Olgu Sunumu
JCAM
Case Report

# Aphasia, Just a Neurological Disorder?

# Afazi Yalnızca Nörolojik bir Bozukluk Mudur?

Afazi / Aphasia

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**Özet**

Hashimoto ensefalopatisi (HE), otoimmün tiroidit ile ilişkili nadir görülen bir hastalıktır. Etiyolojisi tam olarak anlaşılamamıştır. Serum antitiroid antikorları HE'de yüksek seviyelerde görülür. Karakteristik olarak HE Bakteriyel veya viral enfeksiyonların yokluğunda, bilişsel bozukluk, psikiyatrik ve nörolojik belirtiler ile birlikte görülen otoimmun etyolojili bir tablodur. Steroid tedavisine duyarlı bir ensefalopatidir. Altmış yaşında erkek hasta, yaklaşık 9 aydır devam eden unutkanlık ve son 2 gündür olan konuşma kaybı nedeni ile hastaneye kabul edildi. Tiroid fonksiyon testlerinde ileri derece hipotroidi tespit edilmesi üzerine yapılan Elektroensefalografi ve MR görüntülemeleri HE ile uyumlu bulundu. Hashimoto ensefalopatisi nadir görülen ve yeterince tanınmayan bir durum olduğundan diğer nedenler dışlandıktan sonra ayırıcı tanıda düşünülmesi gereken bir hastalıktır. Hashimoto ensefalopatisi doğru tanısı bilişsel ve klinik sunumların farkındalığı ile konulabilir.

**Anahtar Kelimeler**

Hashimoto Ensefalopatisi; Afazi; Nörolojik Bozukluk

**Abstract**

Hashimoto's encephalopathy (HE) is a rare disorder associated with autoimmune thyroiditis. Etiology of HE is not completely understood. High levels of serum antithyroid antibodies are seen in HE. Presentation with otoimmune thyroiditis, cognitive impairment, psychiatric and neurologic symptoms and absence of bacterial or viral enfections are characteristics of HE. HE is a steroid responsive encephalopathy. 60 years old male patient admitted to hospital with forget fulness continuing for 9 months and speech loss starting 2 days ago. Strong positivity of antithyroid antibodies increases the odds for HE. Thyroid function tests showed severe hypothyroidism. Electroencephalography and magnetic resonance imaging results were compatible with HE. HE is diagnosed with differantial diagnosis and exclusion of other reasons. This uncommon disorder is not recognised enough. High titres of serum antithyroid antiboides are always needed for diagnosis. Correct diagnosis requires awareness of wide range of cognitive and clinical presentations of HE.

**Keywords**

Hashimoto's Encephalopathy; Aphasia; Neurological Disorder

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## Introduction

HE is a rare disease which is sensitive to steroids. Diagnosis is made with exclusion of other reasons of encephalopathy and finding positive results for antithyroid antibodies. This syndrome was first described at 1966 by Brain et al[1]. High levels of serum antithyroid antibodies are essential for diagnosis of HE. Autoimmune encefalopathy associated with HE has variable clinical and psychological symptoms reported at case reports[2]. Thyroid pathologies have widespread prevalance at society. At this case report; we present a male patient with HE and aphasia associated with hypothyroidism who had delayed diagnosis.

## Case Report

Our patient is a 60-year-old male retired doorman who doesn't have history of diabetes, hypertension, usage of alcohol and tobacco. He had problems of forgetfulness, impairment of attention and concentration disorder for 9 months. He admitted to neurology clinic due to speech complaints continuing for 2 days. Patient's symptoms include forgetting of familiar names and places of his belongings. For last two days, aphasia added to patient's clinical status. Cognitive examination of patient showed attention deficit, impairment of executive functions, loss of visual memory and computation disorder. Myoclonus was present at both upper and lower extremities. Mini Mental Test evaluation was imposibble due to presence of aphasia. Moderate generalised cortical atrophy was found at cranial MR imaging(Fig.1)..

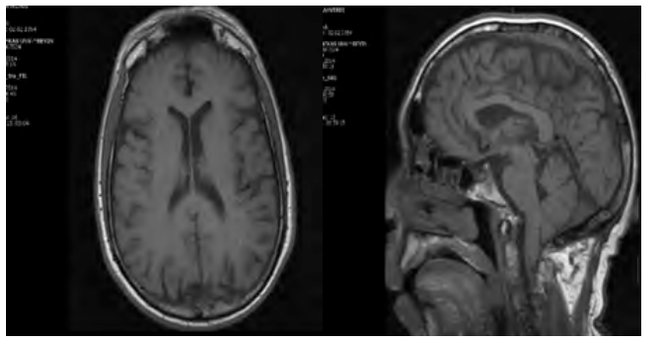


Fig.1. T1- Weighted axial and sagittal cranial MRI showed mild generalized cortical atrophy

Antithyroglobulin antibody level was 600/ml (referance value<34 ml), thyroid function test results were compatible with hypothyroidsm. T3:0,260 ng/ml (2-4,4 ng/ml) T:0,045 ng/ml (0,93-1,70 ng/ml)

Patient's EEG findings at admission were interpreted as generalized slowdown and intermittent rhythmic delta nerve activity . Thyroid ultrasonography showed diffuse heterogeneous and hypoechogen gland.

With these neurological findings and high positivity of anti thyroid antibodies besides the exclusion of another causes of encephalopathy; we have diagnosed the patient with HE and hypothyroidsm. Therapy with 100 mg / day thyroxine and dexamethasone 10 mg / day was started. Patient's symptoms resolved with the treatment of levothyroxin and steroid and he resumed to normal life. Mini mental test score has raised to 29. But his cognitive evaluation showed no development at attention disorder. Aphasia has recovered after two days of steroid admission. Patient's control EEG results 2 months after his hospital discharge were normal.

## Discussion

HE is an uncommon clinical syndrome. Diagnosis of this rare disorder requires positive anti TPO level and exclusion of other cases for encephalopathy. Prevalance is reported 2,1/100.000 and male/female ratio for adults is 1:4[3]. Until present approximately 130 cases were reported and most of these cases were female. In contrast to female predominance of these cases our patient was male[4].

Etiology and pathophysiology of HE could not be understood clearly. These patients have increased sensitivity at different otoimmune processes. Antithyroid antibodies and unidentified antibodies which effects brain functions can be the culprit. Possible mechanisms include cerebral vasculitis, toxic effects of TSH and antibodies to central nervous system[5]. Another opinion suggests that thyroid antibodies bind to cerebellar astrocytes and cause pathojenic effects[6]. Two type of presentation for HE has been reported. Focal neurologic symptoms and cognitive dysfunctions can be seen at type with vasculitis. Other type causes progressive and diffuse deterioration resulting confusion, seizures, psychosis, dementia, fast or slow progressing cognitive impairment and somnolence. Myoclonus, tremor, ataxia, focal or generalised seizures, psychiatric symptoms, visual hallucinations can be seen at diffuse type[4,7].

Both of these types can be seen together and these symptoms are potentially reversible[8]. Plenty of diseases like metabolic, toxic, vasculer and neoplastic disorders have similar characteristics with HE; noticing HE requires careful differantial diagnosis[4]. HE presents itself with clinical, labaratuary and imaging findings. Clinical symptoms are usually progressive[8].Tremor, transient aphasia, seizures, gait ataxia, somnolence, myoclonus, neuropsychiatric symptoms and stroke like condition can occur in 1-7 days of HE[4].

Our patient had expressive aphasia. Focal or tonic-clonic seizures were reported two thirds of the cases. Psychosis is present %85 of the patients[9]. Our patient had myoclonus and depression for one month. Elevated antithyroid antibody levels at serum is an important feature of HE. Thyroid antibodies react with brain tissue and these antibodies effect central nervous system. There is a strong correlation between the titres of antibodies and severity of neurologic findings[4,10]. Serum anti TPO and anti Tg levels are elevated at %100 and %73 of the HE cases respectively[4,7].

Autoantibodies against the NH2-terminal of a-enolase (anti-NAE) are reported positive at % 44 of the cases[11]. We could not assess the level of anti-NAE at our patient. Thyroid function test results show variability at the cases of HE. %35 of the cases have subclinical hypothyroidism, %30 of them have normal thyroid functions, %20 of them have evident hypothyroidism and %7 of them have hyperthyroidism[4,9,10]. Our patient had evident hypothyroidism.

Cranial MR imaging can be normal or can show nonspesific findings at white matter. At some studies, %50 of the HE cases had abnormal CT and MR findings such as white matter abnormalities and subcortical or focal cortical abnormalities[7,12]. Our patient had cerebral atrophy at MR imaging, which is a

nonspesific finding. Abnormal electroencephalography (EEG) results and high total protein levels at Cerebrospinal fluid (CSF) are seen at many of the HE cases[13]. Our patient's EEG results at admisson showed generalized slowdown and intermittent rhythmic delta activity. These results were concordant with literature.

At a study for treatment, patients received hormone replacament therapy and steroids. %92 of the patients who had hypothyroidism and HE had recovered[12]. Our patient received both corticosteroid and levothyroxine. Long term prognosis of HE patients varies, there is a high response to treatment but HE can also be progressive or relapsing[4,10]. There are delays at HE diagnosis because HE demands awareness of its manifestations diagnosis. Treatment delays or no treatment at HE causes poor prognosis. More than %25 of the HE patients have permanent cognitive impairment[14]. Our patient's follow up to present day for ten months showed euthyroidism and mild forgetfulness, all of other cognitive functions of patient is normal.

## Conclusion

HE is an already known disorder. Presentation with mild encephalopathy and aphasia can cause difficulties at diagnosis. A complete and careful neuropsychiatric evaluation is needed. Otoimmune encephalopathy related to hypothyroidism has variable neurologic findings as can be seen at our patient, therefore clinicians should be aware of this.

### *Competing interests*

The authors declare that they have no competing interests.

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