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CASE REPORT

Nocturnal frontal lobe epilepsy presenting with restless leg syndrome-like symptoms

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Abstract We describe the case of a 22-year-old male affected by NFLE reporting paroxysmal RLS-like symptoms. The patient was referred to our Sleep Center due to nocturnal paresthesias and cramps involving the left leg and leading to sleep fragmentation. At age 4, the patient presented with secondary generalized seizures preceded by left leg discomfort, controlled on CBZ. After successive therapy discontinuation, leg symptoms built up in frequency and duration until a secondary generalized seizure re-occurred. On CBZ prompt resumption no further GM seizures occurred albeit persistence of night-time frequent cramps and paraesthesia. Sleep EEG demonstrated asymmetric interictal sharp theta on the right posterior frontal areas, whereas brain MRI results were consistent with a Taylor type right frontal cortical dysplasia. CBZ augmentation and add on therapy with LEV led to further frequency reduction of sensory symptoms.

Keywords NFLE · RLS · Cortical dysplasia Taylor-type

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Introduction

Nocturnal frontal lobe epilepsy (NFLE) is a condition characterized by seizures with a semiology suggesting a frontal lobe origin, occurring predominantly during sleep. Paroxysmal arousal (PA), nocturnal paroxysmal dystonia (NPD) and episodic nocturnal wanderings (ENW) are some of the different seizure types that characterize this condition. NFLE includes both sporadic and familial forms. The age of onset is usually around childhood or adolescence. Interictal/ictal scalp EEG abnormalities are not always detectable and when visible are distributed predominantly over the frontal, fronto-temporal, or temporal regions [1]. Carbamazepine (CBZ) is the first choice antiepileptic drug leading to seizure control in about 80% of patients [2]. Sleep in NFLE is often disturbed by repetitive arousals due to minor motor events besides major seizures. NFLE patients also may report excessive daytime sleepiness [3], probably due to sleep instability expressed by a high cyclic alternating pattern (CAP) rate. Periodic leg movements disorder (PLMD) distinguishes a sleep related motor disorder that may contribute to an increase of CAP rate [4]. PLMS occur in a wide variety of sleep disorders and are also reported in epileptics. They are also found in almost 80–90% of patients presenting with restless legs syndrome (RLS). RLS is a sensory-motor disorder characterized by unpleasant leg (rarely arm) sensations coupled with an irresistible inner urge to move occurring at night and/or in resting position, improved by locomotion. The four essential International Diagnostic Criteria for the diagnosis of RLS [5] are: (1) an urge to move the legs, usually accompanied or caused by uncomfortable and unpleasant sensations in the legs; (2) The urge to move or unpleasant sensations begin or worsen during periods of rest or inactivity such as lying or sitting; (3) The urge to move or

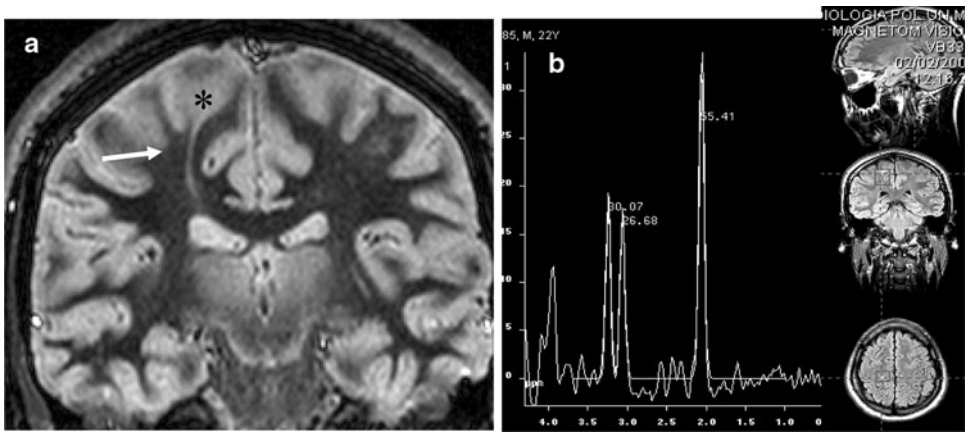

Fig. 1 **a** Brain MRI coronal fast inversion recovery with myelin suppression (FIRMS) sequences: hyperintensity of subcortical white matter (*asterisk*)—radial hyperintensity spreading out to the ventricular ependyma (*arrow*). **b** H-MR Spectroscopy single-voxel SE (TE 135 ms) Normal pattern of main brain metabolites: *N*-acetyl Aspartate (NAA), 2 ppm; creatine (Cr), 3.02 ppm; choline (Cho), 3.22 ppm; second Cr peak, 3.94 ppm

unpleasant sensations are partially or totally relieved by movement, such as walking or stretching, at least as long as the activity continues; and (4) The urge to move or unpleasant sensations are worse in the evening or night than during the day or only occur in the evening or night. We describe the case of a young patient affected by NFLE reporting paroxysmal RLS-like symptoms.

Case report

A 22-year-old man was referred to our Sleep Medicine Centre due to nocturnal paraesthesias and cramps involving the left leg, predominantly the quadriceps, occurring in bouts of 1–2 min of duration several times/week. These symptoms would usually start in the evening inducing motor restlessness and might interfere with sleep initiation or lead to sleep fragmentation in the middle of the night. The patient's familial history was negative for neurologic, psychiatric and sleep disorders. Personal history included nocturnal secondary generalized seizures occurring since age 4, preceded by pain, cramps and weakness of the left leg, lasting 1–2 min. At that time he was started on CBZ 400 mg/day, obtaining seizure complete control. Arbitrary discontinuation of therapy between age 11 and 13 led to a new secondary generalized seizure and therapy resumption, up to age 18, when the drug was successfully discontinued under medical advice. During the following years nocturnal cramps and paraesthesias of the left leg occurred similar to the symptoms presently reported, building up in frequency and duration until the month preceding our evaluation, when a nocturnal secondary generalized seizure re-occurred.

CBZ had been promptly reinstated 400 mg/day with no further generalized seizures albeit persistence of left leg cramps and paraesthetic sensation while at rest, preferentially during the evening. The patient, who had a normal neurological evaluation and blood measurements, underwent a sleep deprived EEG that showed asymmetric interictal sharp theta bursts on the right posterior frontal areas, the same showing poli-spikes on previous paediatric recordings.

The patient successively underwent a brain MRI showing hyperintensity of subcortical right frontal white matter spreading out radially to the ventricular ependyma on coronal fast inversion recovery with myelin suppression (FIRMS) sequences (Fig. 1a). This lesion appeared as a slim area located in the proximity of the precentral gyrus, anterolateral to the paracentral lobulus. HMR spectroscopy (HMRS) proved normal (Fig. 1b) confirming a Taylor type focal dysplasia (FCDT), allowing a differential diagnosis with a low grade glial tumour.

One night video-polysomnography (PSG), performed without a previous adaptation night, confirmed interictal EEG focal abnormalities. Total sleep time (TST) was drastically reduced (262 min) including mostly slow wave sleep (SWS = 40%) with no REM sleep, probably due to patient's discomfort from the unusual sleep setting. No clinical/eegraphic ictal episodes and a low prevalence of PLMs (PLMS index 7) were recorded with an arousal index of 8.3. CBZ was subsequently titrated up to 800 mg/day with progressive reduction of night-time paraesthesias. Ultimately Levetiracetam 500 mg bid was started as add-on therapy with further frequency reduction (one in 3–4 month) of sensory symptoms.

Discussion

Several sleep disorders have been reported to occur in NFLE: either sleep related movements disorders such as bruxism and PLMS or parasomnias such as nightmares or disorder of arousals [6]. Recently Nobili et al. [7] described the relationship of epileptic discharges to arousal instability and PLMS in a case of NFLE studied by stereo-EEG recording, showing the correlation of minor motor events

expressing arousal instability with recurrent epileptic discharges not detectable on scalp EEG. So far no sensory symptoms within the RLS spectrum, in the absence of motor discharges, have been reported as minor events or paroxysmal arousals. Our patient's symptoms were different from the typical RLS spectrum as far as they presented acutely, short in duration and unrelieved by motor activity. Nocturnal paresthesia as well as cramps and strictly unilateral symptoms are neither typical of the syndrome nor are they mentioned in the four diagnostic criteria for RLS, from which this case differ. Nonetheless, it may suggest that a discrete FCDT in an eloquent motor area may elicit primary sensory phenomena within the RLS spectrum as residual symptoms of an overall well controlled symptomatic NFLE.

References

1. Provini F, Plazzi G, Tinuper P, Vandi S, Lugaresi E, Montagna P (1999) Nocturnal frontal lobe epilepsy. A clinical and polygraphic overview of 100 consecutive cases. Brain 122(Pt 6):1017–1031
2. Oldani A, Zucconi M, Ferini-Strambi L (1997) Nocturnal frontal lobe epilepsy: clinical and video-polysomnographic features. Epilepsia 38(Suppl. 3):154–155
3. Vignatelli L, Bisulli F, Naldi I, Ferioli S, Pittau F, Provini F et al (2006) Excessive daytime sleepiness and subjective sleep quality in patients with nocturnal frontal lobe epilepsy: a case control study. Epilepsia 47(Suppl 5):73–77
4. Parrino L, Boselli M, Buccino GP, Spaggiari MC, Di Giovanni G, Terzano MG (1996) The cyclic alternating pattern plays a gate-control on periodic limb movements during non-rapid eye movement sleep. J Clin Neurophysiol 13:314–323
5. Allena RP, Picchietti D, Hening WA, Trenkwalder C, Walters AS, Montplaisir J et al (2003) Restless legs syndrome: diagnostic criteria, special considerations, and epidemiology. A report from the restless legs syndrome diagnosis and epidemiology workshop at the National Institutes of Health. Sleep Med 4(2):101–119
6. Zucconi M, Ferini-Strambi L (2000) NREM parasomnias: arousal disorders and differentiation from nocturnal frontal lobe epilepsy. Clin Neurophysiol 111(Suppl 2):S129–S135
7. Nobili L, Sartori I, Terzaghi M, Stefano F, Mai R, Tassi L, Parrino L, Cossu M, Lo Russo G (2006) Relationship of epileptic discharges to arousal instability and periodic leg movements in a case of nocturnal frontal lobe epilepsy: a stereo-EEG study. Sleep 29(5):701–704