Subacute Sclerosing Panencephalitis (SSPE) - A Preventable Disaster

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Abstract
Subacute sclerosing panencephalitis is a rare chronic progressive encephalitis caused by measles virus. It occurs primarily in children and young adults. Death usually occurs within three years.

Introduction
The disease was first described by Dawson in 1934 under the title inclusion body encephalitis and extensively studied by Van Boqaert who renamed it as subacute sclerosing pancephalitis. It is a result of chronic measles infection. SSPE is a progressive neurological disorder of childhood and early adolescence characterized by inflammation of the brain caused as a complication of the measles virus. It can occur ten years after the initial measles infection and may be due to a defective immune response to the virus or a reactivation of the virus.

Case Report
A seventeen year old male was admitted with twitching movement of body mainly involving right upper limb and right lower limb, decreased verbal output and not recognizing relatives. There was no history of fever, vomiting, convulsions or head injury. He was not vaccinated against measles and had measles at ten years of age.

On examination, patient was vitally stable. General physical examination was unremarkable. On central nervous system examination, patient was conscious but not oriented. He was responding to verbal commands sluggishly. There was decreased verbal output. He was not recognizing his relatives.

There was emotional lability in the form of excessive crying and laughing. He had myoclonic jerks of right half of the body mainly involving right upper limb and right half of face.

Investigations
Routine investigations like CBC, ESR, BSF, RFT’s and LFT’s were within normal limits. CT scan brain was normal. In view of past history of measles, CSF was done and found to have measles IgG antibodies positive. EEG showed periodic bursts of 2-3 seconds of high voltage waves every 5-8 seconds followed by flat pattern suggestive of SSPE (Fig. 1). Patient was treated with sodium valproate and clonazepam. Myoclonic jerks reduced and patient was discharged.

Discussion
SSPE is a rare chronic progressive encephalitis that is caused by a persistent infection by immune resistant virus. The...
incidence of SSPE after measles is 4 in 100,000 cases as compared to a risk after measles vaccine is 0.14 per lac cases. Measles infection before two years of age is common in SSPE. There is latent period of 6-8 years in most SSPE patients. SSPE is incurable disease and usually causes death within 2-4 years of onset.

Pathology
White matter of both the hemispheres of cerebral cortex and brain stems are affected. Eosinophilic inclusion bodies are present in the cytoplasm, nuclei of neurons and glial cells. Parieto-occipital region of the brain is most severely and frequently affected and involvement is generally asymmetric.

Clinical
There is progressive psychoneurological deterioration consisting of personality changes, seizures, myoclonus, ataxia, photosensitivity, ocular abnormality and spasticity. The clinical profiles of the disease leads to various presentation, so early diagnosis and true clinical staging is not always easy. Myoclonic jerk is a key warning sign of SSPE. As the disease progresses, the intensity of spasms and mental deterioration increases. Patient will suffer speech impairment and increasingly deteriorating comprehension with dysphagia. More uncommon presenting features are tremors, dystonia, headache, hemiparkinsonism, choreoretinitis and hallucination.

Diagnosis
EEG shows high amplitude bi or triphasic spikes or slow wave discharge with a normal background initially. With time the background rhythm deteriorates and the myoclonic discharges become less evident. CT scan is rarely helpful but may show low density changes in affected areas.

MRI abnormalities are most commonly seen in the white matter with high signal intensity T₂ and FLAIR images. Progressive cerebral atrophy can be seen on follow up imaging. Diagnosis is confirmed by elevated levels of measles antibodies in the serum and CSF. CSF also shows elevated oligoclonal bands (IgG).

SSPE should be differentiated from subacute measles encephalitis with measles inclusion body encephalitis (MIBE) which occurs in children with immunocompromised cellular immunity. There is no elevation in measles antibody titres though measles virus can be demonstrated in the brain. SSPE should also be differentiated from lipidoses, progressive rubella panencephalitis and various degeneration of white matter.

Outcome measures
Neurologic disability index (NDI) may be the most sensitive to change but is poorly described and complicated to apply. The second measure brief assessment examination (BAE) has been developed specifically for SSPE.

Treatment
No adequate treatment is available for SSPE. Treatment is divided into disease modifying agents and symptomatic therapies. Symptomatic therapies include seizure control. Carbamazepine is recommended for myoclonus induced falling episodes. Inosiplex and interferon have received the most attention. Inosiplex is immune modulating substance that facilitates lymphocyte immune function once transferred by a viral antigen. Interferon activates natural killer cells and directly inhibits viral replication. Typically INF-alpha is administered intraventricularly but INF-beta is used subcutaneously. There is no significant difference outcome if both Inosiplex interferon are used together.
Prognosis

Death usually occurs within three years. If diagnosis is made early then it is possible to cure the disease. However, treatment within Inosiplex combined with interferon can give up to a 50% remission rate.

References


NEUROPATHIC PAIN TREATMENT: A FURTHER STEP FORWARD

Combination of the actions of tricyclic antidepressants and gabapentin adds an additional facet to pain modulation.

The trial did not establish superiority of nortriptyline versus gabapentin, and only slight differences were reported in side-effect profiles, which supports the recommendation that both drug classes be used as first-line treatments.