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The value of video-EEG monitoring to diagnose juvenile myoclonic epilepsy

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ABSTRACT

Objective: A diagnostic accuracy of conventional electroencephalography (EEG) is approximately 50% at best. We aimed to determine the accuracy of video-EEG monitoring (VEM) for a correct diagnosis and the feasibility of its clinical application. The data from all 55 patients (M:F = 31:24) with juvenile myoclonic epilepsy (JME) who underwent VEM were reviewed according to the clinical history, brain imaging and video-EEG findings.

Results: Age at seizure onset ranged from 10 to 25 (15.5 ± 2.7 years). The age at VEM ranged from 15 to 46 (21.8 ± 5.8 years) and 57% (29/51) showed seizures. Of those, 20 patients (69%) showed myoclonic jerks alone, whereas 3 (10%) showed generalized seizures alone. Both of these conditions were observed in 6 patients (21%). Interictal abnormalities alone without clinical seizures were detected in 16 patients (31%). Atypical semiologies such as asymmetric myoclonus or versive seizures were observed in 18 patients (35%) during video monitoring. Interestingly three patients complained of visual aura on history. The duration of VEM ranged from 1 to 6 days (1.8 ± 1.1). Overall, 88% of patients showed an EEG abnormality with/without seizure, concordant with JME. Among 10 patients with a normal conventional EEG before VEM, 9 showed interictal or ictal EEG abnormalities during approximately 1-day of VEM.

Conclusions: VEM for 1 or 2 days is appropriate for making a correct diagnosis of JME, especially in patients having an atypical semiology and a normal result on the conventional EEG.

1. Introduction

Juvenile myoclonic epilepsy (JME) is one of the idiopathic generalized epilepsy syndromes, classically characterized by three semiological features: (1) morning myoclonic jerks in both arms, (2) a brief interruption of consciousness and (3) generalized tonic–clonic seizures. The clinical diagnosis is supported by compatible electroencephalography (EEG) changes, which show generalized polyspike-and-wave patterns without background slow waves, normal brain imaging and a normal cognitive function.[1] Although JME is relatively common and is clearly defined (as listed above),[2–4] it is commonly mis- or under-diagnosed due to the strong reliance on these "typical" three semiologies and EEG results. Simply based on history from patients alone at outpatient clinic, we may regard myoclonic jerking in only one limb[5] and brief unresponsiveness[6] as focal motor and complex partial seizure, respectively. Depending on result of EEG alone to diagnose JME may be troublesome. It is well known that an entirely normal EEG as well as focal or lateralized epileptiform discharges[7–10] can make a diagnosis difficult. Several studies[1,5,11–13] have suggested that even repetitive EEGs do not reveal the suggestive abnormalities of generalized epilepsy in 21–54% of patients. Moreover, accurate diagnosis can be more difficult when we obtain an unreliable or atypical seizure history alone.

Video-EEG monitoring (VEM) system is widely applied to describe the seizure semiology and localization of the seizure focus. In patients with JME, it is held that VEM can demonstrate myoclonic jerks and absence seizures, which might go unrecognized by a patient or doctor; it can also be applicable to detect the typical EEG findings of patients with JME more easily. There have been some observations[1,5,14] that VEM is often helpful to clarify an atypical semiology and electrographical findings, but to date the literature regarding the diagnostic value of VEM in patients with JME has been scarce. Previous studies did not include cases that did not demonstrate clinical seizures and placed more emphasis on ictal recording and semiology. The aim of this study was to determine the feasibility of VEM for making an early and correct diagnosis of JME and if there is a patient subset for which VEM is better indicated.

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2. Materials and methods

VEM was performed on 1935 patients in our center from January 1995 to April 2006. Among them, 55 patients were diagnosed with JME during the entire follow-up period and we included all such patients who underwent VEM in this study. There were 31 men and 24 women, whose ages ranged from 15 to 46 years at VEM (mean 21.8 ± 5.8 years). A diagnosis of JME was made according to the criteria of International League Against Epilepsy.[15] The diagnosis was confirmed from each patient's historical or videotaped semiology, ictal or interictal EEGs, brain imaging and drug response to antiepileptic drugs (AEDs) during the follow-up period. The patients' medical records were reviewed regarding their history, and two epileptologists analyzed the video-EEGs until they reached a consensus. The typical EEG changes compatible with a diagnosis of JME included: (1) a symmetrical generalized polyspike-and-wave, (2) a symmetrical generalized isolated spike-and-wave, and (3) symmetrical generalized spike-and-wave complexes at 3 Hz or over 3 Hz (fast type).[13] Atypical findings such as irregular bursts of slow waves without a discernible generalized epileptiform discharge were not considered to be a characteristic EEG feature of JME.

2.1. Conventional EEG at outpatient clinic

Conventional EEGs (c-EEGs) were performed according to the international 10–20 system at the EEG laboratory in the same hospital without any modification of the patient's AED. Each EEG session lasted for 30 min and any EEG changes occurring during hyperventilation and photic stimulation were also observed.

2.2. Brain imaging

Magnetic resonance imaging (MRI) was performed on 31 patients, and computer tomography (CT) was performed on one. The standard MRI technique was as described.[16] In some cases, 3-mm thick sections of the tentative symptomatogenic regions were also obtained considering the patient's c-EEG and semiology.

2.3. Video-EEG monitoring

In our center, 458 patients were diagnosed as JME from January 1995 to April 2006, of whom 55 patients (12%) performed VEM. Interictal and ictal EEGs and clinical seizures were recorded using a VEM system with the electrodes placed according to the international 10–20 system, including the additional anterior temporal electrodes. VEM was performed for the following reasons: (1) to confirm a diagnosis of JME in patients with drug resistance or a discrepancy between their clinical diagnosis and the c-EEG results performed at an outpatient clinic; (2) to clarify the epilepsy syndrome of patients referred from other centers with an equivocal classification; (3) to identify any vague semiology; or (4) for the first diagnosis of any untreated patient.

VEM was performed after withdrawing the AEDs with the exception of phenobarbital. The EEGs were reviewed using the bipolar and referential montages. All patients were routinely advised not to go to sleep earlier than usual and additional provocations such as photic stimulation or hyperventilation were administered in the morning only to those patients not showing ictal or interictal epileptiform discharges during the previous night. In general, VEM was stopped when the typical EEG pattern of generalized epilepsy or seizures compatible with a diagnosis JME occurred. Occasionally, the length of monitoring was extended when there were inconsistent findings.

2.4. Seizure outcome

All patients included in our study had follow-up duration of at least 1 year after VEM. Their seizure outcomes were measured at last follow-up.

2.5. Statistical analysis

One-way ANOVA was used to examine the statistical significance of any difference between the continuous variables using SPSS for Windows (Version 12.0).

3. Results

Table 1 summarized the patient's clinical data and VEM results. The age of seizure onset ranged from 10 to 25 years (mean 15.5 ± 2.7) and the mean duration between seizure onset and VEM was 6.2 ± 5.8 years. The mean follow-up duration since VEM ranged 12–197 months (mean 57.3 ± 47.2).

3.1. Seizure history

The prior seizure history was taken from a witness or the patients themselves. Among the 55 patients, 41 (75%) had suffered both myoclonic jerks and generalized seizures. Of these, 11 (20%) experienced all three types of seizures, myoclonic jerks, generalized seizure and brief loss of consciousness; two (4%) had had myoclonic jerks plus brief losses of consciousness and one patient (2%) had had only myoclonic jerks.

Table 1
Summary of clinical characteristics and video-EEG results

	No. (%)	Median VEM duration (day) (range)
Seizure history		
M + G	41 (75)	1 (1–6)
M + G + A	11 (20)	2 (1–4)
M + A	2 (4)	2 (2)
Only M	1 (2)	4 (4)
Initial impression before VEM		
JME	36 (65)	1 (1–3)
JME plus	12 (22)	2 (1–6)
Others	7 (13)	3 (1–4)
Lesion on MRI		
Yes	6 (19)	2 (1–6)
No	26[a] (81)	1 (1–5)
Monitored seizures		
Typical[b]	11 (22)	1 (1–5)
Atypical	18 (35)	2 (1–4)
No seizure	22 (43)	1 (1–6)
Interictal EEG abnormality		
Typical[c]	43 (84)	1 (1–5)
Atypical	4 (8)	1.5 (1–3)
Normal	4 (8)	1 (1–6)
Ictal EEG abnormality		
Typical[c]	25 (83)	1 (1–4)
Atypical	5 (17)	2 (1–5)

VEM, video-EEG monitoring; M, myoclonic jerk; G, generalized seizure; A, absence seizure.
[a] Includes one patient undergoing CT.
[b] Denotes a symmetrically involved myoclonic jerk or generalized seizure.
[c] Denotes a symmetrical generalized polyspike-and-wave, a symmetrical generalized isolated spike-and-wave or symmetrical generalized spike-and-wave complexes at 3 Hz or over 3 Hz.