A 53-year-old woman was presented for the evaluation of visual disturbances, generalized and multifocal myoclonus, and progressive ataxia that began at age 30 years (cf. Video 1). Bilateral cherry-red spots in the macula (figure) and a cortical origin in the EEG-EMG coregistration with back-averaging were observed. Reduced neuraminidase activity in fibroblasts and the homozygous mutation c.403G>A in the NEU1 gene confirmed the diagnosis of sialidosis type I.

Sialidosis or cherry-red spot–myoclonus syndrome is classified into normomorphic or type I, beginning usually after 20 years old, whereas dysmorphic or type II begins at birth or in early childhood. In both, generalized myoclonus and ataxia can be found. EEG-EMG coregistration may show cortical potential followed by the myoclonus, proving a cortical origin. Differential diagnosis is necessary for other inherited metabolic disorders such as Tay-Sachs disease or Unverricht-Lundborg disease. Cherry-red spots on the retina, cortical myoclonus, and progressive ataxia are essential keys to suspicion.1,2

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**Disclosure**

B. Vélez Gómez has received honoraria from Sanofi; S. Jesús has received honoraria from AbbVie, Bial, Merz, UCB, Italfarmaco, and Zambon; D. Macías-García has received honoraria from AbbVie, Teva, and Zambon; B. Lechner reports no disclosures relevant to the manuscript; and P. Mir has served in the advisory boards or received honorarium for lecturing from Abbott, Allergan, AbbVie, Bial, Britannia, Italfarmaco, Merz, UCB, Teva, and Zambon. Go to Neurology.org/N for full disclosures.

**Appendix Authors**

<table>
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**References**


Teaching Video NeuroImage: Clues in Myoclonus Evaluation: When to Consider Sialidosis
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