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 syndrome1, peripheral nerve excitability, and Isaac’s syndrome2 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