

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

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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<sup>1</sup> 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.<sup>2</sup>

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<b>Qian Wu, MD, PhD</b>	Department of Neurology First Affiliated Hospital, Kunming Medical University, Kunming, P.R. China	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data

## Appendix (continued)

Name	Location	Contribution
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## References

1. Chazouillères O, Wendum D, Serfaty L, Montebault S, Rosmorduc O, Poupon R. Primary biliary cirrhosis-autoimmune hepatitis overlap syndrome: clinical features and response to therapy. *Hepatology*. 1998;28(2):296-301.
2. Hallett M, Chadwick D, Marsden CD. Cortical reflex myoclonus. *Neurology*. 1979; 29(8):1107-1125.



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