

# An Adolescent with a Suddenly Developed Mask Face

## Ani Maske Yüz Gelişen Bir Adolesan

✉ Pinar Uysal<sup>1</sup>, ✉ Murat Telli<sup>2</sup>, ✉ Yasemin Turan<sup>3</sup>, ✉ Aylin Eryılmaz<sup>4</sup>, ✉ Yasemin Durum<sup>5</sup>, ✉ Ayşe Tosun<sup>6</sup>

<sup>1</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Pediatric Allergy and Immunology, Aydın, Turkey

<sup>2</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Medical Microbiology, Aydın, Turkey

<sup>3</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Physical Therapy and Rehabilitation, Aydın, Turkey

<sup>4</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Otolaryngology, Aydın, Turkey

<sup>5</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Radiology, Aydın, Turkey

<sup>6</sup>Aydın Adnan Menderes University Faculty of Medicine, Department of Pediatric Neurology, Aydın, Turkey



### Keywords

Adolescent, facial palsy, Epstein-Barr virus, sequelae

### Anahtar Kelimeler

Adolesan, fasial palsi, Epstein-Barr virus, sekel

Received/Geliş Tarihi : 24.01.2016

Accepted/Kabul Tarihi : 20.04.2016

doi:10.4274/meandros.galenos.2016.2662

### Address for Correspondence/Yazışma Adresi:

Pinar Uysal MD,  
Aydın Adnan Menderes University Faculty of  
Medicine, Department of Child Health and  
Diseases Allergy and Clinical Immunology,  
Aydın, Turkey

Phone : +90 505 758 89 31

E-mail : druyisal.pinar@gmail.com

ORCID ID: orcid.org/ 0000-0003-0288-608X

© Meandros Medical and Dental Journal, Published by  
Galenos Publishing House.  
This is article distributed under the terms of the  
Creative Commons Attribution NonCommercial 4.0  
International Licence (CC BY-NC 4.0).

### Abstract

A 13-year-old adolescent girl with symptoms of fever, headache, vomiting and suddenly developed stroke at face admitted to emergency department. On her physical examination, exudative tonsillitis, bilateral facial palsy (BFP), and meningeal irritation signs were detected. The diagnosis of meningitis was excluded by normal lumbar puncture. Acute *Epstein-Barr virus (EBV)* infection was diagnosed by a positive Monospot test and high *EBV* early antigen immunoglobulin M titers. In cranial diffusion magnetic resonance imaging pathological contrast enhancement of left facial nerve was detected. Complete axonal degeneration at left half of face and mild axonal degeneration at right half of face were detected in electroneuromyography. She partially recovered from BFP after a year of duration despite extensive physical exercise programme.

### Öz

On üç yaşında adolesan kız hasta ateş, baş ağrısı, kusma ve yüzde ani gelişen felç yakınmaları ile acil servise başvurdu. Olgunun yapılan muayenesinde eksüdatif tonsilit, bilateral fasiyal paralizi (BFP) ve meninks irritasyon bulguları saptandı. Hastada normal bulunan lomber ponksiyon ile menenjit tanısı dışlandı. Hastada akut *Epstein-Barr virus (EBV)* enfeksiyonu tanısı pozitif Monospot testi ve yüksek *EBV* erken antijen immunoglobulin M titreleri ile kondu. Kranial difüzyon manyetik rezonans görüntülemesinde sol fasiyal sinirde patolojik sinyal tutulumu görüldü. Elektromiyografisinde sol yüz yarısında tam aksonal dejenerasyon ve sağ yüz yarısında hafif aksonal dejenerasyon saptandı. Hastamızda yoğun fizik tedavi programına rağmen BFP hastalığın birinci yılından sonra kısmen düzeldi.

### Introduction

While the unilateral facial palsy is mostly idiopathic, the etiological causes of bilateral facial palsy (BFP) includes infectious diseases (meningoencephalitis, Lyme disease, influenza, syphilis, *human immunodeficiency virus (HIV)*, *cytomegalovirus (CMV)*, *Mycoplasma pneumonia*, *herpes zoster*, chickenpox, tuberculous meningitis,

cryptococcal meningitis, neurological diseases (multiple sclerosis, intracranial or cerebellopontine tumours, intracranial hemorrhagia), and other diseases like Miller Fisher syndrome (a variant of Gullian-Barre syndrome), skull fracture, acute leukemia, diabetes mellitus, Bell's palsy and sarcoidosis (1-3). Unlike the unilateral facial palsy, bilateral facial paralysis (BFP) is uncommon. Although, facial palsy has a high rate of spontaneous recovery, some of the cases may need prompt management approach. Besides, steroid usage as a treatment of facial palsy is still controversial in children (4,5).

Herein, a case of a 13-year-old girl having the signs mimicking meningitis and diagnosed as BFP secondary to acute *Epstein-Barr virus (EBV)* infection was presented.

### Case Report

A 13-year-old adolescent girl was admitted to the family physician with a history of sore throat for 10 days, bilateral headache at occipital region, axillary fever around 39-40°C, projectile vomiting and generalized myalgia for 5 days and numbness on face, difficulty in eating, and sleeping with her eyes half open for 3 days. Her past medical history was unremarkable. She was followed with systemic ampicillin-sulbactam for 5 days and referred to us with a prediagnosis of meningitis.

Her axillar body temperature was 39.2°C, pulse 115 bpm, respiratory rate 20/min, and blood pressure 110/60 mmHg at admission to pediatric board. She had a continuous drooling, metallic taste sensation, tonsillar hypertrophy with exudates. At her neurological examination she was conscious and cooperative. She had BFP (grade V and VI facial nerve palsies according to the House-Brackmann scale on the right and left half of face, respectively) (Figure 1).



**Figure 1.** Bilateral facial nerve palsy at the beginning of Epstein Barr virus infection. A) Closing eyes B) Raising eyebrows C) Smiling

Additionally, she had bilateral ptosis and hypomimia. The other cranial nerve examinations were normal. Muscle strength and tonus, as well as, cerebellar tests were normal. She had meningeal irritation signs of neck stiffness with negative Kernig and Brudzinski signs.

Complete blood count, glucose and other biochemical tests were normal. There were no atypical cells or Downey cells on blood smear. The patient's antinuclear antibodies, DNA antibodies, romatoid factor, immunoglobulins (Igs), serum angiotensin converting enzyme, anti-streptolysin O, C-reactive protein and anti-neutrophil cytoplasmic antibody were normal. *HIV* antibody, *Brucella*, *Salmonella*, and *Borrelia burgdorferi* antibody titers were negative. Lumbar puncture was performed. Biochemical, microscopic and cultural analysis of cerebrospinal fluid (CSF) were normal. Serological Ig antibody titres were negative for *HSV-1*, *HSV-2*, *Rubella*, and *CMV*. Polymerase chain reaction analysed in CSF for viral agents including *EBV*, *HSV-1*, *HSV-2*, *CMV*, *HV-6*, *varicella-zoster virus (VZV)* and *Toxoplasma gondii* were negative. *EBV* infection was diagnosed by a positive Monospot test and serological analysis detected by enzyme linked fluorescent assay (VIDAS, Biomerieux, France) as shown in Table 1.

Chest X-ray, conventional cranial magnetic resonance imaging (MRI), cranial diffusion MRI were normal except pathological contrast enhancement of left facial nerve at the admission and after 6 months as shown in Figure 2. The electroneuromyography (EMG) showed mild axonal degeneration at right and complete (at 1<sup>st</sup> and 6<sup>th</sup> months) and then moderate (12<sup>th</sup> month) denervation on left facial nerve.

Intravenous vancomycin 60 mg/kg/day, ceftriaxone 100 mg/kg/day, and acyclovir 500 mg/m<sup>2</sup> every 8

**Table 1. Serological tests for *Epstein-Barr virus* infection during follow-up**

Serological parameters	Date of infection			
	7 <sup>th</sup> day	15 <sup>th</sup> day	42 <sup>th</sup> day	67 <sup>th</sup> day
EBV EA IgM	+	+	-	-
EBV EA IgG	-	-	+	+
EBV VCA IgM	-	-	+	-
EBV VCA IgG	-	-	-	+

EBV: *Epstein-Barr virus*, EA: Early antigen, Ig: Immunoglobulin, VCA: Viral capsid antigen



**Figure 2.** A and B) Cranial magnetic resonance imaging showing pathological contrast enhancement of facial nerve in T1 weighted image magnetic resonance imaging of temporal region at axial section at the admission and after 6 months, respectively. C) Thick left facial nerve within the internal auditory canal (arrowhead) in T2 weighted image



**Figure 3.** Bilateral facial nerve palsy six months later. A) Closing eyes B) Raising eyebrows C) Smiling

hours was commenced immediately until serological confirmation. Fever and generalized myalgia were controlled within 72<sup>nd</sup> hour of the treatment. A course of 60 mg oral prednisolone was included to the therapy on day 3 (8<sup>th</sup> day of onset of palsy) and was gradually stopped by day 21 (29<sup>th</sup> day of onset of palsy). Eye protection measures were taken and lubricants were applied. She presented a slow improvement of the right facial paralysis after six months with a sequel of grade II paresis on the right side. Grade V paresis on the left side continued for 3 months and gradually decreased to grade II after six months despite the extensive physical treatment programme including physical exercise, electrical stimulation and massage as shown in Figure 3. The partial facial weakness on left side continued for a year.

## Discussion

BFP is a rarely seen disease compared to unilateral presentation. The most common cause of the BFP is Lyme disease. Nevertheless, our patient had neither history of tick bite nor recent travel abroad. The *Borrelia burgdorferi* IgM and IgG titres against the spirochete were also negative.

Acute *EBV* infections related to facial nerve palsy have been reported in pediatric age group. *EBV* infection-associated facial nerve palsy are bilateral in around 40% of the cases (6). In literature, the diagnosis of *EBV* infection was determined by serological analysis of antibodies to *EBV* surface proteins. In our case, serum viral capsid antigen (VCA) and early antigen (EA) IgM antibody tests were positive at the end of the first week of symptom onset. Although, most of the *EBV* infections are presented subclinically in previous reports in literature, our case was presented as mimicking meningitis and did not represent the classical components of infectious mononucleosis like hepatitis, lymphadenopathy, organomegaly or atypical lymphocytes in the blood smear. In our case, *EBV* EA IgG and subsequently *EBV* VCA IgG antibodies rised later than a month. We suggest that acute *EBV* infection should be taken into account in children presenting with sudden onset BFP, although they have no specific signs of acute *EBV* infection.

In literature, it has been proposed that facial nerve is mechanically compressed in narrow segment of internal auditory canal secondary to inflammation caused by several etiological agents (7). Our MRI images support the pathological enhancement of the facial nerve. In our case similarly to the previous reports, *EBV* DNA and *EBV*-specific antibodies were negative in CSF fluid. This finding support the hypothesis of the local facial nerve disruption by *EBV* without any meningeal involvement. But contrarily to the previous cases, pleocytosis of CSF could not be detected in the present case (8,9).

The prognosis for BFP seems to be dependent on the underlying aetiology and the treatment is still a dilemma. In the previous case reports, two adult cases with BFP related to acute *EBV* infection were entirely recovered spontaneously within 4 or 6 weeks (8,9).

We suggest that pathological contrast enhancement of facial nerve most probably was related to local inflammatory process during acute *EBV* infection and persistent enhancement after 3 and 6 months might be due to the degeneration of the facial nerve which was concordant with EMG and clinical findings. It was previously reported that, initiation of corticosteroid and antiviral combination treatments within 7 days of onset of BFP might lead a total recovery. Thus, delay in starting steroid treatment might give limited benefit on recovery of the neurological damage even though

the physical stimulation programme. We suggest that, prompt diagnosis and emergent steroid adjustment might have a favourable impact on the prognosis of disease by reducing facial nerve oedema or damage developed secondary to entrapment of the nerve in the auditory canal which leads to compression ischemia or by decreasing inflammatory process in affected nerve at the acute phase of inflammation in our case.

### Conclusion

Although the general acceptance on improvement of *EBV*-associated neurological complications within weeks without any sequelae, in some conditions, full recovery cannot be achieved within weeks even the extensive therapy management programme. In those selected cases, early onset steroid adjustment and physical therapy and rehabilitation programme might have a favourable influence on the prognosis of the BFP.

### Ethics

**Informed Consent:** A written informed consent was received from the patients and the family.

**Peer-review:** Internally peer-reviewed.

### Authorship Contributions

Surgical and medical practices: P.U., M.T., Y.T., A.E., Y.D., A.T., Concept: P.U., Design: P.U., M.T., Y.T., Data Collection or Processing: P.U., Y.D., A.T., Analysis or Interpretation: P.U., M.T., Y.D., Literature search: A.E., A.T., Writing: P.U.

**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study received no financial support.

### References

1. Gevers G, Lemkens P. Bilateral simultaneous facial paralysis-differential diagnosis and treatment options. A case report and review of literature. *Acta Otorhinolaryngol Belg* 2003; 57: 139-46.
2. Teller DC, Murphy TP. Bilateral facial paralysis: a case presentation and literature review. *J Otolaryngol* 1992; 21: 44-7.
3. Pitaro J, Waissbluth S, Daniel SJ. Do children with Bell's palsy benefit from steroid treatment? A systematic review. *Int J Pediatr Otorhinolaryngol* 2012; 76: 921-6.
4. Tiemstra JD, Khatkhate N. Bell's palsy: diagnosis and management. *Am Fam Physician* 2007; 76: 997-1002.
5. Saito H, Takeda T, Kishimoto S. Facial nerve to facial canal cross-sectional area ratio in children. *Laryngoscope* 1992; 102: 1172-6.
6. Terada K, Niizuma T, Kosaka Y, Inoue M, Ogita S, Kataoka N. Bilateral facial nerve palsy associated with Epstein-Barr virus infection with a review of the literature. *Scandinavian Journal of Infectious Diseases* 2004; 36: 75-7.
7. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985; 93: 146-7.
8. Coddington CT, Isaacs JD, Siddiqui AQ, Andrews TC. Neurological picture. Bilateral facial nerve palsy associated with Epstein-Barr virus infection. *J Neurol Neurosurg Psychiatry* 2010; 81: 1155-6.
9. Diedler J, Rieger S, Koch A, Parthe-Peterhans S, Schwaninger M. Bilateral facial palsy: Epstein-Barr virus, not Lyme disease. *Eur J Neurol* 2006; 13: 1029-30.