

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

Giuseppe d'Orsi, Tommaso Martino, Alessandra Lalla,
Maria Teresa Di Claudio, Elena Carapelle, Carlo Avolio

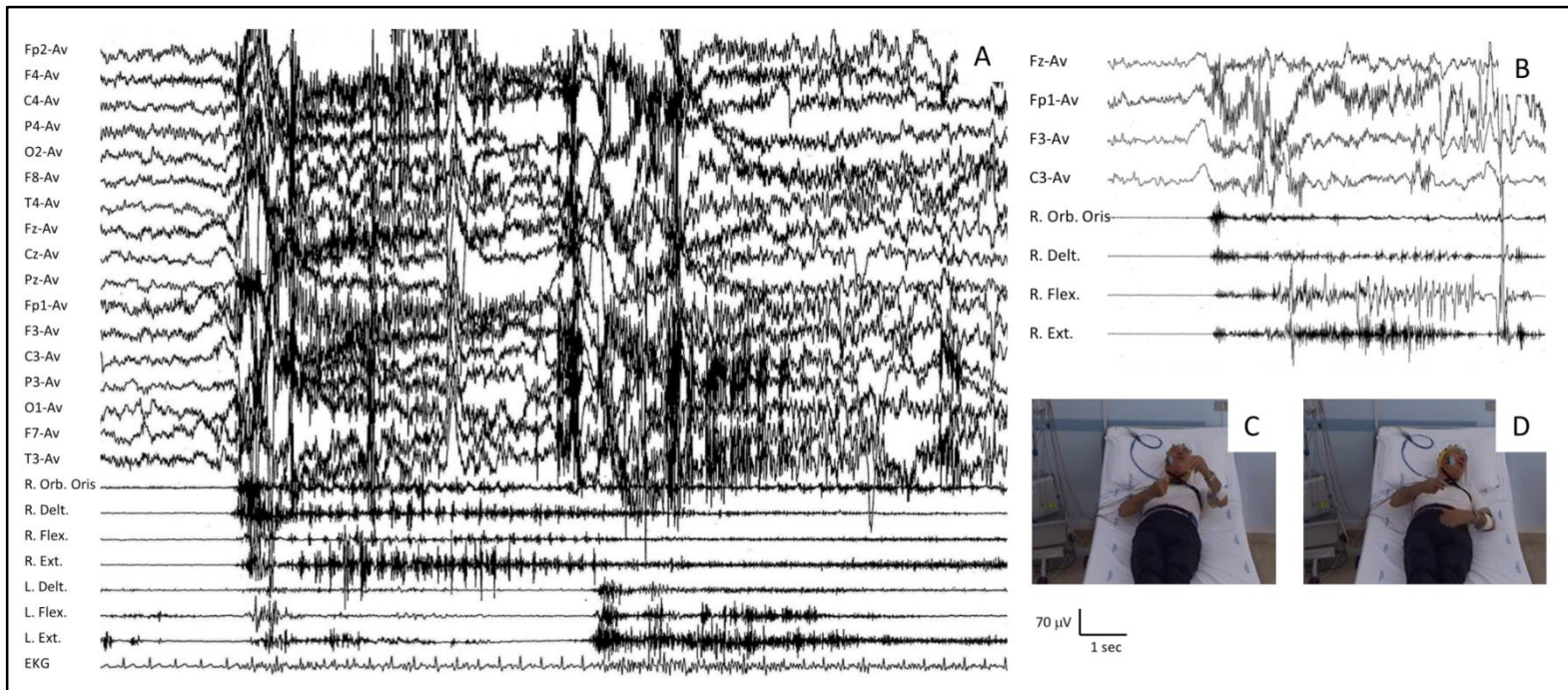
Epilepsy Centre - Clinic of Nervous System Diseases, Riuniti Hospital, Foggia, Italy

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- ❑ We observed a 68-year-old man with a 2-month history of subacute anterograde memory loss and involuntary faciobrachial movements.
- ❑ Video-EEG polygraphic recordings disclosed repetitive events characterized by a sudden, short contraction of the upper limbs and ipsilateral hemiface. A focal contralateral EEG wave was usually detected from frontal or central electrodes, and was accompanied by increased muscle activity, in a crescendo-decrescendo pattern. This EEG/EMG pattern, compatible with asymmetric tonic epileptic spasms, was always followed by oral and gestural automatisms with dystonic posturing of the upper limbs, compatible with temporal lobe seizures.
- ❑ Serum sodium concentration was 127 mmol/l. Brain MRI showed increased FLAIR signal in the bilateral mesial temporal lobes. Serum and CSF contained LGI antibodies at 1:1000 and 1:10 titres, respectively.

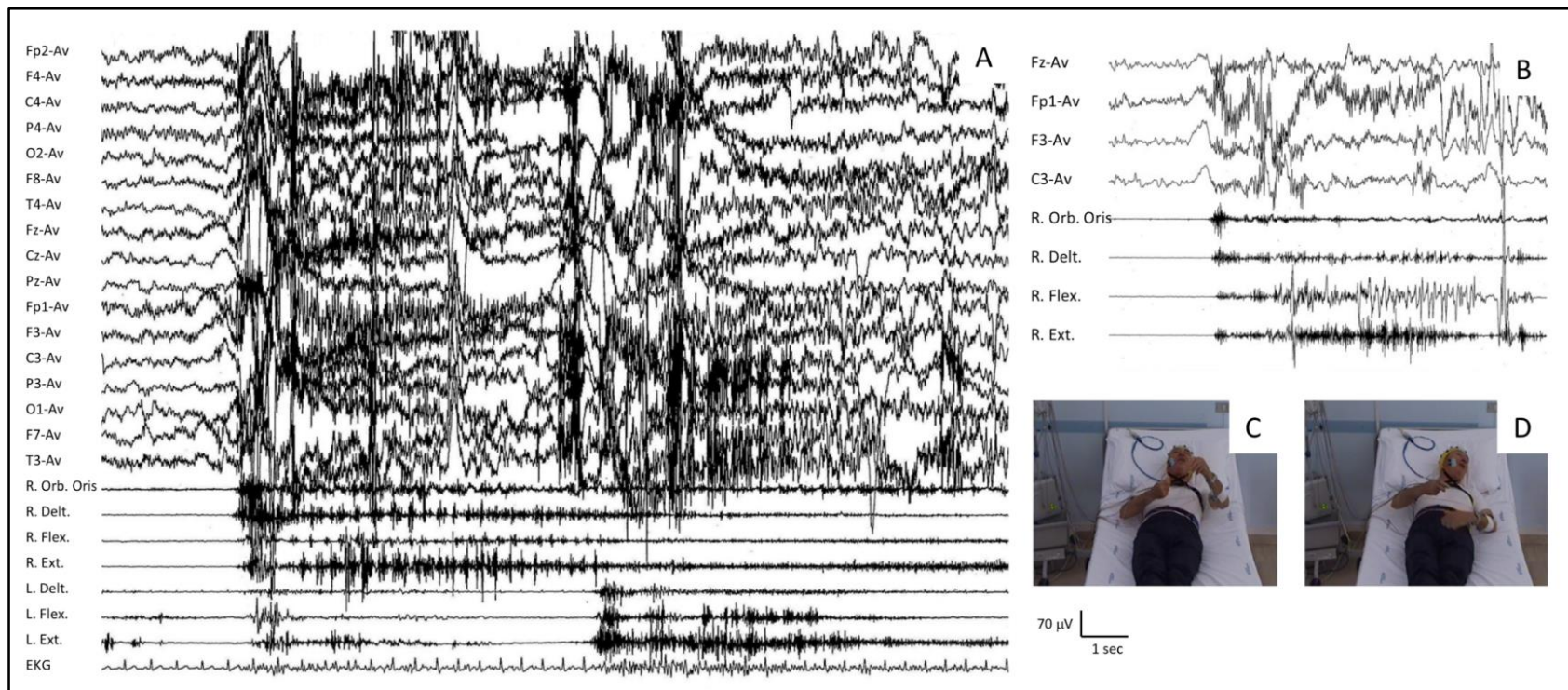
EEG/EMG

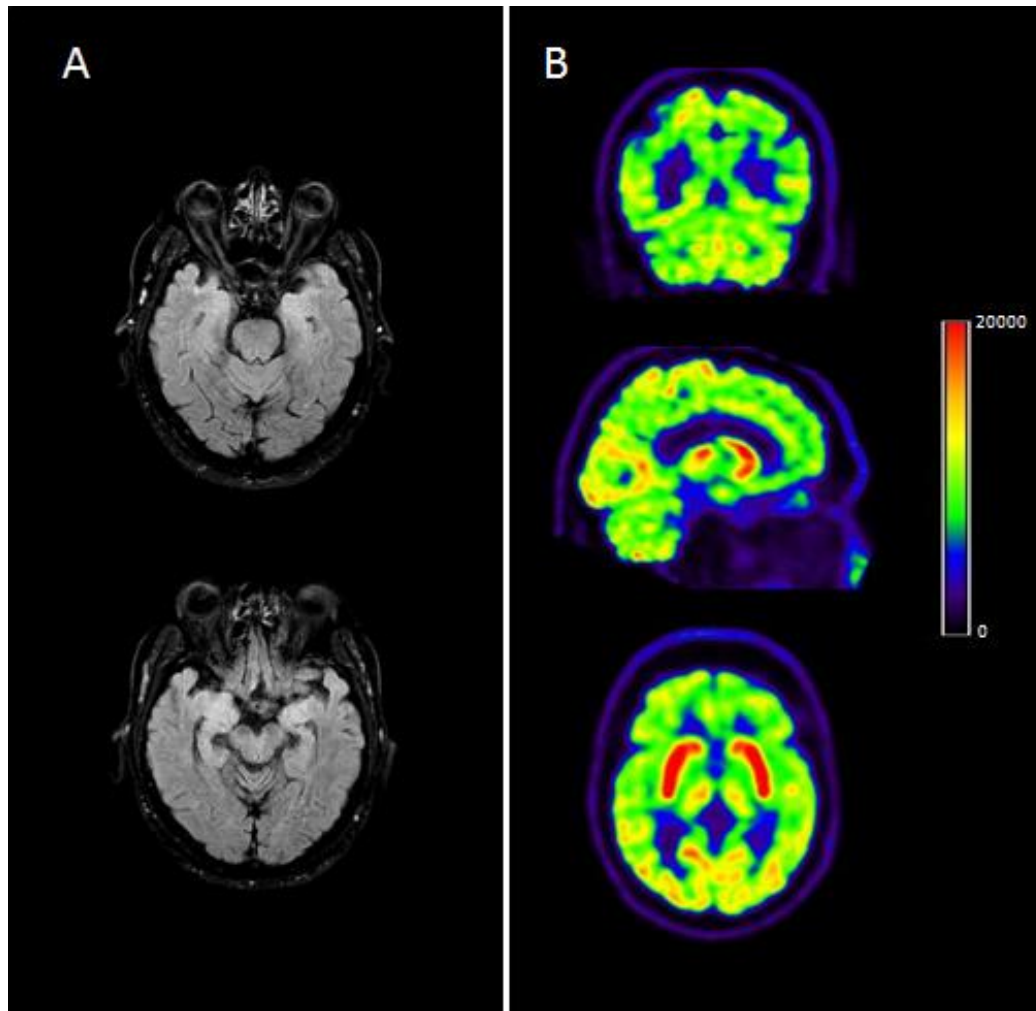
(A, B), a focal contralateral EEG wave, detected from frontal and central electrodes, preceded increased muscle activity, in a crescendo-decrescendo pattern, with a diamond-shaped configuration, especially on the orbicularis oris muscle, deltoid muscle, and extensor muscle of the hand. Latency between wave onset and onset of orbicularis oris spasm, deltoid spasm, extensor spasm, and flexor spasm was 573ms, 707ms, 831ms, and 898ms, respectively. The EEG/EMG pattern usually occurred bilaterally and asynchronously with a short (5-sec) 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.



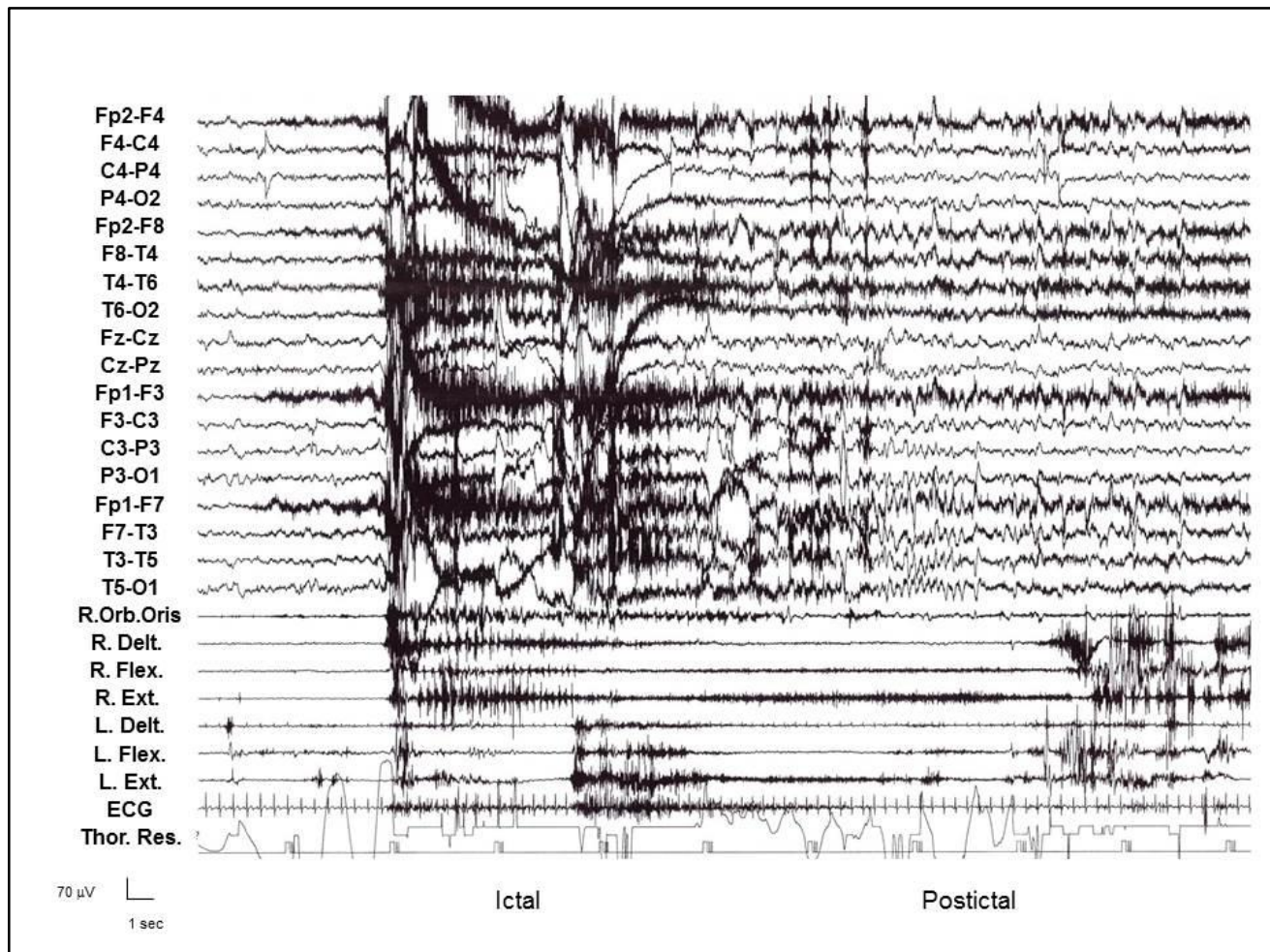
Video-EEG/polygraphic features of faciobrachial epileptic spasms:

(C, D) Clinically, sudden and short (less than 1 sec) contraction of the upper limbs and ipsilateral hemiface was seen.





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).



Video-EEG/polygraphic features of faciobrachial epileptic spasms. Two facio-brachial dystonic seizures occurred bilaterally and asynchronously. A prolonged postictal phase emerged, characterized by ambulatory automatisms, fluent aphasia, visual hallucinations, and scared facial expressions. EEG recording showed ipsilateral temporal rhythmic sharp waves evolving into regional slowing.