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Abstract

We report a family whose members have familial spastic paraplegia (FSP) associated with epilepsy. A man and his sister initially had primary generalized epilepsy with tonic-clonic seizures, but they have had no seizures for years. However, they developed spastic paresis of the lower extremities and presently show features of FSP. Their mother seemed to have suffered from FSP. One son of the female patient has epilepsy. The clinical picture of this family suggests a close relationship between FSP and epilepsy.

KEYWORDS: familial spastic paraplegia, epilepsy

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FAMILIAL SPASTIC PARAPLEGIA WITH EPILEPSY

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Abstract. We report a family whose members have familial spastic paraplegia (FSP) associated with epilepsy. A man and his sister initially had primary generalized epilepsy with tonic-clonic seizures, but they have had no seizures for years. However, they developed spastic paresis of the lower extremities and presently show features of FSP. Their mother seemed to have suffered from FSP. One son of the female patient has epilepsy. The clinical picture of this family suggests a close relationship between FSP and epilepsy.

Key words: familial spastic paraplegia, epilepsy.

Some cases of familial spastic paraplegia (FSP) are associated with other neurological abnormalities, and others are "pure" cases, which show only spastic paraplegia. Among them there have been some, but not many cases with epilepsy (1, 2). We observed a family whose members have FSP, and in whom epilepsy tended to occur.

CASE REPORT

Case 1: 37-Year-Old Male

Family history. His mother (Case II-2), who is dead, had difficulties in walking since about 40 years of age (Fig. 1). She stumbled often, and became more and more disabled. She died of a cerebral infarction at the age of 50. The patient (Case 1) recalled that his mother's walk was just like his own. There were no neurological abnormalities in other members of this family except for one of the patient's sisters (Case 2).

Clinical course. There were no perinatal or developmental abnormalities. When he was twelve, he developed generalized convulsions and was taken to Okayama University Hospital. He was diagnosed as having primary generalized epilepsy with tonic-clonic seizure and has since been taking antiepileptic drugs. At the time of onset, seizures occurred several times a year, but they gradually decreased in frequency and severity, taking on the character of short lapses of consciousness. For the last ten years he has had no seizures, although he often forgot to take his drugs. On the other hand, he developed gait disturbance at seventeen, which gradually progressed. At the age of 30, he could no longer ride a bicycle.

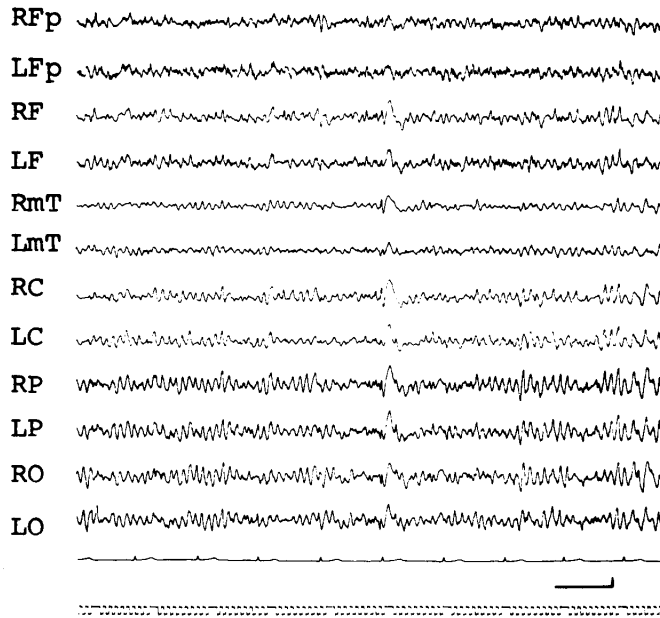


Fig. 2. Case 1. EEG exhibits bilateral, synchronous spike and wave discharges.

and was diagnosed as having primary generalized epilepsy. Seizures gradually reduced in frequency, and finally disappeared eight years ago. She did not take her drugs strictly. At about thirty, gait disturbance appeared and has gradually increased in severity.

Neurologically, there are spastic paraplegia, increased tendon reflexes and positive pathological reflexes in the lower extremities. Her gait is spastic, but is not scissors-like. Her disability is milder than her brother's (Case 1).

EEG shows bilateral, synchronous spike-and-wave complexes. Blood chemistry is normal. Cerebral CT scan and cervical spine roentgenogram show no abnormalities.

Case 3 : 12-Year-Old Boy (Son of Case 2)

There was no abnormality at birth or during development. He developed generalized convulsions at seven years and six months, was diagnosed as having epilepsy and has been taking antiepileptic drugs since. There have been 4 seizures.

At present there is no neurological abnormality, no spasticity of the legs and no gait disturbance. EEG shows frequent bilateral, synchronous spike-and-wave complexes (Fig. 3).

DISCUSSION

The neurological features of Cases 1 and 2 are compatible with FSP on

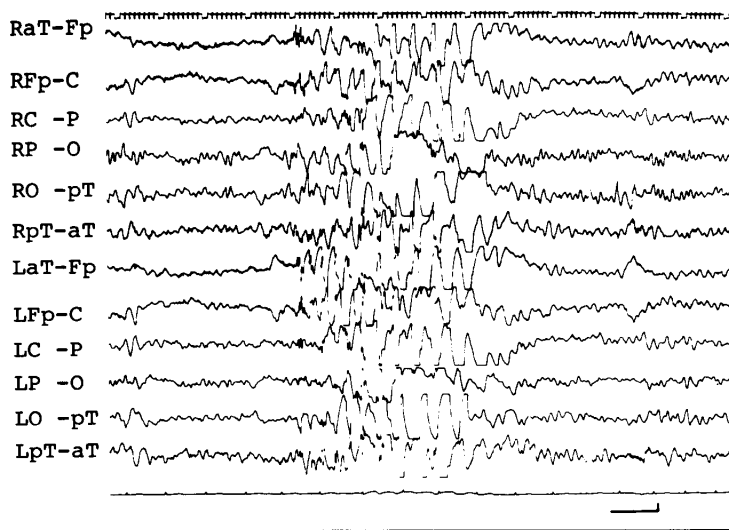


Fig. 3. Case 3. EEG shows diffuse spike and wave bursts.

account of familiarity, spastic paraplegia, negative cerebellar sign, negative sensory disturbance and normal intelligence. Their mother, of whom information is lacking, may have had FSP. The age at onset of FSP, however, was rather advanced in the mother and Case 2. Cases 1 and 2 of this family developed epilepsy first and then had FSP, that is, as the seizure activity decreased in severity, the features of FSP gradually appeared and progressed. Their seizure activity decreased and became very slight over the course of years. There have been no seizures for eight to ten years in spite of irregular drug intake.

The essential feature of FSP is of a spinal lesion. Some cases are known to be associated with diseases of other sites, but relatively little attention has been paid to epilepsy and EEG abnormalities (1-7).

In 1962, Bruyn and Mechelse (8) noticed the association of FSP and epilepsy and insisted this was not a mere coincidence. In Japan, Riku and Koike (9) reported in 1983, two cases in a family very much like our cases.

Epilepsy may be attributed to cerebral or thalamic lesions, and FSP, to spinal cord lesions. Epilepsy and FSP, whose pathological lesions are considered to be different, are closely related in these cases. How they may be related is an interesting problem which requires investigation.

REFERENCES

1. Radermecker, J.: Epilepsy in the degenerative disease. In *Handbook of Clinical Neurology*, ed. P.J. Vinken and G.W. Bruyn, North-Holland, Amsterdam, Vol. 15, pp. 325-372, 1974.

2. Sutherland, J.M. : Familial spastic paraplegia. In *Handbook of Clinical Neurology*, ed. P.J. Vinken and G.W. Bruyn, North-Holland, Amsterdam, Vol. 22, pp. 421-431, 1975.
3. Strümpell, A. : Beiträge zur Pathologie des Rückenmarks. I. Spastische Spinalparalyse. *Arch. Psychiatr. Nervenkr.* **10**, 676-717, 1880.
4. Tyrer, J.H. and Sutherland, J.M. : The primary spino-cerebellar atrophies and their associated defects, with a study of the foot deformity. *Brain* **84**, 289-300, 1961.
5. Malin, J.P. : Zur Differentialdiagnose der hereditären Spastischen Spinalparalyse. Zugleich über einen genetisch selbständigen Typ der rezessiv-erblichen, infantilen spastischen Spinalparalyse. *Nervenarzt* **47**, 661-668, 1976.
6. Okabe, K. : A pedigree with the "spastic type" of hereditary ataxia having noted interfamilial variation and its significance to clinical genetics. *Psychiatr. Neurol. Jpn.* **55**, 692-704, 1953 (in Japanese).
7. Kobayashi, T., Miyasaka, M. and Fukumizu, Y. : Familial spastic spinal paralysis : A family report. *Brain and Nerve* **15**, 475-480, 1963 (in Japanese).
8. Bruyn, G.W. and Mechelse, K. : The association of familial spastic paraplegia and epilepsy in one family. *Psychiatr. Neurol. Neurochir. (Amst.)* **65**, 280-292, 1962.
9. Riku, S. and Koike, Y. : Spastic paraplegia with epilepsy. *Neurol. Med.* **19**, 358-361, 1983 (in Japanese).