Teaching Video NeuroImages: Periodic Alternating Nystagmus in Paraneoplastic KLHL11 Rhomboencephalitis

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Divyanshu Dubey - Dr. Divyanshu Dubey has a patent pending for Kelch-like protein 11 as a marker of neurological autoimmunity and has received research support from Grifols, Center of Multiple Sclerosis and Autoimmune Neurology, Center for Clinical and Translational Science. Dr. Dubey has consulted for UCB and Astellas. He has participated in UCB Advisory Board Meeting in Lyon, France on September 23, 2019. All compensation for consulting activities is paid directly to Mayo Clinic. Dr. Dubey has been a speaker at American Academy of Neurology Annual conference. All compensation for speaking activities is paid directly to Mayo Clinic. Dr. Dubey is on the editorial board of Journal of Clinical Medicine.

Hyun Woo Kim reports no disclosures

Alfonso Sebastian Lopez Chiriboga reports no disclosures
A 58-year-old man developed subacute gait instability, oscillopsia and vertigo. Examination showed gait ataxia and periodic alternating nystagmus (PAN) (video, http://links.lww.com/WNL/B338). 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.² KLHL11-IgG is associated with testicular-germ-cell-tumor paraneoplastic rhomboencephalitis. Early identification is crucial as immunotherapy and cancer treatment can prevent neurologic dysfunction.²
## Appendix 1: Authors

<table>
<thead>
<tr>
<th>Name</th>
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References:


Video: Periodic Alternating Nystagmus

PAN demonstrated by left-beating nystagmus in primary gaze which reverses direction after a transition phase with downbeat nystagmus. This reversal occurred approximately every 90 to 120 seconds. PAN localizes to the cerebellar nodulus and often responds to baclofen.

Video – [http://links.lww.com/WNL/B338](http://links.lww.com/WNL/B338)
**Figure:** Brain MRI in KLHL11 Paraneoplastic Rhomboencephalitis

(A) Axial T2 FLAIR 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).
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