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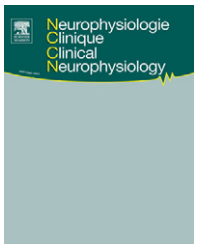

ORIGINAL ARTICLE/ARTICLE ORIGINAL

Transient post-traumatic locked-in syndrome: A case report and a literature review

À propos d'un cas de syndrome de dé-efférentation post-traumatique réversible. Revue de la littérature

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KEYWORDS
Locked-in syndrome;
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Summary
Introduction. — Post-traumatic locked-in syndrome may be particularly difficult to recognize, especially when it follows a state of coma and presents the clinical feature of a ''total'' locked-in syndrome.
Patient and methods. — A 56-year-old male with a closed head injury was admitted in intensive care unit (ICU) with GCS = 4 (V1, M2, E1). Computed tomography (CT) scan disclosed a limited subarachnoid haemorrhage in the sylvian region without any brain oedema or ventricular shift. The GCS did not change until day 6. At the same time EEG showed a reactivity to acoustic stimuli consisting in the paradoxical appearance of a posterior rhythm in alpha range (10—12 c/s), blocked by passive eye opening. Early cortical components (N20—P25) of somatosensory evoked potentials were normal on both hemispheres; middle components were also clearly evident. Magnetic resonance imaging of the brain showed both diffuse and midbrain axonal injuries, particularly in a strategic lesion involving both cerebral peduncles. Event related potentials showed N2 and P3 components to stimulation by rare tones.
Conclusions. — A comprehensive multimodal neurophysiological approach, using the more informative tests and the proper time of recording, should be included in protocols for patients with severe head trauma, in order to establish the actual patient's clinical state and to avoid

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that a locked-in syndrome state be mistaken for prolonged coma, vegetative state, minimally conscious state or akinetic mutism. Neurophysiological evaluation before discharge from ICU can be a baseline evaluation useful for the follow-up of low-responsive patients in the neurorehabilitation unit.

MOTS CLÉS
Traumatisme crânien ;
Syndrome de verrouillage ;
Syndrome de dé-efférentation ;
EEG ;
Potentiels évoqués somesthésiques ;
Potentiels évoqués cognitifs ;
P300

Résumé
Introduction. — La reconnaissance d'un syndrome de dé-efférentiation (*locked-in*) dans le décours d'un traumatisme crânien peut s'avérer difficile, d'autant plus qu'il succède directement à une période comateuse et qu'il se présente sous une forme complète.
Patient et méthodes. — Un homme de 56 ans était admis dans une unité de soins intensifs avec un score de Glasgow de 4 (V1, M2, E1) des suites d'un traumatisme crânien fermé. L'examen tomodensitométrique cérébral objectivait une hémorragie sous-arachnoïdienne limitée dans la région sylvienne droite, sans œdème cérébral ni déviation du système ventriculaire. Le score de Glasgow était inchangé au sixième jour. L'électroencéphalogramme (EEG) montrait une réactivité du tracé aux stimulations auditives, de même que l'apparition paradoxale d'un rythme dans la bande alpha (10—12 c/s) dans les régions postérieures, supprimé par l'ouverture des yeux. Les composantes corticales précoces (N20—P25) des potentiels évoqués somesthésiques étaient normales sur les deux hémisphères ; les composantes moyennes étaient également clairement identifiées. La résonance magnétique cérébrale montrait une atteinte axonale diffuse dans le mésencéphale, et particulièrement une lésion stratégique touchant les deux pédoncules cérébraux. Les potentiels évoqués « cognitifs » montraient la présence des composantes N2 et P3 en réponse aux stimulations rares.
Conclusion. — Une évaluation neurophysiologique multimodale exhaustive, utilisant les tests les plus informatifs et la période d'enregistrement appropriée, devrait faire partie de l'évaluation des traumatisés crâniens, afin d'apprécier correctement la situation clinique du patient et de ne pas confondre un syndrome de dé-efférentiation (*locked-in*) avec un coma prolongé, un état végétatif, un état de conscience minimal ou un mutisme akinétique. Une évaluation neurophysiologique avant le départ des soins intensifs peut constituer un bilan de départ pour l'évaluation ultérieure des patients peu réactifs dans un centre de réadaptation fonctionnelle.

Introduction

The locked-in syndrome (LIS) was first described in 1844 by Dumas in his novel ''Le Comte de Monte-Cristo'' [7] as ''a dead body with living eyes''. It is a state in which selective supranuclear motor de-efferentation produces paralysis of all 4 limbs and the last cranial nerves without interfering with consciousness [28], even if vigilance is often fluctuating, especially in the acute state [12]. Although stroke is the most frequent cause of LIS [18], other aetiologies are known, including traumatic brain injury (TBI) [3], tumor [5], multiple sclerosis [9], pontine abscess [21], brainstem encephalitis, and central pontine myelinolysis [26]. Head trauma is the second most frequent cause of LIS [18]. Post-traumatic LIS is particularly difficult to recognize because it follows a state of coma and may show the clinical feature of ''total'' LIS, in which even eyes and lids movements are lost [18]. The main diagnostic challenge is recognition of the syndrome itself from other clinical states that can mimic LIS, as coma, vegetative state, akinetic mutism, and minimally conscious state. Keeping these pitfalls in mind, we report the case of a man with severe head trauma, in whom the multimodal neurophysiological approach, as an extension of clinical examination, allowed us early identification of a ''total'' LIS.

Case report

A 56-year old male was admitted to our hospital because of closed head injury in a car accident. His medical history did not show any significant disease except for high blood arterial pressure. When he arrived at emergency department, he was comatose (Glasgow Coma Scale [GCS] = 4), with the right pupil larger than the left, both scarcely reactive to light. The first computed tomography (CT) scan showed a limited subarachnoid haemorrhage in sylvian region without any brain oedema or ventricular shift. He was intubated, transferred to the intensive care unit (ICU) and put under mechanical ventilation. Neurosedation (midazolam 0.02 mg/kg per hour) was maintained for the first 2 days. Basing on CT scan, no intracranial pressure (ICP) monitoring was performed. Over the next 6 days GCS did not change (GCS = 4; V1, M2, E1), photomotor response was present, no facial and limb movements were evident spontaneously or in response to pain. A new CT scan did not show any new parenchymal alterations. Electroencephalogram (EEG) recorded on day 7 showed a reactivity to acoustic stimuli with the appearance of an anterior high-voltage rhythmic delta activity and a paradoxical appearance of a posterior rhythm in the alpha range (10—12 c/s), blocked by passive