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BILATERAL SIMULTANEOUS FACIAL PALSY: CLINICAL ANALYSIS OF FIVE CASES RETROSPECTIVE REVIEW

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ABTRACT

Facial palsy is most often unilateral. Simultaneous bilateral palsy is described when the involvement of the opposite side occurs within 30 days of onset of the first side. It is most often a part of a symptom complex of a systemic disease, occurring in 0.3% to 2.0% of all facial palsy cases. Half of all bilateral facial palsies are idiopathic and classified as Bell's palsy. 5 patients have presented to us with simultaneous bilateral facial nerve weakness over the last 40 years. The management of these cases changed over time with more and more investigative facilities available. We discuss the lessons learnt over time with better understanding of the disease and provide a roadmap in the management of these patients.

INTRODUCTION

Facial palsy is a frequently encountered in otorhinolaryngology practice. It is most often unilateral. Simultaneous bilateral facial palsy (BFP) is described when the involvement of the opposite side occurs within 30 days of onset of the first side.[1] The common causes of unilateral facial palsy are idiopathic (Bell's palsy) and traumatic, while numerous other disorders contribute to a minority of the cases. This contrasts with BFP which is most often a part of a systemic disease. The common causes are Lyme disease, Guillian-Barre syndrome, Bell's palsy, leukaemia, sarcoidosis, bacterial meningitis, syphilis, leprosy, Moebius syndrome, infectious mononucleosis and skull fractures.[2]

Bell's Palsy is responsible for about 20% of simultaneous BFP.[3] BFP accounts for 0.3–2% of all cases of facial palsy[4] and has a reported incidence of 1 in 5 million/year.[5] The manifestations of the two sides may vary. This makes it difficult to identify a partial paralysis on either side leading to an under-estimation of the incidence.

CASES

In the last 40 years, 312 patients with facial nerve palsy (FNP) presented to the senior author. 5 of these patients had BFP. Out of five, three patients were diagnosed as bilateral Bell's palsy. One patient had myeloid leukemia and one patient had bilateral Ramsay Hunt syndrome. The patients were managed according to the protocols at that time and the investigative facilities available.

Case 1:- J.S. 75 male came with a 10-day history of inability to close both eyes, dribbling of saliva, difficulty in chewing and holding solid food in his mouth. (Fig 1) There was no history of ear complaints, trauma, fever, diabetes mellitus or hypertension. General physical, cardiovascular, and respiratory examinations were normal. Motors and sensory system examinations did not reveal any abnormality. Lower motor neuron (LMN) type seventh cranial nerve palsy was evident on both the sides. There was change in the taste sensation. Schirmer's test- showed reduced tear formation, bilaterally.

Rest of cranial nerves were normal. Mild sensorineural deafness was present. The patient was investigated. Routine blood examination was normal. Cerebrospinal fluid (CSF) analysis revealed sugar 50 mg/dl, protein-32 mg/dl and chloride-114 mmol/lt. Venereal Disease Research Laboratory test (V.D.R.L.) was non-reactive and X-ray skull and mastoids were normal. The patient was treated as a Bell's palsy and put on oral prednisolone, 20 mg thrice daily for one week, which was tapered off in the next two weeks along with tablet Buphenine (6mg), one tablet thrice daily for 3 weeks, and vitamin B 6+B 12. There was complete bilateral recovery after 3 weeks.

Case 2:- B.S. 55 years male had complaints of pain in the right leg which had gradually progressed to cranially. He consulted a physician for but did not respond to treatment. He was referred for otorhinolaryngology opinion when he developed right sided facial weakness. On examination, there was complete loss of facial muscle tone on the right side of the face. However, there was no history of giddiness, impaired hearing, fever, ear discharge, tuberculosis, diabetes mellitus, or hypertension. The patient was put on corticosteroids,

vasodilators and vitamins. After 3 days he presented with facial weakness of the left side as well. There was no recovery on the right side and the patient had a "mask face". It was associated with loss of taste, dryness of mouth and watering of eyes. Schirmer's test was positive bilaterally. There was no palpable lymphadenopathy or skin lesions. Pure tone audiometry was within normal limit. All other cranial nerves were normal. Complete hemogram, serum electrolytes, liver and renal function tests were normal. V.D.R.L. was nonreactive. X-ray chest and X-ray mastoid were within normal limits. Lumbar puncture and fundus examination did not reveal any abnormality. The presence of right lower limb pain made us consider sarcoidosis and Guillain-Barre syndrome as the etiology but none of the investigations were in favor of either diagnosis. Patient showed improvement in facial with the steroid regimen prescribed earlier and was continued on that. At the end of the first month he had complete recovery on the right side. With follow up of 3 months, the left side still had minor residual weakness which was noticeable only on close inspection.

Case 3:- NS. 65 year old male presented with the history of headache, giddiness, inability to close the left eye and difficulty in mastication for the last 2 days. He had pain around left ear and over the left sub occipital region. He had impairment of hearing and vesicular eruptions around the left external auditory canal opening. Cranial nerve examination revealed an isolated seventh cranial nerve palsy- LMN type. He was diagnosed as a case of Ramsay Hunt syndrome. After three days, he complained of pain and eruptions around the right ear and FNP on the right side. The patient complained of change in the taste sensation and had positional vertigo. Routine blood examination revealed no abnormalities. CSF examination was normal. V.D.R.L. was non-reactive. X-ray skull and mastoids was normal. On fundus examination, the disc margins were blurred. A diagnosis of bilateral Ramsay Hunt syndrome was made. The patient was put on oral prednisolone, 20mg thrice daily for ten days, which was tapered off in the next 20 days. Acyclovir two grams/day in divided doses, vitamins, vasodilators and Prochlorperazine were also given. The patient recovered completely after 30 days but had post herpetic pain around ears, which lasted for three months.

Case 4:- G.R.40 years male had BFP on presentation. He gave a history of pain in the right ear and followed by facial weakness a week later. In the following week he experienced pain in the left ear and developed facial weakness on that side within the next few days. On examination he had bilateral LMN type of facial palsy which was more prominent on the left side. Ear examination revealed middle ear effusion. Audiometry showed an average air-bone of 42.5dB on right side and 50 dB on left side. Blood examination showed haemoglobin-6.2gm %, total leucocyte count was 54,000cu mm and differential count revealed neutrophils- 8%, lymphocytes-4%, monocyte -4%, promyelos-2%, blasts -82%. Erythrocyte sedimentation rate -162mm in first hour and platelet count was 1,40,000/ cu mm. Lumbar puncture was normal. V.D.R.L. was non-reactive. Peripheral blood film showed increased white blood cells with large number of blast cells with large nucleus and Auer rods in occasional myeloblasts. Bone marrow aspiration showed hyper cellularity with markedly increased Myeloid: Erythroid ratio. The features were consistent with that of acute myeloid leukemia (AML)- M1 subtype. Bilateral myringotomy and a course of antibiotics, decongestants and corticosteroids were able to alleviate the earache. Patient was referred to a medical oncologist and he eventually succumbed to the AML.

Case 5:- A 20 year old male patient visited the hospital with sudden onset of inability to close left eye and deviation of angle of mouth to right side for the last three days. There was no history of trauma or travel. On examination he was found to have LMN facial palsy. No history of alteration in the taste. All other cranial nerves and the neuromuscular system examination was found to be normal. Routine blood investigations, VDRL/HIV, liver function, renal function tests were within normal limits. Pure Tone Audiometry was normal. Type As tympanogram with absent reflexes was recorded. He was started on oral prednisolone 40mg thrice day, Acyclovir, vasodilators and vitamins B1+B6+B12. Prednisolone was tapered subsequently. Within 2 weeks, the facial nerve function improved to normal. As the prednisolone was discontinued, the patient came back with right sided facial weakness. There was no lymphadenopathy, fundus was normal. A contrast enhanced magnetic resonance imaging (MRI) of brain and High resolution computerized tomography of the temporal bone was done which revealed no abnormality. The CSF examination was also normal. The patient was documented as bilateral Bell's palsy. He was given another course of oral steroid therapy. He recovered to normal facial nerve function on both sides within 2 weeks.

Patient	Age	Other complaints/	Gap b/w 2	Initial	Positive	Treatment	diagnosis	Final

	& Sex	positive findings	sides presentation	Palsy Grade	investigation results			palsy grade
1	75 years Male	Change in taste	B/L at presentation	R-VI L- VI	Mild SNHL	Oral prednisolone, vasodilator, vitamins.	B/L Bell's Palsy	R- I L- I
2	55 years Male	Right lower limb pain	3 days	R-VI L- VI	-	Oral prednisolone, vasodilator, vitamins.	B/L Bell's Palsy	R- I L- II
3	65 years Male	Headache, giddiness, earache, vesicular eruptions, vertigo	5 days	R-V L- V	Blurred disc margins	Oral prednisolone, Acyclovir, vasodilator, vitamins.	B/L Ramsay Hunt syndrome	R- I L- I
4	40 years Male	Earache	7 days	R- IV L-V	B/L CHL, Abnormal blood counts and peripheral smear	Antibiotics, decongestants and corticosteroids.	AML- M1 subtype	-
5	20 years Male	-	14 days	R- III L-VI	A-s type tympanogram	Oral prednisolone, Acyclovir, vasodilator, vitamins.	B/L Bell's Palsy	R- I L- I

Abbreviations: b/w- between; R- Right; L- Left; SNHL- Sensorineural hearing loss; CHL- Conductive hearing loss; B/L- bilateral; AML- Acute Myeloid Leukemia.

DISCUSSION

The facial nerve carries motor supply to the muscles of the face and stapedius muscle. It carries secretomotor fibers to lacrimal and salivary glands. It carries sensory fibers from tympanic membrane and taste fibers from anterior part of the tongue. A disruption in function of the nerve can lead to weakening of muscles of facial expression, dryness of eye and mouth, taste disturbances and/or hyperacusis.

Bell's Palsy is an acute facial nerve paresis or paralysis with onset in less than 72 hours without any identifiable cause. It accounts for nearly three quarters of all acute unilateral facial palsies.

The differential diagnosis of facial palsy includes congenital, traumatic, neurological, infections, metabolic, neoplastic, toxic, iatrogenic, and idiopathic etiologies. FNP is generally a unilateral entity and it is very uncommon to find a bilateral facial nerve presentation. Less than 2% of FNP cases present as bilateral involvement.[4]

The various causes of bilateral facial palsy are tabled below

Congenital/ Genetic	Sclerosteosis Moebius syndrome Melkersson-Rosenthal syndrome Dystrophia myotonica Thalidomide embryopathy Congenital absence of facial musculature
Trauma	Bilateral temporal bone / Base of skull fracture Electrical injury Forceps delivery Iatrogenic
Infections	Viral: Herpes Zoster; Epstein –Barre virus; Herpes Simplex; Human Immunodeficiency (HIV) Bacterial: Tuberculosis; Tetanus; Diphtheria; Bacterial meningitis Mycoplasma pneumoniae Spirochaete and protozoa: Syphilis; Leptospirosis; Malaria Borreliosis (Lyme disease) Fungi: Cryptococcal meningitis
Neoplastic	Leukemia, Lymphoma Bilateral tumors- Acoustic neuromas Metastatic tumors Brain stem Gliomas
Metabolic	Alcohol Diabetes Acute porphyria
Toxic	Drugs (Vincristine, Paclitaxel)
Autoimmune	Guillain Barre Syndrome; Sarcoidosis; Wagner's Granulomatosis

	Systemic Lupus Erythematosus; Scleroderma; Multiple sclerosis Sjogren's; Polyarteritis Nodosa
Neurological/ neuromuscular disorders	Benign intracranial hypertension; Multiple sclerosis Parkinson's disease; Myaesthenia gravis; Pseudobulbar and bulbar palsy; Bulbospinal neuropathy
Idiopathic	Bell's palsy
Vascular	Anterior circulation Stroke Cerebral aneurysm Intracranial hemorrhage

Simultaneous or subsequent bilateral facial palsy patients need a necessary close observation and diagnostic workup as they may be having systemic diseases which may be life threatening. The workup should include extensive neurological examination, CT/ MRI of brain for tumors, fractures, and other CNS lesions.

In endemic areas Lyme disease is responsible for BFP in 25-36% of the cases.[7-8]

Lyme disease though, is rarely seen in India. It is caused by a tick-borne spirochete, Borrelia burgdorferi and no specific species of ticks causing Lyme borreliosis in India have been recognized. Although the incidence of facial nerve palsy due to an EBV infection is low, upto 40% of the facial palsies associated with EBV are bilateral.[9]

In leukemia, neurological findings are unusual at the time of presentation but may occur frequently during the disease. The cranial nerve palsies may be due to direct infiltration and compression of the nerve as they pass through their osseous foramina or due to hemorrhages in the trunk of the nerve or inflammation.[10]

Ramsay Hunt syndrome is due to the reactivation of varicella-zoster lying dormant in the geniculate ganglion and has been known to cause BFP.[11,12] Immuno-compromised status puts patients at higher risk. Antiviral therapy is effective to control the disease.

The pathogenesis is such cases is not fully understood yet. An immunologically mediated inflammatory polyradiculopathy similar to Guillain Barre Syndrome has been proposed.

Yanagihara et al[13] classified bilateral Bell's palsy into 3 types. a) Simultaneous: when there is no improvement of one side at the time of involvement of the other side. b) Alternating: one side palsy occurs after other side has recovered. c) Recurrent: Bell's palsy recurs independently on each side of the face. Alternating type was the most common in their study. 3 of our patients were diagnosed as bilateral Bell's palsy. Only one of them could be categorized as alternating type according to the classification.

Most FNP patients usually present with a unilateral palsy. If the palsy is sudden in onset, and no cause is apparent on history and a thorough clinical examination, the patient is provisionally diagnosed as a case of Bell's palsy. 84% of these patients achieve near normal facial nerve function[14] and no immediate tests are recommended.[15] There is strong evidence for initial conservative medical treatment in patients with Bell's palsy.[16] In our department, routine blood investigations, and a drug regimen of steroids, anti-virals and vasodilators is the protocol. The importance of protecting the eye and ways to do it, is stressed upon. A high prevalence of Diabetes mellitus type 2 in this part of the world, makes it imperative to check blood sugar levels when starting steroid treatment. Prednisone in the dosage of 1mg/kg body weight and acyclovir 2gm per day in divided doses has shown beneficial effect[17]. Although steroids and antiviral do not change the natural history of the disease, they may diminish the sequelae of faulty degeneration.[18]

If the patient presents with contralateral palsy within the next 4 weeks, the management protocol shifts towards a more aggressive approach to finding the etiology. Investigations ordered may depend on the presence of any concurrent complaints or signs. A lumbar puncture, CE-MRI, fundus examination and an X-ray or computerized tomography (CT) of the chest are done. If the cause can be identified, specific treatment is started.

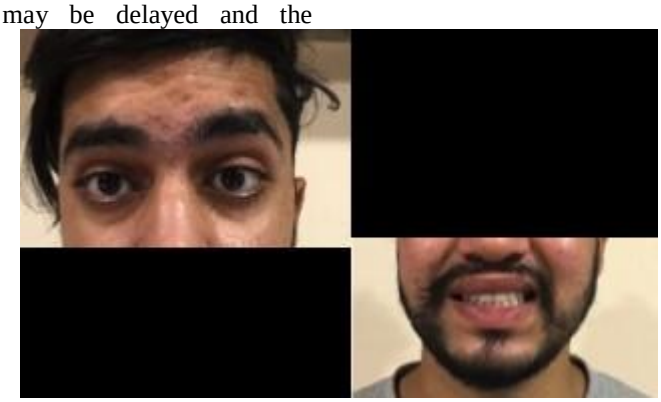
Traumatic skull fractures and cerebello-pontine angle tumors can be excluded with CT scan/ MRI. Contrast-enhanced MRI may even provide a positive radiographic diagnosis of Bell's palsy.[19] Although there is an increasing role of a more extensive investigative panel, primarily of the MRI scan, in the assessment of all unilateral facial palsy, it is not a viable option in clinical practice in all parts of the world at present. Electrodiagnostic testing has a prognostic role and indicated in complete facial paralysis if decompression is being considered. Patients who have greater than 90% degeneration on electroneuronography and have no volitional activity on electromyography are candidates for surgical decompression of the facial nerve and this should be performed within the first 14 days for the best outcome.

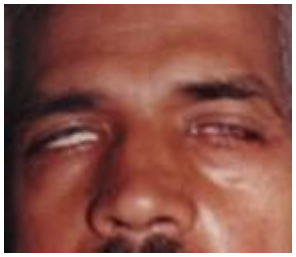
There is some evidence in favour of delayed decompression of the nerve as well.[20]

Keeping in mind the extremely low prevalence of Lyme disease in India, and in the absence of any dermatological findings and we did not consider testing

for it in any of our cases. 4 out of 5 of our cases were followed up and all of them improved to normal or near-normal facial nerve function, with medical management alone.

Since the incidence of BFP is extremely low, clinical diagnosis and treatment may be delayed and the possibility of misdiagnosis is high. BFP can point the treating physician to an underlying systemic disease. It is thus imperative in BFP, that the cause be investigated thoroughly. One of our cases had underlying AML which was diagnosed only after investigating the cause of BFP.





CONCLUSION

The management of unilateral or bilateral facial palsies should be undertaken as per an established protocol. Once bilateral symptoms develop, a systematic approach to unravel the cause should be made. A detailed history, physical examination and neuro-otologic evaluation is necessary. Complete blood and differential counts, renal and liver function tests, viral markers, antibody assays, lumbar puncture, contrast enhanced MRI of brain, and CT scan of chest should be asked for in a sequential manner. This will help rule out any etiologic possibilities. If the diagnosis still falls in idiopathic category and recovery begins on conservative management, a diagnosis of bilateral Bell's palsy is

established. Nerve decompression is indicated in complete paralysis of the nerve, with electrodiagnostic tests predicting poor prognosis. Delayed decompression may be taken up if medical management fails.

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