Teaching Video NeurolImages: Periodic Alternating Nystagmus in Paraneoplastic KLHL11 Rhomboencephalitis

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Figure Brain MRI in KLHL11 Paraneoplastic Rhomboencephalitis

(A) Axial T2 fluid-attenuated inversion recovery MRI demonstrates T2 hyperintensity in the left cerebellar hemisphere predominantly in the dentate nuclei as well as subtle T2 hyperintensity in the cerebellar nodulus (arrow). Seven months after initiation of cancer therapy and immunotherapy, MRI revealed improved T2 signal abnormality and mild atrophy (B).

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. Auto-antibody 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

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nystagmus persisted.² 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.²

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