Acta Paediatrica Hungarica, 26 (2), pp. 97—99 (1985)

Subacute sclerosing panencephalitis in twins

R Michałowicz, Barbara Karkowska, R Ignatowicz, J Michałkiewicz, J Wyszkowski

The Child Health Centre, Warsaw, Poland

Development of subacute sclerosing panencephalitis after measles has been observed in twins although the disease seems to be quite exceptional in members of the same family, and no report has been found on its occurrence in twins.

Subacute sclerosing panencephalitis (SSPE) or Dawson's encephalitis seldom occurs in members of the same family. In spite of this we have observed SSPE in two boys, 8 and 9 years of age, respectively, who were cousins [4]. Since then we observed the disease in twin sisters who earlier had had measles.

Case Reports

Case 1 Girl C. A., born with a birthweight of 2450 g on 24th February 1972 as the third child of a family was admitted to our Institute at the age of 11 years with the suspicion of SSPE. Her two brothers were healthy and had not had measles. The patient at the age of 15 months had had measles with a severe clinical course. After the measles her somatic and psychomotor development had been excellent and unchanged until January 1983, when the first signs of SSPE were observed. Then abnormalities of behaviour, aggression and troubles in school were noted, so far she had been an excellent pupil. At admission psychomotor slow motion, bewilderment and jerks of limbs were observed. Psychological examination showed low spirits and sensitivity, misunderstanding of oral information, weakening of memory and a significant decrease of sight perception.

During her stay in hospital the girl's clinical condition became gradually from bad to worse. The motor signs (tremor, myoclonic jerks) increased considerably and so did the pyramidal signs. Ophthalmological examination did not reveal any abnormality, the EEG showed a typical Rademecker type record (Fig. 1).

The level of anti-measles antibodies, as estimated by the H.I. method, was 1 : 32 in serum and 1 : 8 in CSF. The CSF immunoglobulin level was increased to 33.2%, and the IgG to 28 mg/dl. The gold chloride curve was of paralytic type. Other laboratory tests of the CSF gave normal values.

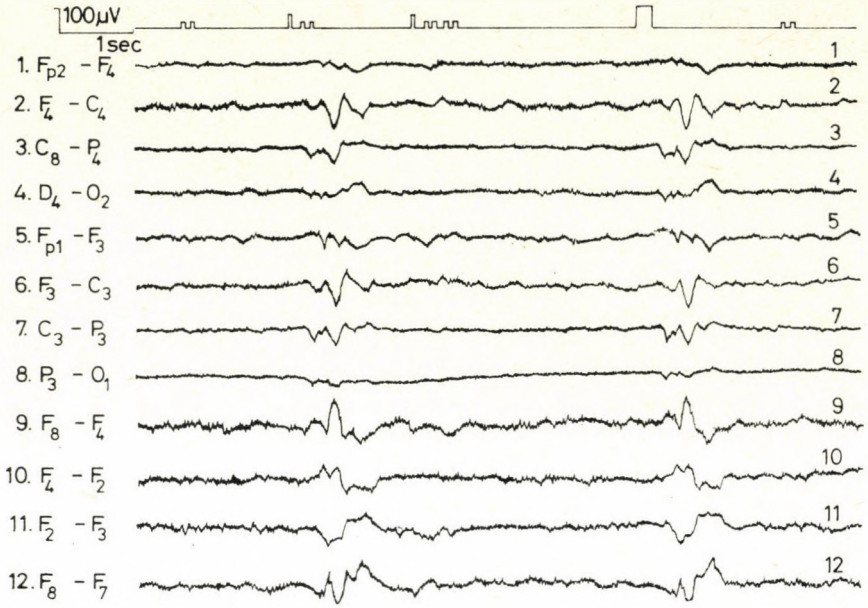

FIG. 1. EEG of Case 1. Typical Rademecker type record

Blast transformation test with PHA and ConA, specific activators of T cells, were normal (66 000 cpm and 45 000 cpm, respectively), in agreement with our currently done studies on other patients with SSPE (data not shown). The serum level of immunoglobulins, estimated by the Mancini technique, was in the normal range (IgG, 1960 mg/dl; IgA, 167 mg/dl). The amount of IgG antibodies in CSF was 128 mg/dl, much higher than normal (2.71 $\pm$ 47 mg/dl). IgM antibodies were not present.

In the brain, computer tomography showed focal demyelination and a narrow ventricle system. All other tests, blood and urine analyses gave normal values. The patient's blood group was A_1Rh^+.

On the basis of the laboratory tests and clinical findings, SSPE in Jabbour's stage II/III [1, 2, 3] was diagnosed.

Case 2 Girl C. Ag. was the twin sister of Case 1. The neonatal and infantile periods had been normal. She had contracted measles at the age of 15 months simultaneously with her sister. The clinical course of the disease was less severe than of Case 1.

In August 1981, psychomotor slow motion, troubles at school, difficulties in memorizing, reading and writing were observed, and she had disturbances of balance and of speech (paraphasia). Somewhat later epileptic seizures, myoclonic jerks and spastic paralysis of the limbs occurred. Ophthalmological examination revealed oculomotor palsy. The EEG showed a lack of basic functions typical of her age and the presence of delta type waves (1—3/sec). The level of anti-

measles antibodies, estimated by the H.I. method, was 1 : 64 in serum and 1 : 8 in CSF. The amount of immunoglobulins in the CSF was increased up to 18% and β-globulins up to 18%. Other laboratory tests were in the range of normal values. She belonged to blood group A Rh^+. On the basis of laboratory tests and the clinical course SSPE in stage II/III was diagnosed.

Both patients were subjected to methisoprinol and amantadine treatment; these drugs had no effect, they even failed to slow down the course of the process. The first child is still alive but in a very poor condition, the second died a year ago.

We have reported these cases of SSPE in cousins and especially those in twins not only because we could not find any publications concerning the problem, but we thought that these observations might prove useful in investigations into slow virus diseases and into the role of genetic factors in the aetiology and pathomechanism of SSPE.

References

1. Jabbour JT, Garcia JH, Lemmi H, Roglaud J, Duenas DA, Sever JL: Subacute sclerosing panencephalitis. JAMA 207:2248—2254, 1969
2. Jabbour JT, Duenas DA, Modlin J: SSPE: clinical staging, course and frequency. Arch Neurol 32:493—494, 1975
3. Jabbour JT, Duenas DA, Modlin J: Treatment of SSPE: Review of current treatments. Arch Neurol 32:496—497 1975
4. Michałowicz R, Karlowska B, Ignatowicz R, Kostrzewska M, Wyszkowski J: Familiar cases of SSPE in children. In: Proc. Symposium on biochemical, immunological and clinical aspects of MS, SSPE and demyelinating processes, Poznań, June 1983. pp 65—66

Received 12 February 1984

Prof R Michałowicz
Child Health Centre,
Al. Dzieci Polskich 20
04-736 Warsaw, Poland