A 58-year-old man developed subacute gait instability, oscillopsia, and vertigo. Examination showed gait ataxia and periodic alternating nystagmus (PAN) (video). MRI revealed T2 hyperintensities in the left cerebellum and nodulus (figure). CSF was inflammatory. Autoantibody evaluation was positive for Kelch-like protein 11 (KLHL11) in serum and CSF by tissue-based immunofluorescence assay and cell-based assay, prompting identification of a retroperitoneal mass by CT imaging consistent with seminoma.1 PAN resolved after plasma exchange, corticosteroids, adjuvant chemotherapy, and symptomatic baclofen; downbeat
nystagmus persisted. KLHL11 immunoglobulin G is associated with testicular germ cell tumor paraneoplastic rhomboencephalitis. Early identification is crucial as immunotherapy and cancer treatment can prevent neurologic dysfunction.

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References
Teaching Video NeuroImages: Periodic Alternating Nystagmus in Paraneoplastic KLHL11 Rhomboencephalitis

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