

LGI1-Antibody Associated Autoimmune Encephalitis Complicated by Primary Polydipsia

Dear Editor

Leucine-rich glioma-inactivated 1 (LGI1) antibody encephalitis is one of the subtypes of the autoimmune encephalitis characterized by antibodies against the LGI1 region of

the voltage-gated potassium channels.^[1] This subtype is characterized by behavioral abnormalities, memory deficits, and classical facio-brachial dystonic seizures besides hyponatremia. We hereby present a typical case of LGI1

associated encephalitis complicated by primary polydipsia previously not reported in the literature.

A 28-year-old female was admitted to our department with complaints of memory impairment, irrelevant talking, and abnormal body movements. As reported by her family members, one month back just one week after her engagement, she started questioning about the Mehendi applied on her hands and feet and could not recollect much about her engagement ceremony. Her family members were also concerned about her not recognizing them properly and asking repeatedly for meals even though she would have eaten already. Over the past three weeks she developed abnormal movements of her right arm and face that would last for few seconds and come repeatedly throughout day and night. On examination she was confused, talking irrelevantly and there was no focal neurological deficit. She had multiple episodes of facio-brachial seizures that would last for few seconds only and come repeatedly after a few minutes. Her baseline investigations were normal except for hyponatremia. Because of a subacute encephalopathy, memory deficits, characteristic facio-brachial seizures, and hyponatremia, a provisional diagnosis of LGII encephalitis were made. Her spinal fluid analysis revealed 5 cells/ μL (all neutrophils) with normal protein (45 mg/dl) and sugar (75 mg/dl with blood sugar of 96 mg/dl). Her MRI brain revealed bilateral symmetrical T2/FLAIR hyperintensities [Figure 1] in mesial temporal lobes and insular cortices with no diffusion restriction or contrast enhancement. Her CSF autoimmune panel was positive for the LGII antibody. She was managed with pulse steroid therapy and antiepileptic drugs. However, she did not show any improvement, and instead, she developed polyuria (average 9L/day) and polydipsia (average 8L/day) without polyphagia. Her blood sugar was normal (88 mg/dl) and the main differentials for polyuria and polydipsia were primary polydipsia and diabetes insipidus. However, her serum sodium was persistently low and the water deprivation test did not show a rise in serum osmolality ruling out diabetes insipidus [Table 1]. Hence a diagnosis of LGII-antibody autoimmune encephalitis complicated by primary polydipsia was made. She was treated by IVIG over 5 days and she showed drastic improvement in encephalopathy, facio-brachial seizures, and osmotic symptoms. She was discharged on steroids and azathioprine and is currently on our follow-up.

Limbic encephalitis associated with LGII antibodies has certain characteristic features that may make clinical diagnosis easy. Facio-brachial seizures are one such feature; these are dystonic movements of the face and ipsilateral arm that may remain for a few seconds and are usually unilateral but can sometimes involve both sides.^[2,3] Prominent psychiatric features like odd behavior, disinhibition, acute psychosis, and memory impairment are other features of this disease.^[4] Another characteristic feature of LGII associated autoimmune encephalitis is hyponatremia that may be resistant to treatment.^[5] Hyponatremia may be seen in 60-80% of

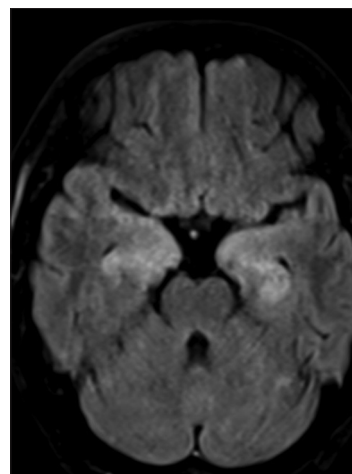


Figure 1: FLAIR MRI sequence showing hyperintensities in bilateral medial temporal lobes

Table 1: Water Deprivation Test

Time	Urine Osmolality	Serum Sodium
At 0 hours	130 mOSm/Kg	127 meq/L
At 2 hours	277 mOSm/Kg	132 meq/L
At 4 hours	397 mOSm/Kg	125 meq/L

patients with LGII associated encephalitis and is usually due to SIADH.^[6] Imaging may be normal in up to 50% of patients but may reveal T2/FLAIR hyperintensities in medial temporal lobes that are usually unilateral but may also be bilateral.^[7] First-line treatment in LGII encephalitis is steroids usually in conjunction with intravenous immunoglobulin and/or plasmapheresis. This therapy is usually effective in 80% of patients although the benefit may take time to appear. Our patient had all the characteristic clinical features of LGII-antibody-associated encephalitis making diagnosis easier further strengthened by a positive CSF LGII antibody test. Our patient however did not respond to initial steroid pulse therapy and developed significant polyuria and polydipsia on the 8th day of admission. On the evaluation of these osmotic symptoms, she had persistent hyponatremia with low urine and serum osmolality. The water deprivation test did not increase serum osmolality although there was an increase in urine osmolality thus confirming the diagnosis of primary polydipsia.

Primary polydipsia is a condition characterized by excessive consumption of water leading to dilute urine and hyponatremia. Primarily found in psychiatric disease patients like schizophrenics, this disorder can also occur in patients with an organic brain disease like sarcoidosis. This is the first case report of primary polydipsia in a patient with autoimmune encephalitis published in the literature. Our patient was managed with IVIG after which she showed a dramatic response in her seizures and polyuria and polydipsia. As more and more cases of LGII encephalitis are reported worldwide newer features of this disease become evident.

Primary polydipsia is one such feature that has not been previously reported.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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