

Faciobrachial dystonic seizures secondary to basal ganglia involvement in anti-LGI1 encephalitis

Dhananjay Gupta¹, Anish Mehta¹, Rangaiah Pradeep¹,
Mahendra Javali¹, Purshottam T. Acharya¹,
Rangasetty Srinivasa¹

¹ Ramaiah Medical College, Department of Neurology, Bangalore, India

Received January 02, 2020; Accepted May 10, 2020

A 60-year-old man presented with a three-month history of memory disturbances and sudden, paroxysmal, involuntary movements of the face and right upper limb, associated with a tendency to fall. He had multiple such events during the day, each lasting for 1-2 seconds. Examination did not reveal any lateralizing neurological deficits. The sudden-onset, short-lasting, paroxysmal events were suggestive of faciobrachial dystonic seizures. A possibility of autoimmune encephalitis was considered and a serum autoimmune panel was positive for anti-LGI1 (leucine-rich-glioma inactivated) antibody at high titre. The routine EEG was normal and we could not capture any "epileptic phenomenon". Brain MRI showed T2/FLAIR hyperintensity in the right putamen, without diffusion restriction (figures 1 and 2). Initially, the patient was treated with levetiracetam and phenytoin, without much benefit. Later, intravenous immunoglobulins (IVIG) were added and the patient showed significant improvement within a few weeks. Follow-up MRI after three months showed resolution of previous changes. Faciobrachial dystonic seizures are pathognomonic of anti-LGI1 encephalitis and are considered an overlap between epileptic spasm and movement disorders. While some authors consider the frontal-temporal lobe as the source of epileptic phenomena, others

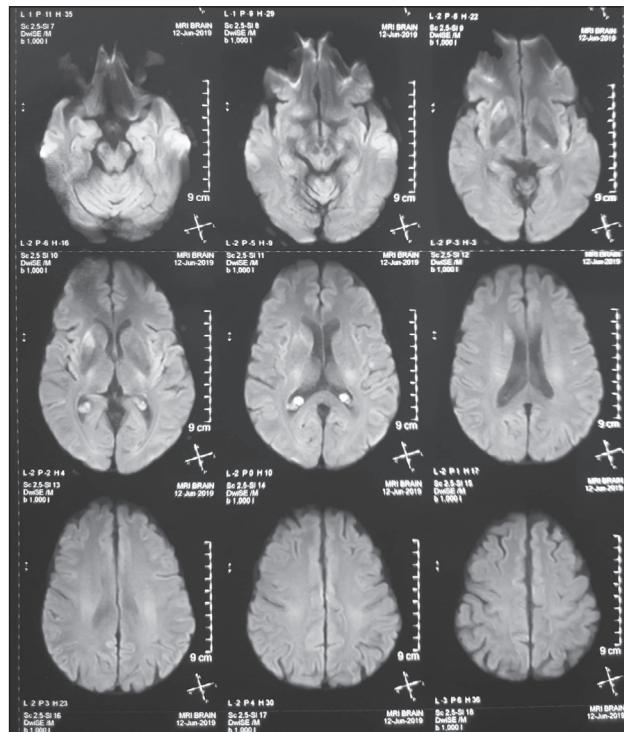


Figure 1. Brain MRI (T2/FLAIR, axial plane) showing hyperintensity in the region of the right basal ganglia/putamen.

have reported a peculiar cortical-subcortical interaction, generated at the basal ganglia (d'Orsi *et al.*, 2018; Iyer *et al.*, 2017). Our patient had neuroimaging changes in the right putamen (basal ganglia), which were reversible after treatment with IVIG and correlated with decreased seizure frequency. Hence, the origin of faciobrachial dystonic seizures is presumably localized to basal ganglia, specifically the putamen. □

Correspondence:

Dr. Rangaiah Pradeep
Department of Neurology,
Ramaiah Medical College and Hospitals,
Bengaluru, 560054 India
<dhananjay.gupta1990@yahoo.com>

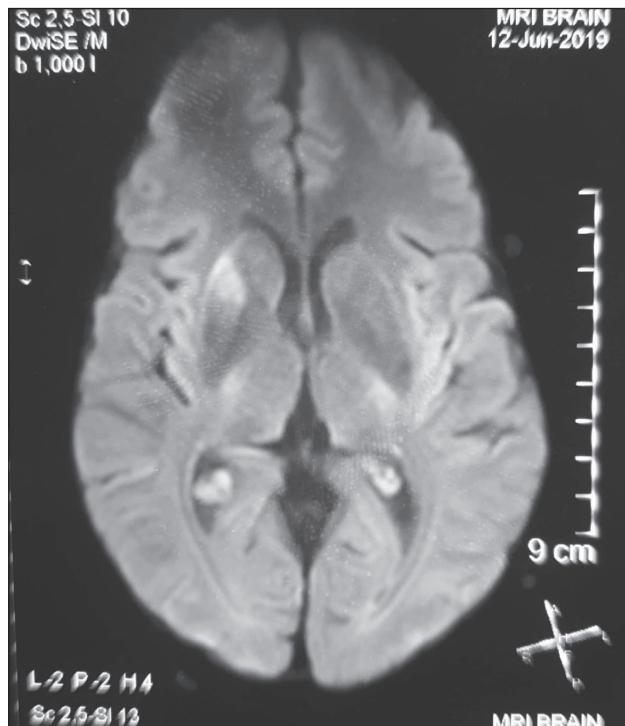


Figure 2. Enlarged view of right putaminal hyperintensity.

Supplementary data.

Summary didactic slides are available on the www.epilepticdisorders.com website.

Disclosures.

None of the authors have any conflict of interest to declare.

References

d'Orsi G, Martino T, Lalla A, Claudio MTD, Carapelle E, Avolio C. Faciobrachial dystonic seizures expressed as epileptic spasms, followed by focal seizures in anti-LGI1 encephalitis: a video-polygraphic study. *Epileptic Disord* 2018; 20(6): 525-9.

Iyer RS, Ramakrishnan TCR, Karunakaran TCR, Shinto A, Kamaleshwaran KK. Faciobrachial dystonic seizures result from fronto-temporo-basalganglial network involvement. *Epilepsy Behav Case Rep* 2017; 8: 47-50.

TEST YOURSELF



- (1) What are faciobrachial dystonic seizures (FBDS)?
- (2) What is the pathophysiology of FBDS?
- (3) What is the treatment for FBDS?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".