, based on a Wernicke's encephalopathy case with neuro-ophthalmological findings they followed (7). In our patient, IHH was a distracting diagnosis for having obesity, headache, papilledema, and increased CSF pressure. However, on the follow-up period, the other signs causing suspicion about WE helped for diagnosis.

The most reliable test in the laboratory is erythrocyte thiamine transketolase level measurement before and after treatment (8). Also, it is possible to measure both serum thiamine and thiamine pyrophosphate levels. But these two are not so reliable since not having exact sensitivity, and specificity for symptomatic patients, do not show brain levels and normal levels don't exclude the diagnosis. In our patient, we could only see the serum thiamine level both before and after treatment, respectively lower than normal and normal levels.

Characteristically, periaqueductal and periventricular symmetrical lesions are seen on MRI (9). Mamillary bodies involving are also frequent (9). Also, it was showed in some patients that there may be lesions in the dorsomedial thalamus, locus ceruleus, cranial nerve nuclei, hippocampus, cerebellum, and cerebral cortex atypically (9). Findings generally show a symmetrical pattern and are as follows; hyperintensity on T2-weighted and fluid-attenuated inversion recovery (FLAIR) images, hypointensity or normal appearance in T1-weighted images, and post-contrast gadolinium enhancement (10). Also, there might be diffusion abnormalities in the lesion areas (9). Although the pathological mechanisms of how brain lesions develop are not fully known, one of the accused mechanisms is glutamate neurotoxicity (11). Although our case's first MRI was normal, the control MRI showed classical findings for the WE.

Until recently, there were no evidence-based recommendations on how to manage thiamine replacement therapy. But, there are some different new regimens for especially severe deficiencies such as advising two days of 3x500 mg IV thiamine with 5 days of 1x500 mg parenteral thiamine and long-term oral 50-100 mg thiamine supplementation (12). Also in patients having bariatric surgery, it is advised to surgeons and endocrinologists to screen for thiamine deficiency for post-bariatric surgery with rapid weight loss and other risk factors. Upon questioning, it was learned that our patient did not receive any protective supplementary medication or diet after the operation. Both the patient and the parents had some psychosocial behavioral issues that could contribute to misunderstanding or not following the recommended dietary supplement. High-dose thiamine replacement was performed in our patient in accordance with the new guidelines, and the treatment yielded good results.

Wernicke encephalopathy is a medical condition requiring immediate attention. Bariatric surgery is one of the causes and as performed more frequently, we are expected to exposure WE more frequently. This diagnosis must be in mind for patients with eating problems and vomiting, not taking vitamin supplements. Since it's sometimes difficult to get thiamine analyzed, empirical treatment is recommended. Sometimes, it might be with some unusual findings like papilledema and polyneuropathy. With effective treatment, it is expected to relieve ocular findings in hours to days, for vestibular functions and ataxia in two weeks. But mental confusion might go on for weeks, even sometimes there might be sequela in cognitive functions.

In the case we reported, the presence of papilledema, obesity, and indistinct characteristics of WE at the beginning caused a delay in diagnosis. Although it is thought that our patient had polyneuropathy, which is one of the uncommon clinical presentations of WE, it could not be confirmed due to technical reasons. In the control one year later, the EMG could not be repeated because the patient did not want it. This is one of the limitations of our article. This case was thought to be important as it would increase awareness of the uncommon findings of Wernicke's encephalopathy.

Table 1. Wernicke's Encephalopathy Etiology

• Chronic alcoholism
• Anorexia nervosa / diet
• Hyperemesis gravidarum
• Long-lasting, poor supported total parenteral nutrition
• Long-lasting fasting, unbalanced feeding (especially refeeding cases)
• Gastrointestinal surgery (especially bariatric surgery)
• Systemic malignancy
• Transplantation
• Hemodialysis or peritoneal dialysis
• AIDS (Acquired Immune Deficiency Syndrome)
• Genetic disorders of thiamine metabolism

(This is one of the patients in the poster presented under the title "Bariatrik Cerrahi Sonrası ve Hiperemesis Sonrası Gelişen İki Wernicke Ensefalopatisi Olgusu" in the 52th National Neurology Congress, which is organized by the Turkish Neurological Society.)

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