Epilepsy of Janz in India: Is it different?

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Objective: Due to certain interesting characteristics, the syndrome of Juvenile Myoclonic Epilepsy (JME) of Janz has attracted plenty of attention. It is important to identify JME early, since it has a favorable prognosis. On the contrary, if the diagnosis is delayed, there may be uncontrolled epilepsy, repeated head injuries, polytherapy and psychosocial trauma. We share our experience about JME in India and its variation from reported observations.

Methods: We selected 108 cases of JME after screening 708 patients of generalized seizures, in an age group of 8-32 years. Diagnosis was established using standard clinical and EEG criteria. Patients who responded satisfactorily (>50% reduction in seizure frequency) with any antiepileptic agent besides sodium valproate (VPA) were advised to continue them as such. If seizures persisted, then substitution by VPA in a dose of 20-40 mg/kg bodyweight was done. Clonazepam was added in a dose of 0.01-0.10 mg/kg bodyweight, only if patients were not seizure free even with maximum dose of VPA (50 mg/kg). Cognitive assessment was done using Mini-Mental Status Examination and it was repeated twice at intervals of 4 months. All subjects were followed up till they were seizures free for a minimum period of 1.5 years. Total duration of study was 6.5 years.

Results: Out of 108 cases of JME, only 5 (5%) patients were referred with this diagnosis, and most of them by neurologists. Majority of them, 62 patients (57%) were referred as intractable seizures. There were 6 patients (6%), referred as subacute sclerosing panencephalitis and 11(10%) as progressive myoclonic epilepsy. A significant number of patients, 24 (22%) presented directly to us, without consulting any physician or neurologist, who later were diagnosed as JME. Lack of clinical suspicion and non-use of activation procedures during EEG recordings were important reasons for diagnostic delay. There were 66% patients who were on polytherapy of phenobarbitone, phenytoin, and carbamazepine, before reporting to our hospital.

We observed many atypical features of JME in our study. They were: (a) The onset of seizures was before 10 years and after 22 years in 24 cases (22%), while in remaining patients it was between 12-18 years; (b) A negative family history in 88 cases (82%); (c) A significant delay in the diagnosis, as 90 patients (83%) presented after more than 2 years of onset of seizures; (d) mild cognitive impairment in 30 patients (28%), and (e) Eight out of 12 patients on phenytoin were seizure free.

Conclusion: Clinical spectrum of JME is different in India. It is grossly underdiagnosed resulting in uncontrolled epilepsy, repeated head injuries, polytherapy and psychosocial trauma, which explain the cognitive decline in them. We suggest that JME should be strongly suspected in juvenile patients of generalized epilepsy not responding to treatment. Activation procedures especially sleep deprivation should be routinely used during EEG recording, before labeling them as intractable epilepsy.

References

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