Steroid-responsive auto-immune encephalitis as a paradigm of neuropsychiatric differential diagnosis: case report and review

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Background: Autoimmune encephalitis (AE) has gained clinical attention over the past years, especially since the identification of autoantibodies directed at brain structures, which led to the emergence of several new clinicopathological entities [1]. These advances have made it possible to identify previously unrecognized clinical conditions in patients with a wide range of unexplained neuropsychiatric symptoms and signs, often leading to psychiatric hospitalization, since AE presentation is often deemed psychiatric [2].

Objectives: We report a clinical case of a patient with a steroid-responsive autoimmune encephalitis (SREAT), a subtype of AE, initially evaluated in a psychiatric setting. We further review the current perspectives on these conditions and their impact on psychiatric practice.

Methods: We carried out a narrative literature review by performing a search on MedLine for English-written articles. The query used was “((autoimmune OR Hashimoto OR limbic) AND encephalitis) AND (psychiatric OR psychiatry))”.

Case Report: A 31 year-old woman, with a medical history of atypical squamous cells of undetermined significance cervical cytology, a previous episode of undefined depressive-anxious disorder, and without any current medication, was admitted in a metropolitan psychiatric emergency department due to a 3-day behavioral disorder. Two days earlier, her mother noticed she was “somewhat different” that day; she left work without warning, something she never did before. Next day, contrary to usual routines, she didn’t prepare dinner for her family and retired to bed. The morning after, she abandoned home, leaving her 2-year-old daughter behind alone, took her car and had a minor car accident, to which she presented post-event amnesia. Then, she was intercepted by some neighbors, when lost, who noticed her speech was incoherent. For that reason, they conducted her to the nearest general ED, where basic tests were performed, including blood alcohol concentration and urinary drug screening, and brain CT – all normal, except a slight increase of aspartate transaminase. Mental State Examination revealed spatiotemporal disorientation, affective incontinence, disorganized speech and persecutory delusions.

Neurologic examination revealed solely deep tendon hyperreflexia of the four limbs. Cerebrospinal fluid analysis was also normal. Blood test revealed thyroiditis. Immunological testing revealed high levels of anti-thyroglobulin (>4) and anti-peroxidase (>5) autoantibodies; other immunology tests were negative (including anti-nuclear and anti-neuronal autoantibodies). Thyroid ultrasound disclosed thyroiditis.

The patient was prescribed IV methylprednisolone 1g/day for 5 days followed by oral prednisolone, with complete remission of neuropsychiatric symptoms around by the 2nd day of treatment, confirming SREAT diagnosis.

Review: SREAT is a rare specific type of AE associated with or without Hashimoto’s thyroiditis. Autoantibodies may be present or not, and thyroid function can be normal. Its prevalence is unestablished and remains underdiagnosed. Women are predominantly affected and the age of presentation varies widely. The clinical picture is pleomorphic, often with neuropsychiatric symptoms and signs, and its presentation may vary from mild cognitive impairment to status epilepticus or coma, challenging diagnosis [4]. Psychotic symptoms are reported to occur in about 25% of cases [5]. Core diagnostic features include high levels of antithyroid autoantibodies and steroid responsiveness (up to 90% of cases) [4]. Its pathophysiology is not well-understood; direct causal relationship between thyroid antibodies and encephalopathy is unlikely [6].

Discussion: We present a clinical report of a case of SREAT, initially suggesting a possible dissociative or psychotic disorder. Atypical clinical features for a purely psychiatric disorder led to a neurologic evaluation, further confirming a neurologic etiology. This report typifies a paradigmatic neuropsychiatric case, demanding thorough psychiatric and neurologic investigation to achieve a reliable diagnosis and effective treatment that, otherwise, could have been missed, with significant consequences.

References