Opercular Syndrome Due to Herpes Encephalitis

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INTRODUCTION

The Foix-Chavany-Marie opercular syndrome is brought on by lesions in the subcortical or cortical regions locations affecting the insula's anterior opercular area created by the parietal, frontal, and temporal lobes' gyri . The syndrome is associated with the opercular strokes, CNS infections such as herpes encephalitis, as well as hereditary, epileptic, and neurodegenerative conditions.

OBJECTIVES, MATERIALS AND METHODS

Herpes simplex encephalitis (HSE) is a rare cause of opercular syndrome. Here we present a patient with complicated febrile seizure, swallowing difficulty and speech distrubance in the acut stage of encephalitis. After the first year of diagnosis she had psychomotor retardation with refractory seizures, MRI and EEG finidngs. Autoimmune encephalitis was suspected due to HSE. The patients MRI and EEG findings were evaluated. The patient was evaluated with Denver developmental screening test II (DDST-II). Speech is either evaluated with Turkısh early Language Development test (TELDT) language skill abnormal with DDST-II test.

CASE PRESENTATION

An 18 month-old girl was presented with two days of fever, vomiting, and recurrent generalized febrile seizures. Ten days ago, the mother had herpetic vesicles on her lip. She was lethargic. She had neck stiffness, aphasia, and swallowing difficulties, speech problems. She had left hemiparesis.She had twitching around lips, sawllowing difficulty. HSV IG M was negative and IG G was positive. The family did not approve the lumbar puncture (LP). Brain MRI with contrast revealed a focal signal increase in T2W, slight heterogeneous enhancement in the right temporal lobe, and diffusion restriction in the right temporoparietal region (Figure 1A, B).

EEG revealed encephalopathy with generalized delta waves and teta discharges in the left hemisphere with attenuation of background rhythm on the right on the fourth day and 12th day of treatment, respectively (Figure 2A, 2B). She was diagnosed with HSE 30 mg/kg/day acyclovir intravenous treatment was initiated for fourteen days. Levetiracetam was added to the treatment for seizures. She had used oral acyclovir 30 mg/kg/day for 14 days after discharge.

In the first year of the diagnosis, she had recurrent oromotor seizures as twitching. Brain MRI revealed a wide encephalomalacia on the effected side (Figure 1C, D). The Denver II development test had warnings in the fine motor area and language area. According to TELDT she had weak in receptive and expressive language. Standart deviation was below -1.5. there was words used but meaningless. Sleep EEG revealed diffuse sharp wave discharges revealing electrical status epilepticus in slepp (ESES) (Figure 2C, 2D). Levetracetam was stopped and valproic acid was added to treatment. Autoimmune encephalitis was suspected due to HSE. Limbic encephalitis panel from serum was negative for NMDAR ,AMPA1, AMPA2, CASPR2, GABARB1/B2, LGI1. The patient was given 400 mg/kg/day IVIG for 5 days . The oromotor seizures ceased . The EEG was the same as the one before treatment on the fifth day of IVIG. The seizures were not seen on the first month of IVIG. The EEG was normal on the first month of IVIG till now.

DISCUSSION

In the course of our patient's hospitalization, there were focal seizures. Early EEG results did not indicate any seizure activity, EEG revaled background rhythm disturbance. She had swallowing difficulty and oromotor seizures, speech distuırbance which was believed to be an active process brought on by insular seizures.

Mastication, swallowing, laryngeal symptoms, speech disturbance, gustatory hallucinations, and autonomic seziures are additional markers of opercular seizures.

Children that developed acute opercular syndrome during HSE were described by Kocak et al. CSF PCR test for HSV is considered to be the gold standard in the diagnosis of HSE. The test result may be negative in the first days and above than tenth day of the disease. The familiy did not approve LP. She had epileptic seizures and MRI findings with ischemic gliotic changes relevant with opercular region. She had recurrent seizures with psychomotor retardation behavioral changes, speech disturbance with EEG findings of ESES. Autuantibody negative immune encephalitis (ANIE) was decided and she had treated with ıntravenous immunoglobulin (IVIG). IVIG is used as the first line immunotherapy in autoimmune encephalitis. Her seizures stopped and EEG became normal but speech disturbance and psychomotor retardation persisted. She had rehabilitaion for language and learning problems. Follow-up is planned for ANIE.

CONCULSION

Here we present a case with opercular syndrome and epilepsy, neurodevelopment delay due to HSE. However, IVIG can be considered as a valuable therapeutic alternative, either as a first- or second-line treatment or as an adjuvant in the treatment of autoimmune neurological diseases also with ESES.

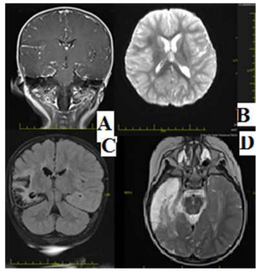

Figure 1 MRI findings A, B. Acute stage of Herpes encephalitis C,D. The ninth month findings after infection

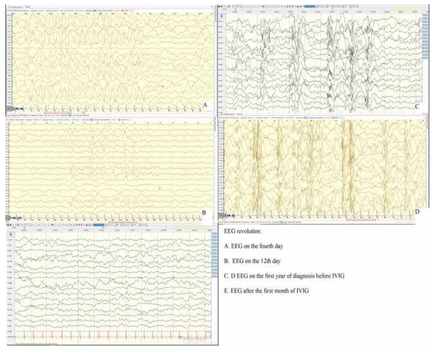

Figure 2
A. EEG on the fourth day, encephalopathy with generalized delta waves and teta discharges. B. EEG on the twelfth day, attenuation of background rhythm on the right hemisphere C, D ESES on the first year of diagnosis.
E. Normal EEG after the first month of IVIG

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