Seizure 2000; **9**: 344–346
doi: 10.1053/seiz.2000.0402, available online at http://www.idealibrary.com on IDEAL

Familial adult onset myoclonic epilepsy associated with migraine

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We report a new type of migraine associated epileptic syndrome in a family: adult onset myoclonic epilepsy with benign course and migraine. Affected members of the family had myoclonic and rare generalised tonic–clonic seizures. Most of the patients, but not all, had a history of migraine. Also, some cases of the family had only migraine. This family will be discussed because of two distinct features. Firstly, in this family a different type of epilepsy, adult onset myoclonic epilepsy was diagnosed that has not been classified in the ILAE 1989, classification(s), but was similar to that previously reported in Japanese families. Secondly, in most of the cases migraine was associated with the epilepsy.

Key words: benign, epilepsy, familial, migraine, myoclonic.

INTRODUCTION

Epilepsy and migraine are common disorders with a wide range of clinical phenotypes. Although an association between these disorders has been suggested, the exact relationship between them is not clear[1,2]. Here we report a new type of migraine associated epileptic syndrome in a family, which is characterised by adult onset myoclonic epilepsy (with a benign course), and migraine.

REPORT OF THE INDEX CASE

A fifty-year-old woman was first seen by one of the authors (S.S) because of myoclonic jerks and unilateral throbbing headache in 1996. She had started having myoclonic jerks at the end of her third decade. Generalised myoclonic jerks had been occurring once or twice a day, usually when resting, especially whilst awake or just before sleep. Myoclonic jerks could be provoked by photic stimulation and clustered during headache attacks. She has had only one generalised tonic–clonic seizure, 20 years ago. Headache attacks have been quite frequent in the past, occurring 6–7 times in a month, and usually accompanied by photosensitivity, nausea, vomiting and visual blurring. She was originally given carbamazepine, later valproic acid (VPA) in a dose of 1000 mgm/day. Myoclonic jerks and migraine attacks lessened in frequency with VPA therapy. Her other previous history was unremarkable.

In her family history, her mother, two elder sisters, uncle and grandfather have suffered from epileptic and headache attacks. There were also pure epileptic and pure migraine cases in the family. There are members of the family who are currently too young to have developed myoclonus which, in this family, seems to start quite late (Fig. 1). The presence of both epileptic and migraine attacks was ascertained in the medical records of the mother and one elder sister. Their myoclonic jerks started in the third or fourth decades also, and their electroencephalograms (EEGs) have shown generalised spikes and multiple spike- and wave complexes. They reported more frequent generalised tonic–clonic seizures. Data for the other affected members of the family were obtained from the patient and her daughter who was well educated and who also suffered from severe migraine attacks. The EEGs of two migraine only cases were normal (patient's daughter and grandson).

The original patient's physical examination was normal except postural tremors on both hands. Laboratory examination showed normal blood count and biochemical analysis. EEG was remarkable with 5–6 Hz rudimentary spike and wave activity in occipital areas induced by hyperventilation and photic stimulation when she was on VPA therapy. Generalised spike

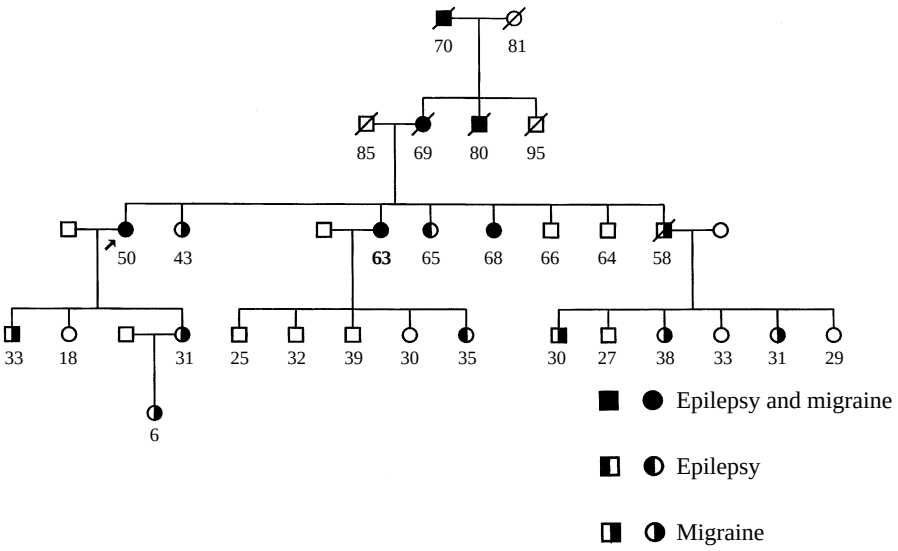

Fig. 1: Pedigree of the family. The arrow shows the index case. Numbers represent ages of the family members. Mother, two sisters, uncle and grand father of the index case had myoclonic epilepsy and migraine. There are also pure epileptic and pure migraine cases in the family.

and multiple spike wave complexes induced by photic stimulation had been reported in some of her previous EEGs. Brain MRI was normal.

She has continued on VPA therapy plus some anti-migraines drugs for the past 3 years. She still reports rare myoclonic jerks: headache attacks occurs at least once a month.

DISCUSSION

The reported epileptic syndrome here shows some different characteristics from the other epileptic syndromes classified by the ILAE[3]. It most closely resembles juvenile myoclonic epilepsy (seizure types, EEG findings, familial occurrence, and benign course) but onset is in adulthood. There is no association with dementia or cerebellar disorder, distinguishing this condition from the progressive myoclonic epilepsies. Benign Adult Familial Myoclonic Epilepsy was previously reported in a few Japanese families[4–6], and the gene locus was assigned to chromosome 8q23.3-q24.11[7]. The characteristics of our patients are very similar to these other Japanese cases: adult onset myoclonus, tremor, non-progressive course, very rare generalised tonic–clonic seizures, autosomal dominant trait, electroencephalographic findings of polyspike and waves and photosensitivity. But there were no migraine attacks reported in these other Japanese families. In our family, some members also suffered from migraine plus epilepsy or just had migraine attacks.

Although migraine and epilepsy are distinct in their pathogenesis and in their genetic basic, both these common disorders share some similarities. Both are episodic in nature and have manifestations suggesting membrane hyperexcitability. In both disorders multifactorial genetic transmission is proposed[8]. The two conditions may coexist or may interrelate in a number of ways. A Genetically determined dysfunction of the occipital lobes, which could lead to both migraine and epilepsy, has been previously proposed[9]. Also there is neurophysiological evidence confirming that the excitability threshold of the occipital cortex in migraine suffers is low compared to normal, thus strongly indicating that the neurones of the occipital cortex can be hyperexcitable in this condition[10]. Interestingly, electrophysiological investigations in the cases with Benign Adult Familial Myoclonic Epilepsy revealed polyspikes in the EEG, giant waves in somatosensory evoked potentials, enhanced long-loop C reflexes and a preceding wave on jerk-locked back averaging of the EEG, suggesting that the myoclonus originated in the cerebral cortex[6]. The cases presented here may give further evidence for this proposed genetic dysfunction. We believe that further genetic studies in these cases will make clear whether this association of epileptic seizures and migraine attacks is just coexistence or whether they occur due to the same genetic defect.

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