Journal of Otolaryngology and Rhinology Research
Received 07 May 2023| Revised 25 May 2023| Accepted 29 May 2023 | Published Online 10 June 2023

DOI: https://doi.org/10.52845/JORR/2023/4.2.1

JORR 04 (02), 131–134 (2023)

ISSN (O) 2693-9282

CASE-REPORT

Case report: Bilateral facial palsy in pediatric acute lymphoblastic leukemia with central nervous system relapse.

Luis Felipe Romero Moreno [1,2] | Juan Darío Jiménez Meléndez [2*] | Gilberto Marrugo Pardo [1,3]

1. Department of Pediatric Otolaryngology, Misercordia Hospital Foundation, Bogotá, Colombia.
2. Department of Otolaryngology Head and Neck Surgery, National University of Colombia, Bogotá, Colombia.
3. Titular Professor, Department of Otolaryngology, National University of Colombia, Bogotá, Colombia

Abstract

Objective: To report the youngest case of a bilateral facial palsy secondary to a central nervous system relapse from acute lymphoblastic leukemia in a female young patient that was treated with intrathecal chemotherapy, systemic corticosteroids, and physical therapy with successful outcomes.

Design: A case report of a 9-years old female patient with bilateral facial palsy.

Results: A 9-years old patient with acute lymphoblastic leukemia presented with a complete right facial nerve palsy that progresses 10 days later to a simultaneous grade IV House-Brackmann left facial nerve palsy. Neuroimaging showed basal pachymeningeal and bilateral facial nerve enhancement, predominantly in the right side. Central nervous system metastasis was confirmed with cerebrospinal fluid cytometry and medical therapy was initiated with intrathecal chemotherapy, systemic corticosteroids, and physical therapy. Three weeks after the onset of treatment, both facial palsies ameliorated to a grade IV and III, left and right facial palsy, respectively. Conclusion: Central nervous system metastasis must be considered as a differential diagnosis of simultaneous bilateral facial palsy is presented. Early treatment with intrathecal chemotherapy, systemic corticosteroids and physical therapy may improve patients' outcomes.

1 | INTRODUCTION

Bilateral facial palsy is an unusual clinical entity that represents 0.3 to 3% of all facial paralysis cases (1). It can occur at any age,but usually affects people between 20 and 59 years, being very rare in children (2). Different etiologies have been studied, these can be classified as acquired , congenital or idiopathic. In the first group causes include trauma, toxic (Linezolid, vincristine), infe -ctious (Lyme disease, Epstein-Barr virus,Syphilis), metabolic (Diabetes mellitus, Wernicke-Korsakoff syndrome), systemic (Sjogren syndrome, systemic lupus erythematous) and neoplasia (Hematological, neurofibromatosis type II), whereas in the second group Moebius syndrome is the most common cause. All this differential diagnosis must be considered to provide prompt medical or surgical

decompression treatment depending on the case.

2 | CASE PRESENTATION

We present a 9-years old female patient with a history of acute lymphoblastic leukemia that was being treated with chemotherapy. During its hospitalization she suddenly developed a right peripheral facial palsy with a VI House-Brackmann (HB) grade that was initially treated with oral prednisolone, acyclovir, and physical therapy. 10 days later she progresses to a grade IV HB left facial nerve palsy. Examination when the left palsy began showed bilateral vision loss related to an optic neuritis. There were no abnormal findings on other cranial nerves examinations. Other sensory or motor system signs or symptoms were absent. Otological findings were normal, without middle ear effusion or vesicular lesions.

At the time of presentation complementary studies were done. Serological tests for Borrelia burgdorferi and syphilis were negative. Neurophysiological evaluation tests showed a bilateral affection of the cortico-retinal pathway, electroneuronography revea -led partial axonal injury in the right side with a 66.7% and a 50% functional loss of the frontal, and nasal/orbital muscles, respectively (Fig 1). Nerve conduction studies of 4 limbs and brainstem auditory evoked response test reported no alterations.

Temporal bone Computed Tomography (CT) scan was normal and showed no space occupying lesions on middle ear or mastoids. Cranial nerves Magnetic Resonance Imaging (MRI) with gadolinium showed T2-weighted basal pachymeningeal and bilateral facial nerve enhancement images, without evidence of acoustic neuroma (Fig 2).

With all these features, Central Nervous System (CNS) metastasis was suspected and confirmed by flow cytometry of cerebrospinal fluid. Systemic and weekly intratechal chemotherapy with BFM REZ 2002 protocolo was started, and systemic corticoste-roids and physical therapy were continued. After three weeks of treatment, the facial palsy ameliorated to a grade IV and III, left and right facial palsy, respectively. The last cerebrospinal fluid cytologic study showed no evidence of malignant cells. Unfortunately, the patient died 1 month later from a septic shock.

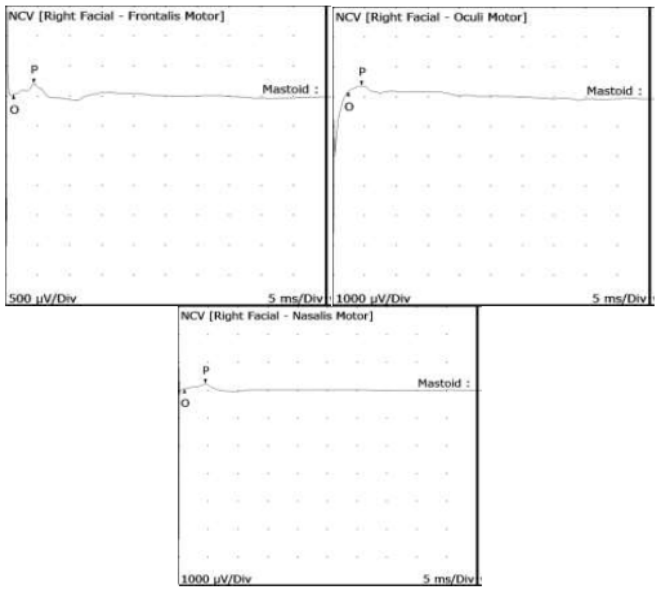

Fig 1. Facial ENoG showed reduced right amplitudes 23 days after the onset of symptoms. Potential action captured by the right frontalis muscle (A), right oculi muscle (B) and right nasali muscle. Left ENoG showed normal results at the moment of the study.

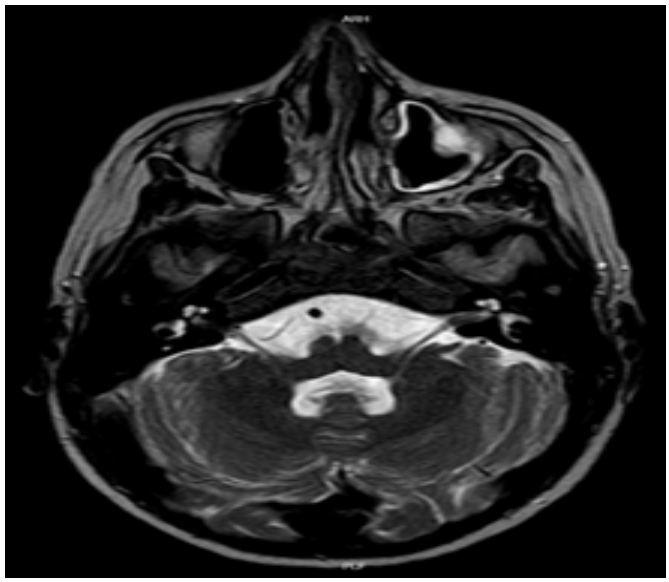

Fig 2. Axial cranial nerves MRI with gadolinium showing T2-weighted basal pachymeningeal and bilateral facial nerve enhancement images.

Supplementary information The online version of this article (https://doi.org/10.52845/JORR/2023/4.2.1) contains supplementary material, which is available to authorized users.

Corresponding Author: *Juan Darío Jiménez Meléndez, Department of Otolaryngology Head and Neck Surgery, National University of Colombia, Bogotá, Colombia. Email: juandariojm@gmail.com*

2 | DISCUSSION

Unlike unilateral facial paralysis, principally attribu -ted to Bell palsy, bilateral facial nerve paralysis most commonly has an identifiable etiology. Gaudin et al, found in a 13-year experience that from 68 patients with bilateral facial nerve palsy that Bell palsy represented 35% of cases, followed by Lyme disease (15%), bilateral vestibular schwannomas (13%) and Moebius syndrome (13%). Hematological malignancy was only documented in a single adult patient that had a CNS lymphoma. No cases of Acute Lymphoblastic Leukemia (ALL) were reported as the case presented in this article.

ALL is the most common hematological malignancy of childhood. Involvement of the CNS is detected in 3-5% of patients at initial diagnosis and in 30-40% of patients at relapse (3). Facial palsy may be a presenting symptom of disease or relapse, but in most cases it is unilateral. Here we present one of the few reported cases of bilateral lower motor neuron facial nerve palsy because of SNC infiltration in a 9-year-old patient with ALL, probably the youngest ever reported (4),(5).

Two main theories have been hypothesized to explain cranial nerve palsies pathogenesis in patients with ALL. The first one is related with the neuropathy created by direct compression and damage of leukemic infiltration on the CNS, which is clinically confirmed by the detection of leukemic cells in the cerebrospinal fluid after lumbar puncture (6), as it was documented in our patient. The second theory implies infectious causes like Epstein-Barr virus and Human T-lymphotropic virus that have been associated with cranial nerve deficits in these patients (7),(8).

Management of bilateral facial nerve palsy in patients with known or suspected CNS implies systemic and intratechal chemotherapy (9). Cranial radiation is being omitted from clinical practice because of its associated forms of damage to normal brain tissue, leaving its use only for patients at highest risk of subsequent CNS relapse, such as those with documented CNS infiltration at the time of diagnosis (10). Up to now, new attempts are being studied, these include tyrosine kinase inhibitors such as dasatinib and imatinib. Other therapies include CXCR4 inhibitors such as plerixafor and ITGA-4 (the ligand of VCAM-1) inhibitors such as TCB3486, whose target is to reduce CNS niche to be not suitable for the survival of ALL cells (11),(12).

In bilateral facial palsy adjuvant therapies must always be considered. Steroids should be administered if not contraindicated for the underlying disease because of its benefits in controlling inflammation and edema of the facial nerve (2). A recent systematic review by Su Jin Kim et al showed that physical therapy may not aid to the regeneration of the injured facial nerve, but it may help in addressing issues with postparetik synkinesis that are commonly associated with severe injuries (13). Finally, permanent facial dysfunction with cosmetic impairment may require restoration surgical treatment with free functioning muscle transplant (14).

Our case highlights a rare presenting symptom in a patient with ALL. CNS infiltration must always be considered in these patients when bilateral facial nerve palsy is documented. An accurate diagnosis is required to start prompt treatment with systemic and intratechal chemotherapy, physiotherapy, and if not contraindicated, systemic steroids. Although, these treatments do not guarantee total illness resolution they may improve prognosis and ameliorate de the palsy as it was documented in our patient. Prospective studies or randomized clinical trials are necessary to demonstrate the effectiveness of new therapeutic interventions.

FINANCING

This research was funded internally by the ENT Department at National University of Colombia.

CONFLICT OF INTEREST.

The authors declare that there is no conflict of interest in the contribution of this article.

BIBLIOGRAPHY

1. Gaudin RA, Jowett N, Banks CA, et al. Bilateral facial paralysis: a 13-year experience. Plast Reconstr Surg 2016;138(4):879–87.
2. Jung J, Park DC, Jung SY, Park MJ, Kim SH, Yeo SG. Bilateral facial palsy. Acta Otolaryngol. 2019 Oct;139(10):934-938.
3. Lenk L, Alsadeq A, Schewe DM. Involvement of the central nervous system in acute lymphoblastic leukemia: opinions on molecular mechanisms and clinical implications based on recent data. Cancer Metastasis Rev. 2020 Mar;39(1):173-187.
4. Lakhotia M, Pahadiya HR, Kumar H, Singh J, Prajapati GR, Sangappa JR. Bilateral Facial Palsy a rare presenting symptom of acute lymphoblastic leukemia with CNS and BM Relapses. J Neurosci Rural Pract. 2015 Oct-Dec;6(4):630-2.
5. Smith V, Traquina DN. Pediatric bilateral facial paralysis. Laryngoscope. 1998 Apr;108(4 Pt 1):519-23.
6. Alsadeq A, Schewe DM. Acute lymphoblastic leukemia of the central nervous system: on the role of PBX1. Haematologica. 2017Apr;102(4):6 11-613.
7. Long CM, Kerschner JE. Parotid mass: Epstein-Barr virus and facial paralysis. Int J Pediatr Otorhinolaryngol 2001;59(2):143-6.
8. Leclercq M, Sauvêtre G, Levesque H, Marie I. Paralysie faciale à bascule révélatrice d'une infection à Human T-lymphotropic virus type 1 (HTLV-1) [Recurrent facial palsy revealing Human T-lymphotropic virus type 1 (HTLV-1) infection]. Rev Med Interne. 2015Oct;36(10):70 1-5.
9. Zhou F, Wen Y, Jin R, Chen H. New attempts for central nervous infiltration of pediatric acut-te lymphoblastic leukemia. Cancer Metastasis Rev. 2019 Dec;38(4):657-671.
10. Sung SH, Jang IS. Isolated Central Nervous System Relapse of Acute Lymphoblastic Leukemia. Brain Tumor Res Treat 2014;2(2):1 14-118.
11. Jeha S, Coustan-Smith E, Pei D, Sandlund JT, Rubnitz JE, Howard SC, et al. Impact of tyrosine kinase inhibitors on minimal residual disease and outcome in childhood Philadelphia chromosome-positive acute lymphoblastic leukemia. Cancer. 2014 May 15;120(10):1514-9.
12. Hsieh YT, Gang EJ, Shishido SN, Kim HN, Pham J, Khazal S, Osborne A, Esguerra ZA, Kwok E, Jang J, Bonig H, Biediger RJ, Vanderslice P, Kim YM. Effects of the small-molecule inhibitor of integrin α4, TBC3486, on pre-B-ALL cells. Leukemia. 2014 Oct;28 (10):2101-4.
13. Kim SJ, Lee HY. Acute Peripheral Facial Palsy: Recent Guidelines and a Systematic Review of the Literature. J Korean Med Sci. 2020 Aug 3;35(30):e245.
14. Roy M, Corkum JP, Shah PS, Borschel GH, Ho ES, Zuker RM, et al. Effectiveness and safety of the use of gracilis muscle for dynamic smile restoration in facial paralysis: a systematic review and meta-analysis. J Plast Reconstr Aesthet Surg 2019;72(8):1254-64.

How to cite this article: Juan Darío Jiménez Meléndez et al. Case report: Bilateral facial palsy in pediatric acute lymphoblastic leukemia with central nervous system relapse. Journal of Otolaryngology and Rhinology Research. 2023;131−134. https:// doi.org/10.52 845/JORR/2023/4.2.1