

## Pseudotümör Serebri Kliniği ile Presente Olan Hashimoto Ensefalopatili Bir Olgu Sunumu

### Hashimoto Encephalitis Presented With Clinical Manifestations of Pseudotumor Cerebri: A Case Report

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#### Özet

Hashimoto ensefalopatisi, otoimmün etyolojiye bağlı olduğu düşünülen ve yüksek antitiroid antikörleri ile birlikte seyreden bir ensefalopati tablosudur. Klinik görünüm global amnezi, psikoz, kognitif etkilenme, epileptik nöbetler şeklinde olabilmektedir. Burada kliniğimize iki yanlı görme bozukluğu, baş ağrısı, davranış değişikliği, uyku düzensizliği şikayeti ile başvuran ve muayenesinde iki yanlı papil ödem saptanıp Pseudotümör serebri (PTS) tanısıyla servisimizde takip edilen 69 yaşında bir kadın hasta sunulacaktır. Etiyolojiye yönelik yapılan tetkiklerinde Hashimoto Tiroiditi harici özellik saptanmayan hastaya iki kez yapılan boşaltıcı lomber ponksiyon (LP) ve 5 gün süreyle 1 gr/gün IV metilprednisolon tedavisi uygulanmış ve hastada klinik düzelleme gözlenmiştir. PTS tablosuna nadiren Hashimoto ensefalopatisi sebep olması nedeniyle olgumuzu sunmayı uygun gördük.

**Anahtar Kelimeler:** Pseudotümör serebri, Hashimoto ensefalopatisi, tiroidit, bilişsel bozulma

#### Abstract

Hashimoto encephalopathy is an encephalopathy associated with high antithyroid antibodies and it is thought to be connected with autoimmune etiology. Clinical presentation may be in the forms of global amnesia, psychosis, cognitive impairment and epileptic seizures. Herein, we will present a case of a 69-year-old female patient who applied to our clinic with bilateral visual impairment, headache, behavioral changes, sleep disorders, bilateral papilledema and established diagnosis of pseudotumor cerebri. As an etiological factor, only Hashimoto thyroiditis was determined. Drainage with lumbar puncture was performed for two times and IV methylprednisolone treatment at daily doses of 1 g was administered for 5 days with resultant clinical improvement. We wanted to present our case of Hashimoto encephalopathy as a rarely seen etiological factor of pseudotumor cerebri.

**Keywords:** Pseudotumor cerebri, Hashimoto encephalopathy, thyroiditis, cognitive dissonance

#### INTRODUCTION

Pseudotumor cerebri is a disease affecting typically obese women in their child-bearing ages. The patients present with daily episodes of widespread headache and visual symptoms. Generally, papilledema is present. All these symptoms may be seen solely (1, 2). They are characterized by increased intracranial pressure without any demonstrable pathology. Although its etiology is not known exactly, its diagnosis can be made by excluding other diagnoses (1). The syndrome has idiopathic and secondary causes. Among frequently seen secondary causes thyroid replacement therapy and hormonal abnormalities including thyroid disorders, hypo- and hyperthyroidism have been detected.

Hashimoto encephalopathy is a rarely seen type

of encephalopathy characterized by normal thyroid functions and antithyroid antibody positivity. It is also called autoimmune dementia. It responds favorably to immunotherapy. It is characterized by subacute or fluctuant cognitive disorder. Its other frequently seen characteristics include tremor, myoclonus, seizures, headache, hypersomnolence and psychiatric symptoms. Lord Brain firstly defined it in the year 1966. Since thyroid function test results are within normal limits, it seems to be directly unrelated to thyroid dysfunction. It is a rarely seen, life-threatening and curable condition (3).

#### CASE REPORT

An otherwise healthy 69-year-old female hypertensive patient who presented with complaints of bilateral vision loss lasting for the

previous 15 days was evaluated in the outpatient clinic of ophthalmology. She was referred to our clinic because of detection of bilateral papilledema and peripheral narrowing of the visual field (tubular vision) during visual field examination. Meanwhile she had frontal headache, nausea and vomiting recurring for 1-2 times. From her intimates and relatives, it has been learnt that for the previous one month she was demonstrating behavioral changes, also especially exaggerated dependency on her grandchild and sleep disorders. She was hospitalized in our service with initial diagnosis of PC in order to investigate the etiology of intracranial hypertension syndrome and with the intention of further examination and treatment.

Neurological examination of the patient with Body Mass Index (BMI) of 27 kg/m<sup>2</sup> was unremarkable except for bilateral tubular vision. Routine biochemical test results were as follows: Erythrocyte sedimentation rate (ESR) 118 mm/h, C reactive protein (CRP) 77 mg/L, T3 2.78 ng/dL, T4 1.50 ng/dL, TSH 0.182 uIU/mL, anti TPO; 45.68 IU/mL, anti-thyroglobulin antibody 700.8 ng/mL, Hg: 9.3 mg/dl and MCV 75 fL. Cranial MR and MR venograms were unremarkable except for an encephalomalasic area in the left frontal lobe. Lumbar puncture (LP) was performed twice. Cerebrospinal fluid (CSF) opening pressure was mildly increased (300 mm H<sub>2</sub>O). CSF was clear and acellular. Its protein level was slightly elevated (45.9 mg/dL). Any atypical cell was not detected. Any bacterial growth was not observed in the culture media. Following LP, the patient expressed alleviation of her headache, while her visual complaints did not change. Tests of the patient who previously suffered from brucellosis were repeated. Tests were negative for brucellosis. Since the clinical status of the patient did not improve despite acetazolamide therapy and LP drainage, also tests for paraneoplastic syndrome and vasculitis were performed. Tumor markers, ENA profile,

ANA, anti dsDNA, coagulation panel were requested. The test results were unremarkable. Abdominal ultrasonography (US), breast US and mammograms were requested in order to search for malignancy. Her gynecologic, endoscopic and colonoscopic examinations were unremarkable. Thyroid US of the patient with increased thyroid autoantibody levels revealed signs suggestive of thyroiditis. Thyroid scanning was interpreted in favor of chronic thyroiditis, Hashimoto thyroiditis. Control thyroid function tests of the previously euthyroid patient demonstrated evidence of hypothyroidism. Occasional episodes of amnesia, temporal confusion and behavioral complaints necessitated psychiatric evaluation which did not suggest any abnormality. Existing symptoms of the patient suggested thyroiditis-related encephalitis. The patient was treated with daily doses of 1 gr IV methylprednisolone administered for 5 days. Ophthalmologic examination revealed development of optic atrophy at the right eye and complete healing of the papilledema of the left eye. At her follow-up visit complete relief of her headache and improvement of her visual complaints, personality changes and symptoms of intracranial hypertension syndrome were detected, while her control thyroid function tests revealed only signs of a mild hypothyroidism.

## DISCUSSION

The beginning complaints of the patient were visual field constriction, blurred vision and changes in behavior. Bilateral papilledema was found in eye examination. High CSF pressure with LP was obtained and this was considered as PTS. The first complaint with PTS patients is visual impairment and papilledema (7). When secondary reasons that may cause PTS was investigated like obesity, cerebral venous thrombosis, metabolic and endocrinological reasons and drug use, autoimmune thyroiditis had been identified in our patient but no other

cause. Behavioral change complaint can be explained by autoimmune thyroiditis (8). In the literature, very few cases have been reported that shows the association with PTS and autoimmune thyroiditis (3-4, 9). In addition, cases developing after treatment with levothyroxine in juvenile and congenital hypothyroidism were defined in the literature (5, 10-11). But it is seen that the described cases are in childhood or adolescence period and there are no cases described in the older age groups. Although there was no evidence of infection, when blood parameters were examined, ESR and CRP were high in our patient. With autoimmune thyroiditis combination of ESR and CRP are common (12). High levels in ESR and CRP may be considered as a cautionary finding in patients diagnosed with PTS when systemic infection is excluded. Behavioral disorders and dementia findings respond well to steroid treatment in autoimmune thyroiditis. In our patient, these findings have improved with steroid treatment and this is a result which supports our diagnosis.

During investigation process for the etiological factors in patients presenting with neuropsychiatric symptoms, headache, visual disturbances, papilledema, most frequently thyroid function tests and thyroid autoantibody analyses are not requested. Since it is a rarely encountered condition which can be treated at the time of diagnosis, in the differential diagnosis of the patients presenting with intracranial hypertension syndrome, cognitive affect and psychiatric symptoms and diagnosis of Hashimoto encephalitis should not be forgotten.

## REFERENCES

1. Dhungana S, Sharrack B, Woodroffe N. Idiopathic intracranial hypertension. *Acta Neurologica Scandinavica* 2010;121: 71-82.
2. Peng K, Fuh J, Wang S. High-pressure headaches: idiopathic intracranial hypertension and its mimics. *Nature Reviews/ Neurology* 2012;8:700-10.
3. Press OW, Ladenson PW. Pseudotumor Cerebri and

- hypothyroidism. *Arch Intern Med.* 1983 Jan;143(1):167-8.
4. Dickman MS, Somasundaram M, Brzozowski L. Pseudotumor cerebri and hyperthyroidism *N Y State J Med.* 1980 Jun;80(7 Pt 1):1118-20.
5. Van Dop C, Conte FA, Koch TK, Clark SJ, Wilson-Davis SL, Grumbach MM. Pseudotumor cerebri associated with initiation of levothyroxine therapy for juvenile hypothyroidism. *N Engl J Med.* 1983 May 5;308(18):1076-80.
6. Castillo P, Woodruff B, Caselli R, Vernino S, Lucchinetti C, Swanson J, et al. Steroid-Responsive Encephalopathy Associated with Autoimmune Thyroiditis. *Arch Neurology* 2006;63(2): 197-202.
7. Friedman DI Papilledema and idiopathic intracranial hypertension. *Continuum (Minneapolis Minn).* 2014 Aug;20(4 Neuro-ophthalmology):857-76.
8. Leyhe T, Müssig K. Cognitive and affective dysfunctions in autoimmune thyroiditis. *Brain Behav Immun.* 2014 Oct;41:261-6.
9. Adams C, Dean HJ, Israels SJ, Patton A, Fewer DH. Primary hypothyroidism with intracranial hypertension and pituitary hyperplasia. *Pediatr Neurol.* 1994 Mar;10(2):166-8.
10. Strickler C, Pilon AF. Presumed levothyroxine-induced pseudotumor cerebri in a pediatric patient being treated for congenital hypothyroidism. *Clin Ophthalmol.* 2007 Dec;1(4):545-9.
11. Huseman CA, Torkelson RD Pseudotumor cerebri following treatment of hypothalamic and primary hypothyroidism. *Am J Dis Child.* 1984 Oct;138(10):927-31.
12. Erden S, Buyukozturk S, Vural P, Değirmencioglu S. Acute-phase reactants in Hashimoto thyroiditis. *Int Immunopharmacol.* 2008 Dec 20;8(13-14):1863-5.