Complex Partial Seizures Evolving into Periodic Spasms

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ABSTRACT. A case of complex partial seizures evolving into periodic spasms is described. A six-month-old boy was referred to us because of complex partial seizures characterized by impairment of consciousness followed by adversion of the eyes and face associated with asymmetric tonic neck reflex-like postures. Interictal electroencephalography (EEG) on admission revealed multiple independent spike foci and clusters of the fast activities, which tended to be pseudoperiodic, during sleep. Ictal EEG showed irregular high voltage slow waves in the left hemisphere which gradually developed into diffuse slow waves followed by periodic sharp and/or slow waves associated with periodic spasms every two or five seconds. He had mild hemiparesis in the right upper limb. Otherwise his development of gross movement was normal. His seizures occurred frequently during the waking state, but sodium valproate was effective. In West syndrome, it is well known that epileptic seizures are characterized by a series of spasms. However, few patients with partial seizures in infancy have been reported to have periodic spasms.

Key words: complex partial seizures—periodic spasms

With recent advances in diagnostic evaluation using closed circuit television and electro-encephalography it has become possible to study the detailed clinical features of complex partial seizures in both children and adults.[1,2] However, the diagnosis of complex partial seizures beginning in infancy is rather difficult because the clinical manifestations are often more atypical than those in other age groups. Variable ictal manifestations exist in complex partial seizures in infancy.[3,4] Recently, Gobbi and his co-workers described a new condition in epileptic patients characterized by episodes of periodic spasms accompanied by unusual EEG complex.[5,6] We report here a case of complex partial seizures in infancy evolving into periodic spasms resembling those of Gobbi's periodic spasms.

CASE REPORT

The patient was a six-month-old boy who was the product of a full term uneventful delivery. With exception of mild clumsiness of the right upper limbs he was normal until six months of age. At that time he experienced his first episodes of motionless staring followed by slow elevation of both arms associated with adversion of the eyes and head toward the right. Each episode

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lasted about 30 seconds, with several episodes each day. Subsequently, his attacks increased and the duration of the episodes tended to become longer. Therefore, he was admitted to our unit. On admission, his general status appeared to be normal. Although he displayed clumsiness of the right hand, his deep tendon reflexes were otherwise normal. His milestones were good until six months of age. Laboratory examinations, including routine hematological and biochemical studies, indicated no abnormalities. Urine and serum aminograms were also within normal range. Neuroradiological studies, including magnetic resonance imaging and computed tomography, revealed no significant abnormalities. However ^{123}iodo-amphetamine (^{123}I-IMP) single photon emission computed tomography (SPECT) disclosed decreased cerebral blood flow in the left hemisphere. There were no abnormalities in short latency somatosensory evoked potentials and auditory brain stem responses. He had frequent complex partial seizures during the waking state. On admission, his interictal waking EEG showed multiple independent spikes which were dominantly in the left hemisphere and irregular slow waves also marked over the left hemisphere (Fig. 1). His sleep EEG records showed clusters of the fast activities which tended to be pseudoperiodic (Fig. 2). The initial ictal phase was characterized by motionless staring and then gradual elevation of the arms presenting with tonic components. There was also adversion of the eyes and face toward the right. Two minutes after the onset of seizures, he began to have periodic spasms with head nodding which involved the axial muscles and resembled those of flexor spasms in West syndrome. These spasms occurred at two or five second intervals, and each cluster was composed of 10 to 20 spasms. During the spasms, he had an impaired level of vigilance and could not follow any commands. Ictal EEG demonstrated focal rhythmic activities including small spike and/or spike-waves in the left frontal-central area followed with irregular high voltage slow waves. These gradually spread to the

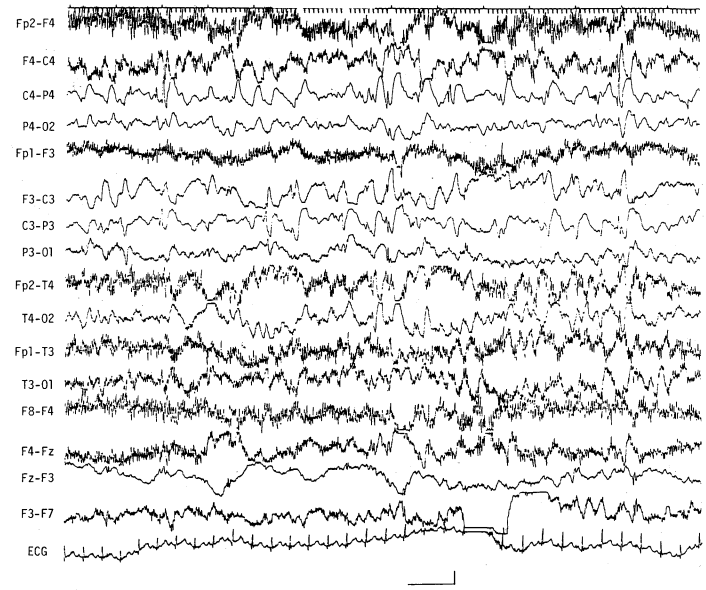

Fig. 1. Interictal waking EEG demonstrating multiple independent spikes foci dominantly in the area of the left hemisphere.

bilateral hemisphere in association with the motionless staring followed by arm elevation and adversion of both the eyes and face, and then they evolved into periodic spasms.

These tonic spasms occurred with diffuse periodic high voltage slow waves on EEG (Fig. 3). Thus there was a one to one relationship between the clinical features and periodic events on EEG. His clinical seizures as well as paroxysmal discharges on EEG disappeared following treatment with 300 mg/day of sodium valproate. Currenty, at three years of age, he shows normal psychomotor development and has had no further complex partial seizures except for a single febrile generalized convulsive seizure.

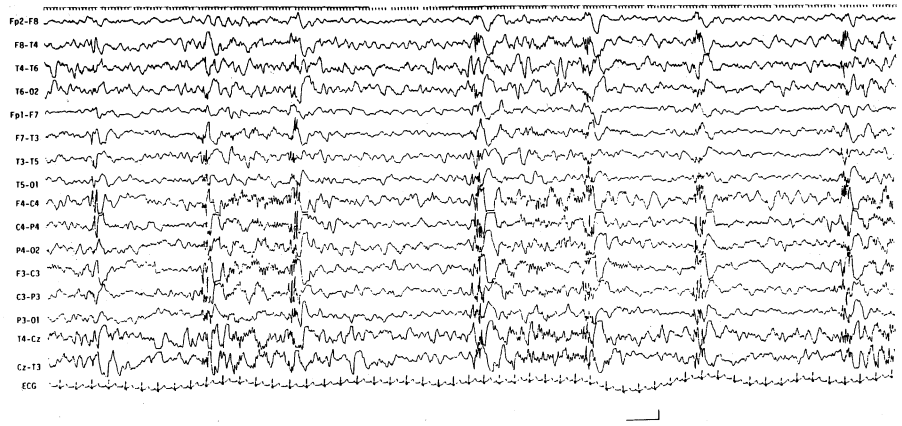

Fig. 2. Sleep EEG demonstrating clusters of the fast activities which tended to be pseudoperiodic.

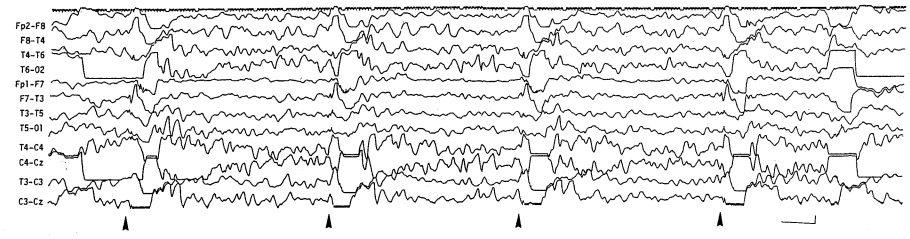

Fig. 3. Ictal EEG of periodic spasms (arrowheads indicating periodic spasms).

DISCUSSION

It is well known that partial seizures beginning in infancy tend to be intractable and are often associated with severe mental handicaps.[4] Recently, EEG/video monitoring systems have come into accepted use, and detailed studies of complex partial seizures in infancy as well as childhood have been reported.[1,3,4,7] Variable clinical features of complex partial seizures exist in infancy. Duchowny reported that seizures in infancy were characterized most frequently by behavioral arrest with forced lateralized deviation of the head and eyes and tonic upper-extremity extensor stiffening.[4] Complex facial automatism, such as chewing, sucking, mouthing and blinking, was also reported

to be relatively common. Our case also showed behavioral arrest followed by asymmetric tonic neck reflex-like postures during the ictal course. Furthermore, his seizures evolved into characteristic periodic spasms that have been previously documented.[5,6] Gobbi *et al.* described seven cases aged between 14 months and 13 years presenting with periodic spasms.[5,6] In their report, two cases had focal EEG abnormalities, while the other five cases showed multifocal EEG abnormalities. They noted that these characteristic events were indicative of bilateral and asymmetric spasms occurring in periodic seqences they were characterized brief complex movements mainly involving the axial muscles, with rising shoulders, adversion of the eyes and head with rectus-type facial grimaces, and stretching of the legs. In addition, four cases had focal clinical seizure and focal EEG features followed by periodic spasms. In all of these cases, each spasm lasted between 0.8 and 3.3 sec and these clinical features were related to EEG complexes characterized by slow waves with a superimposed fast rhythm.[5,6] The presented case also showed periodic spasms resembling those described by Gobbi *et al.*, which were proceeded by complex partial seizures with focal EEG abnormalities. In our case, at the time the periodic spasms occurred, ictal EEG revealed periodic high voltage slow waves exhibiting a one to one relationship between the periodic spasms and ictal EEG events. However, fast rhythm was not seen. The pathophysiology of periodic spasms is still uncertain, but in the description of Gobbi *et al.*, variable brain malformations such as agenesis corpus callosum, lissencephaly, tuberous sclerosis and clastic lesions were recognized and were always associated with impaired psychomotor development. However, cryptogenic cases have also been reported. Our case had no demonstrable abnormalities on either cranial CT or MRI, but SPECT revealed decreased cerebral blood flow in the area of the left hemisphere. Furthermore, at the time of admission, complex partial seizures occurred frequently, and hemiparesis of the right arm was recognized. This latter condition improved, however, after the disappearance of both paroxysmal discharges on EEG and the complex partial seizures. These findings may contribute to the functional abnormalities in the cerebral cortex of our case. The appearance of clusters of the fast activities in sleep EEG records may also suggest the existence of abnormalities is the cortical-subcortical connection. It is known that brief tonic seizures called tonic spasms which tend to be periodic in sequence, also occur in Ohtahara syndrome[8] and West syndrome[9] during infancy. Additionally, it is known that partial seizures may occur in patients who have infantile spasms.[10,11] However, few patients with partial seizures have been reported to have poriodic spasms without hypsarrhythmia on interictal EEG. Therefore, detailed evaluations of ictal manifestations and EEG analysis should be done when diagnosing periodic spasms in infancy.

REFERENCES

1) Holmes, G.L.: Partial complex seizures in children: An analysis of 69 seizures in 24 patients using EEG FM radiotelemetry and videotape recording. Electroenceph. Clin. Neurophysiol. **57**: 13-20, 1984
2) Delgado Escueta, A.V., Bacsal, F.E. and Treiman, D.T.: Complex partial seizures on closed-curcuit television and EEG: A study of 691 attacks in 79 patients. Ann. Neurol.

11: 292-300, 1982
3) Watanabe, K., Yamamoto, N., Negoro, T., Takaesu, E., Aso, K., Furune, S. and Takahashi, I.: Benign complex partial epilepsies in infancy. Pediatr. Neurol. **3:** 208-211, 1987
4) Duchowny, M.S.: Complex partial seizures of infancy. Arch. Neurol. **44:** 911-914, 1987
5) Gobbi, G., Bruno, L., Pini, A., Rossi, P.G. and Tassinari, C.A.: Periodic spasms: An unclassified type of epileptic seizure in childhood. Dev. Med. Child. Neurol. **29:** 766-775, 1987
6) Gobbi, G., Parmeggiani, A., Rossi, P.G. and Tassinari, C.A.: Periodic spasms: A particular type of partial epileptic seizure in infancy and childhood. *In* Satellite meeting of the joint convention of the 5th ICNC and the 3rd AOCCN: New trends in pediatric epileptology. November 11, 1990, in Okayama, Japan.
7) Kotagal, P., Rothner, A.D., Erenberg, G., Cruse, R.P. and Wyllie, E.: Complex partial seizures of childhood onset: A five year follow-up study. Arch. Neurol. **44:** 1177-1180, 1987
8) Ohtahara, S.: Clinicoelectrical delineation of epileptic encephalopathies in childhood. Asian Med. J. **21:** 499-509, 1978
9) King, D.W., Dyken, P.R., Spinks, I.L. and Murvin, A.J.: Infantile spasms: Ictal phenomena. Pediatr. Neurol. **1:** 213-218, 1985
10) Hrachovy, R.A., Frost, J.D., Jr. and Kellaway, P.: Hypsarrhythmia: Variations on the theme. Epilepsia **25:** 317-325, 1984
11) Yamamoto, N., Watanabe, K., Negoro, T., Furune, S., Takahashi, I., Nomura, K. and Matsumoto, A.: Partial seizures evolving to infantile spasms. Epilepsia **29:** 34-40, 1988