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Anti-LGI1 Antibody Autoimmune Encephalitis. Clinical Case Presentation and Literature Review

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Summary. Anti-leucine-rich glioma inactivated 1 (anti-LGI1) encephalitis is a rare autoimmune limbic encephalitis. As the clinical presentation of this disease is similar to other types of encephalitis most often associated with paraneoplastic process or endocrine disorder, a thorough testing for oncological or endocrine disease is necessary to correctly differentiate the diagnosis and administer appropriate treatment.

We present a clinical case of a 63-year-old female patient treated in the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kaunas Clinics for impairment of consciousness, probably of epileptic origin. After identifying memory problems, focal impaired awareness seizures, cognitive dysfunction, and typical brain magnetic resonance imaging changes, the diagnosis of limbic encephalitis was suspected. A thorough testing allowed to exclude paraneoplastic processes or endocrine syndromes, and the results of the cerebrospinal fluid antibody panel confirmed the diagnosis of anti-LGI1 antibody autoimmune encephalitis. Further treatment using glucocorticoids and intravenous immunoglobulin gave good results.

Keywords: autoimmune encephalitis, anti-LGI1 antibody, hyponatremia, seizures.

CLINICAL CASE PRESENTATION

We present a clinical case of a patient treated in the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kaunas Clinics for impairment of consciousness, probably of epileptic origin. The patient was a 63-year-old woman. On August 25, the patient had a motor vehicle collision, possibly due to impaired consciousness, and was immediately admitted to a local hospital to treat several accident-related fractures of the ribs. Also, according to the patient's daughter, since spring her mother had suffered from episodes of disorientation and loss of consciousness. The consultation of the neurologist showed impairment of consciousness, probably of epileptic origin. The recommended treatment was carbamazepine 200 mg 3 times per day (daily dose 600 mg) as the electroencephalography (EEG) showed epileptiform activity in the right temporal and frontotemporal areas and the left temporal area. After the treatment was completed and the patient's clinical status remained stable, it was decided to discharge the patient from the hospital with the recommendation to continue taking carbamazepine.

However, on September 14, the patient returned to the hospital and was consulted by the neurologist for memory impairment and lethargy. Brain computed tomography (CT) was performed which showed no changes. Blood test showed hyponatremia (128 mmol/L). The treatment was adjusted with valproic acid tablets 500 mg 2 times per day.

On September 20, the patient's sister noticed that the patient was disoriented, making it difficult to establish meaningful contact. Recently, the patient was bothered by facial twitches. She was hospitalized in the neurology department of the local hospital. In the ward, the patient had impaired perception, orientation, was difficult to make contact with, and did not follow orders. No other focal neurological symptoms were observed. CT of the brain revealed hypodense areas in parietal dorsal and left occipital areas of the cerebrum, giving suspicion of posterior reversible encephalopathy syndrome (PRES). Analysis of cerebrospinal fluid (CSF) showed no significant changes. Blood biochemistry revealed significant hyponatremia

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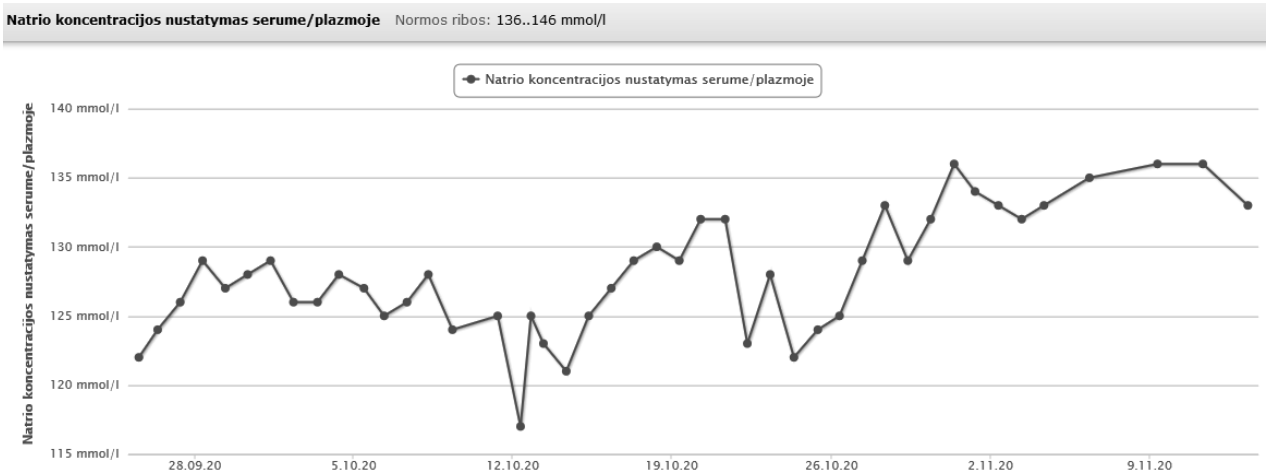

Fig. 1. **Change in serum sodium concentration (mmol/L) during inpatient treatment in the Neurology Department**

(120 mmol/L). Hypertonic saline (NaCl) solution and valproic acid tablets 750 mg/day were prescribed. The patient regained contact but remained disoriented in time and partially in space, had impaired perception and persistent hyponatremia. In other words, the disease was only barely responding to the treatment requiring a rethink of possible diagnosis since epilepsy itself could not explain such phenomena.

On September 25, due to uncertain diagnosis and poor response to antiepileptic drug therapy, the patient was transferred to the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kaunas Clinics for further examination and treatment. Taking the patient's history at the Neurology Department, she could not remember the events of the previous 2–3 months. An objective examination revealed that the patient was conscious, had contact, but was disoriented in time. The questions were not answered meaningfully and the patient could not find appropriate words to describe her condition. During the neurological examination, horizontal nystagmus to the right side was observed with no other focal neurological symptoms. During the entire stay in the Neurology Department, several focal clinically clear epileptic seizures with impaired consciousness lasting ~5 minutes were observed, as well as prolonged post-seizure disorientation.

On October 2, several paroxysms (2 of them encephalographically quite clear, 2 marginal) were recorded in the waking EEG. In the T+/–F domain, initially there was a small episode of depression followed by low-amplitude frequent activity which gradually transitioned to higher-amplitude pseudo-rhythmic slow waves followed by single peak waves. Such episodes would last ~30–40 seconds. In the right temporal field, very slow (delta) waves were recorded. Epileptiform activity in the right frontotemporal and left temporal areas was recorded using EEG between attacks. Valproic acid was prescribed and drug concentrations monitored; the optimal dose was found to be 500 mg 2 times per day. With anti-epileptic treatment administered, seizures were not observed.

During inpatient treatment, treatment-resistant hyponatremia persisted even with hypertonic NaCl infusions (Fig. 1).

On October 1, the patient was consulted by the endocrinologist because of treatment-resistant hyponatremia. The endocrinological cause of hyponatremia (e. g., the syndrome of inappropriate antidiuretic hormone secretion (SIADH)) was excluded after the following biochemical tests:

- urine osmolarity (338 mOsm/kg (50–1400 mOsm/kg))
- serum osmolarity (274 mOsm/kg (280–301 mOsm/kg))
- urine panel (specific gravity 1.004 (1.01–1.025), no other abnormalities)
- sodium concentration in serum (129 mmol/L) and urine (115 mmol/L)
- AKTH (7.5 pmol/L (1.63–14.15 pmol/L))
- radioimmunoassay of morning serum cortisol (579.91 nmol/L)
- Ca (ionised 1.04 mmol/L (1.2–1.43 mmol/L), unionized 2.48 mmol/L (2.2–2.65 mmol/L))
- K (4.25 mmol/L)
- Cl (91 mmol/L (101–109 mmol/L))
- blood glucose (6.26 mmol/L)
- TTH (2.41 mU/L (0.4–3.6 mU/L)

To rule out a possible etiology of the oncological origin of hyponatraemia, thorough tests (chest X-ray, abdominal ultrasound (US), mammography, breast US, thyroid and parathyroid US, esophagogastroduodenoscopy (EGDS), fecal occult blood test, serum cancer markers, cardiac US) were performed and gynecologist's consultation was provided. No evidence of the oncological process was found. Also, serum tests for chronic infections (B. burgdorferi infection, HIV, syphilis) and markers of autoimmune encephalitis (NMDA, CASPR2, AMPA1/2, LGI1, DPPX, GABARB1/B2) were negative.

On October 8, magnetic resonance imaging (MRI) of the brain was performed (Fig. 2). On both sides of the hippocampus in T2W sequences, an unevenly (right>left) higher signal intensity (SI) was detected without signifi-

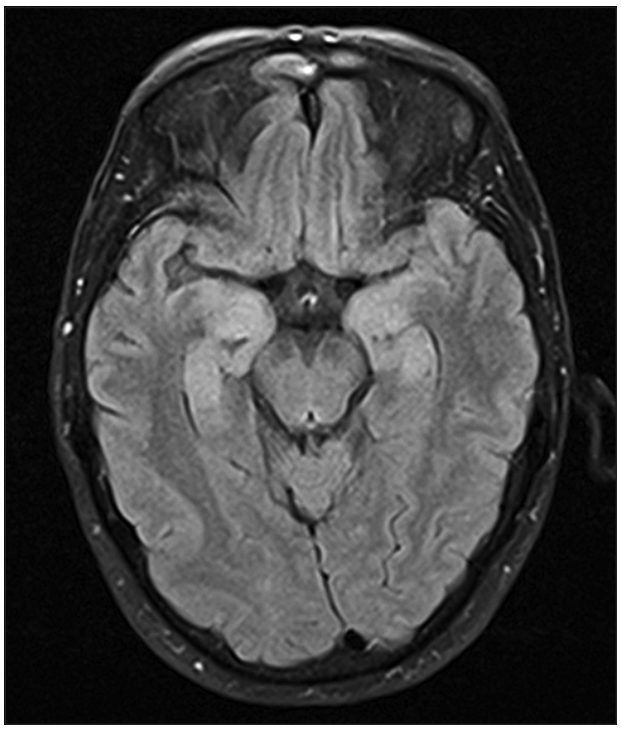

Fig. 2. **Brain MRI scan (T2 FLAIR sequence) of the patient performed on October 8, 2020**

cant decrease in volume. Hippocampal sclerosis could not be excluded as well. Limbic encephalitis was suspected.

On October 12, a lumbar puncture was performed to analyze cerebrospinal fluid which revealed cytosis (3 cells/mm^3, Er 1, Leu 2) and increased protein content 0.61 g/L (prior was 0,45 g/L); antibodies to neuronal surface antigens (NSA-ab) (NMDA, CASPR2, AMPA1/2, LGI1, DPPX, GABARB1/B2) were taken from cerebrospinal fluid. Markers for Ab against antigens associated with paraneoplastic neurological syndromes were negative, with the rest of the autoimmune encephalitis panel expected a few days later.

Based on the examinations performed and a high suspicion of limbic encephalitis reinforced by consultation with radiologists, glucocorticoid pulse therapy with 1 g of methylprednisolone for 3 days was prescribed from October 14. The observed partial effect was a small correction of hyponatremia – 132 mmol/L (October 22). However, significant negative changes in serum sodium concentration (123 mmol/L) were observed the following day (October 23). Meanwhile, the patient's state of consciousness did not change significantly.

On October 15, the full results of the autoimmune encephalitis CSF panel were received which revealed positive anti-LGI1 antibodies, the titer 1:32. Repeated consultation of the endocrinologist showed that the syndrome of inappropriate antidiuretic hormone secretion (SIADH) could not be ruled out. On October 19, tests to exclude limbic encephalitis caused by oncological process (CT of the chest, abdomen, and pelvis) were performed which did not reveal sufficient data for the oncological process. Thus, excluding possible oncological causes, the diagnosis of anti-LGI1 antibody autoimmune encephalitis was confirmed.

Prednisolone (60 mg/day) was prescribed for supportive treatment. Positive changes in hyponatremia were observed – an increase to 136 mmol/L. The patient's condition improved due to the restored consciousness, attention, and memory. On November 3, as the patient's condition improved, prednisolone dose was reduced by 5 mg every 5–7 days. On November 12, when the patient's condition remained significantly improved and hyponatremia was corrected (between 133–136 mmol/L), the patient was discharged for outpatient treatment. On November 30, the patient was contacted, she took prednisolone tablets 10 mg/day. On November 26, sodium serum concentration was 140 mmol/L – well within normal limits. The patient felt well, orientation and perception disorders were not observed. Intravenous IgG was not prescribed during this period.

On January 6, 2021, the patient was re-hospitalized in the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kaunas Clinics due to a relapse. The patient complained of trembling hands for 2 weeks, the right hand being more affected, and "throwing" sideways. Neurological examination showed slow thinking and difficulty in finding words. Horizontal nystagmus to the right was observed. When smiling, a tremor of facial muscles and eyelids was observed. Small intentional hand tremor, right>left. No other neurological symptoms were observed. For relapse treatment, intravenous immunoglobulin 2 g/kg (i. e., 100 g) for 5 days was prescribed. On January 7, awake EEG showed excitability changes and slow waves recorded right frontally. Sodium serum concentration was 135 mmol/L. On January 11, after improvement of clinical condition and normalization of sodium concentration, the patient was referred for further rehabilitation treatment.

LITERATURE REVIEW

Epidemiology

Anti-leucine rich glioma inactivated 1 (anti LG1) encephalitis is a rare autoimmune limbic encephalitis, associated with voltage-gated potassium channels (VGKC). Overall, the prevalence of autoimmune encephalitis increased from 0.4 cases per 100,000 people between 1995–2005 to 1.2 per 100,000 people between 2006–2015 [1]. Anti-LGI1 antibody autoimmune encephalitis is reported to afflict 0.7–1 per 1,000,000 people per year [2–4].

Etiology and pathogenesis

A group of diseases of autoimmune encephalitis is associated with antibodies to neuronal cell surface or synaptic proteins [5]. The target antigens usually play critical roles in synaptic transmission and plasticity. These types of antigens are mainly generated by our immune system or they are cancer-related antibodies. LGI1 together with Caspr2

belong to VGKC-complex antibodies [6]. VGKC adjusts the neuronal excitability of the central and peripheral nervous systems [7]. VGKC antibody was first found in patients with Isaacs' syndrome. Patients suffering from this syndrome complain of progressive muscle stiffness, cramping and weakness, muscle twitching with a rippling appearance (myokymia), delayed muscle relaxation, diminished reflexes, muscle atrophy, difficulty coordinating voluntary movements, and increased sweating which are caused by increased excitability of peripheral nerves [8].

LGI1 is a glycoprotein released by presynaptic membranes and it can interact with presynaptic membrane ADAM metallopeptidase domain 22 and presynaptic membrane ADAM metallopeptidase domain 23 [9]. The binding of anti-LGI1 antibodies may disrupt pre- and postsynaptic LGI1 signalling, resulting in neuronal hyperexcitability [10]. In the hippocampus, LGI1 proteins regulate glutaminergic circulation and interact with ADAM 22/23 transmembrane proteins to regulate AMPA receptor-mediated signal transduction [1, 11]. Immunopathology studies have shown neuronal loss and lymphocyte infiltration around the blood vessels of the hippocampus and amygdala [12].

Since the discovery of anti-LGI1 antibodies in approximately 2010 [13, 14], much has been learned about their role in autoimmune encephalitis. However, its etiology remains unclear. Only 5 to 10% of cases are associated with cancer; the most commonly associated tumor is thymoma [15]. On the other hand, some studies and case reports have shown that anti-LGI1 limbic encephalitis is mainly non-paraneoplastic, tumor markers and radiological examinations are negative [6]. These findings suggest that most of the etiological factors of anti-LGI1 antibody autoimmune encephalitis are still unknown.

Clinical signs

Anti-LGI1 antibodies usually affect neurons in the hippocampus and temporal lobe. Hyperexcitability of the neurons causes clinical signs characteristic of the disease onset. Anti-LGI1 antibody autoimmune encephalitis may present with the impairment of cognitive functions, e. g., memory disturbance. Memory deterioration may be preceded by epileptic seizures, myotonia, mental disorders, and faciobrachial dystonic seizures (FBDS) [16]. Interestingly, hyponatremia and rapid eye movement sleep behavior disorder (REM) are also consistent with this disease.

Some of these symptoms have been shown to have a scientific explanation. For instance, in experiments on mice, it was shown that disruption of the anti-LGI1 protein gene causes temporal lobe epilepsy which is so severe that it can lead to death [14]. After an attack of the disease, patients may experience significant mood and sleep disorders which are mainly related to the damage of the hippocampus and amygdaloid nucleus [17]. In case reports analyzed by Chinese scientists, 60% of autoimmune encephalitis patients with anti-LGI1 antibodies had intractable hyponatremia [6]. Hypotheses state that this hyponatremia may be caused by the syndrome of inappropriate antidiuretic hormone secretion (SIADH) and may be related to the simultaneous LGI1 expression of the hypothalamus and kidney [1, 10].

Diagnostics

As autoimmune encephalitides are quite rare, all possible differentiating diagnostic tools are important for making diagnosis. It is important to carefully study the patient's history and evaluate cognitive functions, as well as to perform blood tests for the assessment of electrolytes (especially sodium which may show hyponatremia) and to analyze CSF for cytosis and protein (can be elevated in neuroinflammatory diseases). Sleep and awake EEG may help differentiate encephalitis from epilepsy. Brain MRI can help find structural changes that may mimic underlying symptoms or reveal characteristic symptoms of limbic encephalitis (high-intensity signal in temporal lobes or hippocampus using T2W/FLAIR sequence). Both blood and CSF should be tested for the markers of autoimmune encephalitides. [18]. Differential diagnosis is provided in the Table.

Treatment

The first-line treatment of anti-LGI1 antibody autoimmune encephalitis is pulse therapy with methylprednisolone, intravenous immunoglobulin, and plasmapheresis [19]. Studies show that treatment with glucocorticoids, IVIG, mycophenolate mofetil, and plasma exchange have resulted in significant clinical improvement in 70 to 80% of patients [20]. Supportive evidence includes a small randomized trial of 17 patients with anti-LGI1 (n=14) or anti-Caspr2 encephalitis and frequent seizures (two or more per week) which suggested that treatment with IVIG was superior to placebo [21]. Other observational data suggest that early initiation of immunotherapy in patients with faciobrachial dystonic seizures may prevent the development of cognitive impairment and improve long-term outcomes [22]. However, treatment with one type of immunosuppressive therapy (e.g., methylprednisolone) might be insufficient and the result may be delayed thus a combination of drugs should be used [23, 24].

Second-line treatments include rituximab, cyclophosphamide, tocilizumab, and interleukin-2 [13]. Australian scientists performed a systematic review [13] where they showed the superiority of the first-line treatment over the second one. Second-line treatment showed higher relapse rate 26.1% vs. 18.7% and lower rates of good outcome (mRS score 0 or seizure freedom) 43.5% vs. 86.7% [13].

Additional symptomatic treatment with antiepileptic drugs to prevent seizures and hypertonic saline solution to adjust sodium concentration is also important [19].

Outcomes

The course of anti-LGI1 antibody encephalitis is subacute, but gradually progressive. Despite the treatment,

Table. **Differential diagnosis**

Disease	Causes	Symptoms	Diagnostics
Anti-LGI1 encephalitis	Autoimmune disease, 5–10% of cases are related to cancer (most commonly – thymoma)	Cognitive impairment (e. g., memory), seizures, hyponatremia, myotonia, FBDS, REM sleep disorders	Anti-LGI1 antibodies in blood serum and CSF
Hyponatremia (rapid correction may result in central pontine myelinolysis due to inadequately changed blood osmolality and dehydration of brain cells)	High blood volume: liver cirrhosis, congestive heart failure, nephrotic syndrome, excessive drinking of fluids Normal blood volume: SIADH, hypothyroidism, pregnancy Low blood volume: vomiting, diarrhea, diuretic use, Addison's disease, pancreatitis	Nausea, vomiting, headache, short term memory loss, confusion, lethargy, fatigue, loss of appetite, irritability, muscle weakness, cramps, seizures, coma	Blood test for sodium ions (normal range 135–145 mmol/L)
Dementia syndrome	Alzheimer's disease, vascular dementia, Lewy's bodies dementia, frontotemporal dementia, progressive supranuclear palsy, corticobasal degeneration, alcohol-related dementia, prion disease	Progressive disturbance of cognitive functions (memory, perception, orientation, language, attention, problem solving), psychological (depression, anxiety) and behavioral symptoms (restlessness, agitation, aggression, sexual)	Careful collection of medical history, including the patient's relatives, clarification of risk factors, MMSE, MoCA, CASI tests, MRI scan to confirm brain atrophy
Epilepsy	Genetic and acquired causes like brain tumor, traumatic brain injury, infections of CNS	Seizures, bite of the tip of the tongue, aura, post-ictal retrograde amnesia	Characteristic EEG changes, clinically observed seizure, responsiveness to anti-seizure therapy
PRES (posterior reversible encephalopathy syndrome)	Certain parts of the brain become swollen because of severely elevated blood pressure, kidney failure (especially in those who receive hemodialysis), sepsis, autoimmune causes (thrombotic thrombocytopenic purpura, primary sclerosing cholangitis, rheumatoid arthritis, Sjögren syndrome, systemic lupus erythematosus, Crohn's disease, neuromyelitis optica)	Seizures, headache, visual disturbances, altered mental state, sometimes limb weakness or inability to speak	Identification of risk factors that result in neurological symptoms. Brain MRI showing symmetrical hyperintensities on T2-weighted imaging in the parietal and occipital lobes
Other autoimmune encephalitides	Anti-NMDA, anti-Caspr2, anti-AMPA, anti-GABA-A, anti-GABA-B, anti-DPPX, and other	Sleep disorders, memory deficits, seizures, decreased level of consciousness, frequent dyskinesias (orofacial, choreoathetoid movements, dystonia), autonomic instability (hyperthermia, tachycardia, bradycardia, blood pressure fluctuations), language dysfunction	Corresponding blood serum and CSF antibodies
SIADH (syndrome of inappropriate antidiuretic hormone secretion)	Dysregulation of ADH secretion may be caused by CNS infections, brain trauma, multiple sclerosis, small cell lung cancer, lymphoma, various drugs (carbamazepine, valproic acid, sertraline, oxytocin, amitriptyline)	Anorexia, nausea, muscle aches, generalized muscle weakness, myoclonus, ataxia, decreased reflexes, tremor, asterixis, Cheyne-Stokes respiration, lethargy, dysarthria, confusion, seizures, coma	Low serum osmolality (<275 mOsm/kg), low serum sodium, high urine sodium, no usage of drugs, no evidence of cirrhosis, nephrosis, thyroid, adrenal dysfunction, or hyperglycemia

the disease can progress or have relapses. After treatment, full recovery of cognitive functions is unlikely, but possible [25]. Deaths from anti-LGI1 antibody autoimmune limbic encephalitis have also been reported [9]. Combination therapy reduces the likelihood of relapse [6].

CONCLUSION

A rare case of anti-LGI1 antibody autoimmune encephalitis is presented in the article. The clinical presentation was similar to several diseases stated in the differential diagnosis. No single underlying cause of treatment-resistant

hyponatremia, seizures, and memory deficit was found during initial clinical examination. These symptoms are seemingly unrelated: epilepsy cannot explain persistent hyponatremia, acute hyponatremia does not cause rapid cognitive decline, and dementia does not cause seizures. A set of unexplained symptoms required a search for a systemic autoimmune disease. Radiological examination revealed no cancer. Cerebrospinal fluid tests for various autoimmune encephalitides revealed positive anti-LGI1 antibodies which confirmed the diagnosis and explained all the symptoms.

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ANTI-LGI1 AUTOIMUNINIS ENCEFALITAS. KLINIKINIS ATVEJIS IR LITERATŪROS APŽVALGA

Santrauka

Anti-LGI1 encefalitas yra reta autoimuninio limbinio encefalito forma. Kadangi klinikiniai šios ligos požymiai panašūs į kitų tipų encefalitus, dažniausiai susijusius su paraneoplastiniu procesu ar endokrininiu sutrikimu, yra būtinas nuodugnus ištyrimas dėl onkologinės ar endokrininės ligos, siekiant teisingai nustatyti diagnozę ir paskirti tinkamą gydymą.

Pristatome klinikinį atvejį apie 63 m. pacientės ligos diagnozės nustatymą ir gydymą Lietuvos sveikatos mokslų universiteto Kauno klinikų Neurologijos klinikoje dėl galimai epileptinės kilmės sąmonės sutrikimo. Dėl atminties sutrikimų, židininių traukulių su sutrikusia sąmone, kognityvinių sutrikimų ir tipiškų pokyčių, stebimų smegenų MRT, yra įtariamas limbinis encefalitas. Išsamus ištyrimas leidžia atmesti paraneoplastinius procesus ar endokrininius sindromus, o likvore esančių antikūnų paletės tyrimas patvirtina autoimuninio anti-LGI1 encefalito diagnozę. Skirtas gydymas gliukokortikoidais ir intraveniniu imunoglobulinu leidžia pasiekti gerų rezultatų.

Raktažodžiai: autoimuninis encefalitas, anti-LGI1 antikūnai, hiponatremija, traukuliai.

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