Arq Neuropsiquiatr 1995, 53(2): 278-280

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

DÉLRIO F. SILVA, MÁRCIA MARQUES LIMA**, RENATO ANGHINAH**, EDMAR ZANOTELI***, JOSÉ GERALDO CAMARGO LIMA*****

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.

Padrão clínico e eletrencefalográfico atípico em um paciente com panencefalite esclerosante subaguda

RESUMO - Descrevemos o quadro clínico e eletrencefalográfico atípicos observados durante a evolução de panencefalite esclerosante subaguda, em um menino de 14 anos. Neste paciente, com história de crises parciais complexas há duas semanas, o padrão eletrencefalográfico atípico foi caracterizado por foco temporal esquerdo que evoluiu para "periodic lateralized epileptiform discharges" (PLEDs), somente durante a 3ª e 4ª semanas surgindo os clássicos complexos periódicos difusos.

PALAVRAS-CHAVE: panencefalite esclerosante subaguda, EEG, complexos periódicos, crise parcial complexa.

Since the early reports of Radermecker (1949)[12] and Cobb and Hill (1950)[2], 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[3] 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[9].

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

* Head of the EEG Sector, Escola Paulista de Medicina (EPM); **Junior Doctor of the EEG Sector; ***Resident in Neurology; ****Full Professor and Head of the Discipline of Neurology, EPM. Aceite: 2-janeiro-1995.

Dr. Délrio Façanha da Silva - Disciplina de Neurologia, Setor de Eletrencefalografia, Escola Paulista de Medicina - Rua Botucatu 740 - 04023-900 São Paulo SP - Brasil.

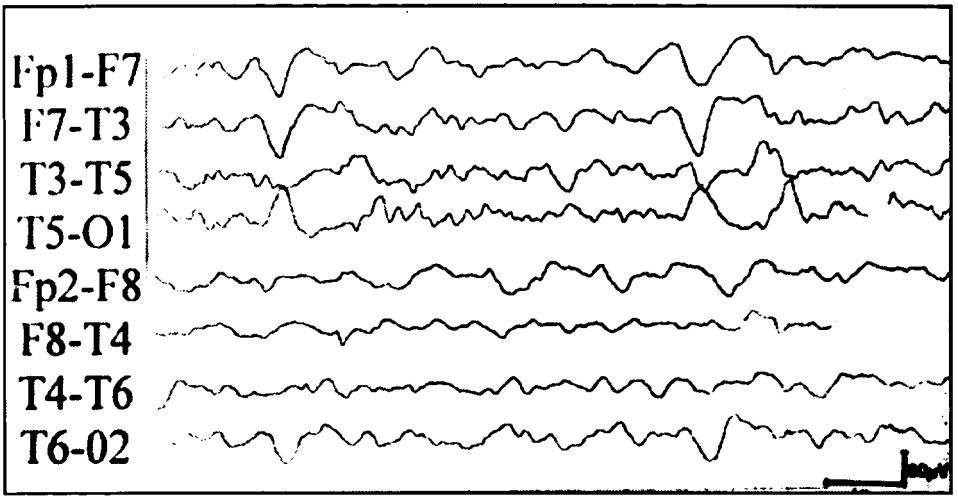

Fig 1. Unilateral left-sided SSPE complexes ocurring pseudo-periodically at intervals of 5-6 sec, and consisting of high voltage slow sharp wave complexes lasting approximately 1 sec and with a slow background activity.

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.

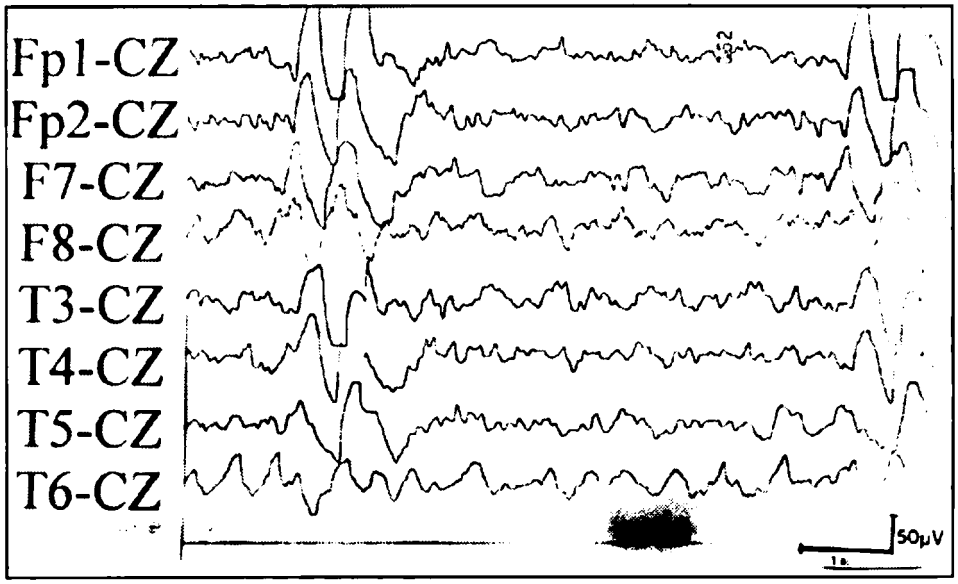

Fig 2. Periodic sharp-and-slow-wave complexes, ocurring every 7-8 sec.

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 occuring every 4-15sec[1]. However, occasionally the EEG may be less typical and thus present a problem in making the diagnosis[8,13]. For example, focal abnormalities are rare in SSPE and may be characterized by frontal[4,6,7], centro-temporal[7], temporal[6,8], temporo-occipital[5] and centro-parietal spikes[4] or by PLEDs[5,7,8,11,14].

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 inittially 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 neurons[5,8,10]. 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

1. Cobb WA. The periodic events of subacute sclerosing leucoencephalitis. Electroencephalogr Clin Neurophysiol 1966, 21:278-294.
2. Cobb W, Hill D. Electroencephalogram in subacute progressive encephalitis. Brain 1950, 73:392-404.
3. Gaches J. Activités périodiques en EEG. Rev EEG Neurophysiol Clin 1971, 1:9-33.
4. Giménez-Roldan S, Martin M, Mateo D, Lopez-Fraile IP. Preclinical EEG abnormalities in SSPE. Neurology 1981, 31:763-767.
5. Lascu O. Cortical focal epilepsy in a case of SSPE: clinical considerations. Neurol Psychiatr 1982, 20:269-273.
6. Lemmi H. The EEG in SSPE (Abstr). Electroencephalogr Clin Neurophysiol 1969, 27:550.
7. Markand ON. Atypical EEG findings in SSPE. Neurol India 1975, 23:70-77.
8. Markand ON, Panszi JG. The EEG in subacute sclerosing panencephalitis. Arch Neurol 1975, 32:719-726.
9. Namer IJ., Tan E, Ciger A, Zileli T. Crises épileptiques partielles complexes: premier symptome d'une panencéphalite subaigue sclérosante. Rev Neurol (Paris) 1986, 142:921-925.
10. Ohya T, Martinez AJ, Jobbour JT. SSPE: correlation of clinical neurophysiologic and neuropathologic findings. Neurology 1974, 24:211-218.
11. Onoda M. Early EEG changes in SSPE. Clin Neurol (Tokyo) 1979, 19:677-686.
12. Radermecker J. Aspects électroencéphalographiques dans un cas d'encéphalite subaigue. Acta Neurol Psychiatr Belg 1949, 49:222-232.
13. Sharp GB, Laney SM, Westmoreland BF, Groover RV. Atypical electroencephalographic pattern in a patient with subacute sclerosing panencephalitis. Electroencephalogr Clin Neurophysiol 1991, 78:311-313.
14. Sisk AM, Griffith JF. Normal electroencephalograms in SSPE. Arch Neurol 1975, 32:575-576.