Movement Disorders
CLINICAL PRACTICE

PHENOMENOLOGY VIDEO

An Adult with Episodic Abnormal Limb Posturing

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Case Description

A previously healthy 37-year-old woman presented with paroxysmal and stereotyped episodes (5–10/day, lasting about 30 seconds) of painful muscle contractions causing abnormal involuntary postures of the left lower limb, rapidly spreading to the homolateral upper limb, along with forearm extension and hand closure while awake (video). Her contorted facial expression was probably due to pain, although facial involvement in the involuntary movements was difficult to exclude. Examination disclosed absent plantar

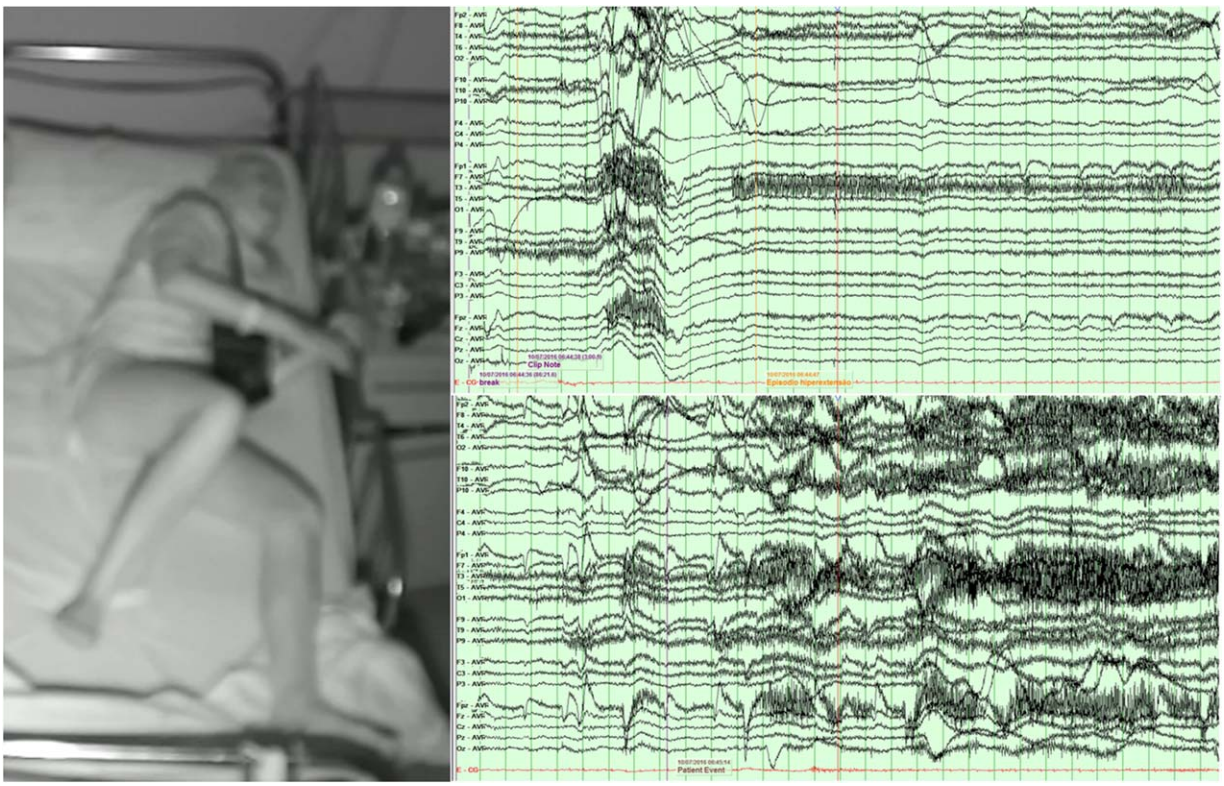

FIG. 1. Video-EEG monitoring during one of the paroxysmal spasms showing no epileptiform activity.

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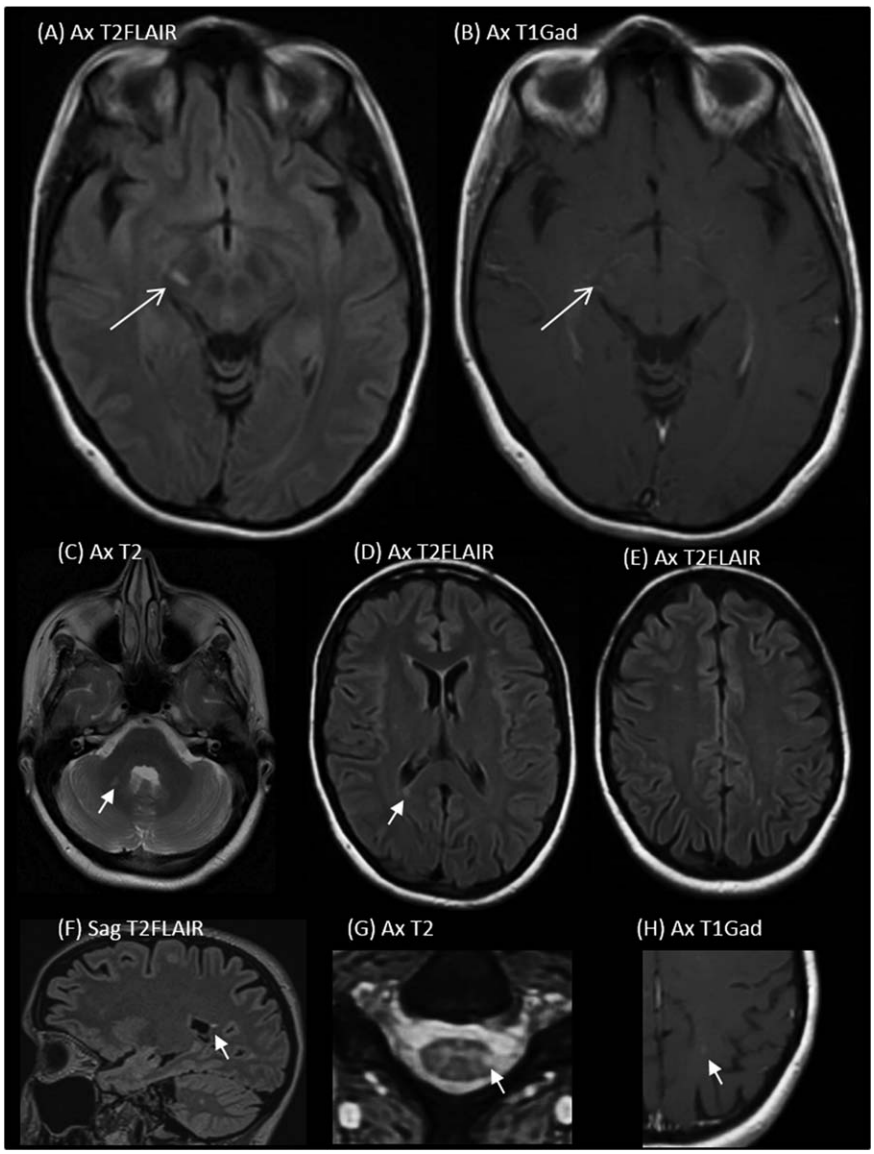

FIG. 2. (**A-B**) Brain MRI (T2) showing a right cerebral peduncle (pyramidal tract) lesion with gadolinium enhancement assumed to cause the tonic spasms; (**C-H**) white matter lesions with 2010 McDonald criteria for dissemination in space and time (one enhancing, asymptomatic lesion, H).

reflexes and lower limb hypopalesthesia. No triggers were identified and family history was unremarkable. Levetiracetam was ineffective.

What is the most likely diagnosis?

- Focal motor epileptic seizures
- Paroxysmal kinesigenic dyskinesia (PKD)
- Paroxysmal non-kinesigenic dyskinesia (PNKD)
- Tonic spasms (TS) due to demyelinating disease

Answer

Focal motor seizures with tonic semiology were first considered. Nevertheless, other epileptic features are usually identified and an ictal video-EEG monitoring was unremarkable (Fig. 1). PKD and PNKD consist of unilateral or bilateral dystonia and chorea attacks, precipitated by movement in PKD, while coffee, alcohol, and fatigue may trigger PNKD episodes.[1] PKD or PNKD would be unlikely due to their typical presentation in childhood or adolescence, positive family history, and absence of pain.[1] This patient's presentation is compatible with TS, an uncommon manifestation of multiple sclerosis (MS), which may constitute a relapse or seldom

the presenting symptom.[2] TS are painful, stereotyped, occur up to 80 times/day, and last about 30 seconds to 2 minutes.[2] They may be preceded by sensory symptoms and triggered by hyperventilation, movements, or tactile stimulation.[2] TS probably result from ephatic neuronal transmission within a corticospinal tract lesion.[2] In fact, MRI showed a right cerebral peduncle enhancing lesion and other features compatible with MS (Fig. 2). CSF oligoclonal bands were detected. A diagnosis of relapsing-remitting MS was established. Methylprednisolone was ineffective. TS remitted under carbamazepine, as described in the literature.[2]

Author Roles

1. Research Project: A. Conception, B. Organization, C. Execution; 2. Statistical Analysis: A. Design, B. Execution, C. Review and Critique; 3. Manuscript Preparation: A. Writing the First Draft, B. Review and Critique.

Mariana Santos: 1A, 1B, 1C, 3A
Leonor Rebordão: 1A, 1B, 1C, 3A
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Cristina Costa: 1A, 1B, 1C, 3A, 3B
Patrícia Pita Lobo: 1A, 1B, 1C, 3A, 3B

Disclosures

Ethical Compliance Statement: We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

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References

1. Edwards MJ, Stamelou M, Quinn N, Bhatia KP. Parkinson's Disease and Other Movement Disorders, second edition, Oxford University Press, 2016, pp 268-275.
2. Compston A, Confavreux C, Lassmann H et al. McAlpine's Multiple Sclerosis, fourth edition, Churchill Livingstone Elsevier, 2006, pp 323-325

Supporting Information

Videos accompanying this article are available in the supporting information here.

Video 1. Paroxysmal dystonia episode: Sudden, painful contraction with abnormal posturing of the left limbs, lasting about 30 seconds.