Atypical presentation of adult-age onset subacute sclerosing panencephalitis

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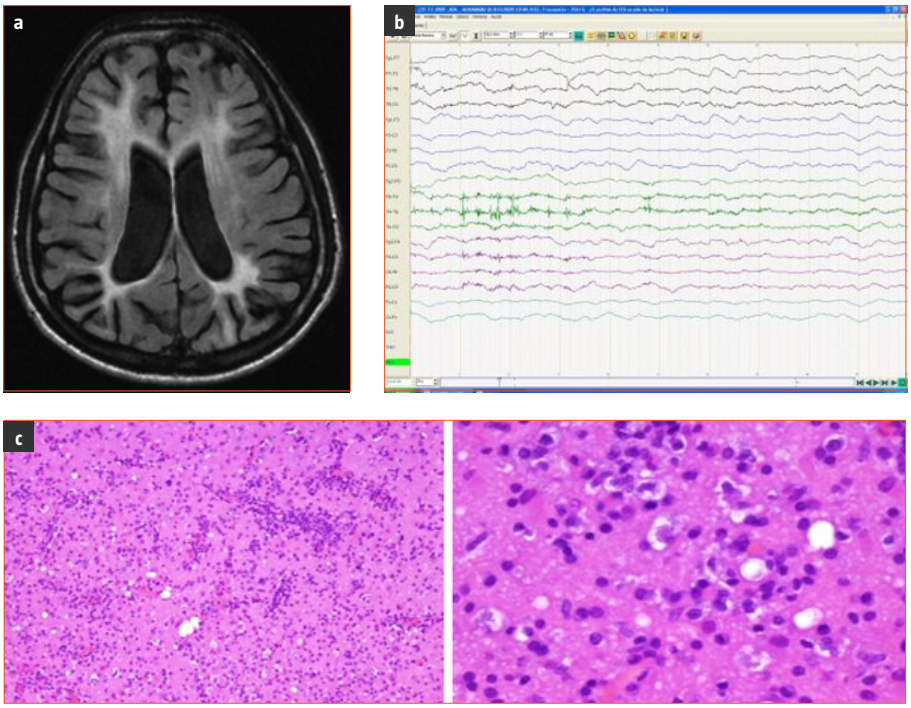

Figure. a) Brain MRI, FLAIR sequence: diffuse white-matter disease and generalized brain atrophy; b) EEG: generalized periodic high amplitude sharp waves (Radermecker complexes); c) Biopsy of the frontal lobe: demyelinated areas, small perivascular cuffs of lymphocytes and moderate microglial proliferation.

Subacute sclerosing panencephalitis (SSPE) is an inflammatory disorder of the central nervous system (CNS) caused by an aberrant measles virus infection [1,2]. Its presentation is rare in developed countries [1] and in adults [3,4]. Visual manifestations are the commonest mode of clinical presentation [4]. The disease apparently has a more aggressive course in adults. We report the case of a 22-year-old man presenting with an atypical adult-onset SSPE in a developed country with a histological confirmed diagnosis.

A 22-year-old man was admitted to our hospital because of behavioral changes. Four days before admission he was found lying on the road. A cranial computed tomography (CT) revealed generalized atrophy. Under the suspicion of psychiatric disorder, neuroleptic treatment was started. While on the emergency room, he had a tonic-clonic seizure.

He was born in Pakistan and had travelled to Spain 3 months before admission. Three years earlier, after his father's death, he suffered a reduction in spontaneous speech and increasing shyness which were interpreted as a reactive depression. Three months before admission to our hospital, he suffered a change in his behavior, with progressive memory impairment.

On neurologic evaluation, he had spontaneous ocular opening and mutism. Pupils were midriatic, reactive. He moved his four extremities symmetrically with no apparent sensitivity disorder. There was increased tone in arms and legs. The deep-tendon reflexes were brisk with bilateral patellar and ankle clonus. The plantar responses were flexor. The results of blood and urine tests were normal. Toxic screening was negative. Testing of blood for antibodies to the human immunodeficiency virus, rapid-plasma-reagin testing, and a paraneoplastic antibody panel were negative.

The analysis of the cerebrospinal fluid (CSF) revealed high levels of protein with absence of cells and normal levels of glucose; no organisms were detected on Gram's staining and PCR testing for JC virus was negative. An electroencephalogram showed diffuse theta-range background slowing without epileptiform activity. Magnetic resonance imaging (MRI) study of the brain showed diffuse white-matter disease and generalized brain atrophy (Figure, a). There was no enhancement after the administration of gadolinium. The determination of arylsulphatase and galactosylceramide β-galactosidase (GALC) were negative. Ophthalmologic and electrophysiology studies were normal.

The patient became quadriparetic with spasticity, akinetic mutism, dysphagia, and autonomic failure with loss of thermoregulation. In order to find a diagnosis, an MRI-guided stereotactic biopsy of the frontal lobe was performed. Microscopical examination showed demyelinated areas, small perivascular cuffs of lymphocytes, which extended into the adjacent parenchyma, with moderate microglial proliferation (Figure, c).

After some weeks, myoclonus appeared. Another EEG showed generalized periodic high-amplitude sharp waves (Figure, b). The measles immunoglobulin G titer was over 1:45,000 on CSF analysis and it was greater than the serum titer (1:450,000). The measles-specific cerebrospinal fluid to serum Ig G antibody index was elevated at 16.1 (normal value < 1.4). Measles specific PCR [6] on a CSF specimen withdrawn 2 years after the initial hospital admission was negative. However, negative results on CSF in cases of subacute sclerosing panencephalitis have been previously reported [7].

Approximately fifty cases of SSPE have been reported in those over 18 years of age [3,4]. Visual manifestations are the commonest clinical presentation [5]. The disease apparently has a more aggressive course in adults and it is rapidly fatal in the majority of patients.

Our case had clinical presentation of this uncommon disease in an unusual age of onset without myoclonus and typical EEG changes until the late stage. The diagnosis was supported by the biopsy. Alternatively, the suspicion of SSPE may be based upon characteristic clinical manifestations, the presence of Radermecker complexes and the demonstration of raised antibody title against measles in the plasma and cerebrospinal fluid [4]. Our patient did not fulfill these criteria until the late stage of the disease when he developed clinical, electric and immunologic criteria.

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