Neurophysiologie Clinique/Clinical Neurophysiology (2016) 46, 119—124

Disponible en ligne sur ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

ORIGINAL ARTICLE/ARTICLE ORIGINAL

Long-term evolution of EEG in Unverricht-Lundborg disease

Évolution à long terme de l'EEG dans la maladie d'Unverricht-Lundborg

Amina Gargouri-Berrechid[a,b], Amina Nasri[a], Imen Kacem[a,b], Youssef Sidhom[a,b], Istabrak Abdelkefi[a], Yosr Hizem[a], Mouna Ben Djebrara[a,b], Riadh Gouider[a,b,*]

[a] *Service de neurologie, UR 12SP21, CHU Razi, Tunis, Tunisia*
[b] *Faculté de médecine de Tunis, université El Manar, Tunis, Tunisia*

Received 6 March 2015; accepted 28 March 2016
Available online 4 May 2016

KEYWORDS
Electroencephalography;
Epilepsy;
Long-term evolution;
Myoclonus;
Unverricht-Lundborg disease

Summary
Objectives. — To describe the EEG characteristics of patients with Unverricht-Lundborg disease (ULD) and their changes during the long-term evolution of the disease.
Methods. — A retrospective study including all patients with ULD confirmed by molecular biology and more than 15 years' duration of disease progression at the time of inclusion. EEGs were recorded at inclusion, 2 years and 5 years of follow-up. Patients who discontinued treatment during follow-up had an EEG monitoring 1 year after reintroduction of therapy.
Results. — Forty-seven EEGs were performed in 17 patients. The mean age at onset was 12.0 ± 5.5 years. The mean duration of follow-up was 26.5 ± 6.9 years. The average background rhythm was 8.2 c/s, and was normal in 30 EEGs (64%), slow in 17 (36%) and disorganized in 11 (23%). Epileptic abnormalities were found in 22 EEGs (47%). Myoclonic jerks were found in 13 EEGs (28%). After re-adaptation of antiepileptic medication in patients who had previously stopped treatment, control EEG showed a normal background rhythm with no epileptic abnormalities throughout the monitoring period.
Conclusion. — This study shows that the progressive disappearance of EEG abnormalities is rather due to antiepileptic treatment than a gradual spontaneous tendency to decrease over time.
© 2016 Elsevier Masson SAS. All rights reserved.

* Corresponding author. 1, rue des Orangers Manouba, 2010 Tunis, Tunisia. Tel.: +216 98321005; fax: +21671601300.
E-mail address: riadh.gouider@gnet.tn (R. Gouider).

http://dx.doi.org/10.1016/j.neucli.2016.03.003
0987-7053/© 2016 Elsevier Masson SAS. All rights reserved.

MOTS CLÉS
Électroencéphalographie ;
Épilepsie ;
Évolution à long terme ;
Maladie d'Unverricht-Lundborg ;
Myoclonie

Résumé
Objectifs. — Décrire les caractéristiques EEG des patients atteints de la maladie d'Unverricht-Lundborg (ULD) et leurs changements au cours de l'évolution à long terme de la maladie.
Méthodes. — Une étude rétrospective a inclus tous les patients atteints d'ULD confirmée par la biologie moléculaire et d'une durée d'évolution de plus de 15 ans. Les EEG ont été enregistrés à l'inclusion et au bout de 2 et 5 ans de suivi. Les patients qui ont interrompu le traitement ont bénéficié d'un EEG réalisé un an après la reprise du traitement.
Résultats. — Quarante-sept EEG ont été réalisés chez 17 patients. L'âge moyen de début était de 12,0 ± 5,5 ans. La durée moyenne de suivi était de 26,5 ± 6,9 ans. Le rythme de fond moyen était de 8,2 c/s, normal dans 30 cas (64 %), lent dans 17 cas (36 %) et désorganisé dans 11 cas (23 %). Des anomalies épileptiques ont été trouvées dans 22 cas (47 %) et des myoclonies dans 13 cas (28 %). Après réajustement du traitement antiépileptique chez les patients qui avaient préalablement arrêté le traitement, les examens EEG de contrôle ont montré un rythme de fond normal sans anomalies épileptiques pendant toute la période de surveillance.
Conclusion. — Cette étude montre que la disparition progressive des anomalies EEG est plutôt due au traitement antiépileptique qu'à une tendance spontanée à diminuer progressivement au cours du temps.
© 2016 Elsevier Masson SAS. Tous droits réservés.

Introduction

Unverricht-Lundborg disease (ULD), also called progressive myoclonic epilepsy type 1 (PME1), represents the purest type of progressive myoclonus epilepsy (PME). It is characterized by the presence of only few symptoms associated with epileptic seizures and myoclonus without progressive cognitive deficit [4]. It is inherited in an autosomal recessive manner. It presents during late childhood and early adolescence, ranging from 8 to 15 years, peaking at around 12 to 13 years old [7]. Diagnosis can be confirmed by the presence of an expansion or less commonly a mutation affecting the gene coding for cystatin B including ''CCCCGCCCCGCG'' dodecamer repeats localized on chromosome 21q22.3 [12]. ULD prevails in the Mediterranean basin, particularly in the Maghreb and in Scandinavian countries, especially in Finland [4]. The electroencephalogram (EEG) is a key diagnostic tool for ULD. It has been commented that the EEG background rhythm initially remains normal in the beginning of the disease with a tendency to show additional slow changes, then becoming altered and slower with disease progression [3]. Paroxysmal discharges with polyspikes and diffuse generalized spike-waves, either spontaneous or precipitated by intermittent photic stimulation (IPS), constituted an inclusion criterion in some series [6,8,11]. Hence, EEG is considered useful for diagnosis, follow-up, prognosis and treatment monitoring. Nevertheless, there have been few systematic studies on long-term EEG outcomes in ULD.

The objective of this study is to describe the characteristics of EEG in patients with ULD in a Tunisian cohort and their modifications during the long-term course of the disease.

Methods

We retrospectively analyzed all patients diagnosed with ULD, in the department of neurology of Razi hospital in Tunis, between January 2005 and January 2013. All cases were confirmed by molecular biology. All patients had at least 15 years of follow-up at the moment of inclusion.

Data concerning clinical features were collected from patients' records: age of the patient, age of onset of the disease and of each sign, type of seizures, current treatment and neurological examination findings, notably specifying cerebellar syndrome and myoclonic features. A simplified score to evaluate myoclonus (Table 1) was used to rate motor handicap [15]. The score was assessed on inclusion and at last follow-up examination.

The EEGs were recorded using surface electrodes placed on the scalp according to the international system 10—20. All patients had at least one interictal waking EEG during at least 20 minutes.

All the records comprised two hyperventilation periods of 5 minutes each and one IPS using flash frequencies varying from 1 to 60 Hz. Each frequency was carried out for 10 seconds followed by a 10-second break with eyes opening and closing. Electromyogram (EMG) was combined with EEG records in all patients. EMG electrodes were placed on the affected muscular group in cases of myoclonus and on the deltoid muscle in their absence. All EEGs were examined specifying background frequency and organization, occurrence of spikes, polyspikes or polyspike-waves, presence of myoclonus on EMG and response to stimulation tests.

EEGs were recorded on inclusion, then at 2 and 5 years of follow-up. Patients who had stopped their antiepileptic medication during the follow-up period underwent an EEG follow-up study 1 year after restarting treatment. The steady state of the disease is defined by the occurrence of at least one seizure per year and a myoclonic score inferior or equal to 2. We divided our study population into two groups: The first group (group 1) comprising patients who had EEG during the period of activity of the disease; and the second group (group 2) comprising patients who had EEG during the steady state of the disease.

Percentages were compared with Chi^2 test and Fisher's exact test. Differences were considered statistically

Download English Version:

https://daneshyari.com/en/article/3082003

Download Persian Version:

https://daneshyari.com/article/3082003

Daneshyari.com