

A VERY YOUNG GIRL LIMBIC ENCEPHALITIS WITH LGI1 ANTIBODIES

GULCAN AKYUZ YUCEL¹, GULTEN OZTURK², OLCAY UNVER², DILSAD TURKDOGAN²

¹UNIVERSITY OF HEALTH SCIENCES, SANCAKTEPE TRAINING AND RESEARCH HOSPITAL

²UNIVERSITY OF MARMARA, PENDIK TRAINING AND RESEARCH HOSPITAL

INTRODUCTION

Anti LGI1 encephalitis is an autoimmune disease characterized by antibodies against leucine-rich glioma inactivated 1 (LGI1) proteins and most commonly seen in male patient at an age range of 30-80 years. Memory disturbances, confusion and seizures are accompanied by dystonic faciobrachial seizures presenting with myoclonus or dystonia often not responsive to anti epileptics. Patients might also have sleep disturbance or hyponatremia.

CASE

We present an 8 years 9 months old girl who presented with tonic seizures, memory disturbances, involuntary dystonic movements, ataxia, sleep disorders. The electroencephalogram (EEG) showed bilateral posterior delta slow wave activity and right fronto-central spike and wave activities and focal status. Her paraneoplastic and limbic encephalitis panels were normal. Her blood autoimmune encephalitis panel had LGI1 antibodies.

She was admitted to hospital after a seizure and memory problems and was given levatiracetam. Pulse steroid, IVIG therapy was given her. Her memory disturbance and ataxia were recovered after the applied immunotherapies. The EEG returned to normal shortly. The patient received azathioprine as a steroid sparing immunomodulatory therapy. She is symptom free since then.

CONCLUSION

Anti LGI1 is the second most common cause of autoimmune encephalitis after anti NMDA encephalitis. To the best of our knowledge this is the youngest patient reported in the literature described so far.

LGI1 encephalitis is well responsive to immunosuppressive therapy and should be considered in strongly suspected autoimmune encephalitis in case of especially dystonic faciobrachial seizures, memory problems and free other autoimmune antibodies.

REFERENCES

- 1- Irani SR, Michell AW, Lang B, et al. Faciobrachial dystonic seizures precede Lgi1 antibody limbic encephalitis. Ann Neurol 2011; 69:892.
- 2- Sen A, Wang J, Laue-Gizzi H, et al. Pathognomonic seizures in limbic encephalitis associated with anti-LGI1 antibodies. Lancet 2014; 383:2018.
- 3- Thompson J, Bi M, Murchison AG, et al. The importance of early immunotherapy in patients with faciobrachial dystonic seizures. Brain 2018; 141:348.
- 4- López-Chiriboga AS, Klein C, Zekeridou A, et al. LGI1 and CASPR2 neurological autoimmunity in children. Ann Neurol 2018; 84:473.

CONTACT

gulcan.akyuz@hotmail.com