

E-Pearl of the Week

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Voltage-gated potassium channel autoimmunity

Clinical manifestations of voltage-gated potassium channel (VGKC) autoimmunity can include subacute cognitive impairment, myoclonus, ataxia, parkinsonism, and diffuse slowing on electroencephalography, among many others. These findings can result in a misdiagnosis of Creutzfeldt-Jakob disease (CJD). Therefore, it is important to consider testing for the presence of serum VGKC antibodies in the initial evaluation of suspected CJD. This distinction is essential because neurologic disease due to VGKC autoimmunity typically has an excellent response to immunotherapy.

Reference

Tan KM, Lennon VA, Klein CJ, Boeve BF, Pittock SJ. Clinical spectrum of voltage-gated potassium channel autoimmunity. *Neurology* 2008;70:1883-1890.