

Article: 0812

Topic: EPW08 - e-Poster Walk Session 08: Research Methodology, Genetics and Molecular Neurobiology, Psychoneuroimmunology

Anti-vgkc Antibody-associated Limbic Encephalitis Presenting with Recurrent Catatonia

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Introduction

Limbic encephalitis is an inflammatory disorder, commonly characterized by psychiatric features and cognitive impairment. It used to be considered a rare disease, usually paraneoplastic, with poor prognosis. Recent findings of membrane-surface directed antibodies, including anti-VGKC (Voltage-Gated Potassium Channels) antibodies, showed a substantial proportion of cases with no association with any malignancy and better prognosis.

Case presentation

A 56-year-old woman was admitted to the psychiatric ward in Nov/2007, with an acute onset of psychomotor retardation and agitation periods, apathy and nihilistic symptoms. She improved very slowly with lorazepam, clomipramine and quetiapine. In Jun/2014, she was admitted again to the psychiatric ward with acute onset of intense headache, confusion, insomnia, agitation and prejudice ideas. There was a clinical deterioration with mnesic deficits, catatonic stupor, autonomic dysfunction and seizures. Complementary exams revealed: ESR>120mm; hyponatraemia; acellular CSF with slightly elevated protein; EEG with slow base activity; Brain MRI with diffuse cortical atrophy and multiples subcortical hyperintensities of frontoparietal regions on FLAIR and T2 sequences; and positive anti-VGKC antibodies. Initially there was a poor transitory improvement with high-dose steroids and intravenous immunoglobulins. After 5 months, she did plasmapheresis with relevant clinical improvement.

Discussion and Conclusions

About 80% of anti-VGKC encephalitis patients have clinical improvement following prompt treatment. In this case, there was not as good improvement as initially expected probably because of the prolonged duration of disease prior to effective treatment. Given the good prognostic outcome with prompt initiation of immunotherapy, early recognition and diagnosis of this rare neuropsychiatric syndrome is crucial.