

RAPID COGNITIVE DECLINE IN A PATIENT WITH MULTIPLE SCLEROSIS: A CHALLENGING DIAGNOSIS TOWARDS SUBACUTE SCLEROSING PANENCEPHALITIS

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BACKGROUND

Subacute sclerosing panencephalitis (SSPE) is a rare, progressive and fatal neurological disorder, caused by the persistence of a mutated measles virus in the central nervous system, years after the primary infection. Early neurological/psychiatric symptoms are often misdiagnosed, making the diagnosis challenging, especially in adults.

MATERIAL AND METHODS

We report a case of a 47 year-old woman with a known diagnosis of multiple sclerosis (MS), never treated with disease-modifying therapy, who presented a progressive cognitive-behavioral deterioration, initially attributed to a possible MS relapse. The brain MRI showed only the known demyelinating lesions (Fig. 1), so the patient was admitted to the Neurology Unit for the diagnostic work-up. The EEG showed periodic triphasic waveforms, supporting the inclusion of prion disease and SSPE among the differential diagnosis (Fig. 3). A follow-up brain MRI (fifteen days later) revealed new bilateral cortical-subcortical T2-hyperintense lesions, without atrophy or contrast enhancement, consistent with an encephalopathic process (Fig. 2). The Anti-channel antibodies and neuronal surface antigens, related to autoimmune/paraneoplastic syndromes, turned out negative in both serum and cerebrospinal fluid (CSF). A negative 14-3-3 and weakly positive total Tau proteins, suggested a non-specific neuronal damage rather than a prion disease.

RESULTS

Intrathecal synthesis of immunoglobulins showed an elevated CSF/serum antibody ratio for measles virus, indicating the diagnosis of SSPE. Although neuroimaging did not evidence atrophy, the extensive white matter involvement also supported the diagnosis. Follow-up neuroimaging showed progressive radiological deterioration, correlating with the clinical fulminant course. The patient died three months after the onset of the neurological deterioration. The coexistence of MS was misleading, since the misinterpretation of the symptoms delayed the admission to hospital. CSF biomarkers, electroencephalographic findings and neuroimaging played a crucial role in guiding the diagnostic process toward such a rare condition, despite the absence of specific treatment options.

CONCLUSIONS

This case underlines the importance of reassessing the diagnosis when the clinical features are not typical for MS. Moreover, although SSPE commonly occurs in children who had measles, it must be considered also in adults with a previous history of measles infection. The fatal course of SSPE underscores the importance of measles vaccination for preventing measles and its potential neurological sequelae.

REFERENCES

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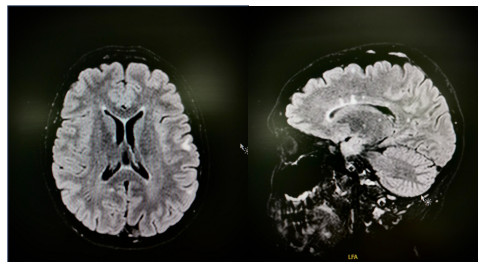


Fig. 1 - Brain MRI on admission

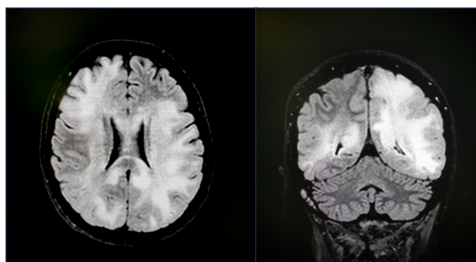


Fig. 2 - Brain MRI after fifteen days

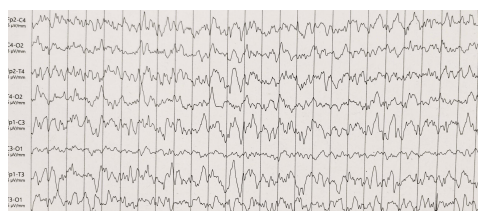


Fig. 3 - EEG showing high-amplitude triphasic waves

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