INTRODUCTION: Subacute Sclerosing Panencephalitis is a chronic complication of measles with delayed onset and outcome that is nearly always fatal.\(^1\) It appears to result from a persistent infection with an altered measles virus that is harbored intracellularly in the central nervous system for several years. After seven to ten years the virus apparently regains virulence and attacks the cells in the central nervous system that offered the virus protection. This slow virus infection results in inflammation and cell death leading to an inoperable neurodegenerative process. Virtually all patients eventually succumb to SSPE.\(^1\)

KEYWORDS: SSPE, Electroencephalography, Myoclonic epilepsy, Magnetic Resonance Imaging.

CASE REPORT: A previously healthy and developmentally normal 14 years old girl born to third degree consanguineous marriage brought with history of behavioral disturbance and decreased school performance since 2 months. Difficulty in standing and walking with involuntary movements of neck, right upper limb and left lower limb since 1 month (fig. 1, fig. 2). Involuntary movements were insidious in onset, slowly progressive repetitive, jerky involving proximal limbs more than distal limbs that are aggravated by anxiety and were absent during sleep. It was also associated with slurring of speech. There were no bowel and bladder disturbances and her sensory system was intact. There was past history of exanthematous illness at the age of 18 months.

She was immunized according to national immunization schedule and there is no significant family history of similar complaints. Her physical examination revealed abnormal sensorium with stable vital parameters. She was conscious but confused with emotional liability. There were persistent jerky involuntary movements involving neck, right upper limb, left lower limb. Her muscle tone was increased in all four limbs with normal deep tendon reflexes and babinski sign positive. Meningeal signs were absent and cranial nerves were normal. Examination of fundus was normal and slit lamp examination excluded presence of kayer Fleisher ring.

Our working differential diagnosis was directed to exclude basal ganglia lesions like Wilsons disease, Functional disorder, Neurodegenerative disorders including SSPE.
A complete blood count, erythrocyte sedimentation rate, biochemical parameters including blood sugar, urea, creatinine, serum ceruloplasmin levels, serum electrolytes and CSF analysis were within normal limits. CT scan brain was normal. Scalp electroencephalography (EEG) showed poly spikes suggestive of generalized myoclonic epilepsy and generalized periodic discharges suggestive of Subacute Sclerosing Panencephalitis. Magnetic resonance imaging (MRI) brain revealed hyper intensities in bilateral periventricular region with mild diffuse cerebral atrophy in the form of prominent Sylvian, fissures, ventricular system, cisternal spaces and sulci.

She was treated with sodium valproate and supportive therapy in the form of passive exercise in all four limbs.

Magnetic resonance imaging (MRI) brain (figure 3) showing hyper intensities in bilateral periventricular region with mild diffuse cerebral atrophy in the form of prominent Sylvian, fissures, ventricular system, cisternal spaces and sulci.

**DISCUSSION:** SSPE is a slowly progressive disease characterized by seizures and progressive deterioration of cognitive and motor functions. The diagnosis of SSPE can be reliably established if at least three of the following five criteria given by Dyken²: (a) Progressive sub-acute mental
deterioration with typical signs like myoclonus; (b) Periodic, Stereotyped, high voltage discharges on EEG; (c) CSF globulin levels greater than 20% of total CSF protein; (d) Raised titers of measles antibodies in blood and/or CSF in the absence of other antibodies, including against Herpes simplex virus (HSV) and Varicella zoster virus (VZV); and (e) typical histopathological findings on brain biopsy or autopsy. However in this case patient with typical clinical features and EEG strongly suggestive of SSPE. We considered the diagnosis of SSPE. The pathogenesis of SSPE is related to defective measles virus maturation in neural cells. Aberrant M (matrix) proteins as well as other envelope proteins interfere with assembly and budding of infectious virus. The virus remains in intracellular form and spreads by cell to cell contact. SSPE accounts for a proportion of childhood neurodegenerative diseases.

Children infected in the first 2 years of life are at greater risk. The median interval between acute measles infection and SSPE is 8 years, with a range from 2-12 years. The early stage is marked by behavioral or personality changes and declining school performance. Myoclonus, seizures, spasticity, choreoathetoid or ballistic movements, ataxia, and chorioretinitis follow in the second stage, Optic atrophy, quadriparesis, autonomic disability, akinetic mutism and coma occur in the final stage of the disease.

Majority of cases follow a progressive downhill course to death within a few years, some temporarily plateau or improve, and possibly 5% remit spontaneously. At the time neurological symptoms occur, neurons and glia contain nuclear and cytoplasmic viral inclusion bodies and high titre antimeasles antibody is found in both serum and CSF. The CSF/serum antibody ratio is consistent with high levels of intrathecal synthesis of measles antibody. CSF pleocytosis is absent, glucose is normal, and total protein is normal or elevated.

Acute symptoms, together with increased intracranial pressure, are poor prognostic signs. The earliest MRI findings are high-signal intensity on T2-weighted images of gray and subcortical white matter in posterior portions of the hemispheres. During the second stage of disease; EEG shows a pattern of generalized slow-wave complexes with a regular periodicity. The complexes may last up to 3 seconds and occur at regular intervals, between 4 and 14 seconds, against a background of depressed activity.

Some patients have improved or stabilized after one or several 6-week treatments with intraventricular INTERFERON-α through an Ommaya reservoir, starting at 10 U/m² body surface area per day, with daily increments, up to 106 U/m per day on the fifth day, 5 days per week, combined with oral ISOPRINOSINE 100 mg/kg per day. There have also been reports of response to intravenous RIBAVIRIN in combination with intrathecal INTERFERON-α. The laboratory endpoint of treatment is the eradication of detectable measles antigen from the CSF. Systemic (subcutaneous) INTERFERON-α in daily dose of up to 5 million units, has been used with intrathecal interferon-α, to simultaneously treat the peripheral reservoirs of measles virus, lymphoid and glandular tissue.

Prolonged or repeated treatments carry the risks of meningitis, interferon-α induced encephalopathy, and interferon-α upper and lower motor neuron toxicity.

CONCLUSION: Immunization with attenuated live measles vaccine is recommended for infants 9 to 12 months of age and MMR vaccine at 15 months of age; measles vaccine is one component of the trivalent measles-mumps-rubella (MMR) vaccine. In areas where measles virus circulates widely, immunization is performed early, at 6 months. Recently MMR booster dose at 5 years is advised by
WHO for developing countries. Fatal infection has followed measles vaccine in severely immune compromised children, but there is no epidemiological evidence that vaccination causes SSPE.

REFERENCES:

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