A Child with Opercular Syndrome as the First Presentation of Herpes Simplex Encephalitis

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Abstract

Opercular Syndrome (OS) is described as a clinical syndrome of bilateral corticobulbar involvement with facio-labio-pharyngo-glosso-laryngo-brachial paralysis. Opercular syndrome occurs due to lesions at cortical or subcortical region involving the anterior opercular area. The causes of OS in adult patients have mostly acquired causes such as head injury, tumors, stroke, vasculitis, meningitis, encephalitis, neurodegenerative diseases, and epileptic disorders; however, the causes of OS in childhood period are most of the time congenital or developmental pathologies. Herpes Simplex Encephalitis (HSE) is known to be a rare acquired cause of OS, which is seen mostly in adult patients. In this paper, a 5-year-old patient presented with opercular syndrome as first presentation of HSE, who showed a favourable response to treatment, is reported in order to underline the importance of early recognition of this presentation of HSE.

Keywords: Childhood; Encephalopathy; Facial paralysis; Foix chavany marie syndrome; Herpes simplex; Opercular syndrome; Rolandic damage

Introduction

The opercular syndrome is known as Foix-Chavany-Marie syndrome, which occurs due to lesions at cortical or subcortical regions involving the anterior opercular area surrounding the insula formed by gyri of the frontal, temporal, and parietal lobes [1]. The causes of OS in adult patients have mostly acquired causes such as head injury, tumors, stroke, vasculitis, meningitis, encephalitis, neurodegenerative diseases, and epileptic disorders; however, the causes of OS in childhood period are most of the time congenital or developmental pathologies [2,3]. Herpes Simplex Encephalitis (HSE) is known to be a rare acquired cause of OS which is seen mostly in adult patients [4,5]. Herpes simplex virus one of the well-known main causes of acute sporadic encephalitis with a significant morbidity and mortality risk [5,6]. In this paper, a 5-year-old patient presented with the opercular syndrome as the first presentation of HSE, who showed a favourable response to treatment, is reported in order to underline the importance of early recognition of this presentation of Herpes Simplex Virus (HSV).

Case Presentation

A 5-year-old previously healthy, right-handed girl presented with a 2-day history of progressive dysphagia, hypersalivation, dysarthria, inability to swallow, numbness of the tongue, and newly occurring intermittent twitching on right side of her mouth. Her medical history revealed that she had throat aches and cough complaints six days before, subsequently developed fever and dysphagia, and was diagnosed with tonsillopharyngitis. One day before admission to our clinic, she was seen by a pediatrician, and she had been diagnosed with peripheral facial paralysis. Her birth history and neurological development were normal. Her family history revealed no neurological disorders. Physical examination findings were normal except fever (39.5°C) and mild tachycardia. Neurological examination showed intermittent twitching on the right side of her mouth. The patient was alert, oriented, and cooperated with no clear cognitive deficits but could not give any answers to the questions due to the inability to speak. She could not swallow, and could not perform voluntary movements of face, tongue, lips, and chin. She had hypersalivation. She was crying and smiling in a way similar to spasticity. Gag reflex was decreased bilaterally. The strength of muscle was found 4/5 in bilaterally lower and upper extremities, but deep tendon reflexes were normal. She had no sensation deficit; no extrapyramidal findings could be elicited, and coordination and gait were intact. She had no sign of meningiomas. HB: 11.4 mg/dL, HTC: 34, WBC: 6250, platelet: 224000, sedimentation: 55 mm/hour, C-reactive protein: 15 mg/dL and biochemistry was totally normal. Intravenous gamma globulin and intravenous acyclovir treatment were given due to suspicion of herpes encephalitis on admission. Biochemistry and microbiology of cerebrospinal fluid was normal (CSF protein: 15 mg/dL, glucose: 53 mg/dL and lactate: 1.37 mmol/L, no cell). Electroencephalography (EEG) showed that periodic lateralized epileptiform discharges in the right frontotemporal region (Figure 1A). Magnetic Resonance Imaging (MRI) of the brain revealed diffusion restriction and linear enchantment on the right Sylvian fissure and left insular cortex (Figure 2A). After the first day, intermittent twitching became more frequent, and she has accepted status epilepticus partials continua, which was treated with phenytoin and carbamazepine. On the second day, she developed a generalized tonic-clonic seizure, which showed partial beginning on right arm and face. Her clinical condition also showed deterioration, and she needed intensive care. She was intubated; infusion of midazolam was started, and intravenous methylprednisolone was added to the treatment (30 mg/kg per
day). She was extubated on the fifth day; she did not have another seizure afterward. The diagnosis of HSE was confirmed by a positive polymerase chain reaction performed on cerebrospinal fluid. The limbic encephalitis panel was found negative. Gliotic changes were observed more significantly on left control cranial MRI in a way that included the bilateral opercular region in the 3rd week of treatment (Figure 2B). On her follow up, steroid treatment was completed to four weeks and acyclovir treatment was completed to three weeks. Three months after the initial symptoms, she was able to say some words, her swallow showed an improvement, and she was able to walk with assistance. Since discharge, she has been seizure-free on carbamazepine therapy, and her EEG check-up was normal in the 3rd month of discharge (Figure 1B). On her sixth month check-up, she could speak and swallow, and she could walk without any assistance.

Discussion

In this paper, a child with OS as the first presentation of HSE is reported in order to underline the importance of early recognition.
of this presentation of HSV. Herpes simplex virus one of the well-known main causes of acute sporadic encephalitis with a significant morbidity and mortality risk [5,6]. The main symptoms of the first presentation of HSE are fever, focal or generalized seizures, and/or somnolence; but clinical findings of OS are rarely seen in HSE [4,5].

The OS is described as a clinical presentation caused by lesions involving the perisylvian opercular cortex, which is the area on the motor homunculus responsible for motor control of the face and mouth [1]. These clinical features are often referred to as facio-labio-pharyngeal-linguo-masticatory paralysis with automate-voluntary dissociation, which helps to clinically differentiate the opercular syndrome from the pseudobulbar palsy associated with bilateral supra-bulbar lesions [2,3]. An automatic-voluntary dissociation is seen in a patient with OS; it can be explained by diverse control mechanisms of voluntary and emotional movements of face, tongue, and pharynx [2,3]. Voluntary control of these muscles originates in the primary motor cortex; however, emotional control of the muscles may go through pathways other than the corticonuclear tracts [3]. Patients with OS are unable to close their eyes, open their mouths, speak, protrude the tongue, or swallow voluntarily; however, they may blink, cough, laugh or yawn spontaneously [1].

The clinical features of OS include anarthria or severe dysarthria, masticatory problems, facial weakness, drooling, dysphagia, a tendency for the mouth to be held half-open, weakness of the tongue, absent movement of the palate, and decreased or absent gag reflexes [2]. Like in this case, patients, especially with facial weakness or one-sided twitching, might wrongly be diagnosed with peripheric facial paralysis, which could cause delays in the diagnosis and treatment of HSE.

The prognosis of OS changes depending on the severity of the clinical disease, the type of etiology, and the time of diagnosis. In disorders like meningoencephalitis, vascular or traumatic insult, the epileptic opercular syndrome may be transient and reversible; but OS due to congenital, developmental causes and perinatal insults is likely
to be permanent [2,3]. Early recognition of clinical features of OS and early treatment of the causes may prevent residual sequelae [3,7]. Despite receiving standard antiviral therapy, an appreciable number of patients with herpes simplex virus encephalitis experiences poor acute outcomes or delayed neurological progression [5-7]. The host immune response during HSE can also lead to brain damage. Even though the effectiveness and pathophysiology of corticosteroids in HSE have not been proven clearly, there is an increasing number of reports in the recent literature favoring steroid use in HSE [6]. Our patient received HSV diagnosis with clinical and EEG findings on admission and acyclovir treatment was given immediately. Nevertheless, her medical condition showed deterioration rapidly. Intravenous steroid therapy was added on her treatment on the second day, and then she showed gradual recovery. Therefore, we think that the favorable clinical outcome of your patient may be related to both antiviral and steroid therapy given early.

In conclusion, OS may be seen as the first presentation of HSE even in the childhood period. Awareness of OS and HSV association may lead to early diagnosis and treatment of patients; therefore, it may be important in order to decrease the mortality and morbidity of HSE.

References