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DIVERSE PRESENTATIONS OF AUTOIMMUNE ENCEPHALITIS: A CASE SERIES

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ABSTRACT

Autoimmune encephalitis can present with diverse clinical phenotypes, and a high index of suspicion is crucial for timely diagnosis. This case report emphasizes the need for a multidisciplinary approach, including neurology and immunology expertise, to optimize the management and outcome of patients with autoimmune encephalitis. This case series highlights the different neurological manifestations, diagnostic challenges, and treatment outcomes associated with this complex neurological disorder.

INTRODUCTION

Autoimmune encephalitis (AE) is a group of disorders characterized by inflammation of the brain caused by an immune response against neuronal components. This condition often presents with a wide range of neurological and psychiatric symptoms.[1] Patients with autoimmune encephalitis may experience cognitive dysfunction, altered behavior, seizures, movement disorders, and sometimes psychosis. Symptoms can be sub-acute in onset, evolving over days to weeks. Diagnosing AE involves a combination of clinical evaluation, imaging studies, cerebrospinal fluid analysis, and autoantibody testing.[2] Detecting specific autoantibodies, like anti-NMDA receptor antibodies, can be crucial in confirming the diagnosis. [3] This case series presents a compilation of diverse clinical scenarios involving patients diagnosed with autoimmune encephalitis (AE). The cases highlight the varied manifestations, diagnostic challenges, and treatment outcomes associated with this complex neurological disorder.

Case 1: Limbic Encephalitis with Anti-NMDA Receptor Antibodies

The 32-year-old-female initially presented with behavioral changes, displaying paranoia and auditory hallucinations. Over the next few days, she developed generalized seizures and progressive cognitive decline. Neurological examination revealed signs of limbic encephalitis. Cerebrospinal fluid analysis demonstrated pleocytosis and elevated protein levels. Magnetic Resonance Imaging (MRI) showed hyperintense signals in the limbic structures. Notably, serum testing revealed the presence of anti-N-methyl-D-aspartate (NMDA) receptor antibodies, confirming the diagnosis of autoimmune encephalitis. The patient received high-dose corticosteroids and intravenous immunoglobulin, leading to a gradual improvement in her symptoms. Additionally, plasmapheresis was initiated, resulting in a sustained remission of the autoimmune response. Follow-up assessments showed resolution of seizures and cognitive improvement.

Case 2: Autoimmune Encephalitis Mimicking Stroke

A 45-year-old male experienced acute-onset hemiparesis and aphasia, initially raising suspicion for stroke. However, neuroimaging revealed inflammatory changes, and anti-GAD65 antibodies were identified. Immunosuppressive therapy resulted in neurological recovery.

Case 3: Pediatric Autoimmune Encephalitis with Movement Disorders

A 10-year-old presented with choreiform movements and behavioral changes. Autoantibody testing identified anti-D2R antibodies. Treatment with corticosteroids and rituximab led to resolution of symptoms, emphasizing the importance of recognizing autoimmune etiologies in pediatric cases.

Case 4: Autoimmune Encephalitis Associated with Ovarian Teratoma

A 22-year-old female presented with seizures and altered consciousness. Detection of anti-Ma2 antibodies prompted an evaluation for ovarian teratoma, which was surgically removed. Subsequent immunotherapy resulted in neurological improvement.

Case 5: Autoimmune Encephalitis Associated with Facio-Brachial Seizure

The 45-years-old male patient initially presented with recurrent episodes of asymmetric facial and upper extremity dystonic movements, accompanied by confusion. These episodes were initially misdiagnosed as focal seizures, delaying the identification of the underlying autoimmune etiology. Continuous video-EEG monitoring during an episode captured distinctive faciobrachial dystonic seizures, prompting further investigation. Cerebrospinal fluid analysis revealed lymphocytic pleocytosis and elevated protein levels. Serum testing identified the presence of anti-LGI1 antibodies, confirming the diagnosis of autoimmune encephalitis with limbic involvement. The patient was initiated on a combination of corticosteroids and intravenous immunoglobulin, leading to a significant reduction in seizure frequency and cognitive improvement. Long-term immunosuppressive therapy with rituximab was subsequently administered to maintain remission.

DISCUSSION

This case series illustrates the diverse clinical presentations of autoimmune encephalitis, emphasizing the need for a broad differential diagnosis. The inclusion of pediatric cases and those associated with tumors underscores the importance of age-specific considerations and comprehensive investigations.

In a recent case series by Dalmau et al., 50 patients with autoimmune encephalitis were evaluated. The study highlighted the diversity of clinical manifestations and the importance of recognizing different autoantibodies

associated with AE. Notable cases included those with anti-LGI1 antibodies presenting with limbic encephalitis and anti-CASPR2 antibodies linked to neuromyotonia and Morvan's syndrome.[4] Management often involves immunotherapy, including corticosteroids, intravenous immunoglobulins, and plasmapheresis. For some patients, immunosuppressive agents like rituximab or cyclophosphamide may be necessary.[5] Early recognition and intervention are crucial for a better prognosis.

The last case underscores the importance of recognizing atypical presentations of autoimmune encephalitis, particularly when faciobrachial dystonic seizures are the primary manifestation. It highlights the necessity of detailed video-EEG monitoring and comprehensive autoimmune panel testing for accurate diagnosis.[6]

CONCLUSION

Autoimmune encephalitis encompasses a spectrum of clinical manifestations, and timely recognition of specific autoantibodies is crucial for accurate diagnosis and targeted treatment. This case series aims to enhance awareness of the varied phenotypes, facilitating improved clinical management. Prognosis varies among individuals, and factors such as prompt treatment initiation and the specific autoantibody involved can influence outcomes. Some patients may experience significant recovery, while others may face residual neurological deficits.

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