Gazi Tıp Dergisi / Gazi Medical Journal

2006: Cilt 17: Sayı 1: 53-55

OLGU SUNUMU - CASE REPORT

FOIX-CHAVANY-MARIE SYNDROME AFTER HERPES ENCEPHALITIS

Şamil HIZLI, Nur PELTEK, M.Akif GÜVEN, Tahsin EDGÜER, Gülşen KÖSE, Gülnar UYSAL

Foix-Chavany-Marie syndrome (FCMS) is a supranuclear (pseudobulbar) palsy caused by congenital or acquired defects of the opercular region and characterized by anarthria or severe dysarthria, loss of functions of the face and tongue muscles, and loss of mastication and swallowing. It is known as an adulthood disease and is very rare in children; operculum syndrome after HSV encephalitis is even rarer. It is a rare cause of anarthria. In this paper, a 17-month-old patient with operculum syndrome secondary to HSV encephalitis is reported.

Key Words: Foix-Chavany-Marie syndrome, child, herpes encephalitis

HERPES ENSEFALİTİNE BAĞLI FOİX-CHAVANY-MARİE SENDROMU

Foix-Chavany-Marie sendromu (FCMS) anterior operküler bölgenin lezyonlarına bağlı gelişen supranükleer(psödobulber) felçdir. Bu sendrom konjenital olabildiği gibi edinilmiş hasara bağlı da gelişebilir. Klinikte anartri / ciddi dizartri , yüz ve dildeki kasların çiğneme ve yutma fonksiyon kaybı ile karakterizedir. Genellikle erişkinlerin hastalığı olarak bilinmekle birlikte çocuklarda da nadiren görülür ancak Herpes simpleks virus ensefalitine sonrasında gelişmesi çok daha nadirdir. Bu yazıda 17 aylık bir vakada HSV ensefalitine bağlı gelişen bir FCMS sunulmuştur.

Anahtar Kelimeler: Foix-Chavany-Marie sendromu, çocuk, herpes ensefaliti

Ankara Çocuk Hastalıkları Eğitim Hastanesi, Ankara

INTRODUCTION

Herpes simplex encephalitis (HSE) is a rare but severe illness with a significant morbidity and mortality risk even after early treatment with antiviral agents. Most of the encephalitis (90%) cases of HSV encephalitis are caused by HSV type 1. Its presentation may be nonspecific and incidence is estimated as two to four cases per million population each year (1,2). Foix-Chavany-Marie syndrome (FCMS) is a rare complication of herpes simplex encephalitis. It is also known as operculum syndrome. The operculum is an area covering the insula and consists of parietal, temporal and frontal gyri. FCMS was described by Foix, Chavany and Marie in 1926 in a patient with facio-linguo-glosso-pharyngo-masticatory diplegia due to a perisylvian or anterior opercular (frontal operculum) lesion (3). Automatic, involuntary, and emotional innervation is preserved in this pathology, while laughing, yawning, and coughing, as well as mimicking movements accompanying emotions, eye closure during sleep, or the blink reflex are unaffected. Findings were related especially with lesions of the anterior operculum. Opercular lesions result from congenital developmental defects (e.g., bilateral cortical dysplasia, bilateral perisylvian polymicrogyria), metabolic diseases (e.g., glutaric aciduria) or acquired lesions such as central nervous system (CNS) infections (herpes simplex, tuberculosis, human immunodeficiency virus, toxoplasmosis) and trauma and vascular events (4). It is known as a disease of adulthood but is also seen in children. Cerebrovascular reasons rank first in adults but CNS infections are most frequent among children.

FCMS may be congenital or acquired and the clinical condition may be persistent or intermittent (5). In this paper a FCMS case is presented since it is a rare cause of anarthria in childhood, and because the patient is the youngest in the literature to suffer from FCMS after herpes encephalitis.

CASE REPORT

This boy was healthy until the age of 17 months, when he had herpes simplex meningoencephalitis, which manifested with afebrile generalized convulsions and somnolence in the emergency department. He had had a fever starting one day before but his temperature was normal upon admittance. He had been hospitalized with suspected meningoencephalitis. At physical examination his body temperature, pulse, respiration rate and arterial tension were as follows: 36.5 °C, 120/min, 40/min, and 90/50 mmHg. His body weight and height were 10 kg (10-25 p) and 84 cm (75 p), respectively. His general condition was poor, with hypoactivity. His oropharynx was hyperemic. A neurological examination showed decreased gross motor strength in the extremities and somnolence. He had no findings of meningeal irritation. The laboratory tests

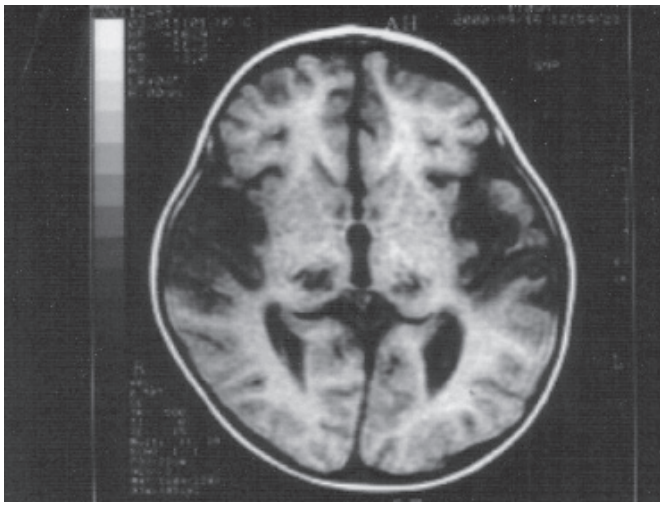

Figure (1a) In the T1W axial image, there are hypointense symmetric encephalomalacic areas in bilateral parietotemporal regions and thalami.

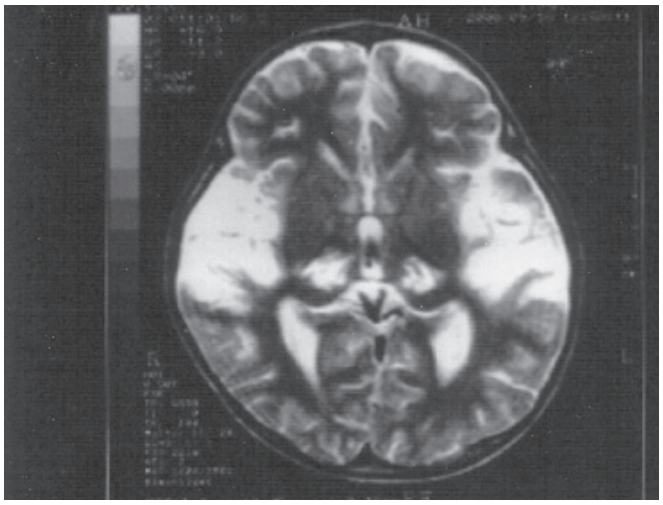

Figure (1b) In the T2 W axial image, hyperintense symmetric encephalomalacic areas in bilateral parietotemporal regions and thalami.

yielded hemoglobin, white blood cell and platelet counts of 9.6 g/dL, 7300/mm3 and 139,000/mm3, respectively. In the peripheral smear 63% lymphocytes and 37% polymorphonuclear leucocytes were seen. Biochemical studies were normal except for an alanine aminotransferase level of 75 U/L. Cerebrospinal fluid (CSF) was clear and showed mild pleocytosis (49 cells/μL). The protein level was 48 mg/dL, glucose was 61 mg/dL and blood glucose level was 97 mg/dL. Ig M antibodies against herpes simplex in serum and CSF were detected by enzyme-linked immunosorbent assay (ELISA). Tuberculosis, human immunodeficiency virus, toxoplasmosis, salmonella, and brucella infections were ruled out by laboratory tests. There was no positivity in the blood, urine, throat or CSF cultures. His immunoglobulin levels were all within the normal range. The EEG revealed a generalized slowing of waves. Cranial tomography of the patient was normal at that time. His clinical condition improved with acyclovir and ceftriaxone treatment. His somnolence decreased and he started to take fluids by nasogastric catheter since he could not swallow. Since his general condition did not improve completely, magnetic resonance imaging was performed, revealing hypodense macrocystic encephalomalacic areas in insular and suprasylvian areas, in precentral gyri and in both thalami (Figure 1a,b). At the control lumbar puncture CSF HSV type I IgM was negative but Ig G was positive; serum HSV type 1 IgM and Ig G antibodies were positive at the same time. Acyclovir treatment was given for 15 days. On day 16, his findings were better and he was discharged during the fourth week of treatment. At a control visit after two weeks, he had severe difficulties in swallowing and took only pureed food. He was unable to sit. His deep tendon reflexes were exaggerated. He was unable to speak or masticate. These clinical findings are consistent with Foix-Chavany-Marie syndrome (anterior operculum syndrome).

DISCUSSION

This FCMS case is the second in the literature diagnosed before two years of age after encephalitis. This patient is the youngest in the literature to suffer from FCMS after herpes encephalitis.

The diagnosis of HSV as the causative agent was based on the results of serology of both blood and cerebrospinal fluid by ELISA (7). The polymerase chain reaction (PCR) technique would have been used in the diagnosis but it was not possible due to the conditions at our hospital at that time (8). An effective and safe treatment, acyclovir can significantly improve outcome if used early. However, considerable diagnostic difficulties, particularly in children, may lead to a delay in treatment since the results may be verified late and an antibody rise is generally detected later during the illness. Therefore, treatment was started early in our case although the diagnosis was confirmed later. Although cranial tomography findings were normal at the beginning, EEG findings were consistent with herpes encephalitis (1) and his other clinical findings did not exclude the possibility of herpes encephalitis. In cranial tomography, there might be low-density areas with contrast and edema/hemorrhage in the temporal region but 57% of patients are expected to have normal cranial tomography findings according to previous studies. Our patient's cranial tomography was normal at the beginning of the disease, but there was multifocal or diffuse involvement of the brain revealed by subsequent MRI.

Hyperintense lesions in the T1W axial image were expected, according to the literature (possibly due to hemorrhage), but there was hypointensity in our patient. This may be explained by infarction in our case secondary to herpes encephalitis vasculitis. This pathophysiology is similar to that in adult FCMS cases with cerebrovascular pathology.

Although acyclovir treatment was started during the early stage, a high percentage of neurologic sequelae was reported (7). In the literature, the prognosis of treated herpes encephalitis cases was reported as 7-30% mortality and 36% sequelae. Prognosis might be affected by factors related to the organism

such as virulence and replication rate as well as the severity of immunologic events leading to tissue destruction. In addition, our patient was below five years of age, which is a negative prognostic factor. There are cases in the literature with better prognoses, showing improvements in swallowing and chewing.

Four months after discharge from hospital, his physical examination revealed diplegia of the facial, pharyngeal and masticatory muscles and learning disability. Enlarged encephalomalacic areas in parietotemporal areas bilaterally and increased temporal lobe atrophy were found at MRI.

Detection of FCMS using clinical findings is also important for determining the etiology of encephalitis since it is mostly infectious in origin among children. FCMS in herpes simplex encephalitis is rare, but must be considered in every case of encephalitis. MRI has the potential to allow a diagnosis early in the course of the disease in parallel with HSV-oligoclonal antibodies and PCR.

Corresponding Address
Şamil HIZLI M.D.
253. Sk.Şahin Apt. No 113 / 11 Hatay / İzmir
Tel : 0 232 245 53 30
e-mail: drsamilhizli@mynet.com

REFERENCES

1. Lahat E, Barr J, Barkai G, Paret G, Brand N, Barzilai A. Long term neurological outcome of herpes encephalitis Arch Dis Child 1999; 80: 69-71.
2. Kohl S, Herpes simplex virus encephalitis in children. Pediatr Clin N Am 1988; 35: 465-83.
3. Foix C, Chavany JA, Marie J. Diplegie faci-linguo-masticatrice d'origine corticosous-cortical sans paralysie desmemres. Rev Neurol 1926; 33: 214-219.
4. Köse G, Yüksel S, Değerliyurt A. Çocukluk çağı konuşma bozukluğunun nadir bir nedeni: Operkulum sendromu, Tıp ve Sağlık Dergisi 1998; 2;3: 32-36.
5. Christen HJ, F Hanefeld F, Kruse E, Imhauser S, Ernst JP, Finkenstaedt M. Foix-Chavany-Marie Syndrome (anterior operculum syndrome in childhood: a reappraisal of Worster-Drought syndrome. Developmental Medicine and Child Neurology 2000; 42: 122-132.
6. Marton R, Gotlieb-Steimatsky T, Klein C, Arlazoroff A. Acute herpes encephalitis: clinical assessment and prognostic data, Acta Neurol Scand 1996; 93: 149-155.
7. Cinque P, Cleator GM, Weber T, Monteyne P, Sindic CJ, Van Loon AM. The role of laboratory investigation in the diagnosis and management of patients with suspected herpes simplex encephalitis: a consensus report. J Neurol Neurosurg Psychiatry 1996; 61: 339-345.