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LUPUS Cerebritis in SLE/SJOGREN'S Overlap Syndrome

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Abstract: ***This article reports on a rare case of lupus cerebritis in an 18 - year - old female, presenting with acute neurological symptoms and diagnosed against the backdrop of systemic lupus erythematosus SLESjogrens overlap syndrome, a type of mixed connective tissue disease MCTD. The patient exhibited symptoms such as seizures, altered sensorium, and autonomic dysregulation, without evident meningeal irritation or infection. Despite normal imaging and basic laboratory findings, cerebrospinal fluid analysis and specific autoantibody profiles confirmed the diagnosis. Treatment with intravenous glucocorticoids led to significant improvement within four days, followed by maintenance therapy. This case underscores the complexity of diagnosing neuropsychiatric manifestations of autoimmune diseases, particularly in the context of MCTD, and emphasizes the importance of comprehensive evaluation and management in such patients. It highlights the critical role of recognizing overlap syndromes in autoimmune diseases for prompt and effective treatment, showcasing the intricacies of autoimmune pathophysiology and the necessity for vigilant follow - up and personalized care.***

Keywords: Systemic Lupus erythematous, Sjogern's Syndrome, Lupus Cerebritis, Mixed Connective tissue Disease, Autoimmune Neuropsychiatric Disorders

1. Introduction

Psychotic events occur less commonly in SLE and usually occur early in the course of disease (within 1 to 3 years). In this case, Lupus cerebritis occurs in background of SLE/Sjogren's overlap syndrome. Autoimmune connective tissue diseases (CTD) include five classical diseases: rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), Sjogren's syndrome, scleroderma, and myositis. Mixed connective tissue disease (MCTD) is a rare systemic autoimmune disease and is referred to as "overlap syndrome" Patients with MCTD have at least two defined CTDs and a distinct antibody known as anti - U1 - ribonucleoprotein (RNP).

2. Case Report

Here: An 18 year old female presented with acute onset of altered sensorium and episodes of seizure like activity associated with involuntary defecation and micturition along with dysphagia and dry mouth.

- Butterfly rash over face with nasolabial sparing was found. •No signs of meningeal irritation and patient was afebrile
- DTRs in B/L UL and LL were normal with sensory system intact • B/L plantar reflex downwards and B/L pupils equal and reactive to light
- ON WORKUP • MRI Brain plain and contract was normal • Routine blood reports such as CBC, LFT, KFT, SERUM ELECTROLYTES were within normal limits and viral markers for hiv 1&2, hbsag, anti HCV were all negative • CSF analyis: cells 80cumm with mostly lymphocytes; protien 90mg/dl; sugar - 44mg/dl and ADA - 7.8 U/L • ANA by IFA (hep2) –positive; pattern - Nuclear (2+), speckled (Ac - 2, 4, 5) • ENA profile - AntiU1 RNP/sm+, Anti - sm+, SS - A/RO - 52, SS - B/La, anti ribosomal P strongly+
- DIAGNOSIS • SLE/Sjogren' s overlap syndrome with Lupus cerebritis was made and patient was started on iv glucocorticoids • Improvement was seen in 4 days • Patient was discharged on HCQS, Azathioprine & oral glucocorticoids.

3. Discussion

Lupus cerebritis is a rare neuropsychiatric manifestation of SLE which can present with seizures, altered mental status, headache, anxiety, depression, psychosis, and pseudodementia. It is a diagnosis of exclusion with complex etiology that could be attributable to various factors such as infections, drug use, brain abnormalities, and metabolic dysfunction [8]. Our patient also has an ongoing history of depression and anxiety. She subsequently presented with recurrent seizures further leading to an episode of status epilepticus that led to the severity of her overall health and hospitalization. Importantly, she was placed on various antiepileptic drugs and glucocorticoids for the management and control of lupus cerebritis. The highlight of this case pertains to MCTD and the way it can present with a focus on only one disorder symptomatically, which in our case was SLE.

4. Conclusion

Autoimmune diseases can present with non - specific symptoms and their prognosis could range from mild to severe. MCTD is a significant autoimmune disease that may present initially with mild symptoms or could progress to develop serious manifestations. This was a rare case of lupus cerebritis, a neuropsychiatric manifestation of SLE, in a patient with previously undiagnosed MCTD. It further highlights the importance of taking a detailed history in addition to accounting for the patient's medical and social history. . In general, any patient with an autoimmune disease

requires regular follow - ups, a prompt work up, and close monitoring to control the progress of underlying fatal complications.

References

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