Teaching Video NeuroImage: Generalized Reflex Myoclonus in Autoimmune Hepatitis-Primary Biliary Cholangitis Overlap Syndrome

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Neurology® 2022;99:82-83. doi:10.1212/WNL.0000000000200733

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Table Hepatic and Autoimmune Workup That Led to the Diagnosis of Autoimmune Hepatitis-Primary Biliary Cholangitis

Laboratory test	Value	Normal range	Laboratory test	Value	Normal range
AST	685.60 IU/L	(13.00–35.00) IU/L	INR	1.11	—
ALT	510.40 IU/L	(7.00–40.00) IU/L	ANA	+	Negative
ALP	729.9 IU/L	(35.0–100.0) IU/L	AMA-M2	+	Negative
GGT	867.0 IU/L	(7.0–45.0) IU/L	AMA-3E	+	Negative
Serum IgG	23.6 g/L	(8.60–17.40) g/L	Ro-52	+	Negative
Ammonia	159.80 μmol/L	(18.00–72.00) μmol/L	SMA	<1:100	<1:100

Abbreviations: ALP = alkaline phosphatase; ALT = alanine aminotransferase; AMA-3E = antimitochondrial 3E antibody; AMA-M2 = antimitochondrial M2 antibody; ANA = antinuclear antibodies; AST = aspartate aminotransferase; GGT = gamma-glutamyltransferase; INR = international normalized ratio; SMA = anti–smooth muscle antibodies.

A 25-year-old woman presented with progressive apathy and disorientation, followed by acute-onset confusion that progressed to stupor. Physical examination revealed generalized reflex myoclonus to both tactile (Video 1, part 1) and visual stimuli (part 2). Hepatic and autoimmune workup was positive for transaminitis, hyperammonemia, and antimitochondrial and anti-smooth muscle antibodies (Table). The rest of her laboratory test results including chemistries were within normal limits. MRI of the brain was likewise unremarkable. EEG showed generalized slowing. She was diagnosed with autoimmune hepatitis-primary biliary cholangitis overlap syndrome[1] with hepatic encephalopathy. She was treated with steroids with full resolution of her myoclonus (Video 1, part 3). Hepatic encephalopathy is usually associated with negative myoclonus (asterixis) rather than reflex myoclonus. Little is known about the mechanism of reflex myoclonus, although small studies suggest cortical and subcortical subtypes reflect the origin of electrical signals leading to the myoclonic jerks.[2]

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Video

Teaching slides
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Acknowledgment

The authors appreciate the help of Dr. Ninad Desai from St. Vincent's Medical Center, Bridgeport, CT, USA, for interpretation of EEG during the photomyoclonic response recording.

Study Funding

The authors report no targeted funding.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Publication History

Received by *Neurology* December 20, 2021. Accepted in final form March 29, 2022. Submitted and externally peer reviewed. The handling editor was Whitley Aamodt, MD, MPH.

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Appendix *(continued)*

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DOI 10.1212/WNL.0000000000200733