DOI:10.4274/tnd.70456
Turk J Neurol 2015;21:171-4

A Rare Variant of Guillain-Barre Syndrome: Facial Diplegia Paresthesia

Nadir Bir Guillain-Barre Sendromu Varyantı: Fasiyal Dipleji Parestezi

Emel Oğuz Akarsu[1], Destina Yalçın[2], Reyhan Sürmeli[2], Ahmet Demir[2], Gülin Sünter[2], Yunus Diler[2]

[1]Ersin Aslan State Hospital, Clinic of Neurology, Gaziantep, Turkey
[2]Ümraniye Training and Research Hospital, Clinic of Neurology, İstanbul, Turkey

Summary

Guillain-Barre syndrome (GBS) is an autoimmune polyneuropathy syndrome with acute onset, characterized by ascending muscle weakness and areflexia. Many rare variants of GBS have been defined. Patients with facial diplegia and paresthesia (FDP) in extremities are classified as a localized GBS variant and called FDP. Muscle weakness is either absent or insignificant in these patients. Atypical presentations with preserved, and at times, brisk reflexes, can be a diagnostic dilemma. Treatment in mild cases is also controversial. We describe a pregnant patient with preserved reflexes who was diagnosed as having FDP, based on the clinical and electrophysiologic features, and highlight the treatment options of this rare variant, particularly during pregnancy.

Keywords: Guillain-Barre syndrome, facial diplegia with paresthesia, pregnancy, treatment

Öz

Guillain-Barre sendromu (GBS) akut başlangıçlı, asendan ekstremite güçsüzlüğü ve arefleksi ile karakterize otoimmün bir polinöropati sendromudur. GBS'nin birçok nadir görülen varyantı tanımlanmıştır. Fasiyal dipleji ve ekstremitelerde parestezi ile seyreden hastalar bir GBS varyantı olan fasiyal dipleji-parestezi (FDP) olarak sınıflanır. Bu hastalarda motor kayıp yoktur veya çok azdır. Refleksleri korunmuş ve bazen artmış olan hastalarda tanısal zorluğa yol açabilir. Hafif olgularda tedavi tartışmalıdır. Bu çalışmada klinik ve elektrofizyolojik özelliklerine dayanarak, FDP tanısı konan, refleksleri korunmuş gebe bir hasta sunulmuş ve bu nadir varyantın, özellikle gebelik sırasındaki tedavi seçenekleri tartışılmıştır.

Anahtar Kelimeler: Guillain-Barre sendromu, fasiyal dipleji ve parestezi, gebelik, tedavi

Introduction

Guillain-Barre syndrome (GBS) is an acute monophasic neuropathic disorder of autoimmune origin characterized by ascending progressive limb weakness and areflexia (1). During the course of the illness, 24-60% of patients develop facial nerve paresis, almost all of which are bilateral (2). The clinical spectrum of GBS has expanded because many variants have been described in recent years (3,4,5,6,7,8). Facial diplegia with paresthesias (FDP) is characterized by prominent facial diplegia and distal limb paresthesia with no or only mild motor deficit, and is a localized form of GBS that can be distinguished by involvement of certain muscle groups or nerves (7,8). In this study, we present a pregnant patient who was diagnosed as having FDP and aim to expand upon the clinical and electrophysiologic aspects of this rare variant and also to discuss GBS complicating pregnancy.

Case Report

A woman aged 28 years in the 12th week of her first pregnancy presented to our emergency department reporting that she could not close her eyes, purse her lips or smile. She noted taste loss ten days ago and one day later she developed paresthesias in her hands and feet, which extended to the level of her wrists and ankles on the same day. On the morning of the day of admission she noted weakness in eye closure. Later that day, she was unable to

Address for Correspondence/Yazışma Adresi: Emel Oğuz Akarsu MD, Ersin Aslan State Hospital, Clinic of Neurology, Gaziantep, Turkey
Phone: +90 505 265 28 66 E-mail: emeloguz@yahoo.com
Received/Geliş Tarihi: 06.02.2015 **Accepted/Kabul Tarihi:** 23.07.2015

smile. She had no history of vaccination, infection, medication intake, trauma or travel abroad. At presentation, her neurologic examination revealed facial diplegia. Other cranial nerves were unaffected. Muscle bulk, tone and power in all limbs were normal. All deep tendon reflexes were present with no clonus and extensor plantar response. Sensory examination revealed no deep or superficial sensory loss or hyperesthesia. Complete blood count (CBC), thyroid function tests, vitamin B12, sedimentation and C-reactive protein (CRP) levels were within normal limits. Vasculitis tests, HIV, syphilis and lyme serology were assessed as negative. Antiganglioside antibody assays were also negative. Cerebrospinal fluid (CSF) examination showed increased protein level (103 mg/dL) with normal glucose (62 mg/dL) and no cells. No abnormalities were detected in the magnetic resonance imaging (MRI) results. The electroneuromyography (ENMG) of the patient, which was taken on 14th day, revealed markedly decreased motor nerve compound muscle action potentials (Table 1) (Figures 1, 2, 3). F-waves were present with no delay in latencies (tibial F-waves 46.72 ms, peroneal F waves 45.52 ms) (Table 2). Conduction velocities of motor and sensory nerves of the upper and lower extremities were normal (upper extremity >50 m/s, lower extremity >45 m/s). There was no conduction block or temporal dispersion. No response was obtained on bilateral blink reflex studies. The needle electrophysiologic study revealed spontaneous active denervation potentials (fibrillation and positive spikes) in orbicularis oculi and orbicularis oris muscles, bilaterally. These results were evaluated to be compatible with axonal motor neuropathy and bilateral facial diplegia. Taken together, the clinical presentation and the electrophysiological findings confirmed the diagnosis of FDP.

We decided to regularly observe her for any progression over a three-week period without treatment because the patient had no motor and autonomic involvement on the 10th day of her symptoms. During the entire hospitalization she developed no motor weakness and remained ambulatory. Intravenous immunoglobulin (IVIG) was withheld as she began to recover. On the second day of her admission her paresthesias resolved and she made a rapid recovery. A follow-up electrophysiologic study (day 29) revealed mild improvement of motor nerve compound muscle action potentials. R1 and R2 responses on both sides

Table 1. Motor and sensory nerve conductions

Nerve	Latency (ms)	Amplitude (mV)	Velocity (m/s)
Right median motor	4.2	1.1	52.3
Right ulnar motor	3.7	5.7	59.7
Right common peroneal motor	8.5	0.7	55.1
Right tibial motor	5.1	0.7	47.7
Right facial motor	4.01	0.1	
Left facial motor	3.6	0.1	
Right median sensory	3.1	13	45.6
Right ulnar sensory	1.93	14	60
Right sural sensory	1.88	20.8	53.3
Right superficial peroneal sensory	2.03	13	46

Table 2. F waves

Nerve	Min F Latency ms	Max F Latency ms	Mean F Latency ms
Right tibial	46.72	49.84	48.53
Left tibial	45.52	49.74	47.3
Right common peroneal	46.13	48.18	47.15
Left common peroneal	45.24	47.12	46.18

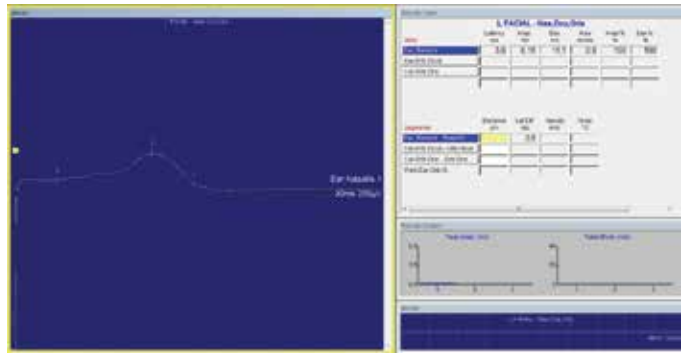

Figure 1. Left facial motor nerve

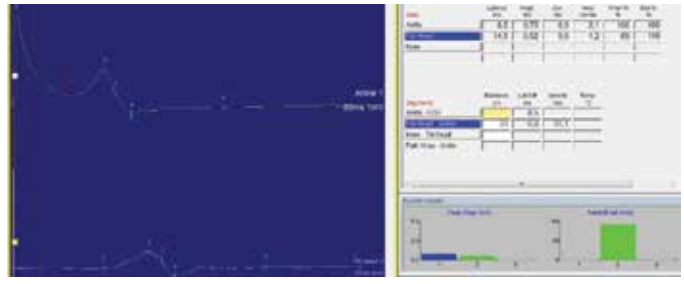

Figure 2. Right common peroneal nerve

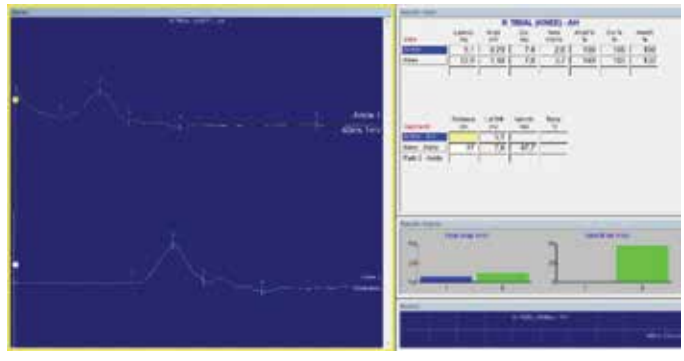

Figure 3. Right tibial motor nerve

were obtained with delayed latencies (Table 3). At the time of discharge, no facial weakness was evident on examination and she was able to close her eyes, whistle, blow, and frown.

Discussion

Simultaneous facial diplegia occurs extremely rarely and can be associated with a variety of neurologic, infectious, neoplastic or degenerative disorders (9,10). Its incidence is estimated as 1 per 5 000 000 population (11). Evaluation of the underlying cause is crucial for the correct and urgent medical treatment (10). FDP, a rare variant of GBS, was first defined in 1994 by Ropper (7). Various case reports have been published since then (6,8,12,13). Susuki et al. (6) screened 8600 patients in a large comprehensive study and FDP was diagnosed in only 22 patients. The hallmark components of the illness are acute onset and rapidly progressive bilateral facial weakness, no or minimal involvement of other cranial nerves, limb weakness or ataxia, paresthesias in the distal dominant limbs, decreased or absent muscle stretch reflexes, the nadir within four weeks, and subsequent recovery (5,6). Our patient showed all these features but her reflexes were preserved during the entire course of the illness. Patients with facial diplegia and hyperreflexia have been reported in the literature and regarded as having a GBS variant (4,14,15).

Taste loss is a common feature of FDP. In contrast, it is rare in typical GBS, but also frequent in patients with Bell's palsy (6,7,10,14,16,17). The presence of taste loss can be a dilemma for distinguishing between FDP and Bell's palsy, but limb paresthesias and hypo- or areflexia, which suggest a systemic polyneuropathy, can help in the differentiation of both conditions (6). Our patient described limb numbness prior to facial diplegia, which was in agreement with other cases. All cases, except those that have anti-CMV antibodies show negative antiganglioside serology, as was the case in our patient, but all have CSF albuminocytologic dissociation, which strongly suggests a GBS variant diagnosis.

In previous reports, nerve conduction studies of the patients with FDP confirmed the presence of systemic demyelinating polyneuropathy, but the electrophysiologic findings of our patient were compatible with axonal polyneuropathy (6,7). To our knowledge, axonal polyneuropathy in patients with FDP has not been described in the literature till now.

There is increasing evidence that supports a good prognosis for this rare variant without treatment. On the other hand, it is hard to predict which patients will deteriorate. Therefore, many studies advise regular observation of patients for two weeks (18). Our patient was admitted on the 10th day of her symptoms. Facial diplegia was present in her first neurologic examination. We decided to observe her regularly for any progression over a three-week period. During this time she developed no motor weakness. At the time of her discharge, no facial weakness was evident on examination.

Table 3. Blink reflex

Nerve	R1 ms	R2 ms	R2-R1
Right supraorbital	13.2	36.4	23.2
Left supraorbital	11.3	33.2	21.9

GBS complicating pregnancy is a rare event. There is no significant difference regarding its incidence in pregnant women and the general population; however, it increases immediately postpartum, during the first 30 days after delivery (19). A comprehensive review of GBS in pregnancy showed that plasmapheresis and IVIG were both effective and safe in the treatment of GBS in pregnancy. IVIG might be a better choice for GBS during pregnancy because it does not involve important alterations in blood volume (9). GBS per se is not an indication for pregnancy termination because there is evidence that it does not shorten disease duration or improve maternal outcomes (19).

Overall, we think that this rare case of FDP expands upon the sparse evidence concerning the clinical course and treatment needs, and adds evidence that polyneuropathy associated with FDP can be axonal. The boundaries of this rare variant remain to be elucidated through further studies.

Authorship Contributions

Informed Consent: A Consent form was filled out by all participants, Concept: Emel Oğuz Akarsu, Destina Yalçın, Design: Emel Oğuz Akarsu, Destina Yalçın, Data Collection or Processing: Emel Oğuz Akarsu, Yunus Diler, Reyhan Sürmeli, Analysis or Interpretation: Gülin Sünter, Ahmet Demir, Literature Search: Gülin Sünter, Ahmet Demir, Reyhan Sürmeli, Writing: Emel Oğuz Akarsu, Destina Yalçın, Peer-review: Externally peer-reviewed. Conflict of Interest: No conflict of interest was declared by the authors, Financial Disclosure: The authors declared that this study has received no financial support.

References

1. Hughes RA, Cornblath DR. Guillain-Barre´ syndrome. Lancet 2005;366:1653-1666.
2. Ropper AH, Wijdicks EFM, Truax BT. Guillain-Barre syndrome. FA Davis: Philadephia; 1991.
3. Ropper AH. Further regional variants of acute immune polyneuropathy. Bifacial weakness or sixth nerve paresis with paraesthesias, lumbar polyradiculopathy and ataxia with pharyngeal-cervical-brachial weakness. Arch Neurol 1994;51:671-675.
4. Lehmann CH, Hartung HP. Variants of Guillain Barre' syndrome: low incidence but high impact. J Neurol 2009;256:1909-1910.
5. Susuki K, Atsumi M, Koga M, Hirata K, Yuki N. Acute facial diplegia and hyperreflexia: a Guillain-Barre´ syndrome variant. Neurology 2004;62:825-827.
6. Susuki K, Koga M, Hirata K, Isogai E, Yuki N. A Guillain-Barre´ syndrome variant with prominent facial diplegia. J Neurol 2009;256:1899-1905.
7. Ropper AH. Further regional variants of acute immune polyneuropathy. Bifacial weakness or sixth nerve paresis with paresthesias, lumbar polyradiculopathy, and ataxia with pharyngeal-cervical-brachial weakness. Arch Neurol 1994;51:671-675.
8. Hozumi A, Yuki N, Yamazaki K, Hirata K. Facial diplegia with paraesthesias: facial nerve enhancement in three dimensional MRI. J Neurol Neurosurg Psychiatry 1999;66:688.
9. Keane JR. Bilateral seventh nerve palsy: analysis of 43 cases and review of the literature. Neurology 1994;44:1198-1202.
10. Peitersen E. Bell's palsy: the spontaneous course of 2,500 peripheral facial nerve palsies of different etiologies. Acta Otolaryngol Suppl 2002:4-30.
11. Price T, Fife DG. Bilateral simultaneous facial nerve palsy. J Laryngol Otol 2002;116:46-48.
12. Takiyama Y, Sato Y, Sawada M, Nishizawa M, Nakano I, Kusunoki S. An unusual case of facial diplegia. Muscle Nerve 1999;22:778-779.
13. Barbi F, Ariatti A, Funakoshi K, Meacci M, Odaka M, Galassi G. Parvovirus B19 infection antedating Guillain-Barre´ syndrome variant with prominent facial diplegia. J Neurol 2011;258:1551-1552.

14. Sethi NK, Torgovnick J, Arsura E, Johnston A, Buescher E. Facial diplegia with hyperreflexia -a mild Guillain-Barre´ syndrome variant, to treat or not to treat? J Brachial Plex Peripher Nerve Inj 2007;2:9.
15. Lehmann HC, Macht S, Jander S, Hartung HP, Methner A. Guillain-Barre´ syndrome variant with prominent facial diplegia, limb paresthesia, and brisk reflexes. J Neurol 2012;259:370-371.
16. Odaka M, Yuki N, Nishimoto Y, Hirata K. Guillain-Barre´ syndrome presenting with loss of taste. Neurology 2002;58:1437-1438.
17. Shuaib A, Becker WJ. Variants of Guillain-Barre´ syndrome: Miller Fisher syndrome, facial diplegia and multiple cranial nerve palsies. Can J Neurol Sci 1987;14:611-616.
18. Green DM, Ropper AH. Mild Guillain-Barre´ syndrome. Arch Neurol 2001;58:1098-1101.
19. Chan LY, Tsui MH, Leung TN. Guillain-Barre syndrome in pregnancy. Acta Obstet Gynecol Scand 2004;83:319-325.