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### **Guillain - Barre Syndrome with Sensory & Bilateral Facial & Glossopharyngeal Nerve involvement - A Case Report**

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#### **ABSTRACT:**

Guillain–Barre Syndrome is an autoimmune type of acute, frequently severe demyelinating polyradiculoneuropathy triggered by an antecedent Infection<sup>1</sup>. Bilateral Facial Nerve paralysis with isolated Glossopharyngeal nerve along with sensory involvement indicates a serious underlying medical condition and is usually atypical with Guillain-Barre Syndrome.

Guillain–Barre Syndrome diagnosis need to be considered in amongst all differential diagnosis. We present here a case of 25-years-female presented with Guillain–Barre Syndrome (areflexic quadriparesis) with Bilateral Facial and Glossopharyngeal nerve and sensory involvement.

#### **KEY WORDS:**

Guillain-Barre Syndrome, Bilateral Facial nerve palsy, Bilateral Glossopharyngeal nerve Palsy, Sensory Involvement

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## **INTRODUCTION:**

Guillain-Barré syndrome (GBS) is also known as acute inflammatory demyelinating polyradiculoneuropathy, is characterized by rapid onset of ascending paralysis accompanied with areflexia or hyporeflexia with occasionally sensory or autonomic involvement. It's occurrence is approximately 1 to 4 cases in 100,000 population annually<sup>1</sup>. Males are at slightly at higher risk than females, while adults are observed to be more affected than children with this disease<sup>1</sup>. 50% of GBS patients are reported to have unilateral facial nerve palsy, while bilateral facial and Glossopharyngeal nerve involvement is considered as an unusual presentation<sup>3</sup>. Patients with bilateral facial nerve palsy, are investigated and sometimes are managed for as Bell's palsy. Differentials such as GBS, diabetes mellitus, bacterial meningitis, infectious mononucleosis, sarcoidosis, human immunodeficiency virus (HIV) infection, Lyme disease, syphilis, and leprosy should be considered. Involvement of Glossopharyngeal Nerve with vagus nerve usually points to bulbar and pseudobulbar palsy. Presence of all with sensory involvement leads one to the diagnosis of spinal cord disease. Different outcomes and the management of these various diagnoses warrants stepwise approach to exclude probable aetiologies such as Atypical GBS.

## **Case Report:**

A 25-year-old female had a history of fever with chills and rashes all over her body 4 days prior to presentation for which she was investigated and diagnosed as Dengue NS1 positive. She presented 4 days later to diagnosis of Dengue NS1 positive with bilateral lower limb weakness which was acute in onset and progressively involved the bilateral upper limb over a duration on 1 day. She progressed to have difficulty in opening mouth, unable to perform complete closure of eyelids and heaviness on both sides of the face. On general examination her vitals were stable. On central nervous system examination, her higher mental functions were intact. Cranial nerves examination showed inability of patient to perform puffing of cheeks, clenching of teeth, frowning on forehead, loss of bilateral nasolabial folds, drooling of saliva and food particles from either side of the angle of mouth and loss of taste sensations on anterior two-third of tongue indicating bilateral Facial Nerve palsy and had a nasal twang which indicated Glossopharyngeal nerve palsy, but Gag reflex was intact. On Motor examination, patient had power of 4/5 at shoulder and elbow joint respectively in both upper limbs, while the power at wrist joint was 3/5 with poor grip bilaterally. Patient had no truncal muscle weakness. Power was 3/5 at hip, knee joint bilaterally respectively and at the ankle joint power for plantar flexion was 1/5, while she could not perform dorsiflexion movement at ankle joint bilaterally. Patient had no bladder or bowel involvement, while sensory system examination

showed loss of joint position below ankle joint. CSF analysis showed raised proteins with albuminocytological dissociation. Nerve Conduction Study done showed Bilateral Symmetrical Severe Sensory Motor, predominantly Demyelinating Polyradiculoneuropathy affecting all four limbs. MRI Brain showed no significant abnormality. She was treated with Plasma Exchange with 20% Albumin done in 5 settings on every alternate day over a duration of 10 days. With the second setting of Plasma Exchange patient started showing improvement with the power. By the end of fifth setting of Plasma Exchange patient had gained power of 5/5 in bilateral upper limb at all the joints with improved hand grip, while in the lower limb the power at hip and knee improved to 5/5 and at the ankle joint the power improved to 4/5 and patient could perform dorsiflexion at ankle joint bilaterally. Gait of the patient improved. She was able to close her eyes and could sense the joint position. Patient was discharged on day 15<sup>th</sup> and was followed up on OPD basis showed up to have a significant clinical improvement.

## **DISCUSSION:**

Bilateral facial palsy with Glossopharyngeal nerve palsy and sensory involvement in Guillain-Barre Syndrome is considered to be as an unusual clinical presentation<sup>2,3</sup>. However, it might appear secondary to systemic diseases such as Lyme<sup>5</sup>. GBS subtypes, Sarcoidosis, HIV infection, Leukaemia, Möbius syndrome, Kawasaki, Mycoplasma infection, Diabetes, Borrelia infection, fracture in base of skull, Syphilis, Pontine Glioma, Pregnancy, Leprosy, Mononucleosis, Linezolid therapy, Systemic Lupus Erythematosus, Cryptococcal Meningitis, Pontine Tegmental Haemorrhage, and Bulbosplinal Muscular Atrophy<sup>3</sup>. In 1994 Ropper, came up with a new variant of GBS with features of facial diplegia, limb paraesthesia, decreased deep tendon reflexes, an elevation in CSF protein but without an increase in white blood cells in more than 75% of cases, and demyelination on nerve conduction studies, as facial diplegia and paresthesia. Most patients with facial diplegia and paresthesia initially present with limb numbness, followed by bilateral facial nerve palsy<sup>4</sup>. Almost all patients have antecedent infection within four weeks prior to the onset of neurological symptoms and many patients have demyelination findings in their limbs on nerve conduction investigations.

GBS being a life threatening disease, considering requirement of immediate medical attention and treatment. Its usual presentation sets with bilateral symmetrical ascending flaccid paralysis but unusual presentations that can manifest such as cranial nerve palsy have been reported, while facial palsy being the most common (24-60%) among the cranial nerve palsy<sup>2</sup>. Bilateral simultaneous facial palsy is increasingly recognized as an atypical variant of GBS in adults. Facial diplegia distal limb paraesthesia, sixth nerve palsy, bilateral lumbar polyradiculopathy, and a

combination of Fisher's syndrome and pharyngeal-cervical-brachial weakness along with facial diplegia with hyporeflexia are considered GBS variants<sup>4</sup>. From 20 to 60% of patients with GBS develop facial palsy that is usually bilateral but mostly associated with limb weakness. Landry Guillain-Barre Syndrome represents a probably infectious polyneuritis that involves primary peripheral nerves that are bulbar, myelitic, and cerebral variants. Bilateral facial diplegia without weakness and paraesthesia is a new but rare variant of GBS that typically presents with rapid progressive acute onset paraesthesia, decreased or absent DTR and albumin-cytological dissociation with, or without extremity weakness<sup>1</sup>. Isolated facial diplegia indicates poor prognosis<sup>2</sup>. Aforementioned case satisfies the criteria for GBS but has Bilateral Facial and Glossopharyngeal nerve involvement along with sensory involvement which is quite unusual. CSF protein does not help to distinguish between the two, because it may be elevated in both. The diagnosis of GBS is important because it helps the physician in planning potential therapies outcomes for the patient, provides useful prognostic information and permits reassurance to the patient and family.

## **CONCLUSION:**

Guillain-Barre Syndrome itself is not a very commonly occurring disease. Presence of Bilateral Facial Nerve Palsy with Glossopharyngeal Nerve involvement usually leads to a suspicion of Bulbar Palsy. Hence a careful evaluation clinically and with Nerve Conduction Studies and Imaging studies coming down to a proper diagnosis is crucial as it will lead to a proper treatment and improvement in the patient.

## **DECLARATION OF PATIENT CONSENT:**

Authors have obtained patient consent to publish the case record and relevant photographs

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