Case Report / Olgu Sunumu

A Reason of Facial Diplegia: Guillain-Barré Syndrome

Bir Fasiyal Dipleji Nedeni: Guillain-Barré Sendromu

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Abstract

Guillain-Barré syndrome (GBS), an acute demyelinating inflammatory poliradiculopathy, is characterised by rapidly progressive, symmetrical, ascending weakness and areflexia. The disease usually occurs after 1-3 weeks of infection, vaccinations, trauma, surgical interventions and immunosuppression. Facial diplegia and paraesthesia which is a rare GBS variant has no weakness whereas, GBS is present with weakness which usually begins from lower extremities. In this article, three cases were presented to emphasise the importance of GBS in patients with facial diplegia.

Keywords
Facial diplegia, Guillain-Barré syndrome, paraesthesia

Öz

Guillain-Barré sendromu (GBS); hızlı progresif, simetrik, asendan yayılımlı güçsüzlük ve arefleksi ile karakterize akut, demiyelinizan, enflamatuvar poliradikülopatidir. Hastalık çoğunlukla 1-3 hafta önce geçirilmiş enfeksiyon, aşılanma, travma, cerrahi girişimler ve immünsupresyon gibi durumlar sonrasında ortaya çıkmaktadır. Çoğunlukla alt ekstremitelerden başlayan güçsüzlükle prezente olurken nadir görülen bir GBS varyantı olan fasiyal dipleji ve parestezide ise motor kayıp yoktur. Bu yazıda; fasiyal dipleji ile gelen hastalarda GBS'nin de önemini vurgulamak için üç olgu sunulmuştur.

Anahtar Kelimeler
Fasiyal dipleji, Guillain-Barré sendromu, parestezi

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Introduction

Guillain-Barré syndrome (GBS); acute demyelinating inflammatory poliradiculopathy, is characterized by rapidly progressive, symmetrical, ascending weakness and areflexia. GBS has become the most common disease causing acute generalized flask paralysis after the decrease in the frequency of poliomyelitis.

The incidence is 1-2/100,000. Generally patients apply to the clinic with a complaint of weakness. Weakness may begin in the lower extremities and spread to the arm, face, and oropharyngeal muscles. Rarely, there may be start an onset of extremity proximals or facial weakness (1). In this article, in order to emphasize the importance of GBS among the differential diagnoses in facial weakness, 3 cases of GBS who presented with facial diplegia were presented.

Case Reports

Case 1

A male aged 33 years had neck pain applied to the another hospital but no significant findings were detected in the examination. Five days later, he presented to our hospital with difficulty in swallowing and speaking. Systemic examination was normal. In the neurological examination of the patient, he was fully orientated. Lack of gaze was not detected. There was significant bilateral facial weakness on the right side of the patient.

Speech of the patient was dysphonic. Neck flexion was 4+/5, extension was 5/5, and lower and upper extremities were 5/5 on the patient's muscle strength examination. Sensory deficit was not detected in the patient. Deep tendon reflexes (DTRs) were hypoactive at the upper and lower extremities and no clonus and extensor plantar response. The patient was hospitalized with a initial diagnosis of GBS. There was no history of infection, trauma and vaccination in the anamnesis of the patient recently. No pathological findings were detected in whole blood tests, biochemistry tests and infectious markers. Cranial magnetic resonance imaging (MRI) was taken to exclude demyelinating disease, mass and vasculitic lesions was evaluated as normal. In the lumbar puncture test, cerebrospinal fluid (CSF) did not have any cells and the CSF protein was found to be 761 mg/dL. In the Electromyography (EMG) test, findings compatible with sensorimotor polyneuropathy with demyelinating feature were obtained. The patient was diagnosed with GBS and intravenous immunoglobulin (IVIG) treatment was administered at a dose of 0.4 g/kg/day for five days. After the treatment, the patient's speech improved, eye closing capacity improved, swallowing difficulties decreased. It was observed that facial weakness on the right side of the patient decreased after 16 months of treatment.

Case 2

A 36-year-old male patient complained of numbness in his hands and feet 15 days ago. After 10 days, the patient was admitted to with us the complaints of weaked eye closing and speech impairment. Systemic examination was normal. In the neurological examination of the patient, he was fully orientated. Lack of gaze was not detected. There was peripheral type facial paralysis on the right side of the patient and eye closing was weak on the left side. In the examination of muscle strength, neck flexion was 4/5, bilateral 4+/5 in the distal upper extremities, proximal and distal 5/5 in the lower extremities. DTRs were not seen at the lower, while biceps and triceps reflexes were not seen at the upper extremities, and styloradial reflex was hypoactive. There was no sense of deficit. No pathological reflex was detected. The patient was hospitalized with a initial diagnosis of GBS. In his anamnesis, it was learned that he had a history of upper respiratory infection about 3 weeks ago. In whole blood tests and biochemistry tests, no pathological findings were detected in infectious markers. Cranial MRI was taken to exclude demyelinating disease, mass and vasculitic lesions was evaluated as normal. In the lumbar puncture test, CSF did not have any cells and the CSF protein was found to be 70 mg/dL. In the EMG test, findings compatible with sensorimotor polyneuropathy with demyelinating feature were obtained. The patient was diagnosed with GBS and IVIG treatment was administered at a dose of 0.4 g/kg/day for five days. After the treatment, the patient's eye closing was better and in the examination of muscle strength the neck flexion became 5/5. At the control of the patient after 12 weeks, it was observed that the examination findings completely improved.

Case 3

A 49-year-old male patient complained of pain in the waist and legs 10 days ago, and steroid therapy was initiated due to peripheral type facial paralysis due to the development of weakness in the left facial half. He applied to our hospital after the patient feel weakness in the right half of the face a few days later. Systemic examination was normal. In the neurological examination of the patient, he was fully orientated. Lack of gaze was not detected. There was significant bilateral facial weakness on the left side of the patient. In the examination of muscle strength, there was no deficit. DTRs were not seen at the upper and lower extremities. Sensory deficit and pathological reflex were not detected. Nerve transmissions were detected normally in EMG. While F responses were normal at the upper extremities, tibial F response was extended at the left lower extremities. The findings were considered significant in terms of early polyneuropathy. Thepatient was hospitalized with a initial diagnosis of GBS. No pathological findings were

detected in whole blood tests, biochemistry tests, Lyme test and infectious markers. Cranial MRI taken to exclude demyelinating disease, mass and vasculitic lesions was evaluated as normal. In the lumbar puncture test, CSF did not have any cells and the CSF protein was found to be 113 mg/dL. It was observed that F responses disappeared in the control EMG performed 10 days later. The patient was diagnosed with GBS and IVIG treatment was administered at a dose of 0.4 g/kg/day for five days. It was observed that the patient's examination findings completely regressed at the 2nd month control after discharge.

Discussion

Peripheral facial paralysis is one of the most common neuropathies, but it is often idiopathic. Bilateral involvement is seen in 0.3% of facial paralysis and there is an underlying cause. In facial diplegia; conditions such as Lyme disease, Mobius syndrome, GBS, sarcoidosis, infectious mononucleosis, leukemia, meningitis, head trauma, multiple sclerosis should be considered. Although GBS is often seen as weakness starting from the lower extremities and spreading upwards, there are varying variants of starting and progression. Facial diplegia and paresthesia is one of the rarely seen GBS variants, and it appears as facial diplegia and paresthesia in the extremities and no motor loss is expected (2).

The majority of patients have a history of infection 1-3 weeks ago as seen in GBS. Less frequently, it can occur after vaccination, trauma, surgical procedures, and immunosuppression. Cytomegalovirus, Epstein-Barr virus, varicella-zoster virus, hepatitis A virus, hepatitis B virus, haemophilus influenza, campylobacter jejuni are some microorganisms that have been linked to GBS. Symptoms reach the highest level in the first 2-4 weeks in GBS. Rapid progression of the disease, bulbar involvement, development of autonomic dysfunction, accompanying facial diplegia are signs of poor prognosis. Approximately 70% of patients with bulbar involvement require a mechanical ventilator. In patients with autonomic dysfunction, deaths due to cardiac arrhythmias can be observed, so if possible, patients should be monitored closely in intensive care conditions. Significant improvement was observed in the prognosis of GBS with the improvement of intensive care conditions. Mortality rates, previously which were up to 33%, it had declined to 5%.

The diagnosis is made with clinical findings. While there is no cell increase in CSF examinations, protein increase is an expected finding. However, the lack of an increase in protein does not exclude the diagnosis of GBS, it may be related to its early stage. The most common electrophysiological abnormalities include prolongation of distal motor responses and F wave latency, absence or loss in F waves, conduction block, decrease in distal compound muscle action potential amplitudes and slowing in motor transmission rates (3).

In treatment, IVIG and plasmapheresis are administered. Studies have shown that neither treatment method is superior to each other. However, because of the experienced center requirement for plasmapheresis and higher risk of developing complications, IVIG; It has become the first choice in GBS treatment due to its ease of use. In patients administered IVIG, complications such as anaphylaxis, aseptic meningitis, congestive heart failure, thrombotic events, and acute renal failure can be observed (4). Therefore, plasmapheresis is recommended in cases such as hyperviscosity, congestive heart failure, chronic kidney failure and congenital IgA failure (5).

As a result, facial diplegia is a rare condition and the underlying causes need to be investigated. In cases such as accompanying paraesthesia or weakness, GBS should be carefully examined and diagnosis should be supported by electrophysiological and CSF examination.

Ethics

Informed Consent: Informed consent was obtained from the patients.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: Z.A., Design: A.A., N.K., Data Collection or Processing: Z.A., A.A., N.K., Analysis or Interpretation: A.A., N.K., Literature Search: Z.A., Critical Review: N.K., Writing: Z.A.

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References

1. Visser LH, van der Meche FG, Meulstee J, Rothbarth PP, Jacobs BC, Schmitz PI, et al. Cytomegalovirus infection and Guillain-Barré syndrome: The clinical, electrophysiologic, and prognostic features. Dutch Guillain-Barré Study Group. Neurology 1996; 47: 668-73.
2. Akarsu EO, Yalçın D, Sürmeli R, Demir A, Sünter G, Diler Y. A Rare Variant of Guillain-Barre Syndrome: Facial Diplegia Paresthesia. Turk J Neurol 2015; 21: 171-4.
3. Cros D, Triggs WJ. Guillain Barre syndrome: clinical neurophysiologic studies, Rev Neurol (Paris) 1996; 152: 339-43.
4. Brannagan TH 3rd. Intravenousgammaglobulinfortreatment of CIDP andrelatedımmune-mediatedneuropathies, Neurology 2002; 59: S33-40.
5. Bradley Walter G. Neurology in Clinical Practice Guillain Barre Syndrome 2012; S2288-96.