Teaching NeuroImages:

Basal ganglia involvement in facio-brachial dystonic seizures associated with LGI1 antibodies

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Figure 1 Brain MRI

Axial (A) and coronal (B) postcontrast T1-weighted images show a lesion involving the left caudate and globus pallidus.

Figure 2 Brain MRI

Axial (A) and coronal (B) postcontrast T1-weighted images with reduction of contrast enhancement of the lesion involving the left caudate and globus pallidus.

A 30-year-old man developed right faciobrachial dystonic seizures (FBDS).¹ Ictal and interictal EEGs were normal. CSF analysis was unremarkable. Brain MRI revealed a gadolinium-enhancing lesion involving the left caudate and globus pallidus (figure 1). Leucine-rich glioma inactivated protein 1 (LGI1) antibodies were detected in the serum. Total-body CT scan revealed no malignancies. The patient underwent 5 cycles of plasmapheresis followed by long-term steroid therapy with complete benefit. A brain MRI performed after 5 months showed reduction of contrast enhancement (figure 2). LGI1, a secreted protein complexed with voltage-gated potassium

channels, is highly expressed in the neocortex and hippocampus.² *LGI1* mutations have been described in patients with autosomal dominant partial epilepsy with auditory features (ADPEAF). Our patient had no clinical features of ADPEAF. Whether FBDS can be classified as epilepsy or dystonia is a matter of debate.³ The involvement of basal ganglia described in our patient can be relevant to the ongoing debate.

AUTHOR CONTRIBUTIONS

Dr. D. Plantone, Dr. Renna and Dr. Iorio: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data, study supervision. Dr. Grossi and Dr. F. Plantone:

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DISCLOSURE

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A COMPARISON OF TAU AND 14-3-3 PROTEIN IN THE DIAGNOSIS OF CREUTZFELDT-JAKOB DISEASE

Andre Karch, Inga Zerr, Göttingen, Germany:

Hamlin et al.¹ compared validities of 2 commonly used CSF tests for sporadic Creutzfeldt-Jakob disease (sCJD)—14-3-3 and total tau—and presented results contrary to previous studies.^{1–4} The present study is restricted to autopsy-confirmed cases, which seems reasonable, since neuropathology is the diagnostic gold standard in sCJD. However, using only autopsy-confirmed cases can also cause severe selection bias in a prospective setting, given that these tests are frequently requested in potentially treatable conditions.^{2,3} In the present study, clinicians were given results of 14-3-3 but not tau tests during the patient's lifetime. Autopsies were obtained in fewer than 10% of all initially referred patients. Thereby, decision about initiation of

postmortem autopsy was directly dependent on 14-3-3 but not tau results. It seems a fair assumption that patients with CJD and negative 14-3-3 as well as patients without CJD and positive 14-3-3 were more likely to be autopsied than patients with consistent clinical history and 14-3-3 test results. Thereby, specificity and sensitivity of 14-3-3 are underestimated in this study design, which could explain differences in 14-3-3 validity when compared to previous studies. A final decision on test accuracy of 14-3-3 and total tau needs careful assessment in a prospective, multicenter study using standardized protocols for laboratory, clinical, and pathologic data.⁵

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CORRECTIONS

Teaching *NeuroImages*: Basal ganglia involvement in facio-brachial dystonic seizures associated with LGI1 antibodies

In the article "Teaching *NeuroImages*: Basal ganglia involvement in facio-brachial dystonic seizures associated with LGI1 antibodies" by D. Plantone et al. (*Neurology*® 2013;80:e183–e184), figures 1 and 2 were transposed and should be reversed. The publisher regrets the error.

Severe congenital *RYR1*-associated myopathy: The expanding clinicopathologic and genetic spectrum

In the article "Severe congenital *RYR1*-associated myopathy: The expanding clinicopathologic and genetic spectrum" by D. Bharucha-Goebel et al. (*Neurology*® 2013;80:1584–1589), there is an error in the author list. The fourth author should have read Kristen Zukosky. The editorial staff regrets the error.

Author disclosures are available upon request (journal@neurology.org).