

Gullian-Barre Syndrome with Bilateral Facial Nerve Palsy. A Rare Entity

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Abstract

Bilateral facial nerve paralysis, as compared to unilateral palsy is an extremely rare presentation with serious aetiologies including post infectious like in Guillain-Barre syndrome, infectious including Lyme disease, Infectious Mononucleosis, autoimmune, malignancy (leukaemia) and traumas. We present a rare case of bilateral facial paralysis due to Guillain-Barré syndrome which was successfully managed at Department of Paediatrics, Liaquat National Hospital. Patient presented in June 2015, she is on regular follow-up with last visit in March 2016.

Keywords: Guillain-Barre syndrome, facial palsy

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Introduction

Bilateral Facial nerve palsy (facial diplegia) is an immensely rare clinical presentation with the incidence of less than 2% of all cases¹. Its incidence in general population is 1 per 5,000,000¹. Majority of these patients have underlying medical conditions, ranging from neurologic, infectious, neoplastic, traumatic, or metabolic disorders. Only 20% of these cases are due to Idiopathic or Bell's palsy². Its most common cause Guillain-Barre Syndrome (GBS) is an immune polyradiculoneuropathy with overall incidence found to be 1.1/100,000/year to 1.8/100,000/year³. Its association with cranial nerve involvement is a known but rare entity.

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Case Report

A 7-year old girl, admitted at department of paediatrics, Liaquat National Hospital through emergency with presenting complaints of acute onset lower limb pain and progressive weakness resulting in difficulty in walking for last 1 week prior to admission. There was a prior history of chicken pox 20 days back. The lower limb weakness progressed to involve upper limb. She gave no history of respiratory difficulty, voice change and dysphagia. There was no other significant history.

On examination she was conscious, oriented and vitally stable. Her facies were placid or mask like. Neurologic examination of lower limbs revealed bilateral decreased tone and power of 2/5 with areflexia and downgoing plantars. The upper limb examination showed loss of grip and fist formation along with decreased power in proximal group of muscles.

he cranial nerve examination revealed bilateral facial nerve palsy. She was unable to close her eyes, frown, and blow and clench teeth. Being a child she was unable to define any altered sense of taste. The other cranial nerves were intact (Fig. 1a & 1b). Rest of the systemic examination was normal.

A clinical diagnosis of GBS with bilateral facial nerve palsy was made confirmed by electromyography and nerve conduction studies. Electromyography (EMG) results showed moderate degree of generalized motor demyelinating polyneuropathy with moderate degree of bilateral facial nerve dysfunction. It was suggestive of acute inflammatory demyelinating polyneuropathy (GBS).



Fig 1a & 1b: Unable To Close Eyes and Frown Suggestive of Bilateral Facial Palsy (with permission from patient).

Patient was immediately started on intravenous immunoglobulin (IVIg) therapy, with significant improvement in power of muscles and progression of weakness was also ceased.

After 5 days of IVIg therapy, patient had significant improvement in gait, facial and limb muscle power and she was advised physiotherapy for facial weakness and was successfully discharged home. Follow up at 3 months showed complete resolution of weakness (Fig. 2a & 2b).

Discussion

Cranial nerve involvement is a known variant of GBS but bilateral facial diplegia is a rare finding requiring thorough physical examination. Bilateral fa-

cial diplegia is a rare neuropathy a more ominous sign with widely varying causes that requires prompt investigation. Our patient presented to us with sign and symptoms of GBS post varicella infection, with bilateral facial weakness detected on clinical examination and confirmed on EMG and nerve conduction studies NCS. A Case reported⁴ bilateral facial palsy with GBS diagnosed on EMG and NCS. Disease manifested as bilateral facial palsy with paraesthesia and numbness in lower extremity with intact muscle power of all four limbs. A similar case was reported by Ramakrishnan et al,⁵ in which bilateral facial palsy was misdiagnosed as bilateral Bell's Palsy. Later, after a detailed clinical examination, it was found to be a variant of Guillain Barre Syndrome and then managed appropriately unlike our case in which muscle power of all four limbs was affected.



Fig 2a & 2b: After 3 months child is able to close eyes with formation of nasolabial folds and can frown (with permission from patient).

Bilateral facial nerve palsy is extremely rare and requires urgent medical attention and evaluation with most common cause being Guillain-Barre Syndrome (GBS), followed by multiple idiopathic cranial neuropathies, Lyme disease, sarcoidosis, meningitis (neoplastic or infectious), brain stem encephalitis, benign intracranial hypertension, leukemia⁵.

Guillain-Barre Syndrome (GBS) is a relatively common cause of neuromuscular weakness. GBS was found in 72.2% patients with AFP⁶. The study

reported annual incidence of GBS in pediatric patients to be 1.37/100,000, more common in boys⁶. GBS leads to a wide variety of deficits, characterized by rapidly evolving, symmetrical and often ascending limb weakness, loss of deep tendon reflexes, variable sensory signs and autonomic dysfunction along with cranial nerve involvement. In a study at AKUH cranial nerve involvement in GBS was seen in 88.2% with facial diparesis in 63.3% of patients⁷.

Cranial nerve involvement, rapid onset of the illness, maximum disability at admission and features of axonal involvement at initial electromyography (EMG) and nerve conduction studies (NCS) are considered poor prognostic factors. Plasma exchange and IVIG remains the mainstay of treatment⁸⁻¹⁰. Our patient showed dramatic response on IVIG therapy with improvement of facial palsy. There was full recovery noted on follow-up at 3 months.

Conclusion

Bilateral facial nerve palsy is extremely uncommon and can be missed on clinical examination due to lack of facial asymmetry; GBS is one of the reasons and should be kept in differentials, as it responds well to early initiation of treatment. Early detection helps in complete recovery or limited weakness.

Conflict of Interest

Authors have no conflict of interests and no grant/ funding from any organization for this study.

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