Bilateral Facial Nerve Palsy in a Child: When the Smile Returns

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INTRODUCTION

Bilateral facial nerve palsy (FNP) is an extremely rare clinical condition, having an incidence of 1 per 5 million population, representing less than 2% of all facial palsy cases, and consisting of the involvement of both the facial nerves simultaneously or sequentially within 30 days.[1] Bilateral FNP is idiopathic in only 23% of cases, and an etiologic factor is often demonstrable,[2,3] and it can also be a rare complication of Epstein–Barr virus (EBV) infection.[4]

We report the case of an 8-year-old girl who presented to our department, 1 month after the onset of bilateral FNP and in which EBV infection was identified as the cause. We discuss the differential diagnosis hypothesis, focusing on outcomes and recovery in case of late referral.

CASE HISTORY

An 8-year-old girl with unremarkable family history was the only child of non-consanguineous parents. Her remote history was negative and her psychomotor development was regular. It was observed that in the last 30 days, the child seemed always sad and unwilling to smile, and the day before the appearance of facial weakness, she presented with headache, vomiting, and diarrhea lasting 1 day and drowsiness for the next 2 days, always without fever. The patient had no finding or recent history of trauma or exposure to tick bites. The baby was sent us by her family physician with the suspect of psychological disturbance. Neurological examination showed bilateral Bell's sign [Figure 1A and B], inability to close the eyes or raise the eyebrows, suggestive for bilateral complete lower motor neuron type of facial palsy (House–Brackmann scale, Grade VI). The patient also had no signs or symptoms of local and systemic infection or current or previous trauma. Blood tests were normal with polymerase chain reaction (PCR), 0.09 mg/dL; neutrophils, 276/µL; and lymphocytes, 273/µL. Serological tests included immunoglobulin (Ig)M–IgG anti-EBV, IgG EBV epstein-barr virus nuclear antigen 1, EBV deoxyribonucleic acid (DNA), IgM–IgG anti-Borrelia burgdorferi, Mosaic sandfly fever IgG–IgM, and IgG–IgM anti-West Nile virus, it was positive exclusively for IgM and IgG anti-EBV and an initial positivity for IgM anti-B. burgdorferi was not confirmed with a second control. Cerebrospinal fluid (CSF) was negative for enterovirus ribonucleic acid (RNA), human herpes virus 6 DNA, varicella zoster virus DNA, cytomegalovirus DNA, herpes simplex virus 1 and 2 DNA, IgG-IgM anti-West Nile virus, and Toscana virus RNA. Polymerase chain reaction on blood for EBV DNA was positive (850 copies/mL). All the PCRs carried out on CSF were negative. The brain contrast-enhanced magnetic resonance imaging (MRI) showed a bilateral inflammation of facial...
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**Discussion**

EBV infection occurs typically in early childhood, at that time, it is mostly asymptomatic. A second peak is observed during late adolescence, when it typically presents as infectious mononucleosis. Neurological manifestations can occur alone or coincidentally with the clinical picture of infectious mononucleosis. Especially in children, neurological involvement such as cranial nerve palsies can be the only clinical sign of EBV infection, and facial nerve is most frequently involved. Furthermore, neurological complications such as cranial nerve palsies, mononeuritis multiplex, meningoencephalitis, and Guillain–Barré syndrome are present in 1%–5% of all patients with acute EBV infection.

Interestingly, approximately 40% of the FNPs associated with EBV were bilateral. Our patient showed bilateral simultaneous FNP associated with EBV infection, probably contracted for at least 30 days. EBV DNA (real-time PCR) was detectable in serum until 30 days after the onset of symptoms, whereas no further positive PCR results were found in CSF after a period of 2 months after the onset of disease.

Detection of EBV DNA is diagnostic of primary EBV infection early in the course of disease and shows a clearer association with the clinical manifestation of disease such as the presence of EBV-specific viral capsid antigen IgG antibodies of low avidity. PCR often fails to detect EBV DNA in CSF even in cases with neurological complications, as in our patient. Because of a polyclonal B lymphocyte stimulation, EBV infection can interfere with serological testing of many diseases. IgM against B. burgdorferi antigens is regularly detected in EBV infection and cannot be discerned by Western blotting from antibodies in real Lyme disease.[3]

Contrast-enhanced MRI in our patient suggested an inflammatory process affecting both facial nerves to confirm the elective target of EBV infection and the opportunity of systemic steroid therapy.

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**Figure 1:** (A) and (B) Total bilateral paralysis of lower motor neuron (House–Brackmann scale, Grade VI). (C) After 3 months, moderately severe bilateral dysfunction of lower motor neuron (House–Brackmann scale, Grade IV). (D) After 6 months, mild severe bilateral dysfunction of lower motor neuron (House–Brackmann scale, Grade II)

**Figure 2:** MRI axial scans, before (A) and after (B) administration of paramagnetic contrast media. Red arrows: bilateral enhancement of the geniculate ganglion and tympanic-mastoid segment of facial nerve
EBV, indeed, was the most likely cause of our patient’s bilateral FNPs in view of the laboratory examinations. Although PCR of CSF for EBV was negative, we have to consider that the examination was performed 1 month after the onset of symptoms and, furthermore, was previously described in patients with neurological complications of EBV[6].

FNP tends to present favorable evolution in childhood with quick and complete recovery, and in any case, resolves within 4 months [Table 1]. Otherwise, our case took longer to heal. This could be explained with the late diagnosis and late therapy or with the pathophysiological mechanism.

The EBV-related nerve damage can occur with two pathophysiological mechanisms: a direct infection of the virus or a postinfectious immunemediated inflammation. In our clinical case, FNP is coincident with the direct detection of the virus in the blood and with normal CSF, so a mechanism of direct, instead of immune-mediated, is hypothesized. In addition to these two basic mechanisms, other factors that determine the degree of affection and its duration can coexist, such as the size of the foramen bone crossed by the facial nerve (the smaller are, the greater the compression of the nerve) and the degree of myelination (the lower the myelination of the nerve, later it will be the recovery of its functionality).

In our patient, we observed the recovery of temporal and zygomatic branches of the facial nerve within 5 months and the recovery of buccal and mandibular branches only after 9 months.

Even though taken late, corticosteroid therapy may have contributed to the child’s full recovery.

**Conclusion**

When a child presents expressionless face and loss of smile, all organic causes must be ruled out before considering psychological problems.

EBV should be routinely tested when a child presents with an apparent neuroinfection as it is a common pathogen that can induce a wide variety of signs and symptoms.

Among the causes of bilateral facial nerve paralysis, EBV infection seems to be associated with slower recovery, despite treatments. The resolution takes place progressively, first in the eyes, then in the mouth, which is probably because of anatomic causes.

Furthermore, even if diagnosis and therapy are late, resolution of EBV-related paralysis can take place completely and the prognosis of this neurological complication remains good.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that

| Author                      | Year of publication | Number of cases | Cause of FNP                  | Outcome            | Recovery timing | Follow-up |
|-----------------------------|---------------------|-----------------|-------------------------------|--------------------|-----------------|----------- |
| Gungor S[7]                 | 2012                | 11-year-old boy | Idiopathic                    | Recovery           | 1 week          | 3 months  |
| Lim TC[8]                   | 2009                | 6-year-old boy  | Kawasaki disease              | Recovery           | 82 days         | 82 days   |
| Sivaswamy L[9]              | 2012                | 15-year-old boy | Guillain–Barré syndrome       | Modest recovery    | 3 months        | 3 months  |
| Francisco T[10]             | 2013                | 13-year-old girl| Neuroborreliosis              | Recovery           | 4 months        | 4 months  |
| Gaudin RA[11]               | 2016                | 68 pz           | Multiple diseases             | Recovery —         | 13 years        | NR        |
| Abitbol C[12]               | 2016                | 8-year-old child| Mastoiditis in myeloid leukemia| Almost completely resolved | 1 month         | 13 years  |
| Quintas E[13]               | 2009                | 16-year-old girl| Meningococcal meningitis and herpetic infection | Recovery | Some weeks later | NR        |
| González Santiago MP[14]    | 2003                | 9-year-old girl | Guillain–Barré syndrome       | Recovery           | 2 months        | NR        |
| Buyukavci M[15]             | 2002                | 13-year-old boy | T cell acute lymphoblastic leukemia | Recovery | 43 days        | NR        |
| Fukuda T[16]                | 1998                | 25-month-old girl | Otorrhea                        | Recovery           | 1 month        | 1 month   |
| Grassin M[17]               | 2017                | 3-year-old boy  | EBV infection                  | Recovery           | <6 weeks        | NR        |

NR = not available; pz = patients
their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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