

In Response To:

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

Letter to the Editor

Hemichorea Associated with CASPR2 AntibodyIryna Vynogradova¹, Vitali Savitski¹, Josef G. Heckmann^{1*}¹ Department of Neurology, Municipal Hospital Landshut, Landshut, Germany**Keywords:** Choreatic syndrome, chorea, hemichorea, paraneoplastic syndrome, CASPR2 antibody**Citation:** Vynogradova I, Savitski V, Heckmann JG. Hemichorea associated with CASPR2 antibody. Tremor Other Hyperkinet Mov 2014; 4. doi: 10.7916/D8VM49C5

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To the editor:

We read with great interest the article “Isolated Chorea Associated with LGI1 Antibody” (LGI1, leucine-rich glioma inactivated 1) by Ramdhani and Frucht.¹ The authors presented an important clinical observation about a treatable etiology of chorea in a 53-year-old male, in whom an underlying tumor could not be detected. By presenting a case vignette of hemichorea associated with antibodies against contactin-associated protein 2 (CASPR2), we wish to emphasize the relationship between a movement disorder and an autoantibody. In our patient, lung cancer was detected.

A 75-year-old female developed choreiform movement disorder, predominantly affecting the left limbs and the left side of the face. She took no neuroleptic drugs and reported that she had been smoking for about 50 years. Her family history was unremarkable. Cranial magnetic resonance images and serum chemistry, including liver enzymes, complete blood count, thyroid function, and cerebrospinal fluid analysis were negative. Her creatine kinase level was slightly elevated, due to the movement disorder (325 U/L, normal <140). Systemic lupus erythematosus and neuroacanthocytosis were not clinically suspected, and tests for these conditions were not performed. Electroencephalography (EEG) revealed normal background activity without signs of epileptic dis-

charges. As a paraneoplastic process was suspected,² computed tomography-positron emission tomography (CT-PET) was performed and revealed a right-side malignant lung tumor. Based on the patient’s history and radiological presentation, it was perceived as smoking-associated lung cancer. Serum analysis for paraneoplastic autoantibodies detected CASPR2 antibodies by indirect immunofluorescence test (titer IgM 1:10; Euroimmun, Lübeck, Germany). Tests for the remainder of the autoantibodies (anti-Hu, -Ri, -Yo, -CV2, -Ma, -Ta, -PCA2, -ANNA, -NMDA, -AMPA-1, and -AMPA-2) were negative. Symptomatic treatment with tiapride (200 mg daily) was introduced and resulted in acceptable symptom control within the following 2 days. Although oncologic therapy was proposed, it was refused by the patient.

Through this case vignette, we wish to complement the observations of Ramdhani and Frucht and Tofaris et al.³ for two reasons. First, autoimmune processes (both paraneoplastic and idiopathic) are well-recognized causes of chorea but are relatively rare.⁴ In most cases, co-existing neurological findings, such as peripheral neuropathy, cognitive decline, epilepsy, or oculomotor disturbances, are present.⁵ In the Ramdhani and Frucht case, and even in our case, isolated chorea and hemichorea, respectively, were the index symptom. Whereas an idiopathic etiology was strongly assumed in the case of Ramdhani and Frucht,¹ in our case, an obvious paraneoplastic etiology has to be considered. The

pronounced CT-PET-findings and the history of excessive smoking led us to suspect a smoking-associated lung cancer. However, a histological tissue diagnosis was refused by the patient, as was subsequent oncologic therapy. Second, the isolated pattern of CASPR2 autoantibody positivity is interesting. Only one case of autoimmune chorea with a positive CASPR2 antibody has been reported, and it was an idiopathic case. This is according to a study on autoimmune chorea in adults,⁴ in which the case of Ramdhani and Frucht can also be grouped. In the paraneoplastic group of autoimmune chorea, CASPR2 has not yet been reported.⁴ On the other hand, CASPR2 is more often associated with neuromyotonia and Morvan syndrome and occurs more often in thymoma,⁶ though these clinical findings were not present in our patient. However, it is informative that LGI1 (the antibody detected in the patients of Ramdhani and Frucht and of Toranis et al.³) and CASPR2 are taken together as voltage-gated potassium channel (VGKC) complex autoantibodies and thus share substantial similarity.^{7,8} To conclude, autoimmune processes must be considered in the differential diagnosis of adult-onset chorea. Besides laboratory and cerebrospinal fluid analysis,^{9,10} a serum panel of autoantibodies, including LGI1 and CASPR2, may contribute to the diagnosis.

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