

Rare presentation of VGKC encephalitis

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Introduction:

Autoimmune encephalitis includes a group of disorders with self-autoantibodies directed to proteins on the different parts of the neuron. VGKC encephalitis occurs due to autoantibodies directed to components of Voltage gated potassium channel (VGKC) protein complex. We present a case of VGKC mediated autoimmune encephalitis in a young female.

Case Presentation:

43-v.o. female presented to the ED with severe headache, photosensitivity, left foot drop. Examination showed no signs of meningeal irritation. Cushing's triad with hypertension, bradycardia and variability in respiratory rate. CT head showed cerebral edema, MRI revealed meningitis with possible cortical infarct. Underwent lumbar puncture, CSF analysis unremarkable. Started on ceftriaxone, vancomycin, acyclovir and dexamethasone empirically prior to the results of CSF analyses. Autoimmune encephalitis was suspected, antibiotics and antivirals discontinued and started on steroids. Her headache and overall clinical condition had improved significantly with steroids. Autoimmune workup positive for Anti- VKGC antibodies, negative for anti NMDA, FISH negative for Miller- Dieker Syndrome antibodies. Patient was continued on steroids with recommendations to follow up with neurology. During follow up, her symptoms improved with no recurrence.

Discussion

Autoimmune encephalitis is a clinical condition in which brain cells are attacked by due to autoimmune antibodies. VGKC autoimmune encephalitis is relatively new clinical entity. In this type, antibodies are present against the Voltage gated potassium channels (VGKC) protein complex clinical presentation varies from cognitive impairment and psychiatric symptoms, chills, seizures to ataxia and cerebellar symptoms. VGKC maintain neuronal activity by maintaining the resting membrane potential and action potentials. Antibodies to these channels effect the limbic system, therefore resulting in symptoms related to behavior and memory. Mainstay of treatment include steroids, plasma exchange, IVIG depending upon the severity. Response to treatment is measured by serial VGKC levels along with clinical, neuropsychological tests, imaging such as MRI or EEG during follow-up.

References

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