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Letter to the Editor

Do not rely on imaging to predict awakening: The value of neurophysiology in a case of Weston-Hurst syndrome

A 68-year-old woman was admitted in ICU 6 days after a common bronchitis. She presented a rapidly progressive headache and a left hemiplegia. The initial CT-scan was inconclusive for a stroke. Lumbar puncture was inflammatory (100 white cells/mm^3, including 91% neutrophils, 412 red cells/mm^3) with an increased level of protein (2.75 g/L) and lactate (4.7 mM) but a normal glucose level. CSF examination for virus and atypical bacteria was negative and the CSF protein electrophoresis was normal. While the disorder of consciousness appeared (GCS: E3M5V1), the MRI showed a diffuse vasogenic edema with subfalcine brain herniation (Fig. 1A, left side). Concomitantly, intra-cranial pressure was monitored and was found initially increased at 32 mmHg.

A post-infectious encephalitis was proposed as a unifying diagnosis. Facing the reported unfavorable outcome of this rare clinical entity, a care withdrawal was rapidly discussed because of the absence of awakening despite a broad-spectrum probabilistic treatment (meningeal antibiotic therapy, 6-day high-dose corticosteroid and 5-day immunoglobulins). Our unit was contacted as a referral center to provide an external opinion in this procedure and the patient was transferred in our specialized neuro-intensive care unit. A stereotactic biopsy was performed. It confirmed the diagnosis of acute hemorrhagic leukoencephalitis (or Weston-Hurst syndrome) with a mixed lymphocytic-macrophagic perivascular infiltrate (Sonneville et al., 2010). The existence of diffuse siderophagic micro-hemorrhage and the absence of demyelination were both compatible with the sub-acute nature of these lesions. No evidence for alternative causes of leukoencephalitis (CMV, JC or lymphoma) was observed. In spite of the expected outcome of this pathology (Sonneville et al., 2010) and the lesion-load observed on T2EG and Susceptibility-Weighted Imaging (SWI) sequences (Fig. 1B), we performed a systematical neurophysiological assessment as routinely used in our center before any care withdrawal decision. The neurophysiological observations were compatible with signs of preserved sub-cortical and cortical (primary cortex and associative cortex) information processing, supporting a favorable awakening outcome (Fischer et al., 1999, 2008). We observed the normality of subcortical conduction and cortical responses using somatosensory (Fig. 2A) and auditory EPs (at the brainstem and cortical levels, Fig. 2B). The EEG was reactive to pain and sound stimulus (Fig. 2C). At a more integrated level, an oddball paradigm demonstrated the presence of a late auditory component (N100) but without Mismatch Negativity (Fig. 2D, upper part). Ultimately a subject own-name P300 response was present (Fig. 2D, lower part). The clinical evolution confirmed this favorable prognostic estimate from the electrophysiological data as she awoke during the following days (she was extubated 7 days after the neurophysiological assessment). She recovered a partial functional autonomy at 1-year as she was able to walk with a technical support despite persistent executive or attention disorders (GOSE: 3.1) with a persistent white matter hyperintensity associated to a diffuse atrophy on MRI (Fig. 1A, right side).

Nosological (AHLE), anamnestic (increased ICP and brain herniation) and radiological (T2EG micro-hemorrhage, magnified by SWI) pejorative data were less accurate than the standard neurophysiological responses, which were dissociated from a dramatic pattern of juxta-cortical lesions. Specifically, in this typically micro-hemorrhagic disease, SWI can be used for diagnostic purposes but should be regarded cautiously to avoid unjustified care withdrawal decision. Despite favorable outcome (Alemdar et al., 2006) and anatomopathological description (Magun et al., 2016) have been previously reported, this case constitutes the first exhaustive report of Weston-Hurst syndrome including both an anatomopathological confirmation and a multimodal analysis using SWI for the morphological description and an extended neurophysiological assessment, beyond a simple EEG for diagnostic purpose (Alemdar et al., 2006). The neurophysiological investigation proved crucial in predicting favorable outcome.

Conflict of interest

No conflict of interest.

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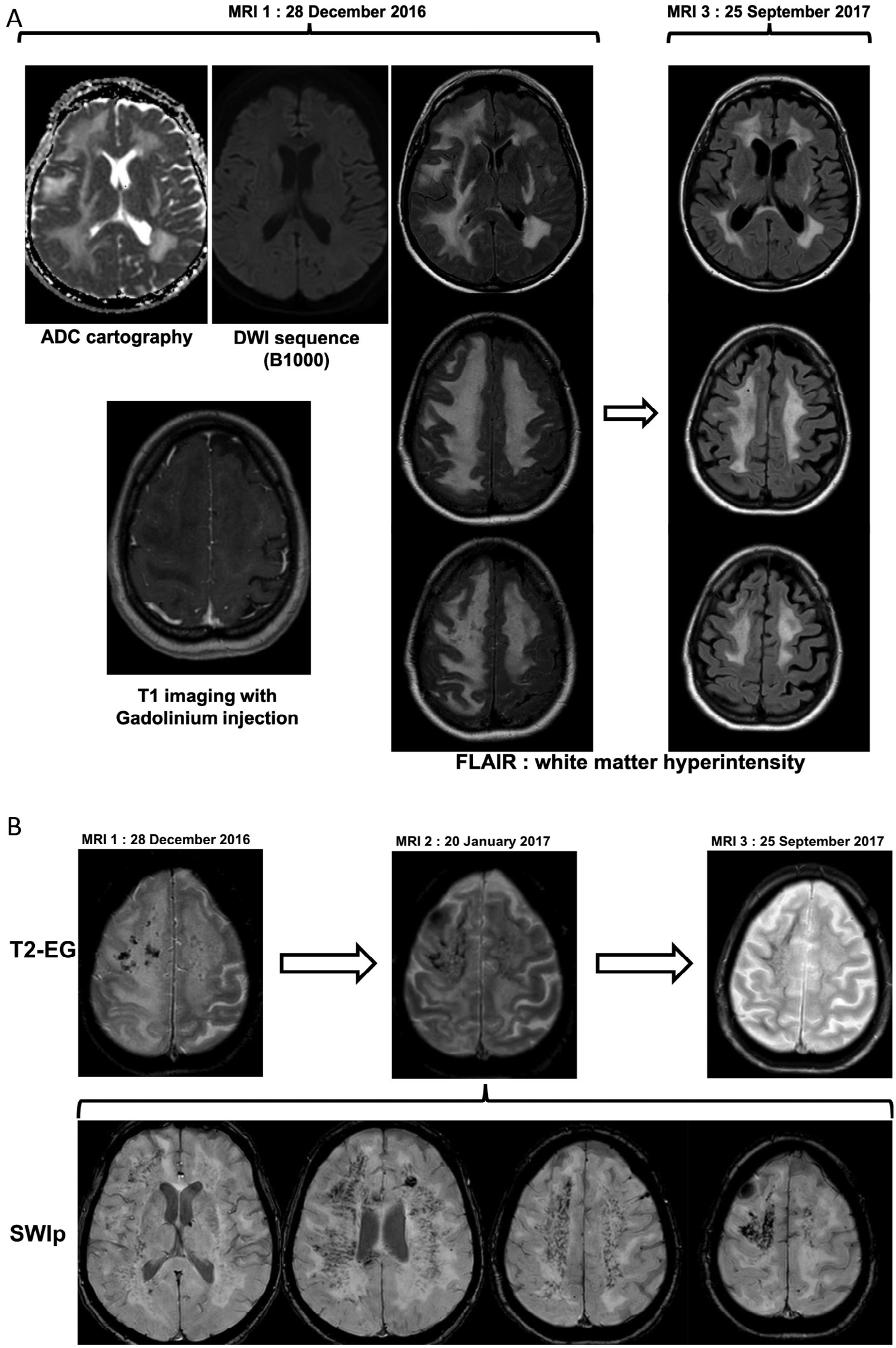

Fig. 1. (A) Evolution of MRI hyperintensity in T2-weighted imaging between initial coma onset (left side) and follow-up after 9 months (right side). Fluid-attenuated inversion recovery (FLAIR) sequences provided an anatomical evaluation of white matter edema. The mechanism is illustrated by Apparent Diffusion Coefficient (ADC) cartography (left-upper corner: vasogenic edema with increased ADC values and normal Diffusion Weighted Imaging image) and T1 sequences after Gadolinium injection (right-lower corner: absence of blood-brain barrier breakdown). (B) Evolution of T2-EG imaging between initial coma onset (left side) and follow-up after 9 months (right side). At the intermediate prognosis evaluation within the first month of coma evolution, T2-EG is compared with Susceptibility-Weighted Imaging sequences illustrating the extension of brain micro-hemorrhages with higher sensitivity.

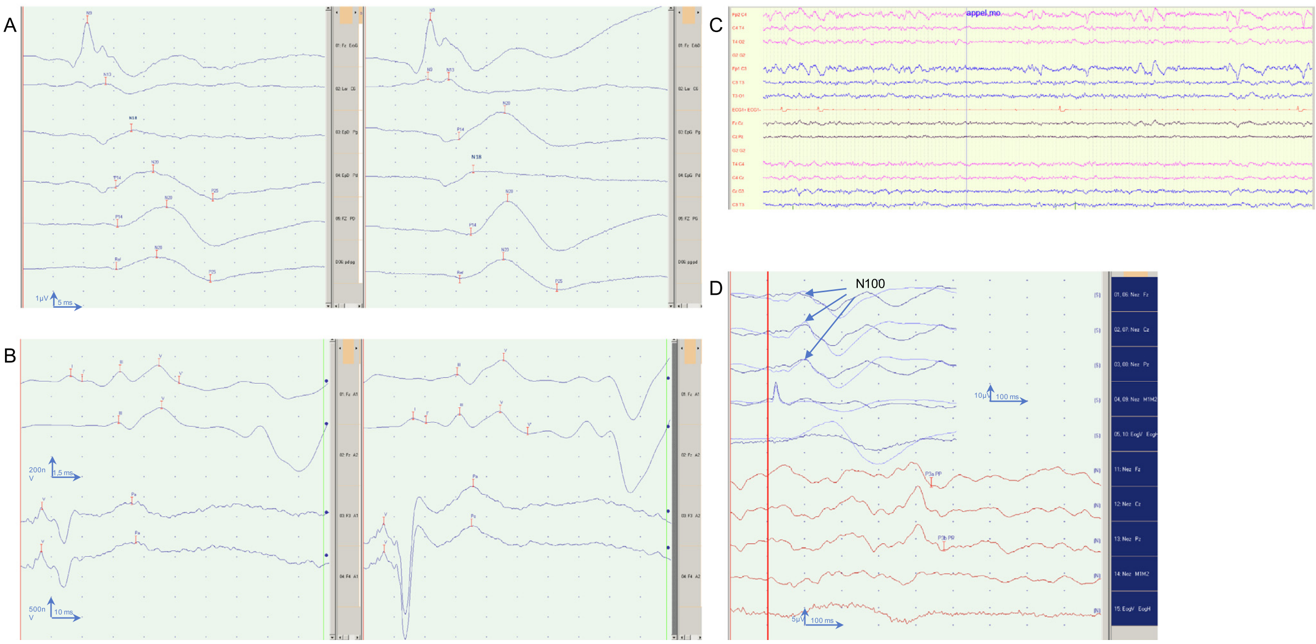

Fig. 2. (A) Somatosensory evoked potentials after right (on the right side) and left (on the left side) median nerve stimulations presenting the integrity of the pathway up to the left (on the right side) and right (on the left side) post-central parietal gyrus (scale: 1 μV/div). (B) Auditory evoked potentials after stimulation of the right ear (on the right side) and the left ear (on the left side) presenting the integrity of the pathway in the brainstem (top part, BAEPs scale: 200 nV/div) and in both superior temporal gyri (bottom part, MLAEPs, scale 500 nV/div). The Na response was not observed because of a concomitant muscular artifact. (C) 30-s page of standard 10–20 system bipolar EEG (from top to bottom: longitudinal right, longitudinal left, median line and transversal derivations; scale: 70 μV/cm) presenting the reactivity to an auditory stimulus (calling the patient's name). (D) Auditory evoked potentials with an oddball mismatch negativity (MMN) paradigm (top part) and P300 (bottom part) paradigm presenting respectively: (i) a N100/P200 complex for standard (light blue) and deviant (dark blue) sounds. No MMN was observed, as N100 curves were not significantly different; (ii) a frontal P3a (up) and a parietal P3b (down) responses. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

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