NEUROIMAGING CHANGES IN PATIENTS WITH SUBACUTE SCLEROSING PANENCEPHALITIS: DYNAMICS OF MRI AND EEG EXAMINATIONS

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a progressive, often fatal disease caused by persistent measles virus infection, typically contracted in early childhood. This study focuses on the neuroimaging findings in SSPE patients, specifically using Magnetic Resonance Imaging (MRI) and Electroencephalography (EEG). These modalities are essential for diagnosing SSPE, tracking disease progression, and monitoring treatment response. This article explores how MRI and EEG findings evolve with the stages of SSPE, highlighting the importance of these tools in clinical decision-making. Advanced neuroimaging methods have become indispensable in understanding SSPE pathophysiology and improving patient outcomes

Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare but devastating progressive neurodegenerative disorder caused by a chronic and persistent measles virus infection in the central nervous system. The disease predominantly affects children and young adults, manifesting years after the initial measles infection, often occurring in infancy or early childhood. While measles itself is preventable through vaccination, SSPE develops in a small fraction of individuals due to an atypical immune response to the virus, resulting in progressive cognitive and motor impairment, seizures, and eventually, death (Jabbour et al., 2017).

Globally, the incidence of SSPE varies, with higher rates in regions where measles outbreaks are more common due to insufficient vaccination coverage. The prevalence of SSPE is closely tied to the effectiveness of measles immunization programs, making the disease largely preventable in countries with strong public health initiatives. However, for individuals affected by SSPE, the prognosis remains grim, with most patients experiencing rapid deterioration over a period of one to three years if untreated (Anlar et al., 2020).

Neuroimaging plays a pivotal role in the diagnosis and management of SSPE, with MRI and EEG being the primary tools for assessing the structural and functional changes in the brain. MRI is particularly valuable for detecting white matter lesions and other structural abnormalities, while EEG provides crucial insights into the electrophysiological disturbances

characteristic of SSPE. Together, these modalities help clinicians not only diagnose SSPE but also monitor its progression and the effects of treatment interventions (Gascon, 2018). This article examines the dynamic neuroimaging changes observed in SSPE patients, focusing on the evolving patterns in MRI and EEG as the disease progresses.

Epidemiology of SSPE

SSPE is rare but occurs with a frequency of approximately 1 in 10,000 to 1 in 100,000 cases of measles. The disease is more common in regions with endemic measles outbreaks, where vaccination rates are low. Several countries, especially in parts of Africa, Asia, and Eastern Europe, have reported higher rates of SSPE due to inadequate measles vaccination coverage. In contrast, countries with well-established vaccination programs, such as the United States and Western Europe, have significantly reduced the incidence of both measles and SSPE (Molnar et al., 2016).

The pathogenesis of SSPE is linked to a persistent infection with a defective form of the measles virus. Unlike typical measles, the virus involved in SSPE does not trigger a full immune response, allowing it to remain dormant within the brain for years. The exact mechanism by which the virus evades the immune system and reactivates years later is not fully understood, but it leads to widespread inflammation, demyelination, and neuronal loss, characteristic of SSPE (Gascon, 2018).

Pathophysiology of SSPE

The hallmark of SSPE pathophysiology is progressive encephalitis caused by the chronic measles virus infection. The virus remains within the central nervous system, where it leads to demyelination, neuronal degeneration, and gliosis, which together cause the characteristic cognitive and motor deficits seen in affected patients (Garg et al., 2020). As the disease progresses, the inflammatory response to the virus exacerbates the damage to neural tissues, leading to widespread destruction of both gray and white matter.

One of the primary pathological features of SSPE is the progressive demyelination of the brain's white matter, particularly in the periventricular and subcortical regions. This demyelination is responsible for the neurological decline observed in SSPE, as the loss of myelin impairs the efficient transmission of nerve signals across the brain. Gliosis, or the proliferation of glial cells in response to the inflammation, is another significant pathological finding, as it contributes to the structural changes seen on MRI (Molnar et al., 2016).

Neuroimaging Techniques in SSPE Diagnosis

Magnetic Resonance Imaging (MRI)

MRI is a non-invasive imaging technique that provides detailed images of brain structures, making it an essential tool in diagnosing and monitoring SSPE. In the early stages of SSPE, MRI may reveal subtle abnormalities, such as small white matter lesions in the periventricular areas or the basal ganglia. These early lesions are often indicative of the demyelination process that characterizes the disease (Molnar et al., 2016).

As the disease progresses, MRI findings become more pronounced. In the intermediate stages of SSPE, MRI typically shows more extensive involvement of the white matter, with hyperintense lesions on T2-weighted and FLAIR sequences. These lesions represent areas of inflammation, demyelination, and gliosis, which contribute to the progressive neurological decline seen in patients (Gascon, 2018).

In advanced SSPE, MRI may reveal widespread cortical atrophy, ventricular enlargement, and diffuse white matter damage. These findings reflect the chronic neurodegenerative process occurring in the brain. In particular, hyperintense lesions in the thalamus and basal ganglia are common in advanced disease and are associated with severe cognitive and motor impairments (Garg et al., 2020). The dynamic changes observed on MRI correspond closely with the clinical progression of SSPE, making it a valuable tool for tracking disease evolution.

Electroencephalography (EEG)

EEG is a critical tool for diagnosing SSPE due to its ability to detect characteristic electrophysiological abnormalities associated with the disease. The hallmark EEG finding in SSPE is periodic slow-wave complexes (PSWCs), which are generalized, high-amplitude discharges that occur every 4 to 10 seconds. These PSWCs are typically present in the early stages of SSPE, even before significant clinical symptoms manifest, making EEG an essential diagnostic tool (Prashanth et al., 2018).

As the disease progresses, the frequency and amplitude of PSWCs increase, correlating with the worsening neurological condition of the patient. In advanced stages of SSPE, EEG may show diffuse cortical suppression, indicating severe neuronal loss and cortical dysfunction. These EEG changes are closely linked to the structural changes observed on MRI, providing a comprehensive view of the disease's impact on both brain structure and function (Prashanth et al., 2018).

Dynamics of MRI and EEG Findings in SSPE

The integration of MRI and EEG findings is crucial for understanding the dynamics of SSPE progression. In the early stages of the disease, MRI may show only minor white matter changes, while EEG demonstrates characteristic PSWCs, often before clinical symptoms become severe. As the disease advances, MRI findings become more pronounced, with extensive demyelination and cortical atrophy, while EEG shows increasingly abnormal electrical activity.

The progression of neuroimaging findings corresponds closely with the clinical stages of SSPE:

1. **Stage I (Initial Stage)**: Patients may present with mild cognitive and behavioral changes. MRI may show minimal abnormalities or subtle white matter lesions, while EEG shows early PSWCs.
2. **Stage II (Intermediate Stage)**: Cognitive decline, motor disturbances, and myoclonus become more prominent. MRI reveals more extensive white matter lesions, and EEG shows more frequent and pronounced PSWCs.
3. **Stage III (Advanced Stage)**: Severe neurological impairment, including motor deficits, seizures, and coma, occurs. MRI shows widespread demyelination, cortical atrophy, and ventricular enlargement, while EEG shows diffuse cortical suppression and less organized electrical activity.
4. **Stage IV (End Stage)**: Patients reach a terminal stage with minimal brain activity. MRI shows advanced cortical atrophy, and EEG may show near-total electrical silence (Gascon, 2018).

Treatment Approaches and Challenges

Despite advances in neuroimaging and early diagnosis, the treatment of SSPE remains challenging. Currently, there is no definitive cure for SSPE, and treatment options are

primarily aimed at slowing disease progression and managing symptoms. Antiviral agents, such as ribavirin and interferon-alpha, have shown some efficacy in reducing the viral load and slowing the progression of SSPE, particularly when administered in the early stages of the disease (Anlar et al., 2020).

Immunomodulatory therapies, including intravenous immunoglobulin (IVIG) and corticosteroids, have also been used to reduce inflammation in the brain, although their long-term effectiveness remains uncertain. These therapies are most effective when combined with antiviral treatment, offering a multifaceted approach to managing SSPE (Molnar et al., 2016).

While early diagnosis through neuroimaging is crucial for maximizing the efficacy of treatment, the prognosis for SSPE remains poor, especially in regions where access to advanced medical care is limited. Future research should focus on developing more effective antiviral and immunomodulatory therapies, as well as improving our understanding of the underlying mechanisms of SSPE.

Conclusion

Subacute sclerosing panencephalitis remains a devastating and largely untreatable disease, with neuroimaging playing a pivotal role in its diagnosis and management. MRI and EEG are essential tools for detecting early brain changes, tracking disease progression, and guiding treatment decisions. The dynamic changes observed in MRI and EEG findings provide valuable insights into the pathophysiology of SSPE, underscoring the importance of continued research into more effective diagnostic and therapeutic approaches

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