Epileptic Disord 2019; 21 (4): 389-90

Brainstem lesion causing paroxysmal ataxia, dysarthria, diplopia and hemifacial spasm (PADDHS)

Enrico Ferrante [1], Maria Raffaella Marazzi [2], Michele Trimboli [1], Davide Dalla Costa [3], Cristina Erminio [2], Lino Nobili [4]

[1] Neurology Department, AOR San Carlo, Potenza
[2] Neurosciences Department, Niguarda Cà Granda Hospital, Milan
[3] Neurorehabilitation Unit, Niguarda Cà Granda Hospital, Milan
[4] Neuropsychiatry Unit, IRCCS, Gaslini Institute, Dinogmi, University of Genoa, Italy

Received January 07, 2019; Accepted June 06, 2019

A 67-year-old man developed paroxysmal ataxia, dysarthria (with a distorted voice that sounded like a 45-rpm record played at 33 rpm), diplopia, and hemifacial spasms (PADDHS) lasting for 15-40 seconds, ~40 times/day. Ictal video-EEGs did not show paroxysmal discharges. Brain MRI revealed a right-midbrain lesion (*figure 1A-C*). Inflammatory/autoimmune/neoplastic work-up was normal. The patient took steroid and broad-spectrum antibiotic/antimycotic therapy empirically. PADDHS attacks lasted for five months and then disappeared, and brain MRI returned to normal (*figure 1D-F*). At 24 months of follow-up, the patient was still asymptomatic. The nature of the lesion was similar to that of a previously described case of Neuro-Behçet disease (Kontzialis and Guryildirim, 2017). The brainstem lesion might have caused PADDHS due to ephatic axonal activation within demyelinated crossed fibre tracts, with interruption of the cerebello-thalamo-cortical pathway. PADDHS is a new syndrome, expanding the clinical spectrum associated with paroxysmal ataxia and dysarthria (PAD) (Li *et al*., 2011). This non-epileptic paroxysmal event should be considered in the differential diagnosis of focal epileptic seizures (Lüders *et al*., 2019). □

VIDEO ONLINE

Correspondence:
Enrico Ferrante
Neurology Department,
AOR San Carlo,
Potenza, Italy
<*enricoferrante@libero.it*>

doi:10.1684/epd.2019.1080

Legend for video sequence

PADDHS attack lasting for 38 seconds, triggered by hyperventilation during EEG. The patient firstly had right facial hemispasm and binocular diplopia, then a distorted voice and upper right limb ataxia become evident before spontaneous and complete resolution of symptoms.

Key words for video research on www.epilepticdisorders.com

Phenomenology: non-epileptic paroxysmal event
Localisation: not applicable
Syndrome: non-epileptic paroxysmal disorder
Aetiology: immune disorder

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

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

References

Kontzialis M, Guryildirim M. Acute/subacute Neuro-Behcet's disease. *Acta Neurol Belg* 2017; 117: 925-6.

Li Y, Zeng C, Luo T. Paroxysmal dysarthria and ataxia in multiple sclerosis and corresponding magnetic resonance imaging findings. *J Neurol* 2011; 258: 273-6.

Lüders H, Vaca GF, Akamatsu N, *et al*. Classification of paroxysmal events and the four-dimensional epilepsy classification system. *Epileptic Disord* 2019; 21: 1-29.

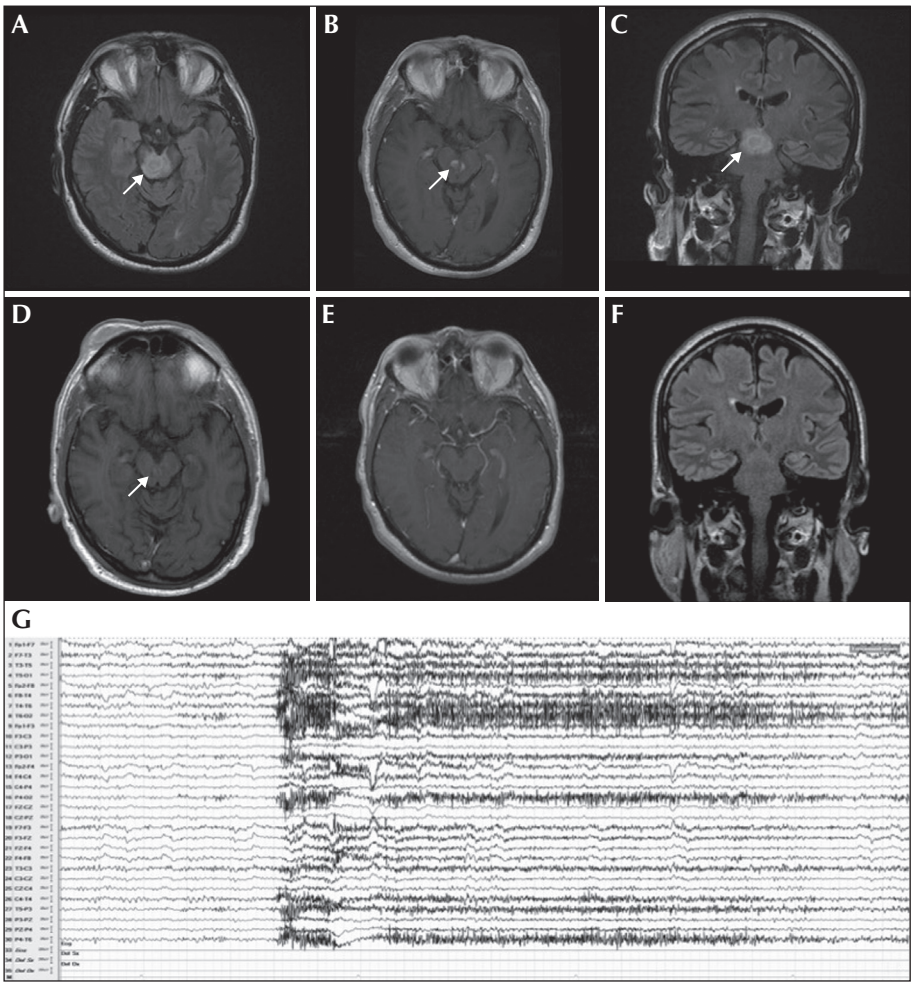

Figure 1. (A-C) Axial and coronal brain MRI showing enhanced right midbrain and a cystic lesion below the level of the red nucleus, extending to the midbrain-pons junction. After five months, MRI showed only a small level of nuanced midbrain flair hyperintensity (D-F) without enhancement (E). Ictal EEG, during hyperventilation, showed movement artefacts and myogenic potentials without paroxysmal discharges (G).

TEST YOURSELF

(1) What disease should be included in the differential diagnosis of a paroxysmal event?
A. Migraine aura
B. Focal epileptic seizure
C. Psychogenic disorder
D. A+B+C

(2) Which of these symptoms is not typical of a pons-midbrain lesion?
A. Ataxia
B. Diplopia
C. Convergence retraction nystagmus
D. Decrease in visual acuity

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".