

A. Sebastian López
Chiriboga, MD
Jason L. Siegel, MD
William O. Tatum, DO
Jerry J. Shih, MD
Eoin P. Flanagan,
MBBCh

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STRIKING BASAL GANGLIA IMAGING ABNORMALITIES IN LGI1 AB FACIOBRACHIAL DYSTONIC SEIZURES

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Autoimmune encephalitis associated with antibodies targeting the neuronal surface antigen leucine-rich, glioma-inactivated 1 (LGI1) protein is accompanied by faciobrachial dystonic seizures (FBDS) in up to half of patients. These hallmark episodes can be mistaken for psychiatric disorders, particularly as the ictal episodes are usually not detectable with surface EEG electrodes. Accompanying imaging abnormalities help exclude psychiatric etiologies, localize the seizures, and improve our understanding of the pathogenesis. Herein, we present striking imaging abnormalities in a patient with FBDS from LGI1-antibody encephalitis.

Case description. A 51-year-old man experienced a generalized tonic-clonic seizure in April 2015. He subsequently developed cognitive changes, visual hallucinations, and severe anxiety and was diagnosed with alcohol withdrawal syndrome at an outside institution; he did not receive immunotherapy. Approximately 12 weeks later, he developed progressive cognitive decline and abnormal movements characterized by posturing-grimacing of the hemiface and the ipsilateral arm, occurring up to 100 times per day (video at Neurology.org/nn). No abnormal discharges were identified during ictal episodes on surface EEG.

Neurologic examination revealed a Mini-Mental State Examination (MMSE) score of 24/30 (points were missed on orientation, calculation, and recall). He experienced multiple, frequent FBDS during the neurologic examination, but it was otherwise nonfocal.

Laboratory abnormalities included serum sodium of 129 mmol/L (normal 135–145 mmol/L) and positive LGI1 antibodies identified via cell-based assay. CSF analysis was normal, and LGI1 antibodies were not detected.

MRI of the brain showed T1 hyperintensities in the basal ganglia, right greater than the left, correlating with the prominent FBDS affecting the left arm more frequently. There were also restricted diffusion changes

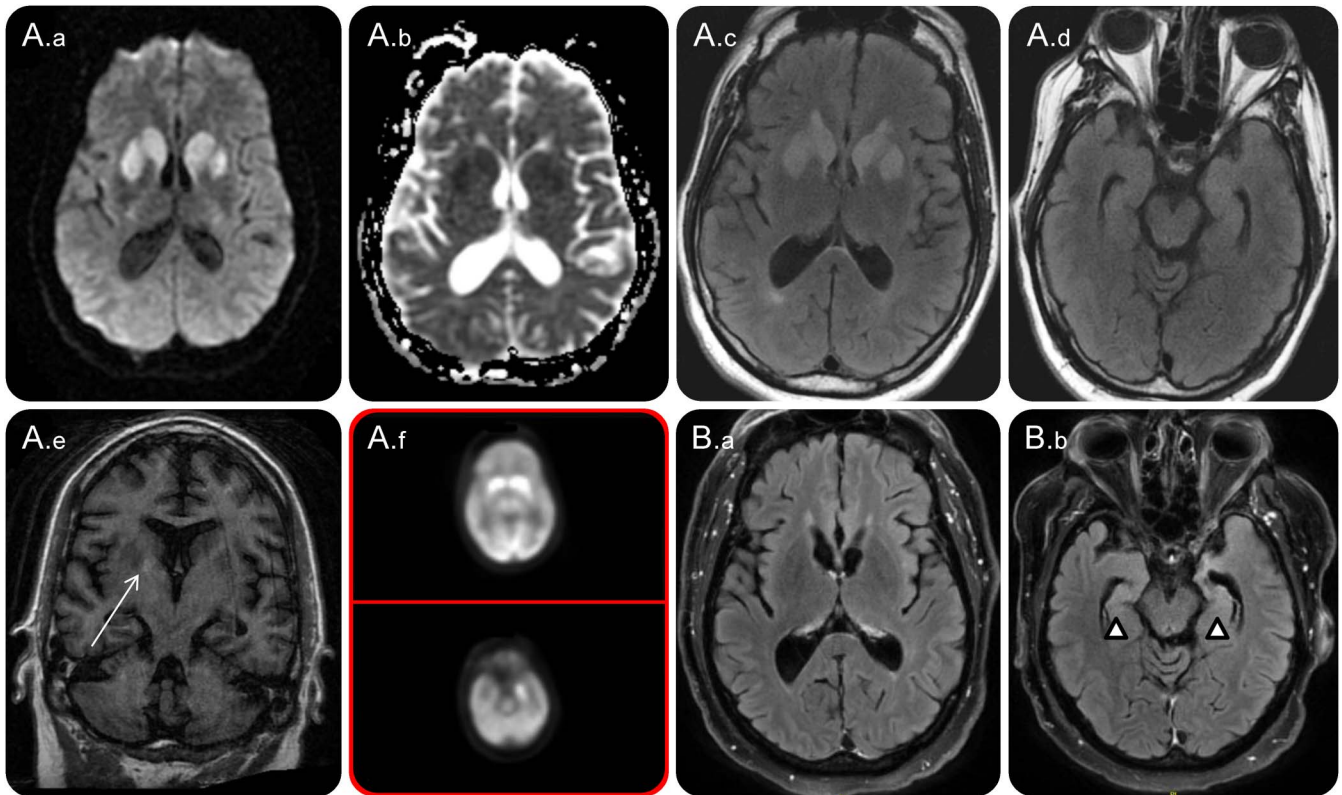
in the bilateral basal ganglia. 18F-fluorodeoxyglucose (18F-FDG) PET demonstrated foci of hypermetabolism in the bilateral basal ganglia and asymmetric hypermetabolism in the left temporal lobe (figure). Follow-up MRI, 16 months after the onset of symptoms, revealed prominent atrophy in the previously described restricted diffusion areas, particularly the caudate nuclei and mesial temporal lobes (figure). Extensive oncologic evaluation was negative.

The FBDS did not improve with antiseizure drugs, including the combination of levetiracetam and lacosamide. Significant improvement of his cognitive dysfunction (confirmed with formal posttreatment neuropsychometric testing and a bedside MMSE score of 29/30) and significant reduction in the frequency (1–2 per week) of FBDS were noted after administration of IV methylprednisolone 1 g for 5 days. He was transitioned to oral steroids and experienced a clinical relapse characterized by increased frequency of FBDS (up to 10 episodes daily) when prednisone was decreased from 20 to 10 mg daily, requiring reinstitution of high-dose prednisone and initiation of mycophenolate mofetil and slow oral prednisone taper. The FBDS frequency decreased after 1 week and continued to improve over time; he has been seizure free for 7 months.

Discussion. This patient presented with classic features of LGI1-antibody encephalitis: subacute cognitive decline, hyponatremia, FBDS, and substantial improvement with immunotherapy.¹ The striking imaging abnormalities illustrated in this report provide further evidence for the basal ganglia as the possible origin of FBDS and suggest evidence that ischemic injury occurs.

Although the pathophysiology of the radiographic finding remains unknown, the prominent restricted diffusion with apparent diffusion coefficient correlate and T1 and T2 hyperintensities, in this case, strongly suggest an ischemic injury from vascular, metabolic, or immune-mediated neuronal damage or due to the high metabolic demand from frequent epileptic activity. The changes observed in 18F-FDG PET/CT^{2,3} characterized by hypermetabolic activity in the same areas corroborate this hypothesis. Because of vascular supply and high glucose and oxygen

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(A) Brain MRI obtained during relapse: (A.a) Prominent restricted diffusion and (A.b) apparent diffusion coefficient correlate with changes in the bilateral basal ganglia. (A.c) T2 fluid-attenuated inversion recovery (FLAIR) hyperintensities in the same “restricted diffusion” distribution. (A.d) FLAIR normal hippocampal size. (A.e) Coronal sequence showing T1 hyperintensities in the right basal ganglia. (A.f) 18F-fluorodeoxyglucose (18F-FDG) PET/CT. Intense 18F-FDG uptake is noted in the bilateral basal ganglia, asymmetric FDG uptake within the left medial temporal lobe. (B) Follow-up brain MRI: (B.a, B.b) 16 months after the onset of symptoms. Axial T2 FLAIR shows prominent caudate nuclei atrophy and bilateral hippocampal atrophy (arrowheads).

utilization, the deep gray nuclei are particularly vulnerable to ischemia or hypoglycemia.⁴

Our patient tested positive for LGI1 in the serum only, illustrating that, in this entity, some patients may have antibodies only detectable in the serum,⁵ whereas others may have antibodies only detectable in the CSF.⁶

This case adds LGI1-antibody encephalitis to the differential diagnosis of bilateral basal ganglia restricted diffusion along with hypoglycemia, hyperammonemia, carbon monoxide poisoning, exposure to cyanide and methanol, osmotic myelinolysis, and venous and arterial infarction.⁴ Given the restricted differential diagnosis of basal ganglia T1-hyperintensity, this recently reported MRI accompaniment of a number of LGI1-antibody associated FBDS is a particularly useful clue to this disorder.⁷

This case also illustrates the progression of MRI changes in LGI1 encephalitis and shows hippocampal and caudate atrophy as a consequence of neuronal loss.

Recognition of the radiologic characteristics of LGI1-antibody FBDS can help support the diagnosis, allow prompt serologic confirmation and early institution of immunotherapy, and thus prevent

severe memory impairment resulting from delays in treatment.^{5,6,8}

From the Department of Neurology (A.S.L.C., J.L.S., W.O.T.), Mayo Clinic, Jacksonville, FL; Department of Neurology (J.J.S.), University of California, San Diego; and Department of Neurology (E.P.F.), Mayo Clinic, Rochester, MN.

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Correspondence to Dr. López Chiriboga: lopez.alfonso@mayo.edu

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