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Case Report

Internal Medicine Section

Facial Diplegia with Paresthesia: An Uncommon Variant of Guillain–Barre Syndrome

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ABSTRACT

Facial nerve palsy (FNP) is a common medical problem and can be unilateral or bilateral. Unilateral facial palsy has an incidence of 25 per 100,000 population and most of them are idiopathic. However, facial diplegia or bilateral facial nerve palsy (B-FNP) is rare with an incidence of just 1 per 5,000,000 population and only 20 percent cases are idiopathic. Facial diplegia is said to be simultaneous if the other side is affected within 30 days of involvement of first side. Guillain-Barre Syndrome (GBS) is a common cause of facial diplegia and almost half of these patients have facial nerve involvement during their illness. Facial Diplegia with Paresthesias (FDP) is a rare localized variant of GBS which is characterized by simultaneous facial diplegia, distal paresthesias and minimal or no motor weakness. We had a patient who presented with simultaneous weakness of bilateral facial nerve and paresthesias. A diagnosis of GBS was made after diligent clinical examination and relevant investigations. Patient responded to IVIG therapy and symptoms resolved within two weeks of therapy.

Keywords: Bell's palsy, Brighton criteria, Polyneuropathy, Sarcoidosis

CASE REPORT

A 37-year-old male, presented to the medical emergency with complaints of tingling sensation over his face and inability to close both eyes for a month. His symptoms started one month back when he noted tingling sensation over left side of his face which subsequently spread to the right side in next three days. This was associated with failure to close both eyes completely, dribbling of saliva from either side of his mouth, with slurring of speech and loss of taste sensation. Over next two weeks, the tingling sensation spread to involve both hands and feet. He also had slight difficulty in getting up from squatting position. For his symptoms he had visited a local doctor and was prescribed oral steroids, but his symptoms didn't abate. He was a teetotaller and there was no history of recent vaccination, travel, infection, trauma or sexual promiscuity.

On examination, his pulse rate was 80/minute regular, normal volume and character. Blood pressure in supine position was 110/80 mmHg with no orthostatic hypotension. He was afebrile, and respiratory rate was 16/minute. Cranial nerve examination revealed bilateral symmetrical lower motor neuron type of facial nerve palsy of grade V severity on House-Brackmann scale [Table/Fig-1,2]. No other cranial nerve involvement was noted. There was loss of vibration sense and proprioception in lower limb. Also, there was mild weakness of proximal muscles (power 4/5) of lower limb. All deep tendon reflexes were absent and bilateral plantar response was flexor. Other systemic examination was essentially normal.

Blood investigations like haemogram, renal function tests, liver function tests and serum electrolytes were normal. HIV, VDRL, Anti Nuclear Antibody (ANA), Anti-Neutrophil Cytoplasmic Antibody (ANCA), Hepatitis B Surface Antigen (HBsAg) and Anti hepatitis C antibody were negative. Vitamin B12, folate and serum ACE (Angiotensin converting enzyme) levels were also normal. Chest and Lumbosacral spine roentgenogram was normal. MRI (non contrast) of brain and cervical spine did not show any abnormality. A lumbar puncture was performed on third day of admission under aseptic precautions that showed albumin-cytological dissociation with a protein of 233 mg/dl and total leucocyte count of 0-5/cu mm. Nerve Conduction Velocity (NCV) study of upper and lower limbs showed sensory-motor polyneuropathy which was both axonal and demyelinating. Prolonged F wave latency was also noted in upper and lower limb nerves. NCV of cranial nerve was suggestive of demyelinating type of facial nerve palsy. A diagnosis of Guillain-Barre Syndrome (GBS) was made based on Brighton GBS diagnostic criteria with level 1 of diagnostic certainty. Intravenous Immunoglobulin (IVIG) was started in view of persistent paresthesias at a dose of 0.4 g/kg/day for 5 days. He showed considerable improvement in his paresthesias after one week of therapy. After two weeks of treatment, his facial diplegia improved with mild residual weakness [Table/Fig-3,4].

[Table/Fig-1]: Loss of wrinkles on forehead and nasolabial fold.
[Table/Fig-2]: Inability to close the eyes completely.

[Table/Fig-3]: Recovery in facial diplegia after treatment.
[Table/Fig-4]: Wrinkles on forehead and nasolabial fold appeared after treatment.

DISCUSSION

Bilateral facial nerve palsy (B-FNP) or facial diplegia is rare with an incidence of 1 per 500,000 population [1]. Unilateral Facial Nerve Palsy (U-FNP) is relatively common and almost half of the cases are idiopathic, called as Bell's palsy. Same is not true for B-FNP and only 20% cases are idiopathic, remaining is secondary to various life threatening illnesses. The common causes of facial diplegia are Lyme's disease, GBS, sarcoidosis, diabetes, acute leukemia, porphyria, HIV, Multiple sclerosis and idiopathic (Bell's palsy).

GBS is an autoimmune mediated polyradiculoneuropathy which manifests as ascending areflexic motor paralysis after a flu like illness. GBS can have varied clinical manifestations, hence, there

are several variants of this disease entity. Acute Inflammatory Demyelinating Polyneuropathy (AIDP) is the most common variant, characterized by symmetrical ascending areflexic paralysis. Acute motor axonal neuropathy (AMAN) is unique as there is no sensory involvement and reflexes are preserved [2]. Acute Motor and Sensory Axonal Neuropathy (AMSAN), Miller Fisher Syndrome (MFS), Bickerstaff encephalitis and Pharyngeal-cervical-brachial weakness are other not so common variants of GBS. Other rare variants are pandysautonomia, pure sensory GBS, sixth nerve palsy with paresthesia and facial diplegia with paresthesia.

Facial diplegia is seen in 50% of affected individuals and is often symmetrical. Many cases of B-FNP who are labelled as Bell's palsy have an underlying etiology for cranial nerve paralysis. In a retrospective study, 356 patients diagnosed as Bell's palsy received an alternative diagnosis in follow up, GBS was a commonly missed diagnosis in these patients [3]. Isolated facial diplegia is a rare variant of GBS with only few cases described so far [4]. Motor weakness is absent or minimal and reflexes are generally absent [5]. However, in another variant of isolated facial diplegia, reflexes were exaggerated [6]. Facial Diplegia with Paresthesias (FDP) is a rare localized variant of GBS in which patient presents with simultaneous facial diplegia, distal limb paresthesias and minimal or no motor weakness [7,8]. Deep tendon reflexes are generally absent in FDP variant but rarely can be present or even exaggerated [9]. Diagnosis of GBS is based upon good clinical examination, Cerebrospinal fluid (CSF) analysis and clinical neurophysiological studies. CSF examination done after first week of disease onset often shows albumin cytological dissociation. Nerve conduction studies and Electromyography are very helpful in establishing diagnosis of GBS. NCV studies in FDP often show demyelinating type of neuropathy but axonal polyneuropathy has been also described [10]. Our patient had simultaneous facial diplegia with paresthesias, mild proximal muscle weakness and absent reflexes. CSF showed albuminocytological dissociation and NCV was suggestive of GBS.

Treatment of GBS includes supportive care and disease modifying therapy with Intravenous Immune Globulin (IVIG) or Plasma Exchange (PE). IVIG or PE therapy should be started within first two weeks of disease onset in patients who are not able to walk without support. There are no definite recommendations for patients with milder disease who can walk without support. FDP variant is a milder form of GBS and patient should be ideally observed for two weeks and disease modifying therapy can be started if limb weakness progresses [11]. However, several patients with severe paresthesias have been successfully treated with IVIG therapy [12]. We also noticed a dramatic response to IVIG in this case, although it was given after a month of symptom onset.

CONCLUSION

There are innumerable causes of bilateral facial nerve palsy and patient often tends to visit several doctors from various specialties like ENT, medicine and neurology. A good clinician should try to look for a secondary cause in all patients before labeling them as bilateral Bell's palsy, which is rare. GBS can present as isolated facial diplegia with or without paresthesias and treating physician should be aware about this uncommon variant.

Consent: Prior consent from the patient was obtained for publication of his images.

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