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Revista de la Facultad de Medicina

CASE REPORT

Anterior opercular syndrome secondary to herpes simplex encephalitis. Case Report

Síndrome opercular anterior secundario a encefalitis herpética. Reporte de Caso

David Sandoval-Rodríguez[1] Jimena Peñaranda-Romero[1] Christian Andrés Rojas[1]

[1] Universidad del Valle - Faculty of Health - School of Medicine - Department of Pediatrics - Cali- Colombia.

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Received: 22/06/2024

Accepted: 21/04/2025

Corresponding author: Jimena Peñaranda-Romero. Departamento de pediatría, Escuela de Medicina, Facultad de Salud, Universidad del Valle. Cali. Colombia. E-mail: jimena.penaranda@correounivalle.edu.co.

Keywords: Encephalitis, Viral; Encephalitis, Herpes Simplex; Deglutition Disorders (MeSH).

Palabras clave: Encefalitis viral; encefalitis por Herpes simple; Trastornos de la deglución (DeCS).

How to cite: Sandoval-Rodríguez D, Peñaranda-Romero J, Rojas CA. Anterior opercular syndrome secondary to herpes simplex encephalitis. Case Report. Rev. Fac. Med. 2025;73:e115219. English. doi: https://doi.org/10.15446/revfacmed.v73.115219.

Cómo citar: Sandoval-Rodríguez D, Peñaranda-Romero J, Rojas CA. [Síndrome opercular anterior secundario a encefalitis herpética. Reporte de Caso]. Rev. Fac. Med. 2025;73:e115219. English. doi: https://doi.org/10.15446/revfacmed.v73.115219.

Abstract

Introduction: Opercular syndrome (OS) is a neurological disorder characterized by paralysis of the orofacial muscles. It may be congenital or acquired, with herpes simplex encephalitis (HSE) being a rare etiology.
Case presentation: A 30-month-old infant girl was referred to the emergency department of a quaternary care hospital in Cali (Colombia) due to sialorrhea, deviation of the lip commissure, decreased strength in the left upper limb, hyporexia, perioral and left eyelid myoclonia compatible with focal seizures and manifestations compatible with HSE (difficulty swallowing fluids, with dental occlusion, phonation, and mastication). Given the clinical suspicion of viral encephalitis, a cerebrospinal fluid analysis was performed, reporting pleocytosis without elevated CSF protein levels and positive isolation for herpes simplex virus, confirming the diagnosis of HSE. Treatment with intravenous acyclovir was started, achieving a favorable clinical outcome, with normal results in the follow-up cerebrospinal fluid analysis performed 21 days after starting antiviral therapy. The patient was discharged after 31 days of hospitalization with indication to attend regular check-ups; however, the patient was not taken to follow-up appointments.
Conclusion: Detecting OS in the pediatric age requires a high level of clinical suspicion upon the appearance of acute-onset dysarthria and dysphagia. This case highlights the need to investigate in depth the etiology of unusual neurological manifestations, since the timely identification of HSE as its underlying cause allows establishing a specific antiviral therapy and improving neurological prognosis.

Resumen

Introducción. El síndrome opercular (SOA) es un trastorno neurológico caracterizado por la parálisis de los músculos orofaciales. Las causas pueden ser congénitas o adquiridas, siendo la encefalitis herpética (HSE) una etiología rara.
Presentación del caso. Niña de 30 meses remitida al servicio de urgencias de un hospital de cuarto nivel de atención en Cali (Colombia) por presencia de sialorrea, desviación de la comisura labial y disminución de la fuerza en miembro superior izquierdo, hiporexia, mioclonías peribucales y palpebrales izquierdas compatibles con crisis epilépticas focales y manifestaciones compatibles con SOA (disfagia a líquidos y dificultad para la oclusión dental, la fonación y la masticación). Dada la sospecha clínica de encefalitis viral, se realizó un análisis de líquido cefalorraquídeo en el que se reportó pleocitosis sin hiperproteinorraquia y aislamiento positivo para virus del herpes simple, confirmándose el diagnóstico de HSE. Se inició manejo con aciclovir intravenoso, logrando una evolución clínica favorable, con resultados normales en el análisis de líquido cefalorraquídeo de control a los 21 días de haberse iniciado el tratamiento antiviral. Luego de 31 días de hospitalización, fue dada de alta con indicación de asistir a controles regulares; sin embargo, la paciente no fue llevada a las citas de seguimiento.
Conclusión. La detección del SOA en la edad pediátrica requiere un alto nivel de sospecha clínica ante la aparición de disartria y disfagia de inicio agudo. Este caso enfatiza la necesidad de profundizar en la búsqueda etiológica frente a manifestaciones neurológicas inusuales, ya que la identificación oportuna de una HSE como causa subyacente permite instaurar un tratamiento antiviral específico y mejorar el pronóstico neurológico.

Introduction

Anterior opercular syndrome (AOS), also known as Foix-Chavany-Marie syndrome or perisylvian syndrome, is a bilateral cortical-subcortical pseudobulbar palsy caused by bilateral paralysis in the brain operculum.[1,2] This syndrome is characterized by voluntary lingual, facial, pharyngeal, and masticatory paralysis with preservation of automatic and involuntary movements.[1,2] The clinical features of AOS are not associated with any specific disorder,[1] and its etiology may be congenital (neuroblastic migration disorders) or acquired (vascular disease, viral encephalitis, epilepsy, head trauma, post-surgical conditions, and neurodegenerative disorders).[3]

The following is the case of a 30-month-old female infant with AOS secondary to herpes simplex virus 1 (HSV-1) encephalitis, whose main clinical manifestation was alteration of the orofacial muscles without fever. The purpose is to contribute to the rapid identification and, therefore, timely treatment of this syndrome.

Case presentation

A 30-month-old female Venezuelan migrant child was taken to the emergency department of a primary care hospital in El Águila (Valle del Cauca, Colombia) due to a 7-day history of sialorrhea, transient deviation of the lip commissure to the right, and decreased strength in the left upper limb. Given these clinical manifestations, she was referred to a secondary care hospital in Tuluá (Valle del Cauca), where she was discharged without any specific treatment since no abnormal findings were detected in the physical examination or in the brain CAT scan performed as part of her assessment. Her legal guardian was instructed to visit the emergency department in case her symptoms returned.

Seventy two hours after discharge, the patient was taken again to the emergency department due to difficulty swallowing fluids and with dental occlusion, phonation, and mastication, manifestations compatible with OAS. Moreover, she presented with paresis of the left upper limb, decreased appetite, asthenia, lethargy, episodes of left perioral and ipsilateral eyelid myoclonia lasting approximately three minutes, consistent with focal motor seizures, and fluctuating level of consciousness. Considering these clinical manifestations, she was referred to a quaternary care center in Cali (Colombia), where treatment with oral levetiracetam (20mg/kg/day) was initiated, achieving adequate control of the epileptic seizures.

Neurological examination on admission revealed absence of the gag reflex, delayed swallowing latency, poor secretion control, impaired tongue mobility, lip incompetence, slight deviation of the left lip commissure, somnolence, and motor aphasia. The patient was afebrile on admission and throughout her hospital stay.

On the second day of hospitalization, a contrast-enhanced MRI of the brain showed extensive hyperintensities on T2 and FLAIR sequences in the frontal lobe and temporal lobes (including the frontal operculum and insular cortex), with areas of diffusion restriction compatible with cortical infarcts (Figure 1).

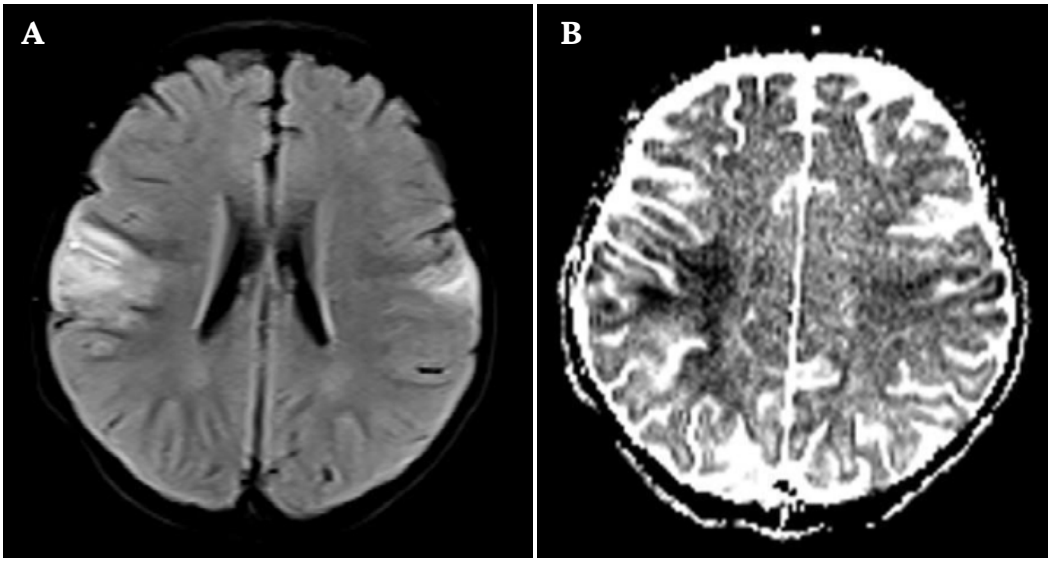

Figure 1. Contrast-enhanced MRI of the brain. A) FLAIR sequence image showing bilateral hyperintense lesions in both frontal lobes, specifically in the superior frontal gyrus with extension to the operculum and insular cortex; B) Diffusion sequence image showing bilateral diffusion restriction areas in the frontal operculum compatible with subacute ischemic infarction.

Considering the clinical manifestations of encephalopathy and focal epileptic seizures, as well as the imaging findings described above, viral encephalitis was suspected. Therefore, an analysis of cerebrospinal fluid obtained by lumbar puncture was performed, showing pleocytosis without elevated CSF protein levels (Table 1) and positive isolation for HSV-1 (Meningitis/Encephalitis FilmArray® panel). Unfortunately, there is no information on the opening pressure. Based on these findings, the patient was diagnosed with herpes simplex encephalitis (HSE), and treatment with intravenous acyclovir (60mg/kg/day) was started on the third day of hospitalization.

Table 1. Analysis of cerebrospinal fluid obtained by lumbar puncture.

Appearance	Transparent
Color	Colorless
Glucose (mg/dL)	56
Protein (mg/dL)	12
Lactate dehydrogenase (U/L)	48
Red blood cells (cells/mm³)	3
Polymorphonuclear cells (cells/mm³)	0
Lymphocytes (cells/mm³)	15
Mononuclear cells (cells/mm³)	0

In addition, during her hospital stay, the patient received speech and language therapy for dysphagia, with gradual improvement. On day 21 of antiviral therapy, a follow-up cerebrospinal fluid analysis was performed, showing normal results. Due to improvement in swallowing function, the patient was discharged 31 days after admission with instructions to attend periodic follow-up appointments. The patient's legal guardian was also advised to continue rehabilitation with occupational and speech therapy; however, at the time this case report was written, no follow-up information was available, as the patient had not attended subsequent follow-up visits or rehabilitation sessions.

Discussion

This article presents the case of a 30-month-old female child with symptoms and clinical signs compatible with OAS (difficulty swallowing fluids and difficulty with dental occlusion, phonation, and mastication), but with no initial clinical evidence of an infectious etiology.

AOS is characterized by severe anarthria or dysarthria, loss of chewing and swallowing functions, and bilateral central paresis affecting the motor functions of the 5th, 7th, 9th, 10th, 11th, and 12th cranial nerves. However, facial expression movements (laughing, crying, yawning, as well as sucking and gag reflexes) are preserved.[1] This syndrome differs from supranuclear palsy in the lack of sphincter disturbances, the rarity of pathological laughing, the decreased tone of the affected muscles, and the suppression of the gag reflex. It also differs from bulbar palsy in the lack of fasciculation, atrophy and denervation, and in the preservation of involuntary innervation and reflexes, except for the gag reflex.[4]

The main cause of AOS in adults is bilateral opercular cerebral infarction, but in the pediatric population, it can be related to both congenital and acquired causes, ranging from neuroblastic migration disorders to epilepsy.[3] Furthermore, it has been described that the most frequent causes in children are neuronal migration disorders and that their main clinical manifestations include pseudobulbar syndrome, epilepsy, and global neurodevelopmental delay, with a tendency to recurrence of seizures and with a generally severe prognosis for the development of spoken language and for the ability to chew and swallow.[5]

Similarly, the reported causes of AOS in children are diverse and include trauma, partial status epilepticus, meningeal tuberculosis, bacterial or viral meningitis, acute disseminated encephalomyelitis, Moyamoya disease,[6] and, rarely, HSE.[6-8] In our patient, AOS was caused by HSE, which was confirmed by isolation of HSV-1 in the cerebrospinal fluid. Infection of the central nervous system by HSV typically presents as encephalitis characterized by fever, seizures, behavioral disturbances, focal neurological signs, and progressive deterioration of consciousness.[9] Notably, our patient did not present with fever, despite antiviral therapy being initiated on the third day of hospitalization after confirming the HSE diagnosis.

Usually, in patients with HSE, CT scans of the skull show unilateral hypodense lesions in the temporal lobe and insular cortex.[10] Additionally, MRI of the brain may show abnormalities such as increased signal intensity on T2-weighted scans, localized edema, and hemorrhages that may range from petechiae to significant bleeding.[11] In our case, diagnostic imaging showed involvement of the frontal operculum, insular cortex, and bilateral parietal region, but no associated bleeding was observed.

As in HSE, HSV-1 AOS has an excellent response to antiviral therapy.[6,9] In our patient, in addition to the antiviral therapy, rehabilitation for the treatment of dysphagia were performed by a multidisciplinary team (physiatry, speech therapy, and physical therapy), achieving a progressive improvement in swallowing and speech.

Conclusion

Detecting AOS in pediatrics requires a high level of clinical suspicion in the presence of acute-onset dysarthria and dysphagia. This case emphasizes the need to investigate in depth the etiology of unusual neurological manifestations, since timely identification of HSE as the underlying cause allows establishing a specific antiviral therapy and improving neurological prognosis. Similarly, early and multidisciplinary rehabilitation plays a

key role in the functional recovery of speech and swallowing, contributing significantly to the minimization of long-term sequelae.

Ethical considerations

Informed consent was obtained from the patient's mother for the preparation of this case report.

Conflicts of interest

None stated by the authors.

Funding

None stated by the authors.

Acknowledgments

None stated by the authors.

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