Teaching Video NeuroImage: Response to IVIg in a Patient With Steroid-Refractory Immune Checkpoint Inhibitor–Related Ocular Myopathy

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Case

A 76-year-old woman with metastatic melanoma presented with a 3-day history of progressive diplopia approximately 3 weeks after her first ipilimumab and nivolumab therapy. Neurologic examination demonstrated bilateral nonfatigable ptosis and severe bilateral ophthalmoplegia without evidence of bulbar or neck weakness. Creatine kinase (CK) was 5213 U/L (normal:26-192). AChR and MuSK antibodies were negative. Electrodiagnostic studies revealed early recruiting, small motor unit potentials with fibrillations in several muscles, and no decrement on repetitive nerve stimulation. Brain and orbit MRIs were normal. CK normalized in 1 week. Immune checkpoint inhibitors (ICIs) were discontinued. She was started on 200 mg of prednisone per day tapered slowly over 6 months. At 3-month follow-up, patient had minimal improvement (Video 1). Monthly IVIg was initiated, and ophthalmoplegia completely resolved (Video 1). ICI myopathy (ICIM) is a drug-induced immune-mediated myopathy that has a predilection for the extraocular muscles.1 ICIM usually has an excellent response to IVIg,1 which should be considered in all steroid-refractory cases.

Author Contributions

S.J. Hooshmand: drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data. C. Aragon Pinto: drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data. T. Liewluck: drafting/revision of the article for content, including medical writing for content; study concept or design. M.V. Pinto: drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data; and study concept or design.

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Reference

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