A 36-year-old man had progressive gait ataxia, diplopia, and hearing loss over 3 years. On examination, he had upper extremity ataxia, truncal ataxia, bradykinesia, and lower extremity spasticity. Brain magnetic resonance imaging revealed a “hot cross bun” sign (Figure). Kelch-like protein 11 (KLHL11) IgG was detected in serum and cerebrospinal fluid.1 Positron emission tomography revealed a hypermetabolic mediastinal mass. Subsequent biopsy revealed seminoma. The hot cross bun sign has been described in patients with multiple system atrophy as well as in a patient with KLHL11 encephalitis.2,3 Some KLHL11 rhombencephalitis cases have been misdiagnosed as multiple system atrophy.1 This case highlights the importance of considering KLHL11 IgG evaluation in
men with progressive brainstem/cerebellar syndrome despite the radiological findings.1

Potential Competing Interests: Dr Dubey has consulted for UCB, Immunovant, and Astellas Pharmaceuticals. All compensation for consulting activities is paid directly to Mayo Clinic. He has patents pending for KLHL11 and LUZP4 as markers of neurological autoimmunity. Dr Pittcock has received grants from Alexion, National Institutes of Health, MedImmune/Viela Bio, Grifols, and Guthy Jackson; all compensation paid directly to Mayo Clinic. He has received consulting fees from Alexion, MedImmune/Viela Bio, and Euroimmun; all compensation paid directly to Mayo Clinic. He has also received consulting fees from Roche/Genentech, Sage Therapeutics, Prime Therapeutics, UCB, and Astellas; all compensation paid directly to Mayo Clinic and as personal compensation. He owns Patent #8,889,102 and Patent #9,891,219B2. He has patents pending for GFAP-IgG, Septin-5-IgG, MAP1B-IgG, Kelch-like protein 11, and PDE10A. Dr Pittock has received personal compensation to attend the UCB Advisory Board Meeting in Stockholm, Sweden, on September 10, 2019. Dr Pittock received personal compensation/honorarium to attend the NMOSD Biomarker Advisory Board Meeting in Munich, Germany, on November 8, 2019. All travel expenses were paid by Hoffman/LaRoche AG. Honorarium was paid directly to the Mayo Clinic. He received no personal compensation. Dr Pittock received personal compensation to attend the virtual Genentech NMOSD Advisory Board Meeting on October 1, 2020. The other authors report no competing interests.

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