

Faciobrachial dystonic seizures expressed as epileptic spasms, followed by focal seizures in anti-LGI1 encephalitis: a video-polygraphic study

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ABSTRACT – The origin of faciobrachial dystonic seizures in anti-LGI1 encephalitis is controversial due to a lack of neurophysiological characterization. We report a 68-year-old man with subacute anterograde memory loss and involuntary faciobrachial movements. Video-polygraphic recordings disclosed repetitive events characterized by sudden, short contraction of the upper limbs and ipsilateral hemiface. A focal contralateral EEG slow wave from frontal or central electrodes was accompanied by increased muscle activity, often with a diamond-shaped configuration, on the orbicularis oris muscle, deltoid muscle, and extensor muscle of the hand. This EEG/EMG pattern (resembling a tonic epileptic spasm) was always followed by oral and gestural automatisms with dystonic posturing of the upper limbs, compatible with a temporal lobe seizure. Brain MRI showed hyperintensity in the bilateral mesial temporal lobes, while 18FDG-PET revealed basal ganglia hypermetabolism with extensive cortical hypometabolism. Serum and CSF were both positive for anti-LGI antibodies. The patient was treated with intravenous methylprednisolone (1 g/day for five days) with seizure freedom within four days after initiation of the immunotherapy. In this case, a video-EEG/polygraphic study disclosed that faciobrachial dystonic seizures may resemble epileptic spasms, and the occurrence in close temporal association with focal seizures as a single ictal event is suggestive of a peculiar cortical-subcortical interaction. [*Published with video sequence on www.epilepticdisorders.com*]

Key words: anti-LGI1 encephalitis, faciobrachial dystonic seizure, video-EEG/polygraphy, epileptic spasm, basal ganglia

Faciobrachial dystonic seizures (FBDS) are motor manifestations pathognomonic of LGI1 antibody encephalitis (Irani *et al.*, 2013). The origin of FBDS is a controversial issue, and FBDS is considered to be a movement disorder (Striano, 2011), an epileptic seizure (Irani *et al.*,



VIDEO ONLINE

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2013), and exhibit features that overlap with those of epileptic and movement disorders (Boesebeck et al., 2013).

Case study

We investigated a 68-year-old man with a two-month history of subacute anterograde memory loss and involuntary faciobrachial movements. Forty-eight hours of continuous video-EEG polygraphic recordings disclosed repetitive (~six per hour) events (see video) characterized by a sudden, short (~one-second) contraction of the upper limbs and ipsilateral hemiface. These episodes occurred indifferently on the right and the left side. A focal contralateral EEG wave (mean duration: 691 ± 260 ms; mean amplitude: 48.8 ± 15.2 μ V; $n=20$ events) was usually detected from frontal or central electrodes, and was accompanied by increased muscle activity, in a crescendo-decrescendo pattern, often with a diamond-shaped configuration, especially on the orbicularis oris muscle, deltoid muscle, and extensor muscle of the hand (mean duration: 967 ± 525 ms; mean amplitude: 38.2 ± 18.8 μ V; $n=20$ events). This EEG/EMG pattern (figure 1), resembling an asymmetric tonic epileptic spasm (Sotero de Menezes and Rho, 2002), was always followed by oral and gestural automatisms with dystonic posturing of the

upper limbs, compatible with a temporal lobe seizure. The ictal EEG was always obscured by muscle or movement artefacts, while a sustained increase in the muscle activity, especially in the extensor muscle of the hand (mean duration: 6325 ± 3054 ms; mean amplitude: 26.8 ± 11.5 μ V; $n=20$ events), concomitant with dystonic posturing, was associated with irregular breathing and tachycardia. For about half of the time, the epileptic spasm followed by a focal seizure occurred bilaterally and asynchronously (with variable side onset, on the left or the right side), separated by a short (~five-second) delay, during wakefulness and NREM sleep Stages 1 and 2. Finally, a prolonged (up to 10-minute) postictal phase emerged, characterized by ambulatory automatisms, fluent aphasia, visual hallucinations, and scared facial expressions (figure 2). EEG recording showed ipsilateral temporal rhythmic sharp waves evolving into regional slowing. We did not find any interictal epileptic activity. Serum sodium concentration was 127 mmol/l. Brain MRI showed increased FLAIR signal in the bilateral mesial temporal lobes, while 18FDG-PET revealed basal ganglia hypermetabolism with extensive cortical hypometabolism (figure 3). No abnormalities were seen in the basal ganglia and mesial temporal lobes on DWI or T1 scans. Serum and CSF contained LGI antibodies at 1:1,000 and 1:10 titres, respectively. The patient was treated with levetiracetam up to 2,000 mg daily for

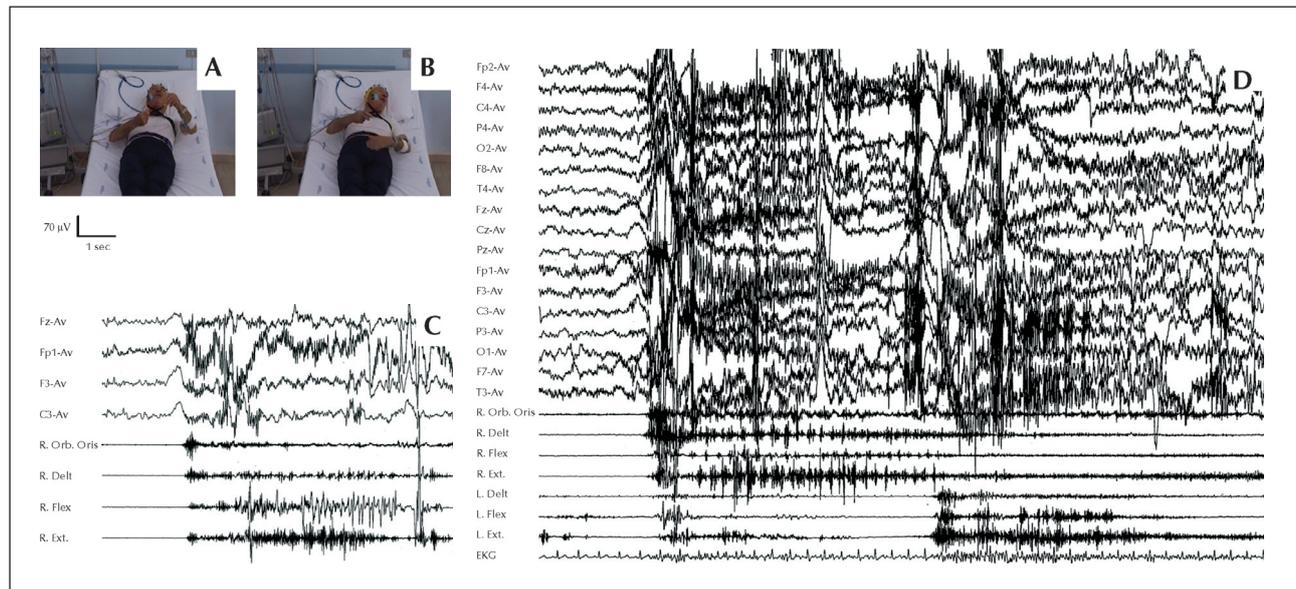


Figure 1. Video-EEG/polygraphic features of faciobrachial epileptic spasms. (A, B) Clinically, sudden and short (lasting for less than one second) contraction of the upper limbs and ipsilateral hemiface was observed. (C, D) Regarding the EEG/EMG, a focal contralateral EEG wave, detected from frontal and central electrodes, preceded increased muscle activity, in a crescendo-decrescendo pattern, with a diamond-shaped configuration, especially on the orbicularis oris muscle, deltoid muscle, and extensor muscle of the hand. Latency between wave onset and onset of orbicularis oris spasm, deltoid spasm, extensor spasm, and flexor spasm was 573 ms, 707 ms, 831 ms, 898 ms, respectively. The EEG/EMG pattern usually occurred bilaterally and asynchronously, with a short (~five-second) delay, and was followed by a focal seizure. EEG of the ictal focal seizure was obscured by muscle or movement artefacts, while a sustained increase in muscle activity, especially in the extensor muscle of the hand, concomitant with dystonic posturing, emerged.

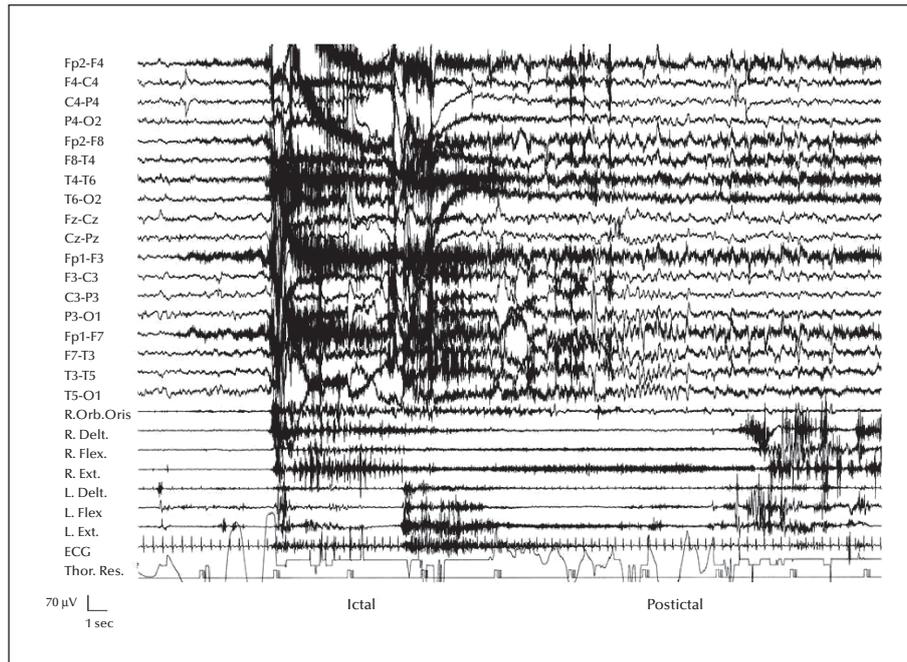


Figure 2. Two faciobrachial dystonic seizures occurred bilaterally and asynchronously (ictal phase). A prolonged postictal phase emerged, characterized by ambulatory automatism, fluent aphasia, visual hallucinations, and scared facial expressions. EEG recording showed ipsilateral temporal rhythmic sharp waves evolving into regional slowing.

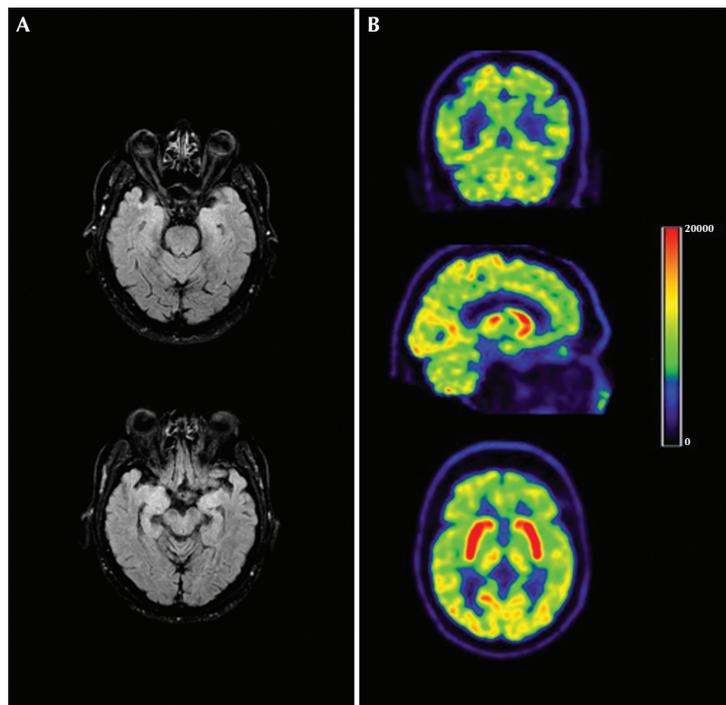


Figure 3. Brain MRI T2/FLAIR showing bilateral hyperintensity in the temporal lobes (A), and 18FDG-PET showing hypermetabolism in the basal ganglia and extensive cortical hypometabolism (B).

one week with no resolution of episodes, and followed by 1 g IV methylprednisolone for five days, with seizure freedom within four days after initiation of the immunotherapy. The patient was followed by a clinical psychologist at yearly intervals; at the last follow-up visit (two years after seizure freedom), the cognitive assessment revealed an amnesic mild cognitive impairment. Brain MRI performed one year after seizure freedom showed a marked reduction in the hyperintensity signal.

Discussion

Briefly, our patient developed anti-LGI1 encephalitis, with classic subacute anterograde memory loss and involuntary faciobrachial movements, clinically characterized by sudden and short contraction of the upper limbs and ipsilateral hemiface. These episodes were preceded by a frontal contralateral slow wave on the EEG recordings, while the EMG pattern of the affected muscles was characterized by a diamond-shaped configuration. Finally, these episodes were followed by oral and gestural automatisms with dystonic posturing of the upper limbs.

Two main observations emerge from our case:

– The difficulty in defining the origin of FBDS was related to the lack of neurophysiological characterization by means of a video-EEG/EMG study (Striano, 2011; Irani *et al.*, 2013). Navarro *et al.* (2016) reported an EEG focal slow wave preceding a contralateral tonic-dystonic seizure, similar to an epileptic spasm, based on simultaneous EEG/EMG recordings. Wennberg *et al.* (2018) demonstrated that the motor spasms are preceded by frontal infraslow activity, based on continuous video-EEG recordings. In our case, a video-EEG/polygraphic study disclosed that FBDS may be expressed as adult-onset epileptic spasms (Sotero de Menezes and Rho, 2002; d'Orsi *et al.*, 2007), as already suggested by others (Irani *et al.*, 2013; Navarro *et al.*, 2016). In contrast to infantile-onset spasms, adult-onset epileptic spasms are less frequent, the intensity of EMG activity is usually milder, and the duration is shorter (one second), while the EEG pattern (slow wave) is not easy to recognize and may be somewhat inconsistent (Bisulli *et al.*, 2002). The occurrence of epileptic spasms and focal seizures in close temporal association as a single ictal event is a rare phenomenon (Pachatz *et al.*, 2003), and a descending electrical volley from cortex to brainstem has been hypothesized. In our case, the sequence of focal seizure preceded by a single faciobrachial violent spasm, which suggests a complex cortical-subcortical interaction, emerged. Moreover, the affected side of FBDS was contralateral to the ictal epileptiform abnormalities, supporting the hypothesis that the cortex could have a critical role

in facilitation or induction regarding the generation of FBDS. Finally, as in previous reports (Irani *et al.*, 2013; Navarro *et al.*, 2016), 18FDG-PET showed prevalent hypermetabolism in the basal ganglia. Therefore, we support the hypothesis that FBDS derive from a peculiar cortical-subcortical interaction, generated at a subcortical level (basal ganglia) and triggered by focal cortical discharges.

– In the International League Against Epilepsy operational classification of seizure types (Fisher *et al.*, 2017), FBDS are not recognized as a seizure type, while epileptic spasms are included. Nevertheless, the recognition of the distinctive faciobrachial motor semiology is particularly relevant for the diagnosis and rapid immunotherapy for seizure control and prevention of cognitive impairment (Irani *et al.*, 2013). Furthermore, recent *in vitro* studies suggest that LGI1 antibodies are epileptogenic (Seagar *et al.*, 2017). Therefore, clinically and neurophysiologically, in our case, we believe that it is more appropriate to use the term “faciobrachial epileptic spasms” (FBES) instead of FBDS. It is possible that FBDS previously described in other reports may also be FBES, however, the lack of a full video-polygraphic study precluded a clear diagnosis. □

Supplementary data.

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

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

Legend for video sequence

Faciobrachial epileptic spasms in anti-LGI1 encephalitis; during wakefulness and sleep.

Key words for video research on www.epilepticdisorders.com

Phenomenology: epileptic spasms
Localisation: basal ganglia, temporal lobes
Syndrome: encephalitis
Aetiology: autoimmune

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TEST YOURSELF



- (1) Faciobrachial dystonic seizures are a typical manifestation of anti-LGI1 encephalitis. What is their origin?
- (2) In anti-LGI1 encephalitis, the typical faciobrachial dystonic seizure may be followed by seizure originating in which lobe?
- (3) What are the characteristic brain MRI findings in anti-LGI1 encephalitis?

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".