Fulminating Adult-Onset Subacute Sclerosing Panencephalitis in a 49-Year-Old Man

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Context: Subacute sclerosing panencephalitis (SSPE) is a rare, slow viral infection caused by a defective measles virus. It is characterized by progressive mental deterioration associated with motor impairment and prominent myoclonus. In about 10% of all cases, the disease can progress rapidly and lead to death within a few months. The oldest previously reported fulminating case was in a 25-year-old man.

Objective: To emphasize the relationship between retinal involvement and acute SSPE by reporting the case of a 49-year-old man with clinical, laboratory, and pathological evidence of acute SSPE.

Setting: Hôpital de l’Enfant-Jésus, Quebec, Quebec.

Report of a Case: This man was referred to the Department of Neurological Sciences on March 21, 2001, because of recent behavioral changes and progressive cognitive impairment over the past few months. Medical history was unremarkable except for an episode of measles in his childhood. Neurological examination showed bilateral myoclonic jerks. Ophthalmic examination revealed bilateral macular swelling and papilledema. Electroencephalography showed periodic sharp and slow-wave discharges. Magnetic resonance imaging showed bilateral diffuse T2-signal hyperintensities in both periventricular and subcortical white matter. Cerebrospinal fluid antimesles antibody titers were highly positive. An Omaya reservoir was inserted and therapy using a combination of high-dose intrathecal interferon alfa and oral isoprinosine were administered for 6 weeks. Despite transient subjective improvement in the patient’s condition, it continued to deteriorate, he became bedridden, and he died on June 26, 2001.

Conclusion: To our knowledge, this patient is the oldest case of SSPE reported in the literature. This patient and other patients with acute SSPE associated with bilateral macular swelling described in the literature raised the possibility of measles virus–acquired virulent neurotropism in the retina before invading the central nervous system.

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SUBACUTE SCLEROSING panencephalitis (SSPE) is a rare subacute inflammatory and degenerative disease of the entire brain, representing a slow viral infection caused by defective measles virus. Subacute sclerosing panencephalitis follows measles infection by several months to several years (mean interval, 7 years) and usually affects children or young adults. It has a gradual, progressive course, leading to death within a few years. In about 10% of all cases, the disease can progress more rapidly and lead to death within a few months. A broad spectrum of visual disorders has been described, including papilledema, papillitis, retinitis, chorioretinitis, and optic nerve pallor as well as homonymous visual field defects and cortical blindness.

The diagnosis of SSPE is based on the characteristic clinical and electroencephalographic findings (periodic complexes) and demonstration of elevated antibody titers against measles in the plasma and cerebrospinal fluid (CSF). High levels of CSF γ-globulin can also help to make the diagnosis. Brismar et al4 evaluated the magnetic resonance imaging scans of 52 patients at different stages of SSPE and found that the earliest findings were focal, intense lesions of white matter.

Few other cases of SSPE with progressive mental deterioration after the age of 35 years have been reported. Furthermore, the oldest previously reported fulminating case was a 25-year-old man, who died within 6 weeks.

REPORT OF A CASE

A 49-year-old, right-handed man was referred to the Department of Neurological Sciences on March 21, 2001, because...
of recent behavioral changes and progressive cognitive impairment over the past few months. Medical history was unremarkable except for an episode of measles in his childhood when he was about 2 years old, according to his mother. No history of encephalitis was reported. On August 1, 2000, his wife began to notice a progressive change of personality and later his efficiency at work deteriorated. During the last month before hospital admission, his wife noticed memory problems, confusion, apathy, and an inability to stay alone without supervision. She also began to observe nocturnal myoclonic jerks.

Results of the initial mental status examination revealed visual hallucinations, poor attention and concentration, disorientation, perseveration, anomia, acalculia, and ideational apraxia. Findings from a basic neurological examination showed bilateral Gegenhalten, bilateral myoclonic jerks with hyperreflexia, and bilateral Babinski sign. Ophthalmic examination revealed bilateral macular swelling and papilledema. The remainder of the neurological examination was unremarkable.

Extensive medical and laboratory workup was done to rule out other diseases; the results of these tests showed no abnormalities. The electroencephalogram showed periodic sharp and slow-wave discharges. The magnetic resonance imaging scans showed bilateral diffuse T2-signal hyperintensities in both periventricular and subcortical white matter. Serum measles IgG antibody titer was positive at 1:18200 and CSF antimeasles antibody titer was highly positive at 1:1800. The antibody titers confirmed the diagnosis. Furthermore, CSF examination findings revealed IgG at 29%. The cerebral biopsy specimen was positive for measles reverse transcriptase–polymerase chain reaction. An Omaya reservoir was inserted and therapy using a combination of high-dose intrathecal interferon alfa and oral isoprinosine were administered for 6 weeks. Despite transient subjective improvement in the patient’s condition, it continued to deteriorate, progressed along the stages described by Risk and Haddad, and he became bedridden, and died on June 2001.

To our knowledge, this the patient herein is the oldest subject in a case of SSPE ever reported in the literature. Furthermore, it is a fulminant case, found in only about 10% of all cases of SSPE. Characteristic clinical and electroencephalographic findings (periodic complexes) were observed and we were able to demonstrate high levels of CSF γ-globulin and mostly the presence of elevated antibody titers against measles in the plasma and CSF. Ophthalmic examination revealed bilateral macular swelling, an observation already described in 2 other cases of acute SSPE. This association raised the question of the relationship between retinal involvement and the more acute forms of SSPE. Perhaps the measles virus acquired virulent neurotropism in the retina before invading the central nervous system.

Dementia with rapid decline is a potential presentation of SSPE, even in middle age. So far treatment has been somewhat disappointing, although some benefits have been reported. The importance of providing an early diagnosis will be particularly relevant as more effective treatments might become available with research.

REFERENCES