Case Report

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Facial Diplegia as an Initial Presentation of Guillain-Barré Syndrome

Internal Medicine Section

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ABSTRACT

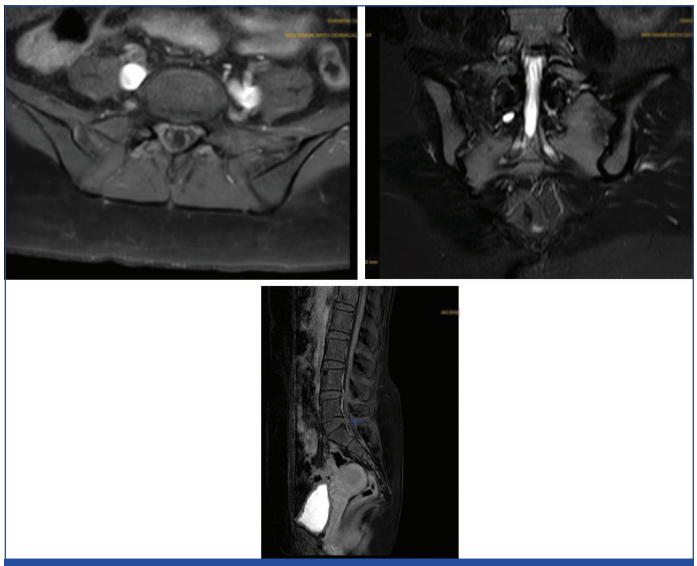
Guillain-Barré Syndrome (GBS) is an acquired immune mediated inflammatory and demyelinating disorder of the peripheral nervous system. This case report is about a 30-year-old female who presented with unilateral facial palsy which progressed to bilateral facial nerve palsy without motor weakness of limbs. Cerebrospinal Fluid (CSF) analysis showed albuminocytological disassociation and Nerve Conduction Studies (NCS) of face showed decreased conduction velocity of nasalis and orbicularis oculi nerves. The patient was treated with intravenous immunoglobulin (0.4 g/kg/day) for five days and showed complete recovery. The present case is reported due to its rarity in presentation i.e. bilateral facial nerve palsy in the initial days of presentation.

Keywords: Albuminocytological dissociation, Bilateral facial nerve palsy, Intravenous immunoglobulin

CASE REPORT

A 30-year-old female with no prior co-morbidities presented to Emergency Room (ER) with complaints of numbness in bilateral soles and palms for three days, difficulty in swallowing to solids and liquids, deviation of angle of mouth to left-side, lower backache for two days. She described a prodromal illness four weeks back including fever and cough. Examination revealed deviation of angle of mouth to left-side and inability to close right eye completely with loss of nasolabial fold on right-side. On detailed neurological examination, it was found that the uvula was in centre and gag reflex was present and there was difficulty in initiation of swallowing due to motor weakness of facial nerve and extraocular muscle movements were normal ruling out the involvement of 3rd, 4th, 6th cranial nerves. No wasting of muscles was noted. Tone and power were normal, diminished knee reflex in right lower limb and loss of bilateral ankle reflexes. Plantar reflex was mute on both sides. Pain and temperature perception were normal in both upper and lower limbs.

All baseline investigations done found to be within the normal limits. Hepatitis B surface antigen (HBsAg) was found to be positive. Ophthalmological workup including fundus evaluation was done and found to be normal. After two days of admission, patient had worsening of numbness over the hands and feet and developed left Lower Motor Neuron (LMN) facial palsy with up rolling of both eyeballs. Nerve conduction study of face showed decreased conduction velocity of nasalis and orbicularis oculi nerve. Magnetic Resonance Imaging (MRI) brain with whole spine showed L5-S1 lumbosacral radiculo-neuropathy and loss of feathery appearance of lumbar nerve roots with marginal increased contrast uptake along the nerve roots [Table/Fig-1]. The patient was diagnosed to have asymmetric Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP). Lumbar puncture done and CSF analysis revealed an increased protein level with normal cell count suggestive of albuminocytological dissociation as shown in the [Table/Fig-2]. She was treated with i.v. methyl prednisone followed by intravenous immunoglobulin (0.4 g/kg/day) for five days led to remarkable clinical recovery. The patient was followed-up in Out Patient Department (OPD) for two months and upto eight months through telephone and she did not have any further episodes of facial weakness.



[Table/Fig-1]: MRI spine of the patient. a) Axial view T1 contrast shows marginal increased contrast uptake in nerves roots. b) Coronal view STIR shows loss of feathery appearance of lumbar nerve roots. c) Sagittal view T1 SAG loss of feathery appearance of lumbar nerve roots.

Cerebrospinal fluid (CSF) profile	Values
CSF colour	Colourless
CSF volume	0.5 mL
CSF appearance	Clear
Coagulum	Absent
RBC's	Nil
Total cells	4 cells/cu.mm (L-100%, N-0%)
CSF glucose (mg/dL)	65
CSF protein (g/dL)	238
CSL chloride (mmol/L)	116
Albuminocytological dissociation	Present

[Table/Fig-2]: Cerebrospinal fluid analysis in a case of GBS on day 2 of presentation.
CSF: Cerebrospinal fluid; RBC's: Red blood cells; L: Lymphocytes; N: Neutrophils

DISCUSSION

The GBS is an acquired immune mediated inflammatory and demyelinating disorder of the peripheral nervous system [1]. The common clinical presentation includes ascending symmetrical weakness, sensory symptoms and areflexia. It uncommonly presents in the atypical forms as brachial pharyngeal variant, Miller fisher and other restricted forms. GBS is a rapidly progressive peripheral polyradiculoneuropathy typically resulting in areflexia and rapidly progressing ascending weakness of atleast two extremities [1,2].

Different subtypes include AIDP, Acute Motor Axonal Neuropathy (AMAN), Acute Motor and Sensory Axonal Neuropathy (AMSAN), acute sensory neuropathy, acute pan dysautonomia and the Fisher syndrome [3].

In the present case, classical presentation of progressive ascending weakness of limbs were not the presenting complaints instead here the patient came with involvement cranial nerve palsy on right-side initially and subsequently on the left-side in due course. Bilateral facial palsy is very rare and accounts for 0.3-2% of facial palsy cases. Fatalities are commonly associated with respiratory failure. Immediate diagnosis and management are very crucial. The most common aetiologies of BFP were Lyme disease, GBS, sarcoidosis and bell's palsy. BFP can be caused by viral diseases like influenza, Cytomegalovirus (CMV), Epstein-Barr Virus (EBV), Herpes Zoster (HZV) and bacterial diseases like tuberculosis, leprosy, tetanus and parasitic manifestations like leptospirosis, malaria, syphilis [4].

Another case report on isolated diplegia also showed a similar presentation with sudden onset of bilateral facial nerve palsy associated with bilateral lagophthalmos without prodromal symptoms, bilaterally absent ankle jerks, albuminocytological dissociation and demyelinating type of facial palsy in NCS and improvement with Intravenous Immune Globulin (IVIG) [5]. Bilateral facial nerve palsy was the initial feature noted in the present case which is rare and expected commonly in later stages of GBS. In the present case, bilateral facial nerve palsy points towards many other differentiating conditions like Multiple Sclerosis (MS) etc., and it was ruled out due to lack of typical MRI brain picture. The index patient was diagnosed with atypical GBS- relative symmetry, numbness and pain at the onset, bilateral facial nerve involvement, prodromal illness, albuminocytological disassociation in CSF, NCS showing demyelination of facial nerve, mild sensory symptoms. These features are strongly supportive of GBS according to modified Asbury criteria and Brighton criteria [6,7].

The pathophysiology of GBS is mainly based on an autoimmune postinfectious process following viral infections like CMV or EBV or bacterial like *Campylobacter jejuni* infections etc., [8]. The main pathology behind occurrence of GBS by *Campylobacter jejuni* is production of antiganglioside antibodies against ganglioside like Lipooligosaccharide (LOS) of bacteria, which acts on gangliosides of peripheral nerves. In the past, a case reported on *Escherichia coli*-associated GBS which presented with lower backache and weakness of lower limbs with history of fever and enteritis, whereas, present case had no history of enteritis, which made *Campylobacter* least likely as a cause. In the present case scenario, exact cause for occurrence of GBS was presumed by supportive evidence of HbsAg positive result [9].

Contemporary studies have reported an association of GBS and conventional influenza, hepatitis B, measles, mumps, rubella, tetanus, polio and gardasil vaccines [10,11], Influenza vaccination was more likely to be associated with development of GBS as compared to other vaccines [12]. A previous study on postvaccination GBS showed bilateral weakness of lower limb as early motor symptom associated with pain, developed within three weeks of vaccination [13]. A meta-analysis on association between influenza vaccination and GBS revealed no risk of GBS in patients who received trivalent influenza vaccine [14]. Bilateral facial palsy may be a specific feature of GBS developed post COVID-19 vaccination [15]. A case was reported in the literature, where patient presented with bilateral facial palsy after 1st dose of COVID-19 vaccination within 10 days and diagnosed as GBS and improved with administration of steroids and IVIG [16]. The present case also had history of COVID-19 vaccination two months prior to admission. But longer duration for onset of symptoms post COVID-19 vaccination ruling out it as a cause.

Cranial nerves are involved in 50% of all cases of GBS, the facial nerves being affected the most. But facial nerve involvement occurs usually following ascending type of limb weakness. Oculomotor involvement might be seen in 10% of cases [17]. The present case had bilateral facial nerve involvement which is most common. More than 50% of GBS patients experience severe pain [18]. However, this symptom is often overlooked because most of the attention is given to limb weakness. Among them, low back pain is a common symptom, but it can cause diagnostic and therapeutic difficulties when patient's experience severe pain in the absence of limb weakness at the early stage of the disease [19]. Pain in GBS may be due to various causes like inflammation of myelinated sensory fibres especially in lower limbs. Sometimes even inflammatory reactions at effected nerve roots may cause radicular nerve pain involving lower back or even radiating to trunk and limbs. The main postulated mechanism for pain is due to excessive production of reactive oxygen species or reactive nitrogen species which in turn resulting in oxidative and nitrosative stress on nerves [20].

Less than 10 cases of sensory predominant GBS are described in literature [21]. The present case had mild sensory symptoms but not sensory predominant. Trauma is one of rarest causes of GBS. Various cause include injury, surgery, brain haemorrhage and fatigue. Exact pathogenesis is still unclear. The present case didn't had any history of trauma [22]. Here, antiGd1a antibody test was not done due to unavoidable reasons. If antiGd1a antibody testing had it been done it would have been more helpful for supporting the diagnosis [23]. In the present case, patient improved well on early identification and initiation of IVIG and other supportive measures. In severe cases, plasmapheresis can be a good option for treatment.

CONCLUSION(S)

The present case report mainly emphasises on the rare presentation of GBS i.e., bilateral facial nerve palsy as an initial presentation. Possibility of atypical presentations of GBS must be kept in mind during initial assessment and early management of patient for better treatment outcome.

REFERENCES

[1] Shahrizaila N, Lehmann HC, Kuwabara S. Guillain-Barré syndrome. The Lancet. 2021;397(10280):1214-28.
[2] Leonhard SE, Mandarakas MR, Gondim FAA, Bateman K, Ferreira MLB, Cornblath DR, et al. Diagnosis and management of Guillain-Barré syndrome in ten steps. Nat Rev Neurol. 2019;15(11):671-83.
[3] Patel K, Nussbaum E, Sico J, Merchant N. Atypical case of Miller-Fisher syndrome presenting with severe dysphagia and weight loss. BMJ Case Reports CP. 2020;13(5):e234316.
[4] Yang A, Dalal V. Bilateral facial palsy: A clinical approach. Cureus. 2021;13(4):e14671.
[5] Sardar S, Sasi S, Menik Arachchige S, Zahid M, Melikyan G. Isolated facial diplegia: A rare presentation of Guillain-Barre syndrome. Clin Case Rep. 2021;9(7):e04473.
[6] Fokke C, van den Berg B, Drenthen J, Walgaard C, van Doorn PA, Jacobs BC. Diagnosis of Guillain-Barré syndrome and validation of Brighton criteria. Brain. 2014;137(1):33-43.
[7] Ghazanfar H, Qazi R, Ghazanfar A, Iftekhar S. Significance of Brighton Criteria in the early diagnosis and management of Guillain-Barré Syndrome. Cureus. 2020;12(5):e8318.
[8] Finsterer J. Triggers of Guillain-Barré Syndrome: *Campylobacter jejuni* Predominates. International Journal of Molecular Sciences. 2022;23(22):14222.
[9] Fujita M, Ueno T, Horiuchi M, Mitsuhashi T, Yamamoto S, Arai A, et al. *Campylobacter coli* infection causes spinal epidural abscess with Guillain-Barré syndrome: A case report. BMC Neurol. 2022;22(1):9.
[10] Principi N, Esposito S. Vaccine-preventable diseases, vaccines and Guillain-Barre' syndrome. Vaccine. 2019;37(37):5544-50.
[11] Chang KH, Lyu RK, Lin WT, Huang YT, Lin HS, Chang SH. Gulllain-Barre Syndrome after trivalent influenza vaccination in adults. Front Neurol. 2019;10:768.
[12] Babazadeh A, Mohseni Afshar Z, Javanian M, Mohammadnia-Afrouzi M, Karkhah A, Masrour-Roudsari J, et al. Influenza vaccination and Guillain-Barré Syndrome: reality or fear. J Transl Int Med. 2019;7(4):137-42.
[13] Park YS, Lee KJ, Kim SW, Kim KM, Suh BC. Clinical features of post-vaccination Guillain-Barré Syndrome (GBS) in Korea. J Korean Med Sci. 2017;32(7):1154-59.
[14] Petráš M, Králová Lesná I, Dáňová J, Čelko AM. Is an increased risk of developing guillain-barré syndrome associated with seasonal influenza vaccination? A systematic review and meta-analysis. Vaccines (Basel). 2020;8(2):150.

[15] Hanson KE, Goddard K, Lewis N, Fireman B, Myers TR, Bakshi N, et al. Incidence of Guillain-Barré Syndrome after COVID-19 vaccination in the vaccine safety datalink. JAMA Network Open. 2022;5(4):e228879.
[16] McKean N, Chircop C. Guillain-Barré syndrome after COVID-19 vaccination. BMJ Case Rep. 2021;14(7):e244125.
[17] Huang X, Lan Z, Zhan Y, Hu Z. Case report and literature analysis: Guillain-Barré Syndrome with delayed unilateral facial palsy. Front Neurol. 2021;12:658266.
[18] Ding X, Jiang H, Hu X, Ren H, Cai H. Guillain-Barré Syndrome and low back pain: Two cases and literature review. Open Med (Wars). 2018;13:503-08.
[19] Hodgeman NT, Lowry LE, Graybill SD. Guillain-Barré Syndrome presenting as an acute back pain. Cureus. 2021;13(8):e17540.
[20] Yao S, Chen H, Zhang Q, Shi Z, Liu J, Lian Z, et al. Pain during the acute phase of Guillain-Barré syndrome. Medicine (Baltimore). 2018;97(34):e11595.
[21] Sharma A, Montesano P, Shah P, Orr L, Kumar M, Stettmeier K, et al. An atypical case of sensory predominant guillain-barre syndrome after influenza a infection. Chest. 2018;154(4):193A.
[22] Huang C, Zhang Y, Deng S, Ren Y, Lu W. Trauma-related Guillain-Barré Syndrome: Systematic review of an emerging concept. Front Neurol. 2020;11:588290.
[23] Zhu J, Zhang Y, Li R, Lin Y, Fu Y, Yan Y, et al. Anti-ganglioside antibodies in Guillain-Barre Syndrome: A novel immunoblotting-panel assay. Front Neurol. 2021;12:760889.

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