ATYPICAL CLINICAL PRESENTATION OF SUBACUTE SCLEROSING PANENCEPHALITIS (SSPE)

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ABSTRACT

SSPE (Subacute sclerosing panencephalitis) is a chronic and slowly progressive inflammatory disease of the central nervous system caused by a persistent measles virus usually affecting the childhood and adolescent age group.

We present the case of a 19 years old adolescent, the product of a normal full-term pregnancy, with a primary measles infection at the age of 7 months. Neurological examination at admission discloses unilateral myoclonic jerks of upper left limb and choreoatetosis of the left hand fingers. *The particularity of our case is the unilaterality of the myoclonic jerks at the onset and relatively old the age of debut.*

The findings of periodic complexes on EEG, elevated gamma globlin and oligiclonal bands in the CSF and elevated measles antibody titers in the serum and CSF were sufficient to make diagnosis of SSPE.

Key words: SSPE (Subacute sclerosing panencephalitis), EEG periodic complexes, measles antibody titers

INTRODUCTION

SSPE (Subacute sclerosing panencephalitis) is the result of a chronic measles infection. SSPE is a chronic and slowly progressive inflammatory disease of the central nervous system caused by a persistent measles virus usually affecting the childhood and adolescent age group. (1)

Typically there is a primary measles infection at very early age, often before 2 years, followed by 6- to 8 years asymptomatic period.

Clinical features at onset are very subtle and nonspecific. The ilness evolves in several stages. The typical clinical course of SSPE is characterized by intellectual deterioration, personality and behavioral changes, myoclonic jerks, visual disturbances and sometimes pyramidal and extrapyramidal symptoms.

Initially there is a decline in school performances and personality changes. Then will be progressive intellectual deterioration in association with focal and generalized seizures, widespread myoclonus, ataxia and sometomes visual sisturbances due to progressive chorioretinitis. As the disease advances, rigidity, hyperactive reflexes, Babinski signs, progressive unresponsiveness, and signs of autonomic dysfunction appear. In the final stage the patient lies insensate, „decorticated". (1,2)

The course is usually steady progressive, death occuring within 1 to 3 years.

Certain atypical features can occur at onset or during the course of illness which can be misleading. Neuroimaging features often are non-specific. (3,4,5)

The diagnostic of SSPE is based upon the prsence of characteristic periodic EEG complex discharges, elevated gamma globulin and oligoclonal bands in CSF and elevated measles antibody titers in the serrum and CSF. (1)

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CASE PRESENTATION

We present the case of a 19 years old adolescent, the product of a normal full-term pregnancy, with a primary measles infection at the age of 7 months.

His family history was non-contributor.

He had a stagewise progression. For some time he had insidious intellectual impairment and behavioural abnormalities. Visual disturbances due to macular chorioretinitis to the left and then to the right eye appeared in the last months. He is presenting at the neurology clinic due to appearance of involuntary movements of the left upper limb.

Physical examination in the emergency room revealed normal vital signs. There was no fever and vomiting. Neurological examination at admission in our hospital discloses unilateral myoclonic jerks of upper left limb and choreoatetosis of the left hand fingers. No seizures were seen.

Cerebral CT scan was unremarkable (Figure 1).

EEG (electroencephalogram) showed typical periodic high amplitude slow waves complexes discharges, synchronous with myoclonic jerks which cannot be suppressed with diazepam. The characteristic abnormality is consisting of periodic (every 5 to 8 s) bursts of 2 to 3 per second high-voltage-waves, followed by a relatively flat pattern (Figures 2-7).

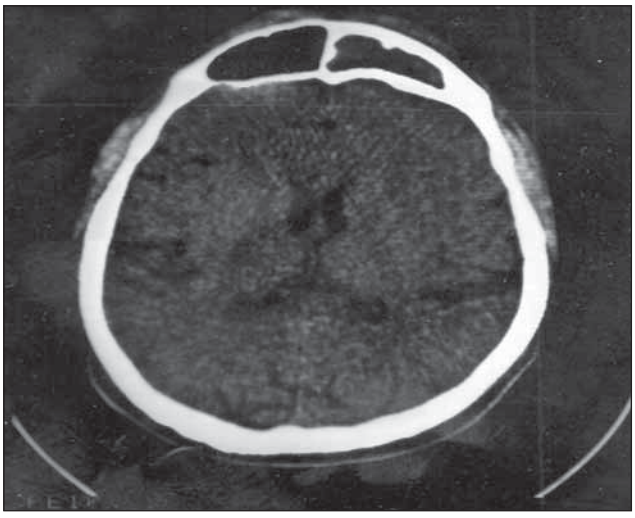


FIGURE 1. *Cerebral CT scan was unremarkable*

FIGURES 2-7. *EEG (electroencephalogram) showed typical periodic high amplitude slow waves complexes discharges, synchronous with myoclonic jerks*

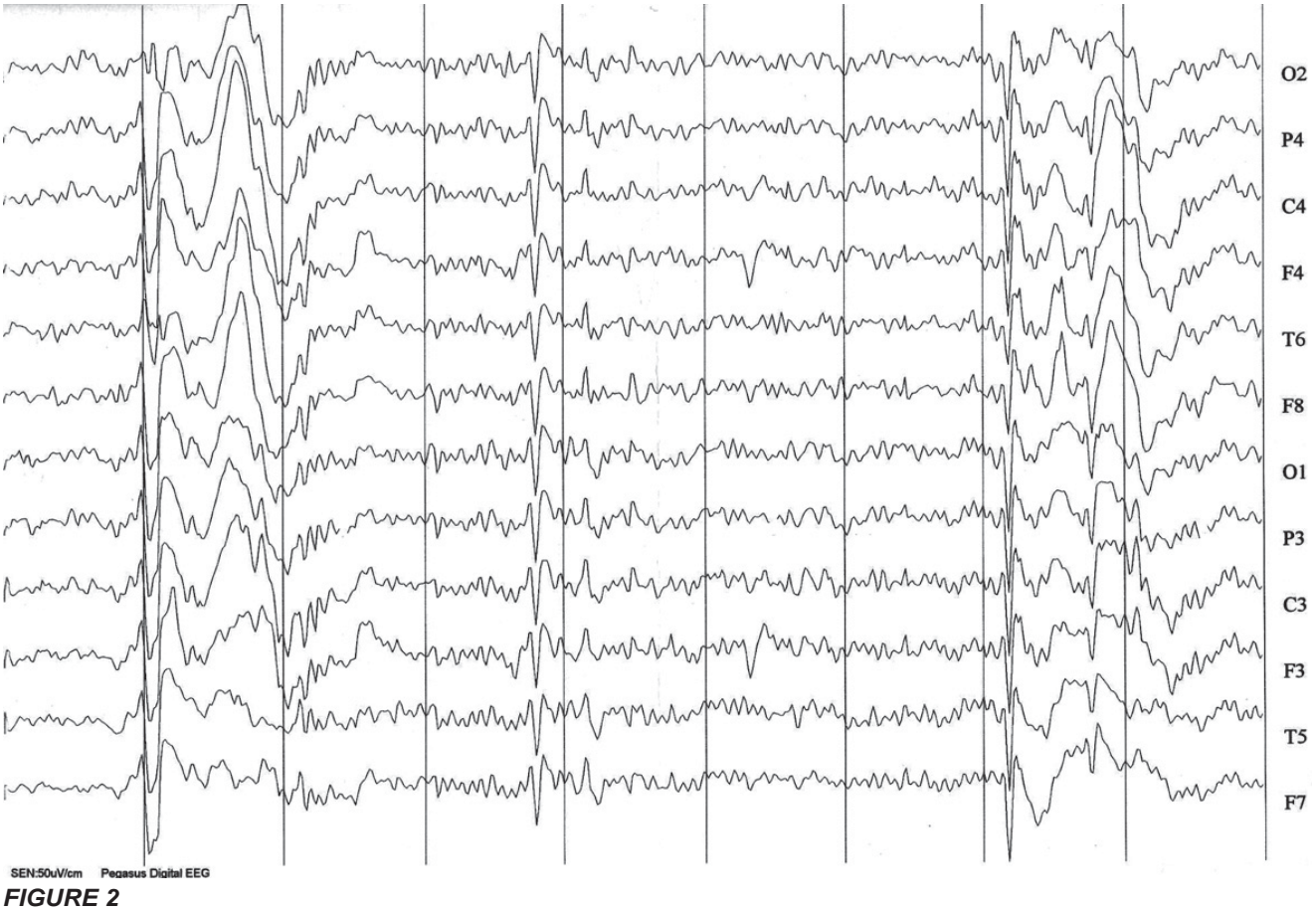
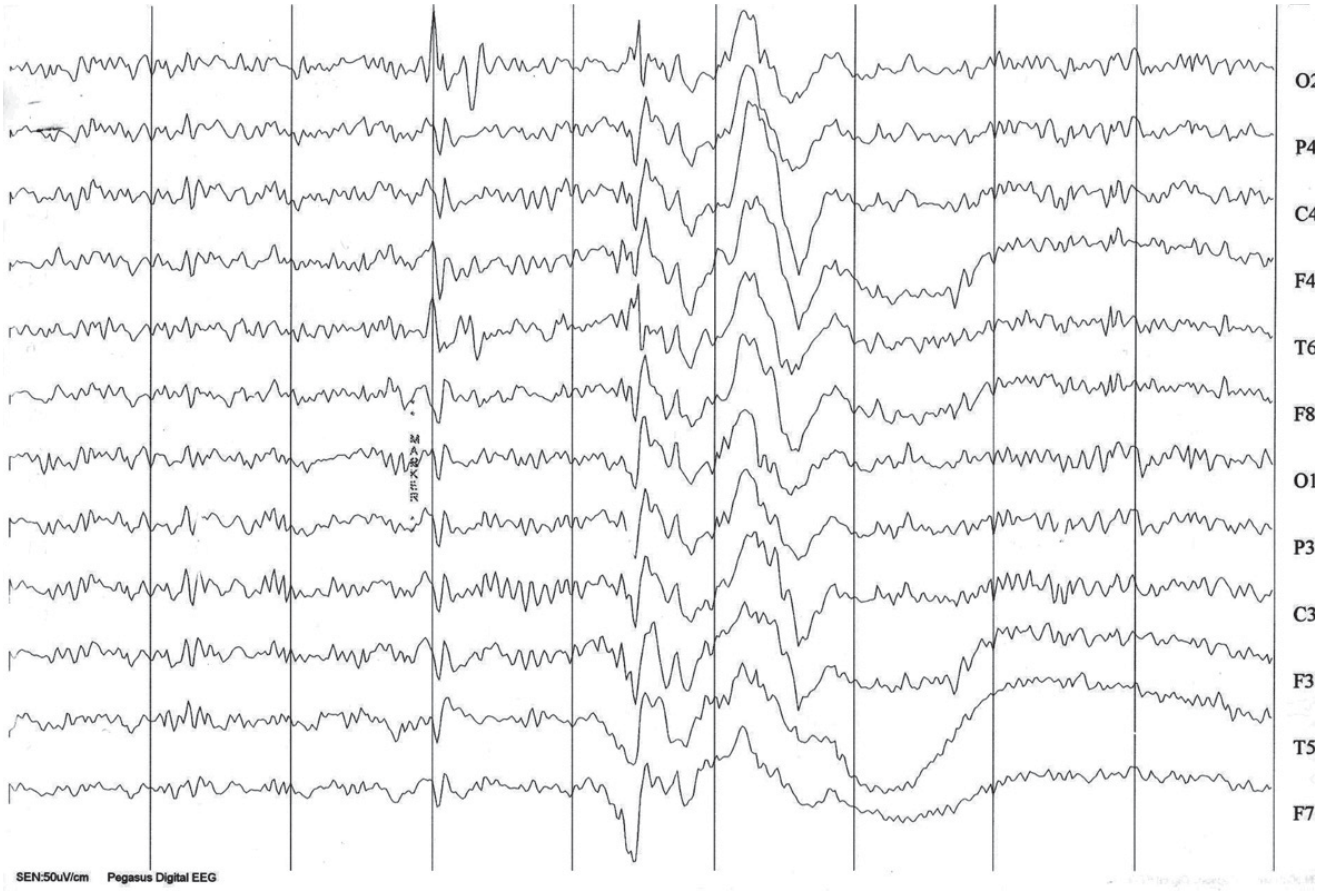


FIGURE 2



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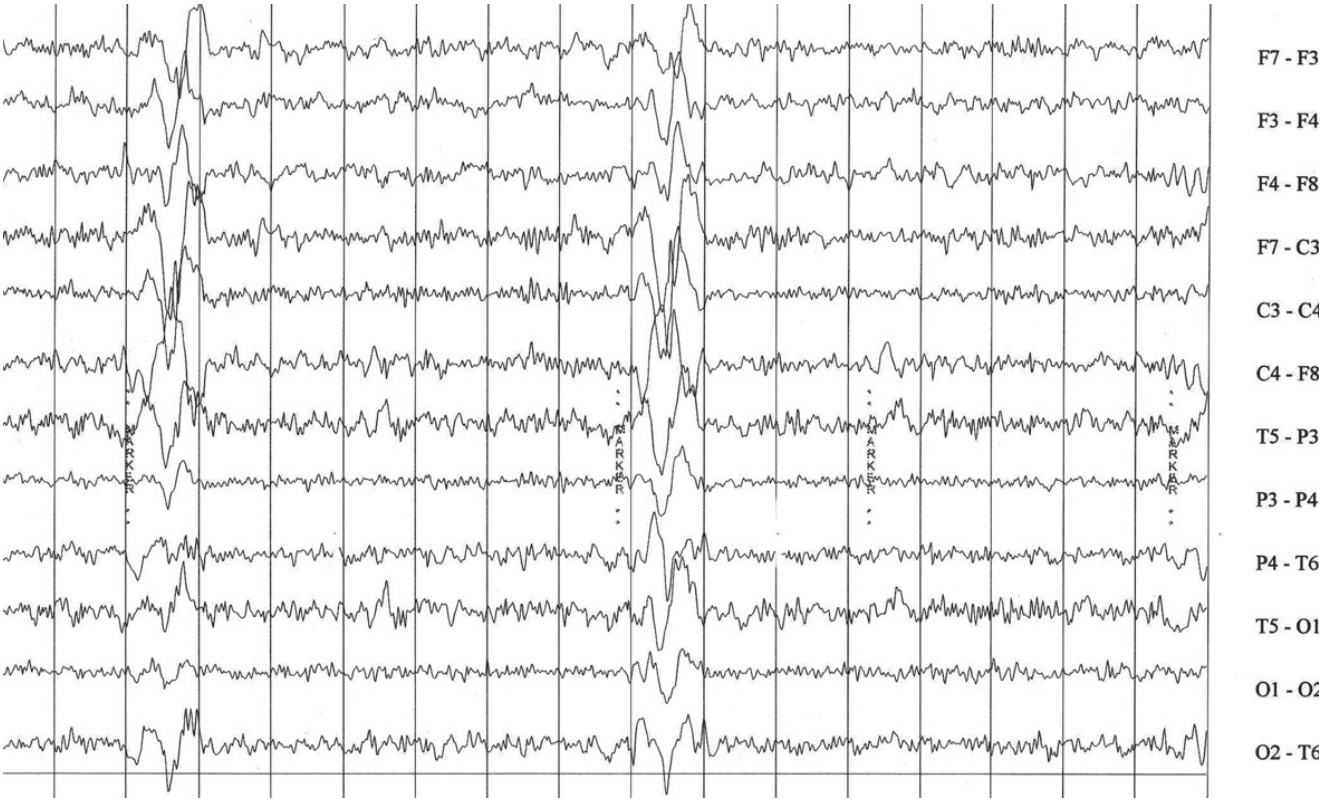
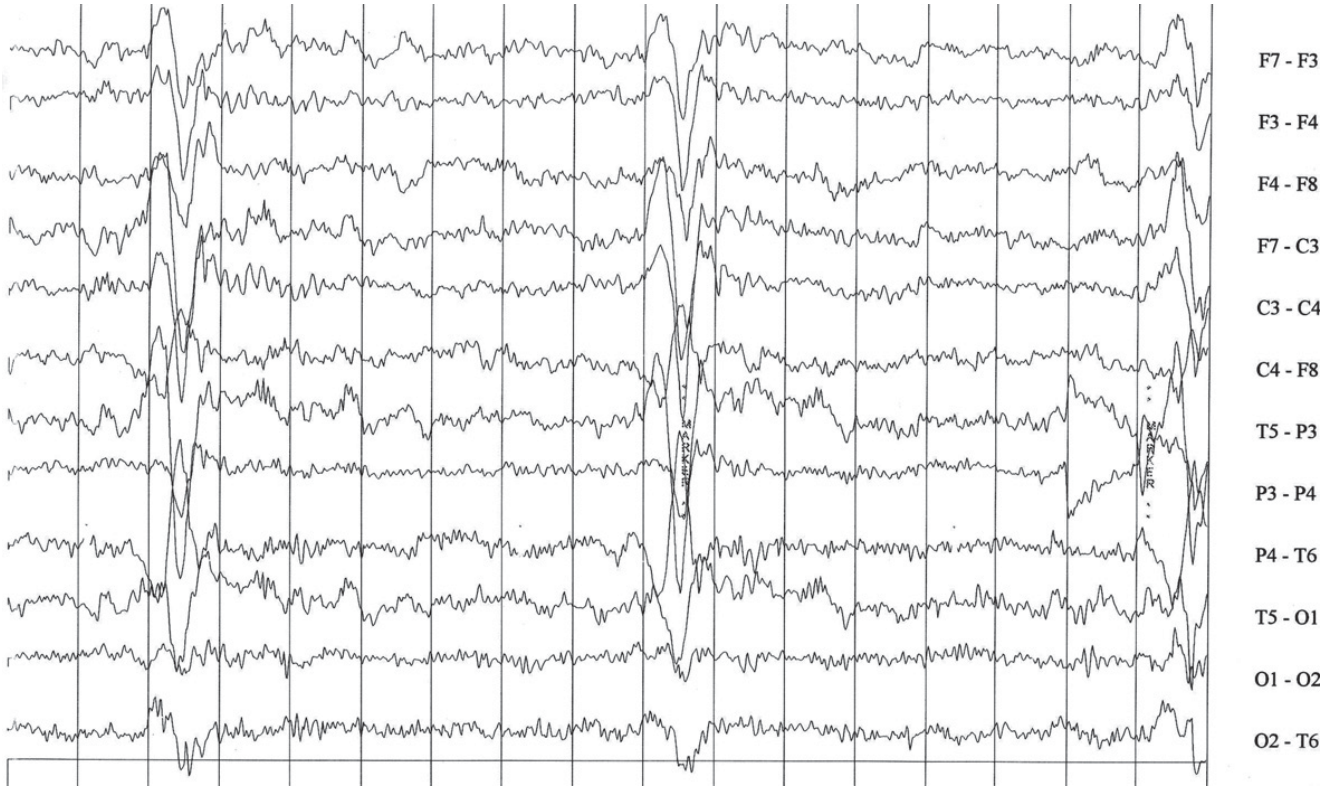


FIGURE 4



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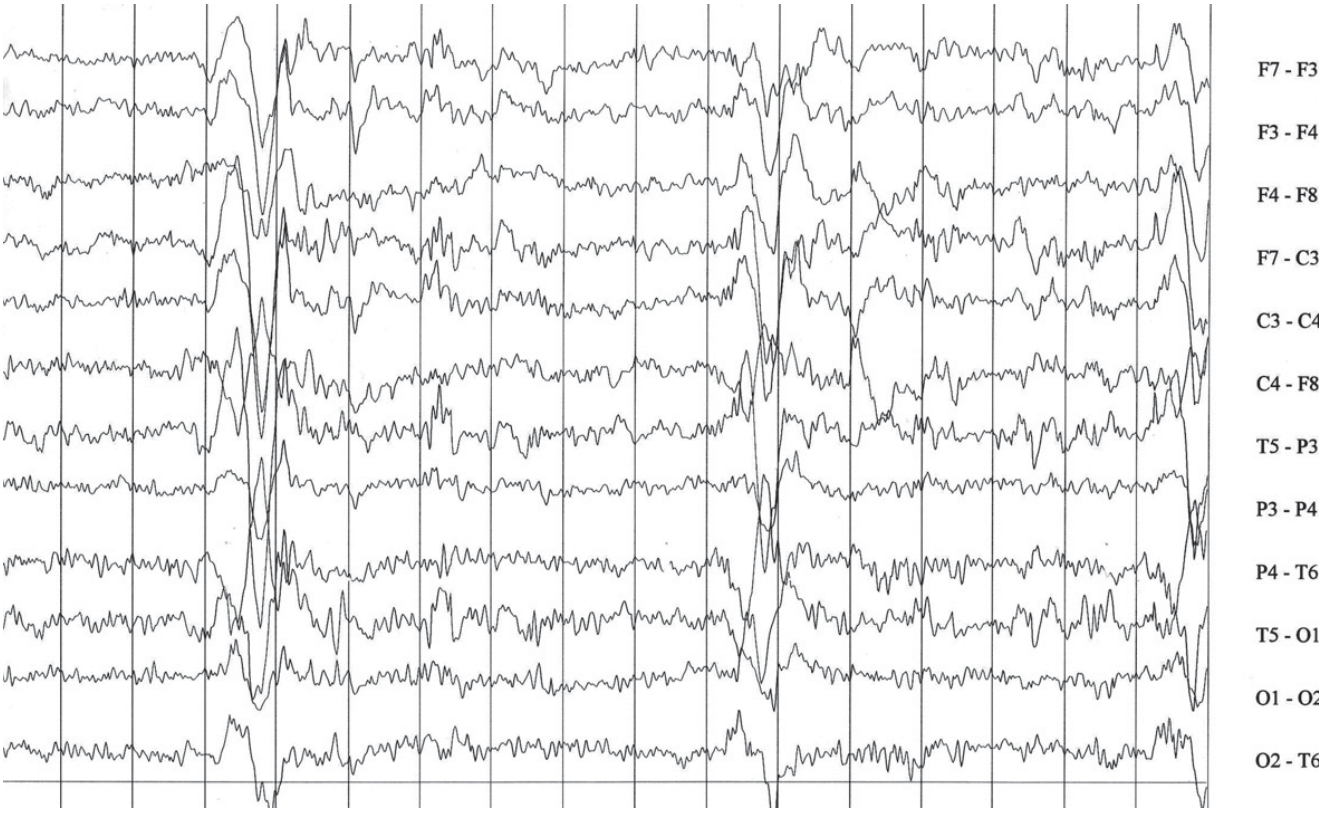

FIGURE 6

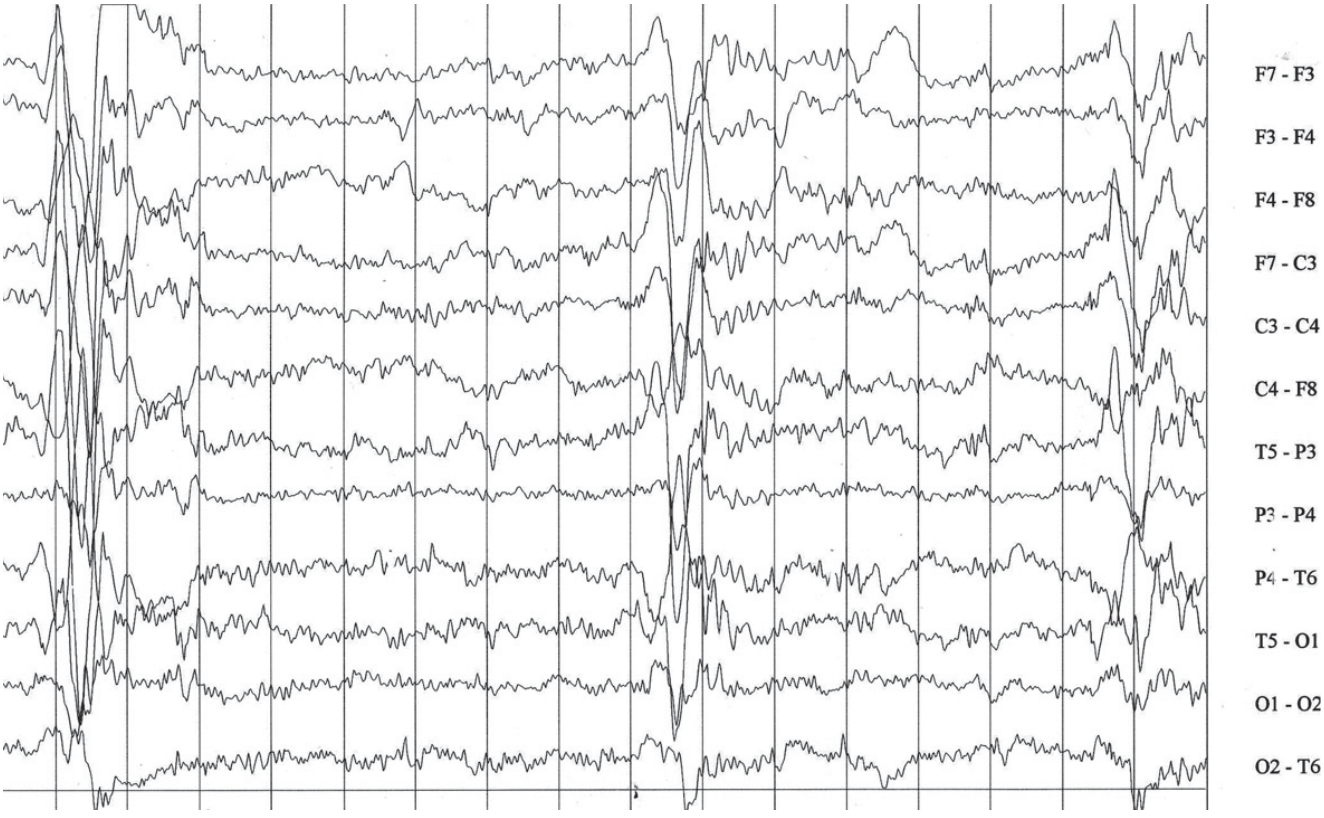

FIGURE 7

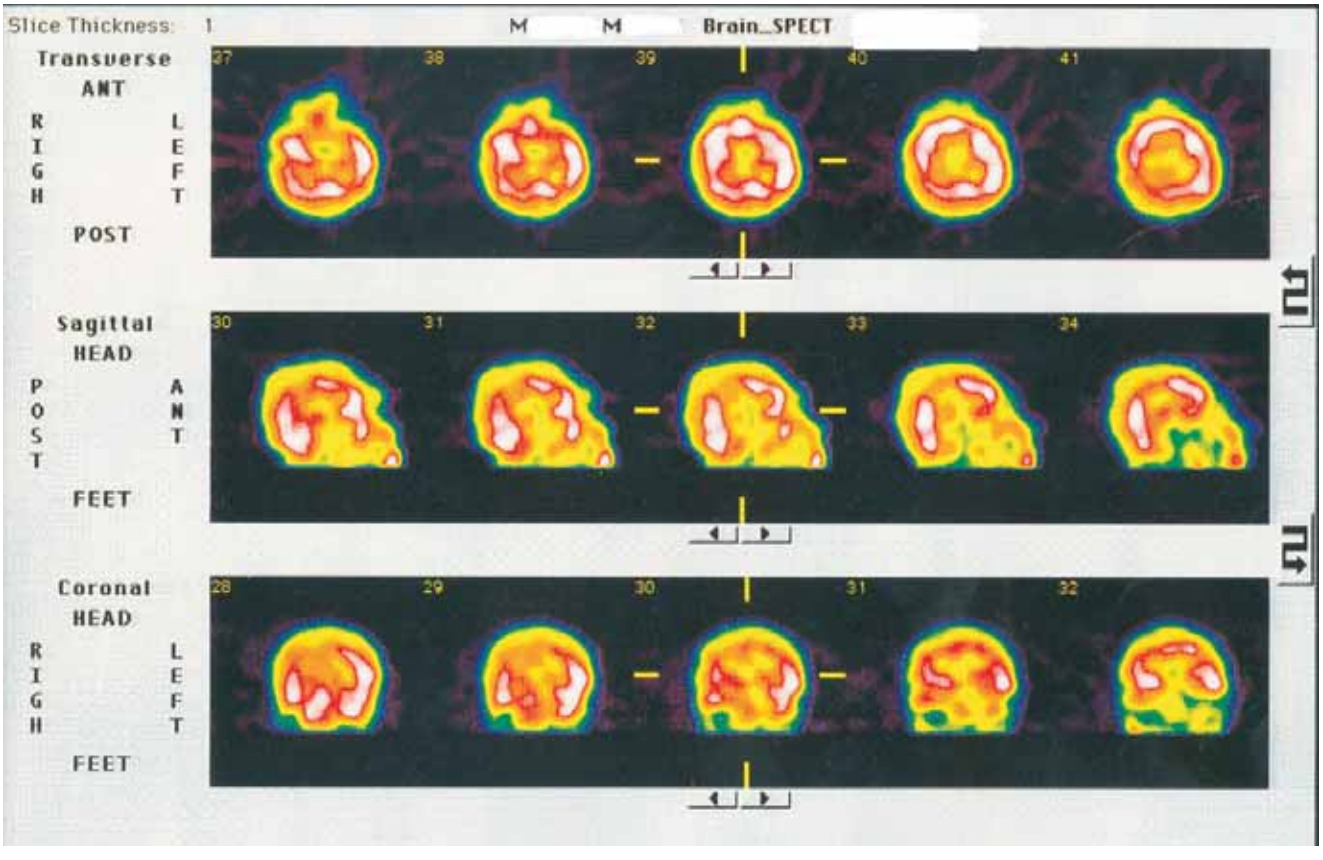

FIGURE 8. *Cerebral mTC-HMPAO SPECT – no anomalies of cerebral blood flow*

We suspected SSPE and we performed lumbar puncture. The CSF contains few cells, but the protein is incresed. Oligoclonal bands were present in CSF.

Both serum and CSF contained high concentrations of neutralizing antibodies to measles (rubeola) – more than 8 times the normal limit - that confirmed the diagnosis.

Cerebral mTC-HMPAO SPECT was also performed, but no anomalies of cerebral blood flow were observed (Figure 8).

DISCUSSION

Subacute sclerosing panencephalitis is a chronic and pregresssive disease caused by defective measles virus in central nervous system.

There is a delay in the development of immune response during the initial infection and a later inadequate immune response that are incapable of clearing the suppressed infection. Another ipothesis is that the brain fall to syntethise a M –protein, wich is essential for the assembly of the viral membrane, related to the extend of viral seeding of the brain during initial infection. Virions (measles nucleocapsides) have been observed in the inclusion-bearing cells examined at electron microscopically (1).

Histologically the lesions involve the cerebral cortex and white matter of both hemispheres and the brainstem, usually the cerebellum is speared. Destruction of nerve cells, neuronophagia and perivenous cuffing by lymphocites and mononuclear cells indicate the viral nature of the infection. In white matter there is degeneration of medullated fibers (myelin and axis cylinders), acompagned by perivascular cuffing with mononuclear cells and fibrous gliosis. The MRI changes begin in the subcortical white matter and spread to the periventricular region. (1)

SSPE affects children and adolescents, rarely apearing beyong age of 10 years old. Typically there is a history of primary measels infection at a very early age, often before 2 years, followed by a 6-to 8 years of asymptomatic period.

The first stage is characterized by insidious intellectual impairment and behavioral abnormalities and visual disturbances due to chorioretinitis.

The second step develops 1-2 months later with acceleration of intellectual decline, repetitive symmetrical myoclonic jerks and specific EEG changes and focal or generalised seizures.

The third stage usually last for 1-4 months, patient become uncommunicative and develop ataxia, spasticity, choreoathetosis and dystonmia, with dissapearance of myoclonus.

The final stage last for monthes or years and is characterised by stupor, coma, autonomic disturbances and death. (6)

The particularity of our case is the unilaterality of the myoclonic jerks. Also, there is a relatively old age of debut of symptoms. (7)

Because of the presence of atypical extrapyramidal sympotoms, the differential diagnosis includes acute disseminated encephalonyelitis, Lafora disease, juvenile neuronal ceroid lipofuscinoses. Also, Schilder demyelinative disease must be ruled out.

The findings of periodic complexes on EEG, elevated gamma globlin and oligiclonal bands in the CSF and elevated measles antibody titers in the serum and CSF are arguments for diagnosis of SSPE.

No effective treatment is available. Amantadine, isoprenosine, intrathechal administration of alpha interferon, intravenous immunoglobulins were tried with no success.

SSPE is one of the most important complications of measles in children and adolescents. SSPE still occurs frequently in countries with insufficient measles immunization, and unfortunately, there is no cure for SSPE.

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