



Evaluation of LGI1-antibody Encephalitis, a Rare Cause of Limbic Encephalitis, from Diagnosis to Treatment

Limbik Ensefalitin Nadir Bir Nedeni Olan LGI1-antikor Ensefalitinin Tanıdan Tedaviye Kadar Değerlendirilmesi

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Anahtar Kelimeler: LGI1-antikor ensefaliti, prognoz, tedavi

Dear Editor,

Anti-leucine-rich glioma inactivated-1 (LGI1)-associated limbic encephalitis is a rare neurological disease presenting with subacute progressive encephalopathy and fasciobrachial dystonic seizures (FBDS) (1). LGI1-associated limbic encephalitis is characterized by a poor prognosis, although focal-onset epileptic seizures and cognitive impairment often respond to immunotherapy (2). The prevalence of autoimmune encephalitis is 13/100000, and the incidence rate is 1.2/100000 (3). A case of LGI1-related autoimmune encephalitis responding to immunotherapy with subacute onset cognitive impairment and seizures resistant to antiepileptic therapy was planned to be discussed in the light of the literature. A 67-year-old female patient was admitted to the neurology service for further examination and treatment due to complaints of worsening episodic memory, disorientation, apathy in the last few weeks, aimless wandering around the house and startle on the right side in the last 3 months. After she was hospitalized, it was observed that she had difficulty in fulfilling two-step orders. In the mini-mental state examination, the patient scored 21/30. Serum sodium level was 128 meq/ml, which recovered spontaneously without treatment. In her clinical follow-up, fasciobrachial dystonic seizures were observed. They occurred many times during the day, lasting for 10 seconds, accompanied by a clonic beat on the right side of the face, in which the head turned to the right, awareness was preserved, and dystonic posture on the right arm was observed. Interictal wakefulness electroencephalography revealed left frontotemporal weighted 5-6 Hz 100-110 μ V sharp

wave activity together with left frontocentrottemporal 6-7 Hz 40-45 μ V theta activity (Figure 1). Levetiracetam 1000 mg/day was started and the dose was gradually increased to 3000 mg/day. In the cranial magnetic resonance imaging (MRI) of the patient, on

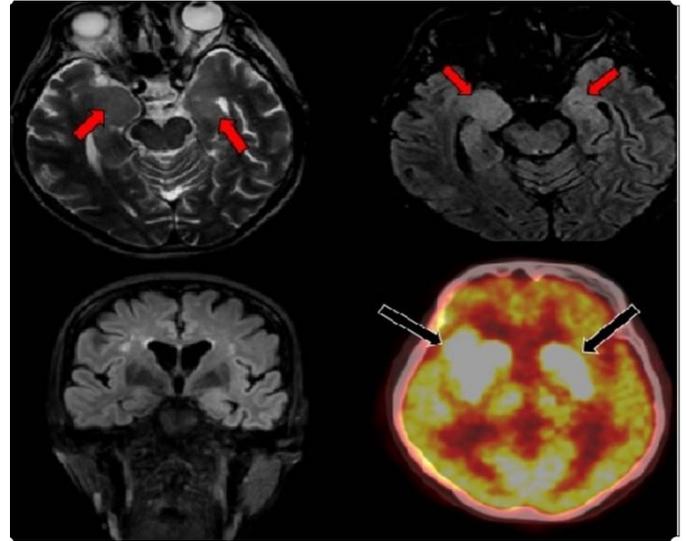


Figure 1. In the cranial MRI, T2W hyperintense bilateral mesial temporal lesions are seen. On the brain PET, bilateral hypermetabolism in the basal ganglia are seen

MRI: Magnetic resonance imaging, PET: Positron emission tomography

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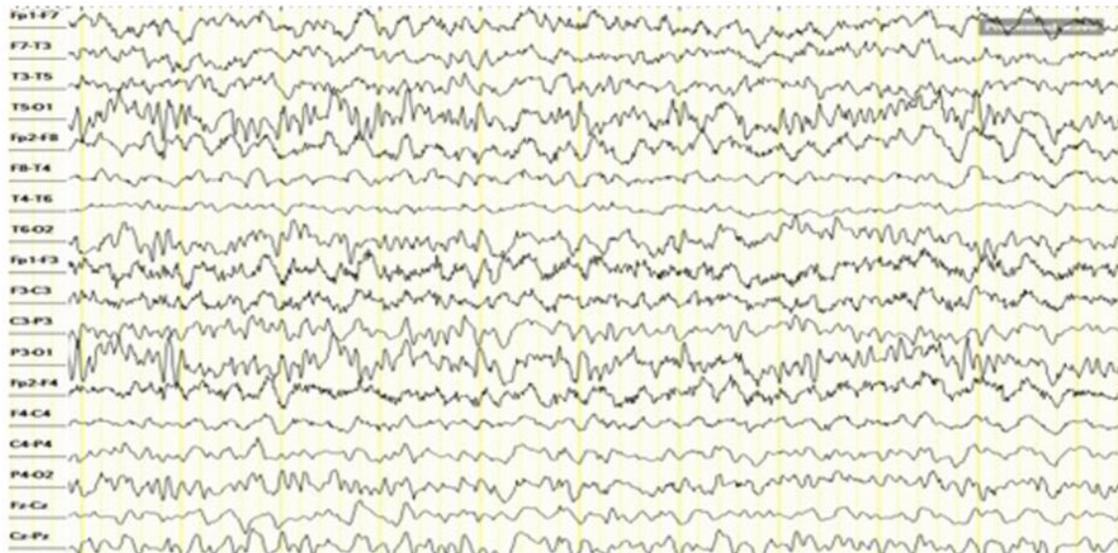


Figure 2. Interictal wakefulness electroencephalography shows left frontotemporal sharp wave activity together with left frontocentrotemporal theta activity

T2W and FLAIR sequences, there were hyperintensities in bilateral mesial temporal lobes (Figure 2). Glucose and protein levels, and cell counts were normal in the cerebrospinal fluid. As the patient's seizures continued, lacosamide 100 mg/day was started and the dose was increased to 400 mg/day due to ongoing seizures. Among the serological markers studied in serum sample, anti-LGI1 was seropositive. Thorax computed tomography (CT), abdominal CT, and whole-body positron emission tomography (PET) did not show any malignancy. On the brain PET, bilateral hypermetabolism was observed in the basal ganglia (Figure 2). Intravenous immune globulin (IVIG) was started at a dose of 2 g/kg for 5 days, it was observed that FBDS decreased in the first week after treatment, but there was no significant improvement in cognition. Serum sodium level recovered spontaneously. In the first month after discharge, 1000 mg/day IV methylprednisolone treatment was administered due to sleepiness and frequent seizures. In the first month follow-up, her seizures had disappeared completely, but the patient was still describing episodic memory problems. The patient was treated with 0.5 g/kg IVIG per month and she was stable in the first year. In our patient, limbic encephalitis was suspected due to hyperintensity in the bilateral hippocampal structures in T2W and FLAIR sequences in cranial MRI in a patient with subacute onset dementia and focal onset epileptic seizures. For LGI1-related autoimmune encephalitis, it can be said that FBDSs are typical with subclinical seizures. These seizures can repeat many times during the day, lasting for a few seconds, with dystonia, spasm, and contractions involving the ipsilateral face in which awareness is affected or preserved. Ictal extremity dystonia indicates involvement at the level of the basal ganglia, amnesia with ipsilateral automatism indicates involvement in the temporal region, and the forced and involuntary turning of the trunk and head (versive seizure) indicates involvement in the frontal region (4). LGI-1-associated encephalitis often presents with resistance to antiepileptic drug therapy. It exhibits a dramatic response to immunotherapy and corticosteroids. Currently the optimal duration of immunotherapy is not known. In animal studies, LGI1 was found to be highly expressed in the hypothalamus and kidney

tubules, so it was assumed that the resistant hyponatremia seen in LGI1-related autoimmune encephalitis was a result of autoimmune mechanisms in the hypothalamic-pituitary axis (5). Its association with malignancy is rare in the literature and it is emphasized that malignancy should be excluded. LGI1 encephalitis should be kept in mind in patients with rapidly progressive cognitive impairment with an age peak between 61-64 years of age.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: H.İ.U., Y.D., A.B.D., Concept: H.İ.U., Y.D., A.B.D., Design: H.İ.U., Y.D., A.B.D., Data Collection or Processing: H.İ.U., Y.D., A.B.D., Analysis or Interpretation: H.İ.U., Y.D., A.B.D., Literature Search: H.İ.U., Y.D., A.B.D., Writing: H.İ.U., Y.D., A.B.D.

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