ATYPICAL CLINICAL AND ELECTROENCEPHALOGRAPHIC PATTERN IN A PATIENT WITH SUBACUTE SCLEROSING PANENCEPHALITIS

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SUMMARY - We describe an atypical clinical and electroencephalographic (EEG) pattern observed during the course of subacute sclerosing panencephalitis in a 14 year-old boy. In this patient with a two weeks history of partial complex seizures, the atypical EEG pattern was characterized by an initial left temporal focus which evolved to periodic lateralized epileptiform discharges (PLEDs) and, only during the 3rd and 4th weeks the typical bilateral and generalized periodic complexes appeared.

KEY WORDS: subacute sclerosing panencephalitis, EEG, periodic complexes, partial complex seizure.

Since the early reports of Radermecker (1949) and Cobb and Hill (1950), the great diagnostic value of electroencephalographic (EEG) findings in subacute sclerosing panencephalitis (SSPE) has been outlined by numerous workers. As is well known, the characteristic EEG pattern consists of high-voltage and polymorphic periodic complexes, recurring at long intervals, grossly widespread throughout both hemispheres that are often associated with motor spasms.

In the case reported here, the patient presented partial complex seizures and an atypical EEG finding, making the diagnosis more difficult. We are aware of only one study of SSPE with partial complex seizures.

REPORT OF CASE

HJ, a 14-year-old boy had partial complex seizures two weeks before being seen at the Escola Paulista de Medicina, that were characterized by loss of consciousness, staring, verbal and gestual automatisms, several times per day. Also, cognitive and intellectual deterioration had developed. A diagnosis of partial complex seizures and behavioural disturbance were made. Despite treatment with anticonvulsants (carbamazepine) the
number of seizures was still the same. His past history showed that he had measles between the ages of 5 and 6 years. On examination, we found him to be a demented boy with decreased recent memory and confusion as to time and place. During the third week he has developed a tetraparesis with a bilateral Babinski response and periodic myoclonus-like jerks. The cerebrospinal fluid (CSF) protein concentration was 28mg/100ml with immunoglobulin at 5.4mg/100ml and a pleocytosis of 18 cells. The CSF measles titer was 1:4. Computed tomography (CT) scan was normal.

Fig 2. Periodic sharp-and-slow-wave complexes, occurring every 7-8 sec.
The first EEG made during the first week of hospitalization, when the child had partial complex seizures, showed paroxysms of sharp and slow waves at the left fronto-temporal region. His second EEG at the 2nd. week, showed periodic lateralized epileptiform discharges (PLEDs) (Fig 1) at the same region. The two last EEG carried out during the third and fourth week, when the patient had myoclonic jerks, showed the typical EEG pattern of SSPE (Fig 2). He died after five days of his last EEG.

COMMENTS

One of the diagnostic features of SSPE is the characteristic EEG pattern consisting of periodic complexes usually occurring every 4-15 sec1. However, occasionally the EEG may be less typical and thus present a problem in making the diagnosis6,13. For example, focal abnormalities are rare in SSPE and may be characterized by frontal4,6,7, centro-temporal7, temporal6,8, temporo-occipital5 and centro-parietal spikes4 or by PLEDs5,7,8,11,14.

Our patient had an atypical clinical and EEG patterns for SSPE. This atypical feature made the diagnosis difficult by the clinical picture and the EEG which initially showed a focal paroxysmal abnormality and a PLEDs pattern. It is of interest that such lateralization should occur in a disease process whose pathology is known to be diffuse.

The pathophysiology of epileptiform abnormalities in SSPE is unclear. This may, in part, reflect involvement of cortical and subcortical neurons5,8,10. It is possible that in our patient the disease had an initial involvement of a limited cortical area with posterior extension to subcortical and brain stem regions.

REFERENCES

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