ISSN: 2643-4539

Çetin et al. Int J Neurodegener Dis 2022, 5:024
DOI: 10.23937/2643-4539/1710024
Volume 5 | Issue 1
Open Access

International Journal of
Neurodegenerative Disorders

CASE REPORT

The Management of LG1-Associated Encephalitis that Presents with Acute Impaired Consciousness

Güldeniz Çetin[1*], Nazire Çelem[2], Özdem Ertürk Çetin[2], Şevki Şahin[2] and Serkan Demir[2]

[1]*Kartal Kızılay Hospital, Turkey*

[2]*Dr. Ilhan Varank Sancaktepe Training and Research Hospital, Turkey*

***Corresponding author:** *Güldeniz Çetin, Ilhan Varank Sancaktepe Training and Research Hospital, Turkey*

Summary

Anti-LGI1-associated autoimmune encephalitis is a limbic system pathology characterized by persistent seizures, cognitive and neuropsychiatric symptoms and is treated with immunomodulators depending on the autoimmune etiology. Our case is an example of an 88-year-old male patient who came with autoimmune encephalitis without typical characteristics.In this article, a case of limbic encephalitis who presented with the complaint of confusion will be discussed, and the clinical approach will be discussed regarding the diagnosis and treatment of cases with atypical clinical findings.

Introduction

An autoimmune encephalitis is a group of syndromes that have a paraneoplastic or immunological etiology characterized by subacute onset memory disorder and seizures. Early diagnosis and treatment management of it is difficult because viral encephalitis can be confused with conditions such as psychiatric diseases, dementia, and delirium. Although it responds well to immune treatment, treatment-resistant cases may also be seen rarely. Early diagnosis and treatment are very important because the disease can cause irreversible cognitive destruction, epilepsy, and coma when delayed. The purpose of this case report is to bring to mind autoimmune encephalitis in acute-onset consciousness disorders differentfrom its subacute and epileptic forms and to emphasize the contribution of early diagnosis and immunosuppressive treatment to cognitive deterioration and prevention of epilepsy.

Case Report

An 88-year-old male patient was brought to our neurology clinic with complaints of perseverations not exceeding 5 minutes, echolalia, and incomprehension once or twice a day that lasted for 4-5 days. His neurological examination and laboratory examination results were normal, and he did not have a fever. The Mini-Mental Test Battery result was 27/30. It was learned that the patient, who had a history of hepatic encephalopathy, received 800 mg/day carbamazepine treatment, and he had been neglecting the treatment for the last few days although he paid attention to regular use. Epileptic discharge was not observed in routine EEG, but it was consistent with diffuse background rhythm slowing. He was admitted to our ward for examination and treatment.

Diazepam½ ampoule was administered with the preliminary diagnosis of nonconvulsive status epilepticus (NCSE) for the patient whose orientation and cooperation deteriorated immediately after admission to the ward, and his complaints regressed completely afterthe diazepam administration. Considering the status, levetiracetam 3000 mg/day infusion was started. A Valproic acid infusion was added at a dose of 20-40 mg/kg/day to the patient, who had deteriorated consciousness after 12 hours of well-being. However, the patient, whose complaints recurred in less that 12 hours, and whose attack durations were prolonged, underwent contrast-enhanced CR MRI with the preliminary diagnosis of Subcortical infarct (?) Encephalitis (?) Meningitis (?). No pathology was detected in the imaging (Figure 1).

Citation: Çetin G, Çelem N, Çetin OE, Şahin S, Demir S (2022) The Management of LG1-Associated Encephalitis that Presents with Acute Impaired Consciousness. Int J Neurodegener Dis 5:024. doi.org/10.23937/2643-4539/1710024

Accepted: December 20, 2022: **Published:** December 22, 2022

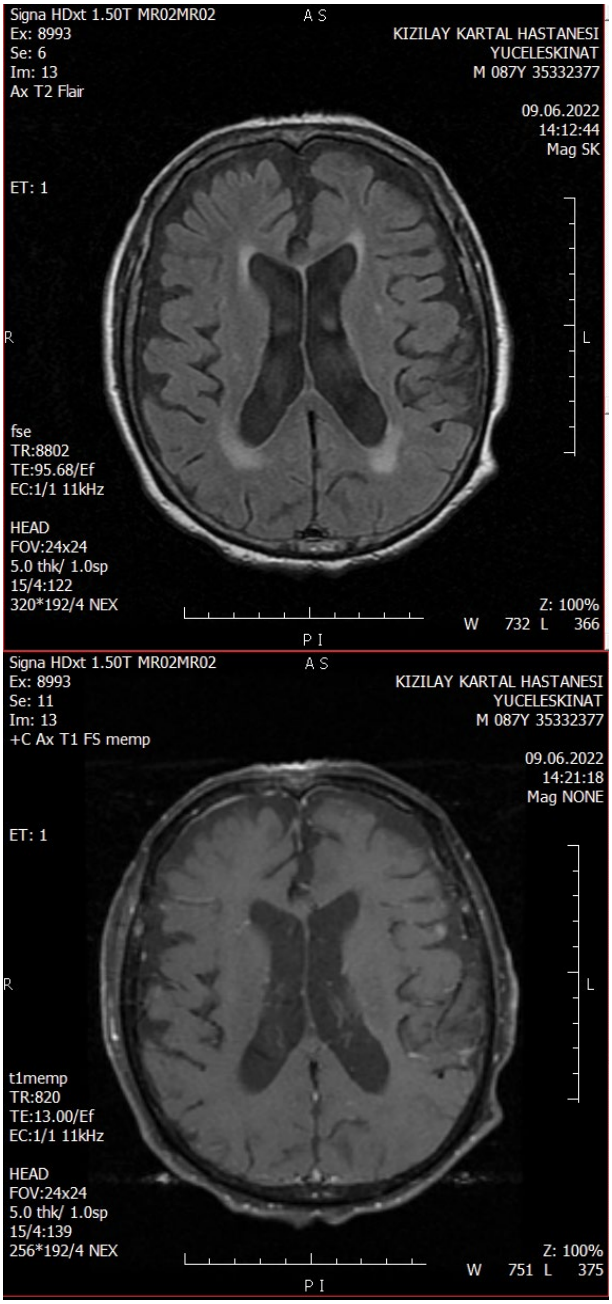

Figure 1: CR MRI with the preliminary diagnosis of Subcortical infarct (?) Encephalitis (?) Meningitis (?).

Laboratory test results were normal during the follow-ups in inpatient treatment. Since the patient had a history of hepatic encephalopathy, his ammonia level was found to be normal. Autoimmune-paraneoplastic encephalitis was included in the differential diagnosis because of the new-onset epileptic seizures of the patient and the absence of biochemical and central structural pathologies related to the etiology of the diffuse background slowing detected in the EEG. Abdominal USI and abdominal CT for malignancy screening revealed no pathology except for non-cirrhotic hepatic shrinkage andthoracic CT was normal.

A lumbar puncture was performed with the suspicion of autoimmune-paraneoplastic encephalitis. CSF findings were normal except for total protein elevation. The CSF viral meningitis and encephalitis panel were negative. A panel of autoimmune and paraneoplastic encephalitis was sent from the serum. No pathology was detected in the paraneoplasia panel, but anti-LGI-1 autoantibody was found +1 positive in the autoimmune encephalitis panel scan. Intravenous immunoglobulin (IVIG) treatment was given for immunosuppressive treatment for 5 days. Since his agitation continued during the follow-up examinations, aripiprazole and haloperidol were added to the treatment with support from psychiatry. Although there was partial seizure control, 1 gr/day IV methylprednisolone treatment was administered for 10 days after 3 weeks to achieve complete remission. Seizure control was achieved after 5 days of loading and monthly IVIG treatment after IV methylprednisolone, the patient did not have seizures. The Mini-Mental Test Battery was the same as on arrival at discharge.

Discussion

LGI1-associated encephalitis, which was first described in 2001, is an autoimmune disease characterized by faciobrachial dystonic seizures (FBDS) accompanying cognitive impairment, and persistent hyponatremia and increased T2 FLAIR signal in the medial temporal lobes on magnetic resonance imaging (MRI) [1]. It is usually detected in middle-aged men, and the mean age of onset is 60 [2].

Patients often present with subacute onset of consciousness disorders. It was shown in neuropsychiatric tests that there is deteriorated verbal and visuospatial episodic memory performance, impaired verbal fluency and storage, delayed free recall, and difficulty in recalling. Although short-term memory impairment is observed in the early period, permanent memory problems develop with hippocampus changes in the chronic period [3,4]. In addition to memory problems, many psychiatric disorders such as agitation, depression, and hallucinations might accompany the clinical manifestation and be confused with the delirium-dementia complex [5]. The development of rapidly progressive cognitive disorders in 15% of patients complicates the differential diagnosis of neurodegenerative diseases [6].

Seizures accompany more than 80% of cases. Faciobrachial dystonic seizures (FBDS), which are described recently as a phenomenon, are characteristic of LGI1-LE [7]. Although this type of seizure is semiologically suggestive of frontal lobe involvement, it is found in basal ganglia changes, unlike usual frontal or temporal lobe seizures [8].

Hyponatremia is another important finding of LGI1-related encephalitis, with an incidence of 60-88% [9,10]. Inappropriate ADH syndrome develops because of excessive secretion of LG1 antibodies in the hypothalamus and kidney [9,11].

Demonstration of LG1 autoantibodies in CSF is specific for the diagnosis but might not always be detected. The reason for this may be a 1-10% difference between CSF VGKC antibody values and serum values [10].

PET-CT can be used as an aid in examining brain activity and monitoring treatment response. PET finding, which is characterized by hypo-metabolism in the parietal and occipital cortex and hyper-metabolism in the basal ganglia, can be evaluated in favor of autoimmune encephalitis [12].

There is no fixed protocol for immunotreatment, which is the gold standard. Depending on the patient's clinical manifestation, intravenous or oral corticosteroids, plasmapheresis, IVIG, or combinations can be applied [13].

There are several significant points in the presentation of our case. The first of these was the acute state of consciousness change, whichis not usual in limbic encephalitis. Although the fluctuating distraction, persevere speech and echolalia, and difficulty in understanding the words developing in the last 1 week were evaluated primarily in favor of subcortical infarction, the normal imaging, the control of antiepileptics for some time, and the complete disappearance of the complaints with immunotreatment showed that this change was associated with limbic encephalitis.

Another characteristic is the absence of convulsive seizures. Although no epileptic wave activity was detected in EEG, changes in consciousness, which is a partial response to antiepileptics, suggested the presence of accompanying complex partial seizures.

The inability to show the mesial temporal lobe and basal ganglia inflammation characteristic of the diagnosis of limbic encephalitis in our patient is one of the most challenging parts of the diagnosis process. Although no pathology was detected except for a minimal protein elevation in the CSF examination, the presence of LG1 autoantibody in the serum established the diagnosis.

Conclusion

Although few cases were detected in the literature regarding LGI1, which is a rare encephalitis type, the importance of early diagnosis still maintains its seriousness. The purpose of this case study was to emphasize that the devastating effects of the disease can be eliminated with early diagnosis and immunotreatment in patients who do not have clinically characteristic autoimmune encephalitis manifestation. The diagnosis of autoimmune encephalitis should be kept in mind in acute consciousness disorders even without seizures, hyponatremia, pathological CSF, and MRI findings. The importance of close follow-up of the patient and differential diagnosis can determine the progression of a curable neurodegenerative disease.

References

1. Krastinova E, Vigneron M, Le Bras P, Gasnault J, Goujard C (2012) Treatment of limbic encephalitis with anti-glioma-inactivated 1 (LGI1) antibodies. J Clin Neurosci 19: 1580-1582.
2. Li Z, Cui T, Shi W, Wang Q (2016) Clinical analysis of leucinerich glioma inactivated-1 protein antibody associated with limbic encephalitis onset with seizures. Medicine (Baltimore) 95: e4244.
3. Griffith SP, Malpas CB, Alpitsis R, O'Brien TJ, Monif M (2020) The neuropsychological spectrum of antiLGI1 antibody mediated autoimmune encephalitis. J Neuroimmunol 345: 577271.
4. Van Sonderen A, Schreurs MW, Wirtz PW, Sillevis Smitt PA, Titulaer MJ (2016) From VGKC to LGI1 and Caspr2 encephalitis: The evolution of a disease entity over time. Autoimmun Rev 15: 970-974.
5. Gao L, Liu A, Zhan S, Wang L, Li L, et al. (2016) Clinical characterization of autoimmune LGI1 antibody limbic encephalitis. Epilepsy Behav 56: 165-169.
6. Escudero D, Guasp M, Arino H, Gaig C, Martinez-Hernandez E, et al. (2017) Antibody-associated CNS syndromes without signs of inflammation in the elderly. Neurology 89: 1471-1475.
7. Irani SR, Stagg CJ, Schott JM, Rosenthal CR, Schneider SA, et al. (2013) Faciobrachial dystonic seizures: The influence of immunotherapy on seizure control and prevention of cognitive impairment in a broadening phenotype. Brain 136: 3151-3162.
8. Szots M, Marton A, Kover F, Tunde K, Timea B, et al. (2014) Natural course of LGI1 encephalitis: 3-5 years of follow-up without immunotherapy. J Neurol Sci 343: 198-202.
9. Lai M, Huijbers MG, Lancaster E, Francesc G, Luis B, et al. (2010) Investigation of LGI1 as the antigen in limbic encephalitis previously attributed to potassium channels: A case series. Lancet Neurol 9: 776-785.
10. Vincent A, Buckley C, Schott JM, Ian B, Bonnie KD, et al. (2004) Potassium channel antibody-associated encephalopathy: A potentially immunotherapy-responsive form of limbic encephalitis. Brain 127: 701-712.
11. Ellison DH, Berl T (2007) The syndrome of inappropriate antidiuresis. N Engl J Med 356: 2064-2072.
12. Tripathi M, Tripathi M, Roy SG, Parida GK, Ihtisham K, et al. (2018) Metabolic topography of autoimmune non-paraneoplastic encephalitis. Neuroradiology 60: 189-198.
13. Shin YW, Lee ST, Shin JW, Moon J, Lim JA, et al. (2013) VGKC-complex/LGI1-antibody encephalitis: Clinical manifestations and response to immunotherapy. J Neuroimmunol 265: 75-81.