Tremor and Other Hyperkinetic Movements

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In Response To:

Vynogradova I, Savitski V, Heckmann JG. Hemichorea associated with CASPR2 antibody. Tremor Other Hyperkinet Mov. 2014; 4. doi: 10.7916/D8VM49C5

Original Article:

Ramdhani RA, Frucht SJ. Isolated Chorea Associated with LGI1 Antibody. Tremor Other Hyperkinet Mov. 2014; 4. doi: 10.7916/D8MG7MFC

Letter to the Editor

Reply to: Hemichorea Associated with CASPR2 Antibody

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To the editor:

The case presented by Vynogradova and colleagues adds to the ever-expanding etiological spectrum of autoimmune chorea. This is the first reported case of isolated paraneoplastic chorea associated with anti-CASPR2, paralleling our report of isolated idiopathic chorea associated with anti-LGI1. Both cases share an asymmetric predominance of the chorea as well as the absence of limbic encephalitis. LGI1 and CASPR2 antibodies are specific to the VGKC-protein complex and each are associated with a number of distinct clinical phenotypes: Faciobrachial dystonic seizures (FBDS), hyponatremia and limbic encephalitis (LE) in the former; and Morvan's syndrome[1], peripheral nerve excitability, and Isaac's syndrome[2] in the latter. Despite their phenotypic differences, these two cases highlight that phenotypically-similar late-onset isolated chorea can be a result of distinct pathophysiological mechanisms which will result in a different clinical course. This is illustrated by the fact that, five months after having a complete remission following pulse steroid therapy, our patient's chorea relapsed. A second course of steroids did not engender as robust a response as the initial treatment. Seizure, metabolic disturbances, and LE remain absent and a steroid-sparing therapy (i.e., rituximab, IVIG) will be considered next. These cases emphasize the importance of antibody screening in late-onset chorea due to the diagnostic and treatment implications.

References

1. Irani SR, Pettingill P, Kleopa KA, et al. Morvan syndrome: clinical and serological observations in 29 cases. *Ann Neurol* 2012;72:241-255.

2. Panzer J, Dalmau J. Movement disorders in paraneoplastic and autoimmune disease. *Curr Opin Neurol* 2011;24:346-353.