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Autoimmune Encephalitis: Onset, Diagnosis and Management of A Hidden Disease

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Background: Autoimmune encephalitis (AE) represents a syndrome characterized by brain inflammation (1.2/100,000 person-year). Most cases present specific autoantibodies (autoAbs) in the serum and/or cerebrospinal fluid (CSF). Some patients do not show autoAbs, representing further clinical challenge. We describe the history of a patient, from atypical onset to therapeutic management.

Case History: A 62-year-old man referred to emergency department for hyponatremia and confusional state. He had hystory of hypertension, recent major depressive episode (3-4 months before) treated with antidepressants (including sertraline), subsequent finding of marked hyponatremia and doubtful generalized critical episodes treated with anticomitial therapy. On neurological evaluation, he appeared alert, confabulating, disoriented. Multiple EEGs, cerebral MRI and Holter ECG were normal. Hyponatremia in probable SIADH was managed with Tolvaptan. CSF showed mirror pattern (barrier damage); mature lymphocytes to cytology; increase in B cells and NK cells to blood flow cytometric exam. Screening for specific antineuron autoAbs was positive. He was treated with intravenous immunoglobulins 30 gr for 3 days, followed by Rituximab, with a progressive neurologic improvement.

Discussion: Diagnosis of AE is difficult, and needs awareness, rapid search for clinical, imaging and laboratory data, like extensive antibody screening. Adequate therapy determined progressive improvement of neurological sequelae of our patient.

References

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