

# Neurology<sup>®</sup>

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

Domenico Plantone, Rosaria Renna, Dario Grossi, et al.

*Neurology* 2013;80:e183-e184

DOI 10.1212/WNL.0b013e31828f17fa

**This information is current as of April 22, 2013**

The online version of this article, along with updated information and services, is located on the World Wide Web at:

<http://www.neurology.org/content/80/17/e183.full.html>

*Neurology*® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2013 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.



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

Domenico Plantone, Rosaria Renna, Dario Grossi, et al.

*Neurology* 2013;80:e183-e184

DOI 10.1212/WNL.0b013e31828f17fa

**This information is current as of April 22, 2013**

<b>Updated Information &amp; Services</b>	including high resolution figures, can be found at: <a href="http://www.neurology.org/content/80/17/e183.full.html">http://www.neurology.org/content/80/17/e183.full.html</a>
<b>References</b>	This article cites 3 articles, 0 of which you can access for free at: <a href="http://www.neurology.org/content/80/17/e183.full.html##ref-list-1">http://www.neurology.org/content/80/17/e183.full.html##ref-list-1</a>
<b>Subspecialty Collections</b>	This article, along with others on similar topics, appears in the following collection(s): <b>All Immunology</b> <a href="http://www.neurology.org/cgi/collection/all_immunology">http://www.neurology.org/cgi/collection/all_immunology</a> <b>Autoimmune diseases</b> <a href="http://www.neurology.org/cgi/collection/autoimmune_diseases">http://www.neurology.org/cgi/collection/autoimmune_diseases</a> <b>MRI</b> <a href="http://www.neurology.org/cgi/collection/mri">http://www.neurology.org/cgi/collection/mri</a>
<b>Permissions &amp; Licensing</b>	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: <a href="http://www.neurology.org/misc/about.xhtml#permissions">http://www.neurology.org/misc/about.xhtml#permissions</a>
<b>Reprints</b>	Information about ordering reprints can be found online: <a href="http://www.neurology.org/misc/addir.xhtml#reprintsus">http://www.neurology.org/misc/addir.xhtml#reprintsus</a>

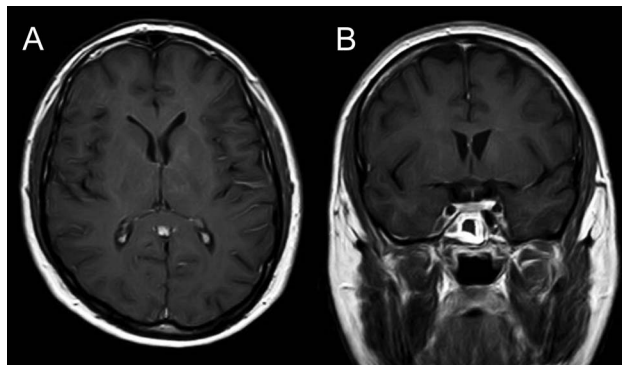


# Teaching NeuroImages: Basal ganglia involvement in facio-brachial dystonic seizures associated with LGI1 antibodies

Domenico Plantone, MD  
Rosaria Renna, MD  
Dario Grossi, MD  
Francesca Plantone, MD  
Raffaele Iorio, MD

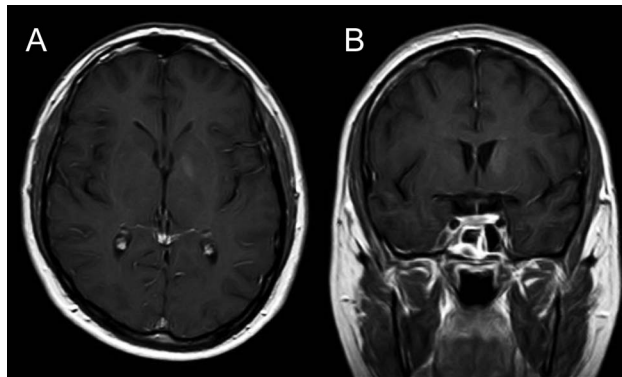
Correspondence to  
Dr. Plantone:  
domenicoplantone@hotmail.com

**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).<sup>1</sup> 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.<sup>2</sup> *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.<sup>3</sup> 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:

From the Department of Neurosciences (D.P., R.R., R.I.), Catholic University of Sacred Heart, Rome; Struttura Complessa di Neurologia (D.G.), Stabilimento Ospedaliero S. Giacomo, Monopoli; and Department of Surgical Sciences (F.P.), Parma University Medical School, Parma, Italy. Go to [Neurology.org](http://Neurology.org) for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

drafting/revising the manuscript, study concept or design, analysis or interpretation of data.

### STUDY FUNDING

No targeted funding reported.

### DISCLOSURE

The authors report no disclosures. Go to [Neurology.org](http://Neurology.org) for full disclosures.

### REFERENCES

1. Irani SR, Michell AW, Lang B, et al. Faciobrachial dystonic seizures precede Lgi1 antibody limbic encephalitis. *Ann Neurol* 2011;69:892–900.
2. Iorio R, Lennon VA. Neural antigen-specific autoimmune disorders. *Immunol Rev* 2012;248:104–121.
3. Striano P. Faciobrachial dystonic attacks: seizures or movement disorder? *Ann Neurol* 2011;70:179–180.