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Fronto-polar epilepsy masquerading as juvenile myoclonic epilepsy

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ABSTRACT – A woman diagnosed with juvenile myoclonic epilepsy for over 30 years presented with stereotyped episodes of abnormal sensations in both arms. Continuous video-EEG monitoring for 14 days and MRI brain revealed that the patient's somatosensory events with associated postictal aphasia, as well as her myoclonic and generalised tonic-clonic seizures, were likely due to a symptomatic left fronto-polar epilepsy. Our case emphasizes the need for clinicians to consider fronto-polar epilepsy as a potential cause of myoclonic seizures, particularly when associated with other semiologic features suggestive of frontal lobe epilepsy. *[Published with video sequences]*

Key words: myoclonus, epilepsy, frontal lobe, juvenile myoclonic epilepsy

Myoclonus is an important manifestation of several diseases of the central nervous system including the myoclonic epilepsies, the most important of which is juvenile myoclonic epilepsy (JME) (Leppik, 2003). In an effort to contribute to our evolving understanding of myoclonic seizures and their possible localising value, we present a patient whose initial clinical presentation and investigations were consistent with JME, but was later found to most likely suffer from a symptomatic left fronto-polar epilepsy.

Case study

A 49-year-old woman was referred to our epilepsy service for refractory seizures that she developed at age 14. Her peri-natal and developmental histories were unremarkable. Her family history included a grandmother who suffered from recurrent epileptic seizures which remitted at the end of adolescence and two sisters who possibly experienced isolated seizures during childhood. Our patient's typical convulsive seizure began with a myoclonic jerk of the legs, arms or trunk which was soon followed by a decrease in her level of consciousness and invariably evolved into a generalised tonic-clonic seizure (GTCS). There was an additional long-standing history of the patient running forward just prior to the onset of her GTCS, although the significance of this was not initially clear. According to the patient and

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her family, the postictal period was without any localising symptoms or signs. Her seizures recurred as frequently as every two months during adolescence but eventually waned to a handful per year (including a two-year seizure-free period). The patient was also known to have isolated myoclonic jerks which occurred several times per week but without any particular diurnal pattern. Prior routine EEGs had demonstrated bursts of 4-7-Hz generalised spike-and-wave activity which occasionally had a left temporal predominance. A previous CT scan of the brain was reportedly normal. Given the prominent history of myoclonus and convulsions, lack of aura, age of onset and initial EEG findings, the patient was diagnosed with JME.

In 2006 (coinciding with menopause), her seizure condition worsened with almost monthly convulsions prompting several antiepileptic drug trials (valproate, topiramate, clobazam, phenytoin, phenobarbital and carbamazepine). She also reported a novel type of spell characterised by abnormal sensations in both arms, extending from the fingers to the elbows without an associated alteration in her level of consciousness. These were quite rare except for a five-day period during which she experienced these sensations daily. She was eventually admitted to our video-EEG telemetry unit to better define these spells as well as her epileptic syndrome.

Continuous video-EEG monitoring for 14 days revealed a normal background rhythm but frequent interictal epileptiform discharges in the form of two-second bursts of generalised 2-3-Hz spike/polyspike-and-wave activity which was increased by hyperventilation and intermittent photic stimulation. Frequent myoclonus of the legs, arms and trunk was observed and electrographically associated with two to three-second bursts of 2.5-3-Hz generalised spike-and-wave activity. On two occasions, this myoclonus was followed by an alteration in the patient's level of consciousness and a brief attempt to lunge or run forward which was quickly interrupted by a GTCS. These seizures lasted approximately 60 seconds and were electrographically associated with the same 2.5-3-Hz generalized spike-and-wave activity. Towards the end of her hospitalisation, and only after the discontinuation of all of her antiepileptic medications, a third type of clinical event was observed. On this occasion the patient had a myoclonic jerk of the right leg associated with a three-second burst of generalised spike-and-wave activity. Seven seconds later, the patient experienced a second myoclonic jerk but also dysesthesias of both hands which was soon followed by an expressive aphasia. Electrographically, this event was associated with left fronto-temporo-central sharply contoured rhythmic theta activity which soon generalised but maintained left hemispheric predominance *(figure 1, see video sequence*).

MRI brain revealed a cystic lesion of the left fronto-marginal and inferior transverse fronto-polar gyri with surrounding gliosis *(figure 2)*. Ictal SPECT was unremarkable although the interictal SPECT demonstrated a mild hypoperfusion of the left frontal and temporal lobes.

Based on the clinical, EEG and neuroimaging findings, the patient was diagnosed with a left fronto-polar epilepsy syndrome presenting with prominent myoclonus as well as stereotyped motor agitation and somatosensory phenomena with postictal aphasia. Her seizures were eventually controlled with topiramate at 200 mg per day.

Discussion

In the case we have presented, the initial clinical presentation, EEG findings and normal brain scan were strongly suggestive of JME. It was the recent exacerbation of her seizure condition along with the development of new and atypical somatosensory events which prompted admission to hospital for prolonged video-EEG telemetry and high-resolution MR imaging. Only then was it suspected that the patient most likely suffered from a symptomatic left fronto-polar epilepsy syndrome. Of course, definitive proof in the form of invasive electrographic recordings or seizure freedom following lesionectomy is not available due to the patient's good response to medical therapy.

We consider it unlikely that our patient had simultaneous JME and symptomatic frontal lobe epilepsy. The co-occurrence of the myoclonus, focal semiologic features and electrographic discharges strongly suggest that they are due to a single epileptic process. We also consider it unlikely that all of her findings could be accounted for by an idiopathic generalised epilepsy. Firstly, while focal electrographic discharges (Usui *et al*., 2005), even evolving from an initial generalised epileptiform discharge (Williamson *et al*., 2009), have been described in JME, the observed left fronto-temporo-central discharges and the postictal aphasia corresponded very well with the location of the described lesion. In addition, although focal semiologies have been described in JME, they are typically simple versive head movements or posturing (Usui *et al*., 2005) and do not include stereotyped motor agitation, somatosensory phenomena or postictal aphasia. An alternate hypothesis would be that our patient did in fact have JME but that it had been clinically and electrographically modified by the fronto-polar lesion, therefore allowing for the focal semiologic and electrographic findings. We believe that such network plasticity in JME is not well established and therefore do not support this hypothesis. Finally, although rare, photoparoxysmal responses have been described in occipital, parietal as well as frontal lobe epilepsy

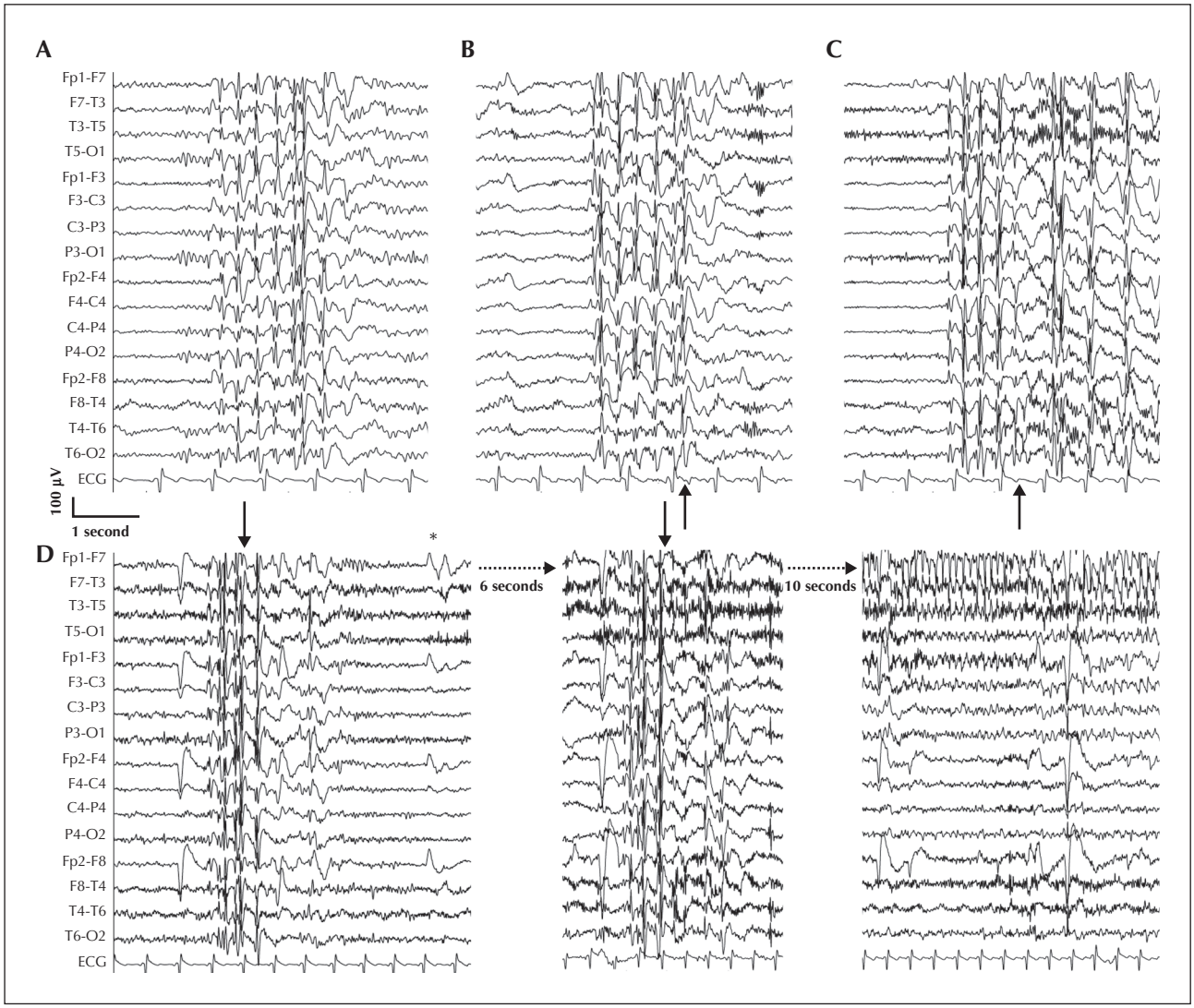

Figure 1. Electroencephalogram with (**A**) interictal generalised 2-3-Hz spike-and-wave discharges which were increased by hyperventilation and intermittent photic stimulation; (**B**) myoclonus (black arrow) with associated generalised spike-and-wave discharges; (**C**) the onset of a GTCS preceded by myoclonus (black arrow) and lunging with associated generalised spike-and-wave discharges; (**D**) an epileptic seizure characterised by recurrent myoclonus with associated generalised spike-and-wave discharges (black arrows) followed by hand dysesthesias (*) and postictal aphasia with associated left fronto-temporo-central sharply contoured rhythmic theta activity. Low and high frequency filters were set at 0.5 and 35 Hz, respectively.

(Seshia and Carmant, 2005) and, therefore, we do not believe that its presence challenges the diagnosis of a symptomatic frontal lobe epilepsy.

There has been only one previous report of myoclonic seizures occurring in the context of a symptomatic frontal lobe epilepsy (Cho *et al*., 2010). It is perhaps of some interest that frontal lobe dysfunction in patients with JME has been demonstrated in a number of recent neuropsychological, FDG-PET and dense array electroencephalography studies (Holmes *et al*., 2010). In fact, it has been previously suggested that asymmetric activation *via* the frontal lobes of the basal ganglia, supplementary motor area or prefrontal area can result in myoclonus, although this remains to be proven (Cho *et al*., 2010).

In summary, we present a patient who initially presented with a syndrome suggestive of JME but was later found to suffer from a symptomatic fronto-polar epilepsy. Definitive proof in the form of invasive electrographic recordings or post-surgical seizure freedom is not available. Our case emphasizes the need for clinicians to consider fronto-polar epilepsy as a potential cause of myoclonic seizures when associated with other semiologic or electrographic features suggestive of frontal lobe epilepsy. □

Disclosure.
None of the authors has any conflict of interest or financial support to disclose.

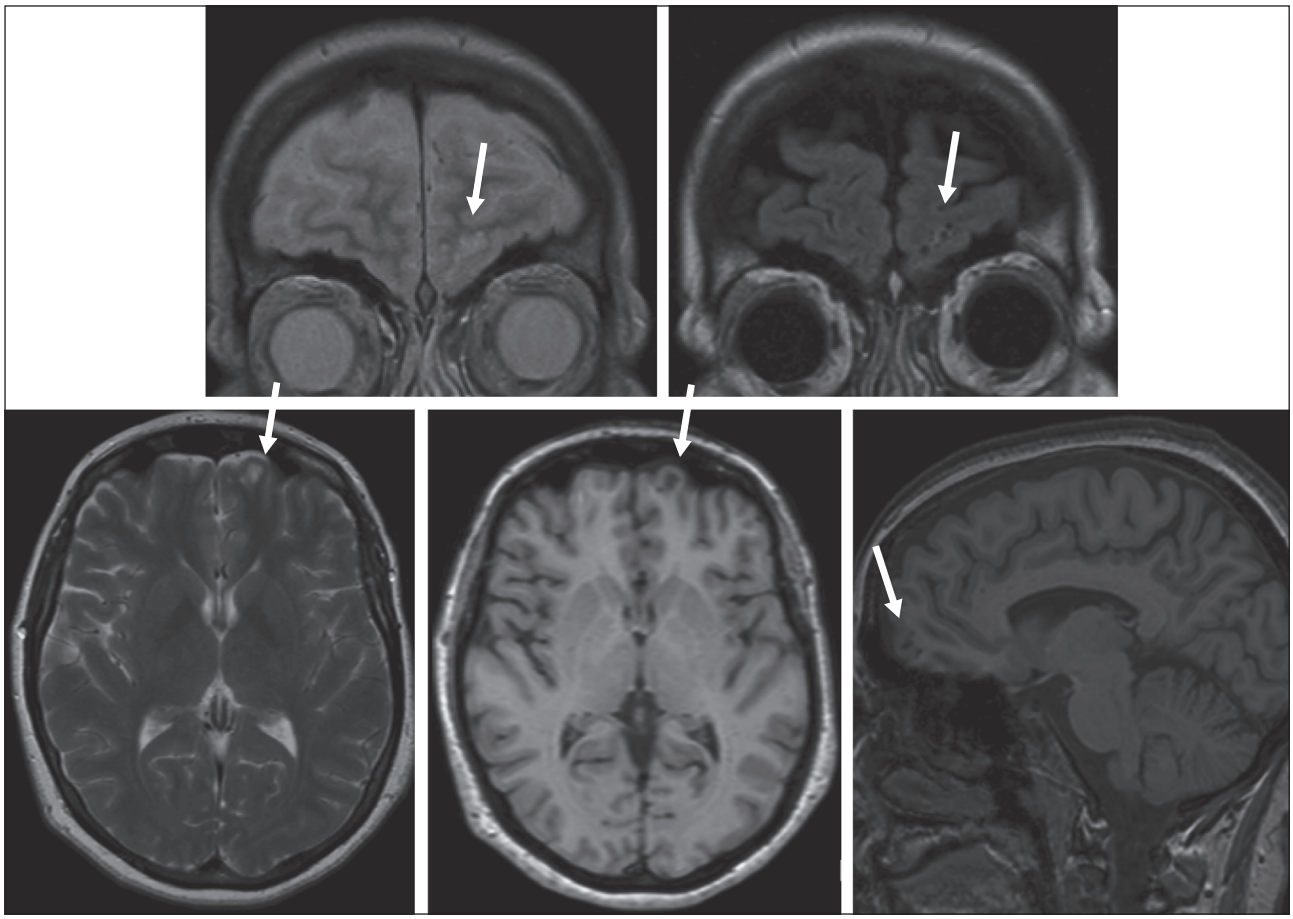

Figure 2. Brain MRI demonstrating a cystic lesion of the left fronto-marginal and inferior transverse fronto-polar gyri (white arrows).

Legend for video sequence

Video sequences corresponding to the EEG recordings illustrated in *figure 1*, sections (**B**), (**C**) and (**D**).
B) Myoclonus with associated generalised spike-and-wave discharges.
C) Onset of a GTCS preceded by myoclonus and lunging with associated generalised spike-and-wave discharges.
D) An epileptic seizure characterised by recurrent myoclonus with associated generalised spike-and-wave discharges followed by hand dysesthesias and postictal aphasia with associated left fronto-temporo-central sharply contoured rhythmic theta activity.

Key words for video research on www.epilepticdisorders.com
Etiology: gliosis
Phenomenology: myoclonic seizure, focal seizure not otherwise specified, tonic-clonic seizure
Localization: frontal lobe (left)
Syndrome: focal non-idiopathic frontal (FLE)

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