Current trends in the management of subacute sclerosing panencephalitis

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Abstract

Subacute sclerosing panencephalitis (SSPE) is slow viral encephalitic sequelae occurring secondary to measles infection. Symptoms occur insidiously and progresses to coma and death. This condition carries a very high mortality. This article discusses the various drug therapies tried in this condition. The currently proven drug regimens include Isoprinosine, Interferon alpha and their combination.

Introduction

Subacute sclerosing panencephalitis (SSPE) is slow viral encephalitic sequelae occurring years after infection with measles virus. The prevalence proportionately follows the incidence of measles. The exact pathophysiology of the disease is not completely known; however it is believed that factors causing a selective humoral over cellular immune response against the virus may play a role. A defective virus structure secondary to immature immune response mounted is also believed to play a role. The clinical features occur on an average of 6 years after measles virus infection and includes insidious onset of behavioral changes, myoclonus and involuntary movements progress sing to coma and death. The diagnosis is made clinically, supported by periodic complexes on electroencephalography, brain imaging suggestive of demyelination, and immunological evidence of measles infection.

Review

The pharmacologic management of SSPE includes symptomatic therapies and disease modifying agents. Many different anticonvulsants have been tried and no single agent has emerged as the best. With current evidence carbamazepine has been effective in few settings. There are case reports of successful treatment of myoclonus with trihexyphenidil and ketogenic diet. In the disease modifying drugs, Isoprinosine (inosiplex) and interferon have been studied in detail. Isoprinosine is an immune modulating substance which promotes lymphocyte proliferation, production of immunoglobulin and lymphokines that facilitates lymphocyte immune function once triggered by a viral antigen. It has been found to be effective in numerous trials. Interferon acts by activating Natural Killer cells and directly inhibiting virus replication. IFN alpha is administered intraventricularly and has found to be effective. Beta-interferon has also been successfully used in treating seven patients in a case series. Combined use of both Isoprinosine and IFN alpha has been found to be effective in a large multicentric trial, although conflicting reports of it being ineffective in the early stages of SSPE are present.

Other immune-modulating medications tried in SSPE include Cimetidine and thymus extract in combination with Isoprinosine, but not found to be effective. Ribavirin has been used as adjunct therapy, in addition to intra-ventricular IFN alpha, with minimal success. There are isolated reports claiming success with IVIF, amantadine, steroids and acyclovir, but none proven in multicentric trials. Rituximab (anti CD 20 antibody) has been tried in SSPE with no success. Flupirtine, an anti apoptotic agent which has been used with limited success in Alzheimer’s and prion diseases has been hypothesized to halt the disease progression in SSPE, but no clinical data is available yet.

Conclusion(s)

With the current available evidence only Isoprinosine, Interferon alpha and their combination appears to be effective in slowing the progression of SSPE. Irrespective of recent insights into the possible treatment options, prognosis is uniformly dismal. Therefore, the most successful strategy in the management of SSPE is the prevention of the primary disease by means of immunization.

Abbreviation(s)

SSPE -- Subacute sclerosing panencephalitis.
References

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