Atypical petit mal epilepsy in twins

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Atypical petit mal epilepsy with the clinical and electroencephalog- 
graphic findings in a child; with EEG changes alone in the identical twin 
are discussed.

Petit mal epilepsy has relative incidence of 3% and the peak age of onset 
is 5 years. It has been reported in identical twins by Lennox. The present 
communication describes atypical petit mal epilepsy, in one of the identical 
twin sisters, with characteristic EEG changes alone in the other.

Report of case

Patient, M., 10-year-old Hindu girl 
was admitted to the Neurology Depart-
ment with 2 years history of transient 
attacks of unawareness and smacking 
movements of the lips as if chewing some-
thing. At times she dropped things which 
she was holding in the hands. The dura-
tion of each attack was 20 to 25 seconds 
and the frequency 15 to 20 times a day. 
She never had tonic-clonic convulsions, 
tongue bite, fall, micturition or defeca-
tion during the attack. There was no 
history of febrile convulsions, head 
injury or symptoms of raised intracranial 
tension.

Patient was first-born of a normal 
twin delivery, antenatal history being 
insignificant. Developmental milestones 
were achieved normally. She was studying 
in class V. Intelligence quotient was 
110. There was no history of consanguini-
ity, syphilis, diabetes, epilepsy or 
migraine in the family. Two brothers aged 
14 years and 8 years respectively, are 
normal and healthy.

Physical examination was within 
limits. Fundus oculi were normal. 
Hyperventilation for two minutes repro-
duced the seizure pattern described above.

EEG showed normal alpha activity. 
Hyperventilation produced a generalised 
burst of typical 3 cps spike and slow 
wave pattern (Fig.1) suggestive of 
petit mal epilepsy. Photic stimulation did 
not show any abnormality.

The patient's twin sister had identi-
cal morphological features and blood 
groups suggestive of monovular twinning. 
She had no history of epilepsy. Detailed 
examination revealed no positive finding. 
Hyperventilation for 2 minutes did not 
produce a fit. Her EEG revealed normal 
alpha activity. Hyperventilation produced 
a generalised burst of 3 cps spike and
Fig. 1: Eeg of the patient showing typical 3 cps spike and slow wave pattern.

slow wave discharge lasting for 2 seconds initially preceded by polyspikes (Fig. 2). The EEG of both brothers and the parents were normal and did not reveal any seizure discharge.

Discussion

The present case had certain atypical features eg. smacking of lips, duration attack being 20-25 seconds suggestive of atypical petit mal epilepsy or complex absence seizures. The EEG of the unaffected identical twin was abnormal although not exactly identical to the other sister. The atypical manifestations of petit mal epilepsy are not common, and their presence in identical twins as reported here is still rarer.

Twin-pair data indicate a strong inheritable predisposition for epilepsy. In the cryptogenic variety there is about 85% concordance for epilepsy in the monozygotic twins (versus 5% for the dizygotic twins) while in symptomatic type there is 14% concordance among monozygotic twins (versus 0% for dizygotic twins). Genetic analysis of the clinical and electroencephalographic aspects of petit mal epilepsy has shown a significantly higher (13%) than normal (2.5%) incidence of convulsions in the parents and siblings of centrencephalic epileptics. The centrencephalic type of EEG is the expression of an autosomal dominant gene with the highest penetration between the age of 4 to 16 years and 18 months to 6 years.

Inheritance is not the only possible explanation for the combination of familial aggregation and high concordance among monozygotic twins. A familial tendency to an epileptogenic trauma occurring before or during birth might also be held responsible for it.