

## Guillain Barré syndrome variant with facial diplegia and paresthesias: A case report and review of the literature

Guillain Barré sendromu varyantı fasiyal dipleji ve parestezi: Olgu sunumu ve literatürün gözden geçirilmesi

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### Abstract

Guillain Barré syndrome (GBS) is a disorder characterized by areflexia and progressive ascending lower extremity weakness. Bilateral peripheral facial diplegia and paresthesia is a rare GBS variant in which the preceding infection, auto-antibody positivity and treatment models are still unclear. In this rare variant, no case with an etiology of varicella infection has been reported yet. Here, we report a case with facial diplegia and paresthesia secondary to varicella infection and discuss clinical, electrophysiological and laboratory findings, and review the literature focusing on the latest reports on this disease.

**Keywords:** Facial diplegia, Guillain-Barré syndrome, varicella.

### Öz

*Guillain Barré sendromu (GBS), arefleksi ve ilerleyici alt ekstremite güçsüzlüğü ile karakterize bir hastalıktır. Nadir bir GBS varyantı olan bilateral periferik fasiyal dipleji ve parestezinin, öncül enfeksiyonu, oto-antikorları ve tedavi şekilleri hala bilinmemektedir. Bu nadir varyantın etyolojisinde varisella enfeksiyonu saptanan olgu henüz bildirilmemiştir. Bu yazıda, varisella enfeksiyonuna sekonder gelişen fasiyal dipleji ve parestezi olgusunun klinik, elektrofizyolojik ve laboratuvar bulguları, bu hastalıkla ilgili son yayınlara odaklanarak gözden geçirilmektedir.*

**Anahtar Sözcükler:** Fasiyal dipleji, Guillain Barré sendromu, suçiçeği.

### Introduction

Guillain Barré Syndrome (GBS) is a disorder characterized by areflexia and progressive ascending lower extremity weakness (1). There is always a preceding viral infection. Diagnosis relies on normal cell numbers in the cerebrospinal fluid (CSF) despite clinical findings, electrophysiological investigations and increased CSF protein (1,2). Atypical presentations may cause diagnostic confusion (2). In the course of the disease, 24-60% of cases develop peripheral facial paralysis and of these 0.3-2% are facial diplegia (3,4). In the literature, few GBS variant cases with isolated peripheral facial diplegia, albuminocytologic dissociation in the CSF and normal electro-physiological examination have been reported (4).

In our case, we aimed to discuss the clinical, electrophysiological and laboratory findings of a rare case of facial diplegia and paresthesias, a variant of GBS, with a review of the literature.

### Case Report

The patient was a 64-year-old right-handed male presenting with difficulty in closing his eyes, whistling and drinking water for three days. He had referred to the hospital with pain in his left arm spreading to the right and numbness in fingertips of this side. His pain and numbness in the upper extremities had disappeared about twenty days following the initial complaints. The patient described no preceding surgery, immunization or infection history. He had diabetes for three years. Neurological examination revealed bilateral peripheral facial palsy (Figure-1a). The corneal reflex was lost bilaterally. Deep tendon reflexes were normoactive. The patient has given written informed consent for publication of this case report and the accompanying images.

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**Figure-1. a.** Neurological examination revealed a bilateral peripheral facial palsy. **b.** At the ninth month follow-up, facial palsy on the right side was partially recovered.

Superficial and deep sensation was in the normal range. According to the laboratory findings; fasting glucose was 254 mg/dl (70-110), HbA1c was 10.4% (normal range: 4.8-5.9), sedimentation rate was 24 mm/h, CRP was 11.3 mg/L, anti-TPO was 13.3 and anti-thyroglobulin Ab was found to be 10.8. Brucellosis, syphilis and other markers of infection (EBV, HSV, CMV, toxoplasmosis, rubella, measles) were negative in the CSF examination. Varicella IgM and IgG were borderline positive in the serum. Vasculitic and hepatitis markers were negative. Lyme serology was negative and angiotensin converting enzyme level in the CSF was 10 U/L (normal range 8-52 U/L) in the lumbar puncture performed on the fifth day of facial paralysis. Thorax CT was normal. In the electrophysiological examination performed on the seventh day of facial paralysis, the upper and lower extremity nerve conduction studies and needle EMG findings were normal. Severe axonal degeneration was detected in the branches of bilateral facial nerve passing through the frontalis, orbicularis oculi and orbicularis oris muscles. Since CSF protein levels were high and did not contain any cells, serum antiganglioside antibodies were sent to test a possible GBS variant with facial diplegia and paresthesias. Levels of antiGM1 IgM and IgG were borderline high, while the levels of other antibodies were within normal range. Anti-GM1 IgM antibody in CSF was positive. One month later, control laboratory findings revealed normal varicella IgM and positive IgG levels. The patient, whose clinical status was stable except for peripheral facial paralysis, was not initiated a treatment with intravenous immunoglobulin (IVIg) due to the potential side effects of IVIg.

Spontaneous minimal improvement in closing the left eyelid was observed two weeks after the beginning of the complaints. Exercise program was continued. Due to an improvement in his complaints at the 20th day of hospital follow-up, IVIg treatment at a dose of 0.4 g/kg/day was administered for five days. At the ninth month follow-up, facial paralysis on the right side had partially recovered (Figure-1b).

## Discussion

Guillain Barré syndrome is thought to be an autoimmune disease that results in the production of antibodies against antigenic proteins. It may be an autoimmune disease in which antibodies are directed against the peripheral myelin proteins but in some cases axonal structures can become the main target of immune-mediated damage (5-7). Therefore, although GBS is known as an acute inflammatory demyelinating polyneuropathy (AIDP), axonal damage can also occur as a primary pathology. In several studies, presence of antibodies against GQ1b, a glycolipid expressed on axolemmal membrane of the cranial nerves in Miller-Fisher syndrome and polyneuritis, and presence of antibodies against GM1 ganglioside in axonal GBS occurring after *Campylobacter jejuni* infection were shown (5-7).

Bilateral synchronous facial diplegia and paresthesias developing as a variant of GBS are rare neurological findings (2-4). Presence of anti-ganglioside serology supports the diagnosis of GBS variant. GD1a and GQ1b are both present in human facial nerve. Whether GT1a is present in human facial nerve is not known. Patients with GBS and antiGT1a antibody frequently develop facial paralysis (1,6). In our patient, antiGT1a, antiGD1a and antiGQ1b levels were negative. The reason why facial nerve ascribes too much importance to other cranial nerves is not clear yet. In Kuwabara et al.'s study on 54 GBS patients, positive anti-GM1 antibodies were found in 28 (52%) of the patients; 21 had only IgG, four only IgM, and three had both positive. In our case, IgM was high. In his control tests after two months, high antiGM1 IgM levels were also observed in both the serum and CSF.

Normal or increased deep tendon reflexes were observed in some patients with GBS (7). The preserved deep tendon reflexes in our patient may be associated with anti-GM1 positivity. Anti-GM1 antibody was detected in 7 hyperreflexia GBS patients (7). The mechanism causing hyperreflexia in GBS patients is still unclear. It was hypothesized that it may be due to the increased motor neuron excitability, preserved senses, affected motor fibers and dysfunction of spinal inhibitory neurons (1,7).

Patients presenting with progressive bilateral facial weakness, distal dominant paresthesia in the extremities, and hypo- or areflexia are also considered to have GBS variant with facial diplegia and paresthesia. The cases of variant GBS, also called as facial diplegia and limb paraesthesia (FDP), have been rarely reported. Susuki et al. (4) made a 7-year retrospective analysis of 8600 patients with GBS. They detected 22 (0.26%) patients whose clinical features were similar to variant GBS facial diplegia and paresthesia. They found

accompanying findings such as ataxia, limb weakness, ophthalmoplegia, bulbar involvement and dysarthria in 12 of the 22 patients. Numbness in the extremities was the initial finding in 19 patients. No concomitant infection was detected in four patients. Significant proportion of antibodies against CMV and EBV were detected in those accompanying an infection. Nerve conduction studies were normal in two cases. Seven patients had also developed facial paralysis. One patient had no concomitant infection but normal nerve conduction studies. Anti-GM2 and GD1b as anti-ganglioside antibody was detected in this patient. All patients had albuminocytologic dissociation (4). Sethi et al. (8) described a GBS variant accompanied by bilateral peripheral facial paralysis and hyperreflexia. Similarly, our case had admitted to the clinic with bilateral simultaneous facial weakness following paresthesia in the arms. CSF findings showed albuminocytologic dissociation. The electrophysiological findings were normal except axonal involvement detected in facial nerves.

In the literature, no case with normal electrophysiological findings in extremities and positive varicella antibody and antiGM1 has been reported. From this point of view, our

case may be the first one representing these features. Bilateral peripheral facial paralysis and paresthesia, a variant of GBS, is a disease whose features such as preceding infection, autoantibody status and treatment are still unclear. Molecular similarities between human ganglioside and microbial lipo-oligosaccharide lead to the development of different types of GBS (1). Whether GBS variants accompanied by isolated facial paralysis need to be treated remains controversial. Anecdotal experiences have been reported in the literature. Worsening in complaints following temporary improvement in weakness and paresthesia after immunotherapy may be associated with immunotherapy (8). Initially, IVIG was not considered in our case but due to progression, IVIG treatment was started after minimal improvement.

In the differential diagnosis of cases admitted with synchronous facial paralysis with no accompanying extremity weakness, facial diplegia and paresthesia variant with GBS should be considered. A detailed electrophysiological investigation and CSF examination should be performed and the patient should be monitored for the risk of progression.

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