Commentary

Subacute sclerosing panencephalitis (SSPE), also known as Dawson's encephalitis, is a rare and severe disease caused by chronic and persistent immunoresistant measles virus infection, which originates as inflammation and progressive demyelinization of the central nervous system. SSPE typically presents in children and adolescents, but there are a very few cases reported in adulthood.[1]

The occurrence of SSPE has decreased in Western countries as a result of generalized measles vaccination coverage, with an incidence, for example, in the United States of 0.6 cases per 1,000,000 inhabitants in 1980, whereas the annual incidence in populations who are not immunized is 5-10 cases per 1,000,000 inhabitants per year.[1,2]

Adult-onset SSPE usually presents in subjects between 20 and 35 years of age, with a mean of 25 years, and the duration of disease ranges from 8 months to 6 years (mean 24 months). The presenting features and the clinical course are very variable[3]; however, the initial symptoms usually include conductual and behavioral changes followed, after weeks or months, by myoclonus symptoms and progressive cognitive impairment. In a later phase, other symptoms reported are seizures, visual alterations, cerebellum-related symptoms, alterations of the pyramidal and extrapyramidal systems and, finally, after a period of vegetative status, the patient dies. In a small percentage of cases, stabilization or rare improvement of the patient's clinical condition may be observed,[4] with negative responses to attempts of symptomatic treatment, such as oral isoprinosine alone or associated with intraventricular interferon.[5]

Standard diagnostic methods reveal hyperintense lesions in T2-weighted sequences at cortisubcortical level and periventricular white matter substance, with certain preference for the occipitotemporal regions, in the brain magnetic resonance scan. The electroencephalogram (EEG) may typically disclose synchronous and bilateral periodic complexes of slow waves recurring at intervals of 5-7 s (Radermecker complexes). During the course of the disease, complexes precede myoclonus spasms, but SSPE patients with completely nonspecific EEG manifestations have been reported.

The definitive diagnosis is based on the demonstration of high titers of antimeasles antibodies in the cerebrospinal

fluid (CSF).[1] The presence of a high CSF: Serum ratio of specific IgG antibodies against measles supports the diagnosis. Histological findings in sterotactic biopsies or necropsy studies are characterized by a chronic leptomeningeal, perivascular, and parenchymatous chronic inflammatory infiltrate, with neuronal degeneration, gliosis, demyelinization, and astrocyte proliferation. Crowdy type A inclusion bodies are intranuclear or intracytoplasmatic viral particles found both in the neurons and glial cells. Neurofibrillary tangles is another characteristic finding, particularly when the disease has been present for some years.[2,6]

In adult patients with SSPE, clinical manifestations may be atypical and heterogeneous, with absence of myoclonus or EEG periodic complexes, as we can observe in Mahendra *et al*'s[7] case report; for this reason, a high clinical suspicion index for the diagnosis is required, and an extensive differential diagnosis should be made to exclude metabolic, demyelinating, genetic, infectious, and paraneoplastic diseases.[8] In some cases, brain biopsy has been the only method to establish a definitive diagnosis of the disease.[2,6]

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References

1. Jagtap SA, Nair MD, Kambale HJ. Subacute sclerosing panencephalitis: A clinical appraisal. Ann Indian Acad Neurol 2013;16:631-3.
2. González de la Aleja J, Posada IJ, Sepúlveda-Sánchez JM, Galán L, Conde-Gallego E, Ricoy-Campo JR. Adult-onset subacute sclerosing panencephalitis: Clinicopathological findings. Rev Neurol 2005;40:729-32.
3. Valenzuela V, Miranda M, Aguilera L, Fabres L, Galdames D. Adult onset subacute sclerosing panencephalitis. Report of two cases with atypical presentation. Rev Med Chil 1999;127:589-94.
4. Sharma V, Biswas D. Steroid-responsive myoclonus in adult-onset subacute sclerosing panencephalitis. J Neuropsychiatry Clin Neurosci 2013;25:E30-2.
5. Eroglu E, Gokcil Z, Bek S, Ulas UH, Ozdag MF, Odabasi Z. Long-term follow-up of patients with adult-onset subacute sclerosing panencephalitis. J Neurol Sci 2008;275:113-6.
6. Vilas D, Becerra JL, Lozano M, Soriano A, Matas L, Martro E, *et al*. Atypical presentation of adult-age onset subacute sclerosing panencephalitis. Rev Neurol 2012;54:60-1.
7. Javali M, Menon R, Chakor R. Adult onset subacute sclerosing panencepahlitis: Lessons learnt from an atypical presentation. J Neurosci Rural Pract 2014 2014;5:310-3.
8. Arboix A, Bechich S, Oliveres M, Garcia-Eroles L, Massons J, Targa C. Ischemic stroke of unusual cause: Clinical features, etiology and outcome. Eur J Neurol 2001;8:133-9.