Short Communication

Idiopathic paroxysmal kinesigenic dyskinesia in Malaysia, a multi-racial Southeast Asian country

Mei-Ling Sharon Tai *, Shen-Yang Lim, Chong Tin Tan
Division of Neurology, Department of Medicine, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia

ABSTRACT

Paroxysmal kinesigenic dyskinesia is a rare disorder, and there are few reports of Asian patients with this condition. We reviewed the clinical features of all patients with idiopathic paroxysmal kinesigenic dyskinesia (PKD) seen at a major neurological centre in Malaysia. The charts of 11 patients with idiopathic PKD seen between 1995 and 2008 were reviewed retrospectively. The male:female ratio was 9:2. Ten patients were of Chinese ethnicity, and one was Malay. Three patients (from two families) had a family history of PKD. The involuntary movement was dystonia in 73% of patients. In one patient, attacks were precipitated by vestibular stimulation. One patient had generalized epilepsy. Another patient who did not have epilepsy demonstrated epileptiform discharges. Only slightly over one-quarter of patients had a positive family history. Males, and people of Chinese ancestry, seem to be affected more frequently by PKD in certain Asian populations.

© 2010 Elsevier Ltd. All rights reserved.

1. Introduction

Paroxysmal kinesigenic dyskinesia (PKD) is a rare disorder that typically involves frequent, brief episodes of dyskinesias that are triggered by sudden movement. The condition can be sporadic or familial (autosomal dominant inheritance with incomplete penetrance). Diagnostic criteria for idiopathic PKD have been proposed. We analysed the clinical features of patients with idiopathic PKD seen at a major neurological centre in Malaysia, a multi-racial Southeast Asian country.

2. Methods

We conducted a retrospective review of the medical charts of 11 patients with idiopathic PKD seen between 1995 and 2008 at the outpatient neurology clinic of the University of Malaya Medical Centre, Kuala Lumpur. We had no patients with symptomatic PKD. The diagnosis of PKD was confirmed, however, using the criteria proposed by Bruno et al.

3. Results

Ten patients were of Chinese, and one was Malay, ethnicity. There were no Indian patients. Nine patients were male, two female. The mean age of onset was 13.5 years (±standard deviation [SD] 4.5 years; range: 8–21 years). All our patients had their attacks triggered by sudden movement. In four patients, startle was also a trigger. In one patient, attacks were precipitated by vestibular stimulation; this patient has been reported in detail elsewhere. Episodes lasted less than 5 minutes. Emotional stress was reported to lower the threshold for attacks in two patients. No patient reported a sensory aura (the absence of this symptom was specifically documented in eight patients). One patient also had generalised epilepsy, 14 years after the onset of PKD. Three patients (27%) had a family history of at least one family member being similarly affected by PKD. Two patients were from the same family (mother and son). The mother’s brother had PKD and later developed motor neuron disease, and his two children also had PKD; however, the latter three individuals were not seen by us. The Malay patient had a son with PKD, who was not seen by us; there were no other people with PKD in this family.

The electroencephalogram (EEG) of one patient (without epilepsy) showed frequent generalised (frontal-predominant) bursts of slow waves and sharp waves. The EEG was normal for the other patients, including the patient with clinical seizures. All other investigations were normal, including brain imaging, which was performed in four patients. Seven patients responded well to pheynytoin (100–400 mg daily) or carbamazepine (200–400 mg daily). One patient improved while taking sodium valproate, and another partially responded to levodopa. Most patients (6/11) experienced complete remission of symptoms. Three other patients improved, but two patients showed no change over follow-up (one of whom was non-compliant with treatment).

* Corresponding author. Tel.: +60 379492742; fax: +60 379494613. E-mail address: sharon@ummc.edu.my (M.-L.S. Tai).
4. Discussion

One major finding of this study is the preponderance of Chinese patients with idiopathic PKD (91% of patients). Malaysia has a multi-racial population, comprising three major groups: (i) Malays (approximately 65%), Chinese (26%), and Indians (8%). Patients attending our neurology clinic, from which this series of patients with PKD was derived, are of the following racial distribution (based on an audit conducted by M.-L.S. Tai in May 2009): 53% Chinese, 26% Malay, and 21% Indian, reflecting a tendency for ethnic Chinese to live primarily in urban areas, such as Kuala Lumpur. A series from Singapore, another multi-racial Southeast Asian country, reported a similar racial distribution of patients with idiopathic PKD (Chinese:Malay:Indian = 13:1:1; that is, Chinese patients accounting for 87% of patients). We also found a male preponderance (9:2), similar to that seen in the Singaporean series (14:1). However, other investigators found the male preponderance to be less pronounced, including Jung et al. (reporting on Taiwanese–Chinese patients) and Zhou et al. (mainland Chinese population) (male:female ratios of 2.3–2.5:1). Furthermore, only 27% of our patients with idiopathic PKD had a similar family history, comparable again to the findings of Tan et al. (positive family history in 21% of patients), but lower than the rates typically reported in the world literature (with most patients having a positive family history).

The clinical characteristics of Chinese patients were previously reported to be “essentially similar” to those of patients reported elsewhere. Our findings were also typical of those described in the literature. Interestingly, one patient had both PKD and epilepsy, and another had an abnormal EEG but without a history of clinical seizures. Two patients (13.3%) in the series by Tan et al. had PKD and epilepsy; other studies have also reported on a link between PKD and epilepsy.

In conclusion, we found a preponderance of patients who were Chinese and male, and most patients did not report a family history of PKD. These findings support previous observations in a similar patient population. The underlying reasons for these differences, compared to the literature from other populations, deserve further study.

References