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Editorial

Subacute sclerosing panencephalitis: A preventable terminal neurological illness

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1. Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare progressive neurodegenerative disease that affects the central nervous system, primarily the brain. It is caused by a chronic infection with a mutated form of the measles virus.[1] The disease usually occurs years after the initial measles infection and is often fatal. SSPE affects primarily children and young adults with an incidence of 4-11/1 lakh population.[2]

2. Pathogenesis

The pathogenesis of SSPE is not fully understood, but it is believed to be related to a persistent measles virus infection in the brain. The measles virus enters the body through the respiratory tract and then spreads to other organs, including the brain. The virus can infect and destroy brain cells, leading to the development of SSPE. It is thought that a mutation in the virus may allow it to persist in the brain for years after the initial infection. Multiple alterations in M (matrix) protein of envelope membrane in measles virus have been described in SSPE because of extensive point mutations in viral genome, might result in failure of immune system to eliminate the viral infection.[3] In addition, there is a deficient response of interferon to infected neurons in SSPE.[4] In addition, high levels of neutralising antibody are present in the serum and cerebrospinal fluid of SSPE patients. Recently studies have suggested that apoptosis of various cell types may contribute to the neuropathogenesis of measles virus infection, either as a direct effect of viral infection or of cytokine mediated responses, resulting in oligodendroglial and neuronal cell death in SSPE. Focal or diffuse perivascular infiltrates of lymphocytes, plasma cells, and phagocytes are present in the meninges and in the brain parenchyma of SSPE patients. Studies of inflammatory cell infiltrate in brain tissue from patients with SSPE have shown that the perivascular cells are predominantly CD4+T cells, with B cells seen more frequently in the parenchymal inflammatory infiltrate. Immune dysfunction, inflammatory mediators, and genetics have been hypothesized to play a role in the pathogenesis of SSPE. There is often evidence of neuronal degeneration, gliosis, proliferation of astrocytes, perivascular cuffing, lymphocytic and plasma cell infiltration, and demyelination. The parieto-occipital region of the brain is most severely affected, subsequently; pathological involvement spreads to the anterior portions of cerebral hemispheres, subcortical structures, brainstem, and spinal cord.

3. Clinical Features and Diagnosis

The onset of SSPE is usually insidious, with the initial symptoms being behavioral changes, cognitive decline, and motor disturbances. Patients may experience seizures, muscle spasms, and difficulty walking. As the disease progresses, patients may develop myoclonic jerks, rigidity, and dystonia. In the later stages, patients may become bedridden and unresponsive.[5] The diagnosis of SSPE is based on clinical presentation, cerebrospinal fluid (CSF) analysis, electroencephalography (EEG), and magnetic resonance imaging (MRI). The CSF analysis usually shows elevated levels of measles-specific antibodies and increased protein levels. EEG often shows periodic sharp wave complexes, which are characteristic of SSPE. MRI may show white matter changes and brain atrophy.[6]

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4. Management

The course of illness in SSPE is inevitably fatal in 95% cases within 1-3 years in majority of cases even with best possible treatment. There have been no definitive eradicative treatment for SSPE at present, and treatment is primarily supportive. Anti-seizure medications are used to control seizures, and muscle relaxants may be used to manage muscle spasms. Physical therapy may also be helpful in maintaining mobility and preventing contractures. Immunomodulatory therapies, such as interferon alpha and ribavirin, have been used in some cases, but their efficacy is unclear.[7,8] Immunoglobulin therapy which may be beneficial, is not well explored in SSPE.[9] There is a surge in SSPE in developing countries like India where there are barriers to vaccination against measles virus due to various sociocultural reasons. Prevention of SSPE is possible through vaccination against measles. There is an unmet urgent need among Indian neurologists for more well-defined treatment regimens for individuals with SSPE, even if the disease is still avoidable with vaccination.[10]

5. Source of Funding

None.

6. Conflict of Interest

None.

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