

Case Report

Bilateral Facial Paralysis

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Abstract

This is a retrospective analysis of the records of patients with bilateral facial palsy (BFP) encountered in the clinic between 2012 and 2017. Of the 12 cases of BFP, 3 were due to Bell's palsy, 3 to Guillain Barre syndrome, 2 to trauma, 2 to sarcoidosis, 1 to Miller-Fisher syndrome, and 1 to Lyme disease. The differential diagnosis of BFP includes systemic, infectious, traumatic, neuromuscular, vascular, and toxic causes, as well as idiopathic causes.

Keywords: Bilateral facial paralysis

Facial palsy is a common condition in neurologic practice. Such palsies are usually idiopathic and unilateral. However, a rarer bilateral form is occasionally encountered and comprises 0.3 to 2 percent of all cases of facial palsy. The differential diagnosis of bilateral facial palsy includes systemic, infectious, traumatic, neuromuscular, vascular and toxic as well as idiopathic causes.^[1-4] We want to report twelve patients with unusual presentations of bilateral facial palsy and discuss possible etiologies.

Materials and Methods

We performed a retrospective analysis of medical records of twelve cases with bilateral facial nerve palsy presenting to Neurology Clinic of Kocaeli University Hospital between 2012 and 2017. Main eligibility criterion was presence of bilateral seventh nerve palsy. Simultaneous onset was not required, and involvement of other cranial nerves and minor long-tract signs were accepted exclusion criteria were simultaneous extremity weakness in Guillain-Barre syndrome (GBS) and facial weakness due to lesions of the upper motor neuron, the neuromuscular junction, or the muscle.

The clinical data such as age, sex, antecedent events, date of onset, initial symptoms, duration of disease progression, and time between onset and improvement, other neurological symptoms and signs, results of lumbar puncture, electrophysiology and serological test, neuroimage, treatments and outcome were recorded. All patient were examined within 3 weeks. EMG was performed 8 to 42 days after the onset of facial palsy.

The patient sat on an examination chair and was instructed to close the eyes tightly, wrinkle the forehead, show the teeth, smile, stick the tongue out. Facial paralysis of the patients was evaluated with the House-Brackman (H-B) grading scale. The 6- scale House-Brackman facial function into normal (I), mild dysfunction (II), moderate dysfunction (III), moderately severe dysfunction (IV), severe dysfunction (V), and total paralysis (VI).

Initial laboratory studies were examined a complete blood count, erythrocyte sedimentation rate, blood glucose, liver function studies, FTA-ABS, Angiotensin converting enzyme level, Serum calcium level, Lyme titer, a chest radiography, lumbar puncture, an enhanced CT or MRI and audiometry.

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Table 1. The clinical data of patients with BFP

| Diagnosis | Number | Age | Grade | CSF protein |
|--------------|--------|------------|----------|-------------|
| GBS | 3 | 53, 47, 26 | V, IV, V | High |
| BP | 3 | 40, 72, 68 | V, IV, V | Normal |
| Sarcoidosis | 2 | 65, 33 | V, IV | Normal |
| Trauma | 2 | 22, 25 | III, VI | Normal |
| MFS | 1 | 18 | V | High |
| Lyme disease | 1 | 61 | V | High |

GBS: 25%; BP: 25%; MFS: 8.3%.

Results

Twelve patients were diagnosed as Bilateral Facial Paralysis (BFP), three Guillain Barre Syndrome (GBS), three Bells palsy, two Sarcoidosis, two trauma, one Miller Fisher Syndrome (MFS), one Lyme Disease the clinical data of patients with BFP were summarized in table 1 and analyzed separately as follows.

Clinical Features

Ages of the patients ranged from 18-72 years 7 were female and 5 male. All of the antecedent events in our three cases who two were GBS and one was Bells palsy were upper respiratory tract infection. The interval between antecedent events and disease onset ranged from 7 to 25 days. Sudden onset of paralysis was presented 8 patients of GBS, trauma, Bells palsy, MFS, Lyme disease, while progressive onset was presented 4 patients of DM and Sarcoidosis patients. The occurrence of previous symptoms in this study was no previous symptoms 6 cases, ear pain 3 cases (Bells palsy, Lyme disease, DM), headache 2 cases (DM, Sarcoidosis), facial paresthesia one case (Sarcoidosis). In six cases was admitted the hospital with the complaint of facial weakness first on the one side and later within two- three weeks on the other side. In ten cases, physical examination demonstrated bilateral facial nerve palsies with one side being worse than the other side.

Other Neurological Signs

Three patients in whom bifacial weakness was preceded by upper extremity weakness, with bulbar involvement and areflexia were considered to have GBS. One patient with bifacial weakness, ophthalmoplegia, ataxia, and areflexia was considered to have Miller Fisher syndrome. One patient with Sarcoidosis was accompanied by multiple cranial neuropathy (including abducens, glossopharyngeal nerve paralysis). One year ago, the another patient with sarcoidosis developed bilateral anterior uveitis, which responded to local steroid treatment.

Laboratory Findings

CSF protein values was elevated in the GBS patients and Lyme patient. In one patient with sarcoidosis was accompanied by multiple cranial neuropathy, MRI demonstrated multiple small areas of high signal intensity on T2-weighted images with pathological contrast enhancement in the brainstem. The ACE level was 24 mg/ml (normal: 2.8-8.0 mg/ml).

In the another patient with sarcoidosis, chest MRI revealed bilateral hilar lymphadenopathy with reticulo-nodular shadowing in both lung fields. In one patient with Lyme disease, Lyme Ab (IgG+IgM) was 3.6 mg/dl (normal limit was <1 mg/dl). A young woman developed immediate onset bilateral facial paralysis from closed head injury. CT scan demonstrated a longitudinal temporal bone fracture.

Treatment and Outcome

In GBS Patients, intravenous immunoglobulin was given for five days at a dosage of 0.4 g/kg/ day. Six months later, one patient had a slight weakness in all facial muscles and in the proximal muscles of his arms. One patient had a moderate weakness in all facial muscles and a slight weakness in bulbar and all muscles of her arms. One patient gradually improved over the next six weeks. In patients with sarcoidosis, after three days of intravenous therapy, the patients continued with oral prednisolone, which gradually improved after four months. A corticosteroid therapy 1 mg/kg/day for 20 days were administered in case with Bells palsy. The patient's symptoms gradually improved first on the left side and then on the right side after two months. In two cases with Bells palsy and diabetes mellitus, it regulated only hyperglycemia and was given supportive therapy. Six months later, the patients was still presenting a moderate-severe weakness in all facial muscles with a mild amount of synkinesis Oral doxycycline was given a dosage 200 mg twice a day for 4 weeks in case with Lyme disease. Six months later, the patient was still presenting a slight weakness in all facial muscles with a moderate amount of synkinesis. It didn't start any medication for treatment of MFS. Ataxia and facial palsy disappeared on one month. The patient who suffered from compression had a fracture of the left temporal bone and showed poor improvement with severe sequelae of facial muscles. The patient who suffered from a car accident improved after three months.

In two patients with GBS, one patient with MFS, one patient with idiopathic Bell's palsy, one patient with sarcoidosis, one patient with trauma, the nerve conduction studies of the facial nerve showed bilaterally moderate prolonged distal latencies and moderate reduction of compound muscle action potential amplitude. EMG revealed a few spontaneous

activity and bilaterally mild- moderately polyphasic motor units at all sites. In one patient with GBS, one patient with Lyme disease, one patient with Bell's palsy and diabetic, the nerve conduction studies of the facial nerve revealed bilaterally severe prolonged distal latencies and severe decreased of compound muscle action potential amplitude. EMG revealed some spontaneous activity and long duration, high amplitude, and dense polyphasic motor units at all sites. In one patient with closed head injury and a longitudinal temporal bone fracture, one patient with multiple cranial neuropathy due to sarcoidosis, one patient with Bell's palsy and diabetic, all branches of the facial nerve were inexcitable with electrical stimulation. EMG revealed dense spontaneous activity and bilaterally a few polyphasic large motor units at all sites.

Discussion

Bilateral simultaneous facial palsy is an extreme clinical entity. The incidence is approximately one per five million per year. The most common causes of bilateral facial nerve palsy are Lyme disease; Guillain-Barre syndrome; idiopathic (Bell's) palsy, leukaemia, sarcoidosis, bacterial meningitis, syphilis, leprosy, Moebius syndrome, infectious mononucleosis and skull fracture.

In our series, three patients with isolated bifacial weakness were considered to have bilateral Bell's palsy. Two of these patients were old and diabetic. Our cases presented with bilateral pain in the mastoid bone area, facial numbness, change in taste and numbness of the tongue. The etiology of Bell's palsy has been argued about for many years.^[6] Four possible mechanisms have been proposed: (i) genetic, because hereditary factors have proven to be important; (ii) vascular, where edema and compression are result of insufficient blood supply; (iii) infective causes of yet undetermined etiology, and (iv) an autoimmune process.^[5] Bilateral Bell's palsy appears to have the same favourable prognosis as unilateral paralysis.^[7,8] One side appears to recover function quickly with the opposite side taking several weeks to fully recover. We recommend combined treatment with prednisone, which should be initiated as soon as possible. It is well known that with increasing age there is an increase in vascular degeneration, which leads to decrease of the peripheral blood supply. This may be the reason for the decrease in recovery of facial nerve in older patients with Bells palsy.

It is generally agreed that the incidence of diabetes is even higher in elderly patients with Bells palsy and in those patients with bilateral or recurrent palsies. It has been suggested that the occurrence of a facial palsy in a diabetic should eliminate the diagnosis of Bells palsy. A casual re-

lationship has not been established yet, which may mean that diabetics are rarely more prone to developing idiopathic facial palsies.^[18] Subclinical involvement of the contralateral side has been reported in diabetic patients with unilateral facial paralysis.^[19]

Three patients in whom bifacial weakness was preceded by extremity weakness, with involvement of other cranial nerves and elevated CSF protein values were considered to have GBS. Electrophysiological testing in these patients were mildly prolonged distal motor latency, and F response in the extremities. Guillain-Barre syndrome is an immune mediated peripheral neuropathy characterised by acute onset of symmetric limb weakness and areflexia. Patients with typical Guillain-Barre syndrome experience greater weakness of the leg than the arm with an ascending progression. Some patients with Guillain-Barre syndrome however, present muscle weakness only in the oropharynx, neck, and proximal upper limb muscles and a descending pattern of weakness appears as illness progresses. The next common cause proved to be the 'descending' variant of GBS.^[9]

In our series, one patient with bifacial weakness, ophthalmoplegia, ataxia, and areflexia was considered to have Miller Fisher syndrome. In Miller Fisher syndrome, which is characterised by ophthalmoplegia, ataxia, and areflexia and is considered a variant of Guillain-Barre syndrome, facial palsy, independently of limb weakness, is also frequently encountered (in about 50%).^[9, 10] Therefore, facial palsy and the cardinal neurological signs in Miller Fisher syndrome may result from a similar pathomechanism.^[11]

We presented two cases of bilateral facial palsy due to head trauma. The patient who suffered from compression had a fracture of the left temporal bone and showed poor improvement with severe sequelae of facial muscles. The patient who suffered from a car accident had no fracture of the skull and improved after three weeks. Both, unilateral and bilateral facial palsy may result from skull trauma.^[12] Bilateral traumatic facial nerve palsies associated with a head injury usually result from a longitudinal petrous fracture across the skull base and are thus tangential injuries to the facial nerve. They are however rare and, because of the severity of the injury, death probably occurs in many without identification of the facial weakness.^[13]

We presented two cases of bilateral facial palsy due to Sarcoidosis, one of which had facial palsy only. There was a history of polyneuritis cranialis and anterior uveitis, involvement of the lungs was documented in chest MRI scans. The second case presented with elevated ACE levels, there were no other systemic abnormal findings. Bilateral facial palsy was accompanied by multiple cranial neuropathy.

Sarcoidosis is a granulomatous disease of undetermined etiology. Involvement of the nervous system can range from peripheral or cranial neuropathy to central nervous system. Cranial neuropathies are the most common manifestation of neurosarcoidosis. Facial nerve palsies are a classical manifestation of neurosarcoidosis.^[14] The facial nerve is the most commonly affected cranial nerve, with unilateral paralysis present in 1-3% and bilateral paralysis in 0.1-0.2% of patients with sarcoidosis.^[15] The pathogenesis is thought to be infiltration of the nerve by sarcoid granuloma, but clinicopathologic corection is difficult to obtain. The onset of the palsy is often sudden and the palsy may be incomplete. Although the course is variable, the palsy often spontaneously resolves. The response to steroids is thought to be favorable.^[17]

Lyme disease is a multisystemic illness caused by a spirochetal infection transmitted by tick bites. Facial nerve palsy has been found in 11% of patients with Lyme disease, being bilateral in 30-40% of these cases.^[3,4] Erythema and regional lymphadenopathy develop after a tick bite. Some weeks later, myalgia and a lymphocytic meningitis may develop with cranial nerve palsies. Immuneassay using antibody titres to *Borrelia* IgG is the investigation of choice. Isolated facial palsy without evidence of cerebrospinal fluid infection can be treated with oral antibiotics such as doxycycline. The prognosis of borreliac facial paralysis is known to be good.^[16] But in our patient, facial weakness resolved with mild sequelae after four weeks with antibiotic treatment.

Facial nerve conduction studies revealed findings for demyelination that distal latencies were prolonged and for axonal degeneration that compound motor action potential were decreased. Results of needle EMG in facial muscles were supported predictions regarding prognosis and recovery. We conclude that electromyography is an objective physiological method for evaluating facial nerve function during the early and late stages of facial palsy.

Diagnostic measures should aim to exclude conditions requiring the immediate initiation of specific therapy. In this effort, detailed neurological and otological examination appear imperative, supplemented by the appropriate radiologic imaging, electrophysiologic tests and laboratory procedures.

Disclosures

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