Subacute sclerosing panencephalitis in a Roma adolescent

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Background:

Subacute sclerosing panencephalitis (SSPE) is a rare and fatal complication of measles characterized by cognitive and motor deterioration, with death typically occurring within one to three years of symptom onset. Prior to measles vaccination programs, the incidence of SSPE in developed countries was estimated at 1 case per 100,000 population, but was 100 times higher in some developing nations suggesting that environmental or genetic factors, or specific measles genotypes might be risk factors. A consistent SSPE risk factor has been measles infection before the second birthday.

Case presentation:

- ❖ Patient’s history: 13-year-old boy presented with frequent myoclonus started a month ago leading to falls on the ground.
- ❖ Medical history: totally unvaccinated and infected with measles at the age of two years.
- ❖ Clinical examination: dysarthria, gait disorder, personality changes and intellectual deterioration.
- ❖ EEG: periodic bursts of high-amplitude, slow-wave complexes every 6 seconds correlated with myoclonous (**Figure 1**).
- ❖ Brain MRI: atypical findings on the left lobe (**Figure 2**).
- ❖ CSF analysis: elevated measles IgM and IgG antibody titers, elevated IgG index and presence of oligoclonal bands (**Table**).
- ❖ Treatment approach:
 - 1st step: valproic acid, levetiracetam and intrathecal interferon α
 - 2nd step: intraventricular infusions through an ommaya reservoir
 - 3rd step: isoprinoside
- ❖ Patient’s follow-up:
 - serious complications (convulsions, high fever, progressive weakness, rigidity) followed each intraventricular infusion
 - temporary discontinuation of interferon α .
 - without clinical deterioration after a 6-month follow-up period

Figure 2. MRI T2w revealed increased signal on the left temporal lobe

	Results
Abs IgG against measles on serum	48,3 NTU(NV < 9)
Abs IgM against measles on serum	0,02 (index)(NV <0,1)
CSF	cells = 3, protein = 38 mgr/dL, glucose = 61mgr/dL
Abs IgG against measles on CSF	34,8 NTU(NV < 9)
Abs IgM against measles on CSF	0,01 (index)(NV <0,1)
IgG index	2,75 (NV <0,66)
Oligoclonal bands on CSF	positive (type 3)
PCR on CSF sample	measles: slightly positive VZV, AdV, enterovirus, parvovirus B19, HSV1,2: negative

Table. Serum and CSF analysis

Figure 1. Patient’s EEG revealed a highly characteristic pattern of SSPE

Learning Points/ Discussion:

- SSPE remains a devastating condition.
- Intraventricular Interferon α infusions were associated with serious side effects in our patient and led to treatment discontinuation.
- Prevention through vaccination is the only way to eradicate this fatal complication..