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Juvenile Myoclonic Epilepsy among Sudanese Patients: Prevalence and EEG Characterizations

Abstract

Introduction: Juvenile myoclonic epilepsy (JME) is well recognised, age-related electroclinical, generalised, epileptic syndrome. JME exceeds the 5% of all epilepsies and around 20% of all idiopathic generalised epilepsies. The classical presentations consist of myoclonic jerks (MJs), occur early in the morning and precipitated by sleep deprivation. JME is well documented under diagnosed.

Aims: The aims were to identify the prevalence and EEGs characteristics among Sudanese patients with JME.

Methods and Patients: A cross-sectional clinical based study that consists of EEGs recording and history reviewing of the all epileptic patients presented to EEG unit of the National Ribat University, and El magzoub neurosciences centre, from March 2003 to May 2012.

Results: Forty-four patients with JME have been diagnosed, with an overall prevalence of 2.13%, based on EEG finding of generalised 3.5-6 Hz single, bifid and polyspikes - slow wave complexes on normal brain background activity. The mean age of JME patients at diagnosis was 19.55 ± 8.98 years; the majority of patients (75%) were between 10 and 30 years. Myoclonic jerks were detected in 90.9% of the patients with a mean age of onset 10.48 ± 4.81 years. Absence attacks were confirmed in 77.27% of JME patients, and generalised tonic-clonic seizures in 84.1% of patients with a mean age of onset 13.92 ± 5.65 years, convulsions were detected in 77.3% of all patients experienced GTCs.

Conclusion: JME among Sudanese epileptic patients has a low prevalence, thus a high rate of under diagnosis. The most important misdiagnosis pitfalls are the low awareness among doctors, who fail to ask about MJs, underestimation of the MJs by parents, who do not seek advice before the occurrence of fits, and the empirical anti-convulsion drugs that may blackout the MJs. EEGs asymmetries led to missing interpretation and misdiagnosis.

Keywords: JME; MJs; EEGs

Research Article

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Abbreviations: AEDs: Anti-Epileptic Drugs; EEGs: Encephalography; GTCS: Generalised Tonic-Clonic Seizures; ILAE: International League Against Epilepsy; JME: Juvenile Myoclonic Epilepsy; Mjs: Myoclonic Jerks; PSW: Poly Spikes-Wave; SW: Spike-Wave

Introduction

JME is a syndrome that appears around puberty and is characterized by seizures with bilateral, single or repetitive, arrhythmic, irregular myoclonic jerks, predominantly in the arms. Some of the patients may fall due to jerks, but usually not associated with loss of consciousness. Often, there are generalized tonic - clonic seizures (GTCS) and less frequent absences. The seizures usually occur shortly after awakening, and are often precipitated by sleep deprivation [1]. The prevalence of JME estimated to be 5 %- 10 % of all epilepsies, and 18% of idiopathic generalized epilepsy [2] similar to Obeid (10.7%) [3], but many studies revealed lower prevalence, particularly those conducted in specialized clinics as it was 4.1 by Genton et al. [4]. Females have been found to outnumbered males with JME [5]. Typically the JME appears in the second decade, with a peak between 12 and 18 years [6]. JME is an electroclinical epileptic syndrome thus its diagnosis should be supported by clinical and electrophysiological findings. generally there are A large number of experts agree on two sets of diagnostic criteria , class I and class II, both agreed on jerks that aggravated by sleep deprivation or /and stress [7]. The major EEG features in JME are short discharges of generalized spike-wave (SW) or poly spikes-wave (PSW) complexes associated with epileptic myoclonic [8]. During a myoclonic seizure, EEG shows irregular 3 to 4 Hz polyspikes-waves with front central predominance. The EEGs of Seventy four to eighty-one percent of patients with JME displayed 4 to 6 Hz bilateral polyspikes and slow wave discharges with frontal predominance over a normal background activity [3,8].

The diagnosis of JME is often missed at the onset of seizures [9]. In a series studies the mean delay between the onset of symptoms and correct diagnosis was close to 8 years: the only diagnosis proposed by the referring physician was JME in 22 (16%) patients

only [4], decreased to 3.3% in Indian study , with mean delay of diagnosis reaches 8.6±7.0 years [10]. To the best of our knowledge, there are no concrete epidemiological studies for JME among Sudanese epileptic patients, despite the common agreement among the Sudanese neurologists and neurophysiologists about the respectful number of patients with JME, they deal routinely within their clinics and EEG centers. Therefore our major aim is to detect the prevalence of JME and correlate the EEGs findings with clinical presentation among Sudanese patients.

Patients and Methods

Based on the most frequent patterns of EEGs and clinical features, 44 patients with JME were assembled from 3523 patients, presented to EEG unit of the National Ribat University, and El magzoub neurosciences centre, from March 2003 to May 2012. Any patient who had diagnosed as JME have been recruited and reviewed personally or with informants through a preformed questionnaire and the EEGs have been repeated for all of them. The EEG machine we used to record was the Medtronic machine. The EEGs traces were interpreted by an expert clinical neurophysiologist and neurologist. Objectives of the study have been explained for all patients through a phone call and informed consents have been signed by patients or their legally authorized persons.

Results

Based on the characteristic EEGs changes in JME (the generalised 3.5 - 6 Hz single, bifid and polyspike-slow wave complexes) [3,8], we found that the prevalence of JME among abnormal EEGs was found to be 44 patients (2.13%). Associated asymmetries of EEGs localizations and / or patterns have been observed in 45.5% of JME patients. Gender distribution revealed moderate female predominance (56.81%), while the male was 43.2%. The mean age of JME patients at diagnosis was 19.55 ± 8.98 years; the majority of patients (75%) were between 10 and 30 years. Myoclonic jerks were detected in 90.9% of the patients with a mean age of onset 10.48 ± 4.81 years. As far as time is concerned, jerks occurred during early morning and in the day time (59.1%, 50%). Sleep deprivation appears to be the major trigger for MJs in 61.4% of the patients, other important triggering factors distributed as follow; emotional stress accounted 50% of the causes whereas, the early morning awakening was 36.4%. Absence attacks were confirmed in 77.27% of JME patients, and generalised tonic-clonic seizures in 84.1% of patients with a mean age of onset 13.92 ± 5.65 years, convulsions were detected in 77.3% of all patients experienced GTCs. Only five doctors (11.4%), from all doctors who referred their patients for EEG test, included the JME among their differential diagnoses. Thirty-eight patients (86.4%) were delivered normally and 6 patients (13.6%) by caesarian sections, those who showed the positive history of febrile convulsions were 27.3%. Sleep disturbance was found in 40.9 %.Memory capacities declined in 79.5% and school or work performance deteriorated in 86.4% of JME patients.

Discussion

The present JME prevalence result among epilepsy patients in Sudan is 2.13%. A similar estimate was determined by the previous work of Fong group [11], but a higher estimate was obtained by Ali [12]. This low prevalence can be explained in the light of the report of Bureau [13], who found that the patients with JME are easily controlled, thus most of them will not be referred to the neurologic centers, and they will be lost without the precise diagnosis. Inconsistent with the result obtained by Murthy et al. [14], we found that 77.3% of patients presented with the classic triad symptoms at the time of the diagnosis; he found it only in 17.5% of patients [14]. Ninety present of Sudanese JME had MJs lesser than Ali et al. [12] observation [12], the four patients (10%) who denied the occurrence of MJs, were already on AEDs, at the time of referral, that may obscure their jerks, as Panyiotobolous & Loncman [15,16] stated in two different studies [15,16]. Myoclonic jerks in Sudanese patients triggered by sleep deprivation in 61.4% and emotional stress in 52.3%, partially accorded with da Silva et al. [17] results. Photic stimulation and television watching were known stimulators of MJs in 13.5% of Sudanese patients, consistent with the observation of da Saliva et al. [17]. All our patients denied alcohol consumption, although Janz & da Silva [17] included it among the major triggers (Figure 1 & 2).

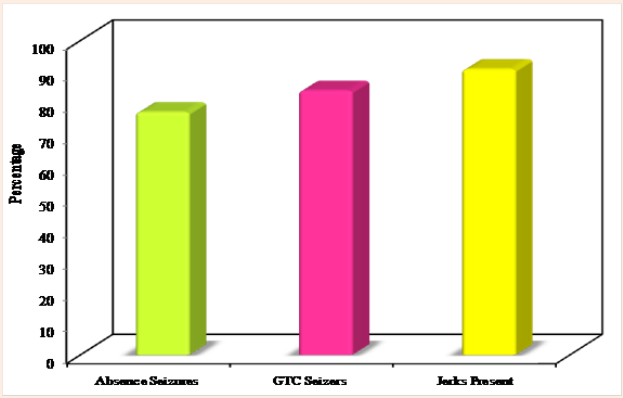

Figure 1: Frequency of Myoclonic Jerks, GTC and Absences.

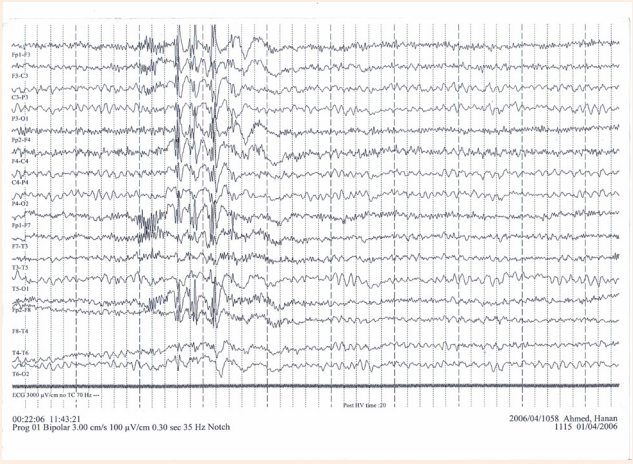

Figure 2: Generalized 3.5-6 Hz single, bifid and polyspikes – slow wave complexes on normal brain background activity.

All patients with JME showed the characteristic generalized 3.5 - 6 Hz single, bifid and polyspike-slow wave complexes, on normal background activity. EEG asymmetries have detected in 45.5% of patients in present study, higher than Montaleni [18] (38.1%) and lower than Letourneau [19] (53.8%) [18,19]. The EEG records of 18.8% of our patients showed focal discharges, accorded with Murthy results [14]. These EEG asymmetries, particularly the focal discharges are not uncommon in JME patients and many investigators accused as the major pitfall of misdiagnosis and may be the same explanation of low prevalence of JME among Sudanese patients. Only five doctors (11.4%) who referred their patients to our centres were labelled their referring report with JME diagnosis, something between Genton & Vijai [9,10] results but the mean delay of diagnosis of all patients was 4.35 years, in partial accordance with the both studies [9,10]. It is apparently that the poor awareness about JME syndrome among doctors had an immense role in JME misdiagnosis, reflected in the form of the low prevalence and long means delay time. This premise is confirmed by the success of increasing Spanish doctors awareness in decreasing the mean delay of JME diagnosis from 10.6 years in 1994 [20] to 2.4 years in 2001 [21]. The females represent 56.8% of Sudanese JME patients, against the declaration of ILAE (1985) [22], that states equal male to female ratio, but this female predominance has been accorded with most recent observations by Camfield & Ali et al. [5,12]. An 88.6% Sudanese of patients with JME were diagnosed in their second decade of life, compatibles with the conclusion reached by Ali et al. [12] and the classic characterization of Janz [23]. The patients in the current study have wide age range (5- 55 years), it is in line with Panyiotobolous [15] results (2 - 40 years) [15], but the minimum and maximum ages were higher in Sudanese patients, embodies more delay of diagnosis in Sudanese patients (Table 1-3).

Table 1: The Prevalence of JME among Patients with Abnormal EEGs.

Abnormal EEGs	No.	%
Juvenile myoclonic epilepsy (JME)	44	2.13
Patients of abnormal EEGs do not include JME patients	2019	97.87

Table 2: The EEGs Findings in patients with JME.

Patients Number (2063)	EEG Findings
44	Generalized 3.5-6 Hz single, bifid and polyspike – slow wave complexes on normal cortical background activity

Table 3: Age at Diagnosis of JME.

Age in Years	Number of Patients	Percentage
<10	5	11.4
10 - <20	22	50
20 - <30	11	25
≥ 30	6	13.6
Min. – Max.	5.0 – 55.0	
Mean ± SD.	19.55 ± 8.98	
Median	18	

Conclusion

It appears that there is no difference between Sudanese patients with JME and the international patients regarding the common EEGs findings.

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