

Bilateral facial nerve palsy secondary to an atypical presentation of Gullain-Barré syndrome

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ABSTRACT:

Bilateral simultaneous facial nerve palsy is an extremely rare clinical entity and may occur in association with a variety of neurological, infectious, neoplastic or degenerative disorders. We describe a patient, who presented with facial diplegia and normal reflexes on examination. During the entire hospitalization, he developed no motor weakness and remained ambulatory. Whether treatment is warranted for this and other milder variants of Gullain-Barré syndrome is also discussed. Atypical presentations with preserved or brisk reflexes, can be a diagnostic dilemma.

KEYWORDS: Bilateral facial nerve palsy, Gullain-Barré syndrome, Aetiology.

INTRODUCTION:

Unilateral facial nerve palsy is relatively common, with an incidence of around 25 per 100,000 population. The underlying cause is found in only around 20% of cases with the majority of cases attributable to Bell's palsy.¹ Bilateral facial nerve palsy (B-FNP) however, is exceedingly rare with an incidence of 1 per 5,000,000 population. This may represent an underestimate due to the uneven manifestation of the B-FNP.¹ Furthermore the aetiology also differs from that of unilateral facial nerve palsy, often heralding the presence of a serious underlying medical illness.

B-FNP constitutes a medical urgency and all patients need to be evaluated further for the underlying cause and institution of medical treatment. There are many causes of B-FNP and the most common are idiopathic (Bell's) palsy, Guillain-Barré syndrome (GBS), diabetes mellitus, bacterial meningitis, infectious mononucleosis, sarcoidosis and Human Immunodeficiency Virus (HIV) infection.¹ Other causes such as Lyme disease, syphilis and leprosy are well documented in the literature but are less important in our setting.

The management of patients with B-FNP differs only slightly from that of patients with unilateral facial nerve palsy. The main difference is in the diagnostic evaluation of these patients and exclusion of life-threatening conditions. Differential diagnosis is important because the treatment outcome depends on the cause. Unpredictably rapid deterioration (within

hours) in respiratory capacity and progression of muscle weakness can occur in some patients with GBS. If no serious cause is found, the prognosis remains equally favorable to that of unilateral facial nerve palsy.¹ We discuss the approach to diagnosing B-FNP, the steps needed to rule out serious illnesses and a brief summary of our management of our patient.

CASE REPORT

A 39-year-old male cook with no previous medical illness presented to the casualty with acute slurring of speech, and dribbling from the mouth on eating. He was unable to close his eyes and mouth and the tongue had altered taste sensation. He was otherwise ambulant with no peripheral limb weakness. Further history revealed an episode of transient febrile illness two weeks prior to admission with no obvious focus of infection. There was no history of trauma, travel abroad or jungle trekking. He was a teetotaler and lifelong non-smoker and denied any history of illicit drug use, chemical exposure or inhalation. He was a widower and had a history of sexual promiscuity and unprotected sex, the last incident being 4 - 5 months ago.

Examination revealed peripheral (lower motor neuron) paresis of bilateral facial nerves (House-Brackmann grade V) with right sided ptosis, bilateral Bell's phenomenon and inability to purse his lips or smile. (Figure 1a and 1b). Examination of other cranial nerves was normal. Muscle bulk, tone and power in all the limbs were normal. All deep tendon jerks were present and no peripheral nerve thickening was present. Sensory examination did not reveal any superficial or deep sensory loss or hyperaesthesia. There were no cerebellar signs. He had marked symptomatic postural hypotension (> 20 mmHg drop in systolic blood pressure on standing) unaccompanied by cardioacceleration. The heart rate remained unchanged at 90 beats / min irrespective of the postural alterations. There was no further progression in his symptoms and signs throughout his 5-day stay in the hospital.

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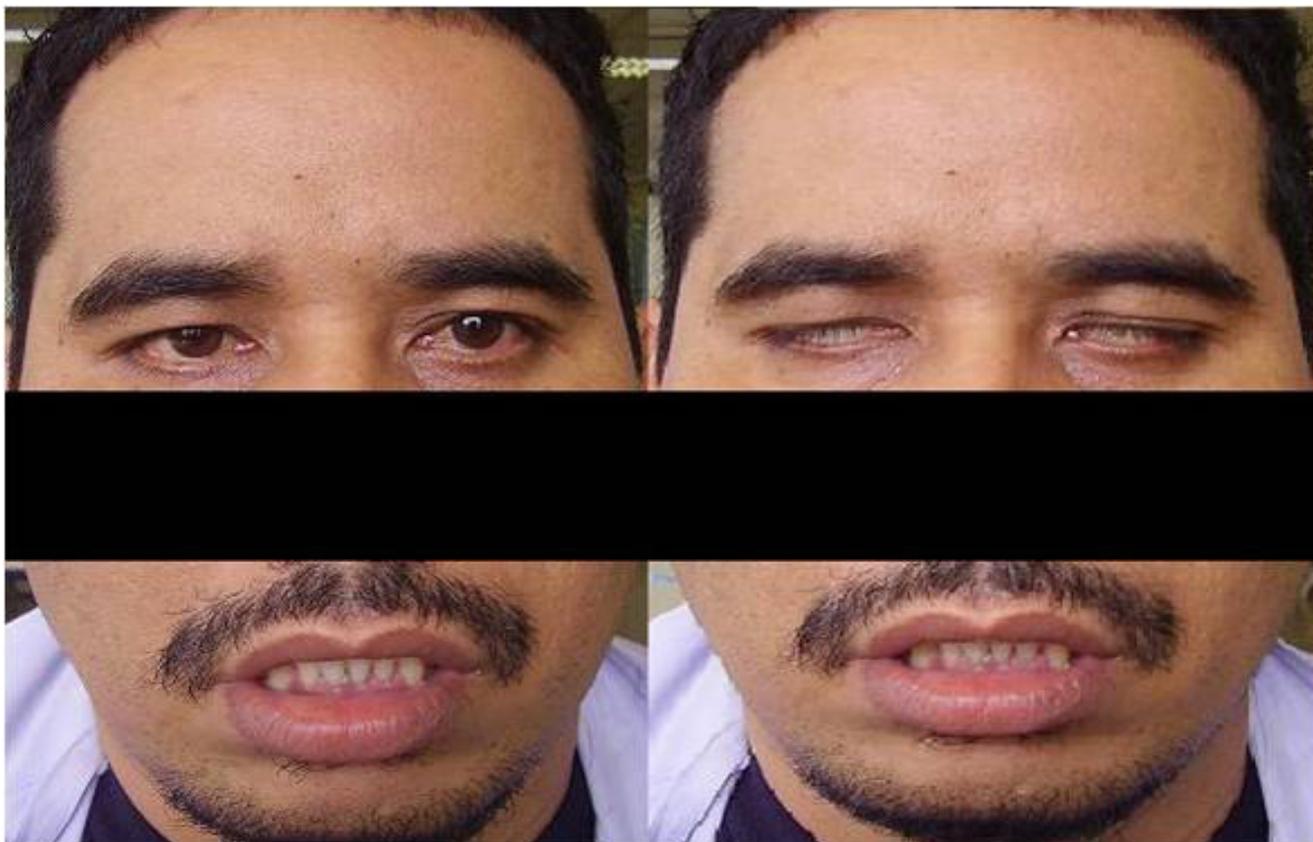


Figure 1a. Bilateral facial nerve palsy

Figure 1b. Bilateral Bell's phenomenon

Pictures courtesy of Dr. Ngiu Chai Soon, National University Hospital, Malaysia.

His full blood picture showed moderate leucocytosis with absolute lymphocytosis. His biochemistry and liver function tests were normal and he had negative VDRL, autoantibody, retroviral, hepatitis and serum angiotensin converting enzyme (ACE) screen. His ESR was mildly raised but the mantoux test was negative. CSF examination revealed raised total proteins with hypocellularity denoting characteristic albumino-cytological dissociation associated with GBS. The results of the investigations are summarized in Table 1.

His chest X-ray and computed tomography of the brain revealed no obvious abnormality (Figure 2a and 2b). The facial nerve conduction study (NCS) on Day 5, revealed reduced conduction velocity, prolonged F-waves and distal latencies.

This patient was observed regularly for any progression in the symptoms. During the entire hospitalization, he developed no motor weakness and remained ambulatory. Intravenous immunoglobulin (IVIg) was withheld as the patient was clinically stable. He was not started on steroids. He was only managed with extensive physiotherapy and symptomatic treatment. He showed extreme unease at our probing questions regarding drug abuse and promiscuity, as he was a Muslim and the notion of promiscuity is frowned upon in the Muslim culture. He eventually took an at-own-risk discharge from the ward and was never seen again despite being called for follow up 2 weeks later.

Table 1 Investigation findings

FBC		
Total White Count	14.8 x 10⁹/L (4.0 - 10.0)	
Lymphocytes	5.3 x 10⁹/L (1.0 - 3.0)	
Haemoglobin	16.4 g/dL (14.0 - 17.0)	
Platelet	356 x 10⁹/L (150 - 400)	
ESR	18 mm/hr	
FBS	4.9 mmol/L (3.0 - 6.7)	
HbA_{1c}	6.6 % (4.4 - 6.4)	
CSF		
Total Protein	598 mg/L (150 - 450)	
Glucose	3.5 mmol/L (2.2 - 4.2)	
Cells	Negative	
C + S	No Growth	
Viral Screen	IgM	IgG
Epstein-Barr virus	Negative	Positive
Cytomegalovirus	Negative	Positive
Herpes Simplex 1 & 2	Negative	Positive
Varicella Zoster	Negative	Positive
Measles	Negative	Positive
Mumps	Negative	Positive
Rubella	Negative	Positive

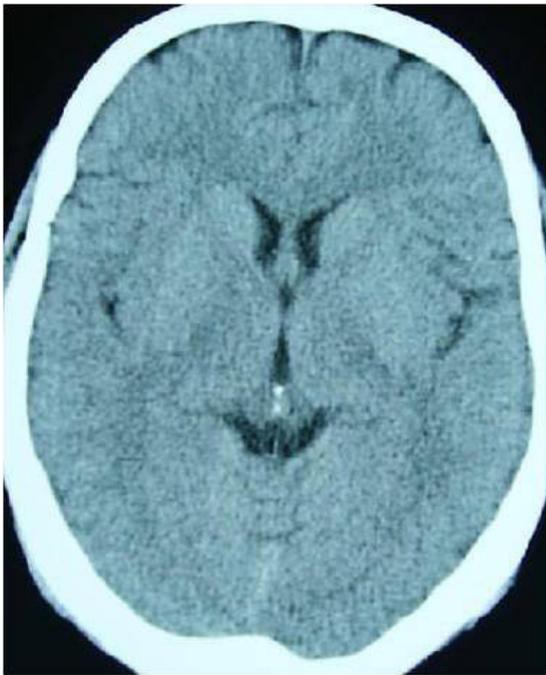


Figure 2a. Normal Brain CT



Figure 2b. CXR showing no hilar or apical lesions

DISCUSSION

In Malaysia the causes of B-FNP are similar to the causes in other areas of the world. Anecdotally the most common causes are idiopathic Bell's palsy, GBS, diabetes mellitus and bacterial meningitis although data is lacking due to the rarity of this condition.² Lyme disease, sarcoidosis, leprosy and syphilis are far less common and thus rarely seen. Isolated facial diplegia with minimal to no limb weakness has been described as a GBS variant.^{3, 4}

Usually in these cases, areflexia helps in distinguishing GBS as the underlying etiology. Our patient presented with isolated facial diplegia and autonomic dysfunction. Ocular muscle palsy is very uncommon, only occurring in approximately 10% of patients. In our patient only partial oculomotor palsy with ptosis on right side was present. The clinical indicators as to the diagnosis of GBS were: acute presentation preceded by a flu-like illness, autonomic dysfunction and albumino-cytological dissociation on CSF examination.⁵ The only atypical finding was the intact reflexes. Preserved reflexes in GBS have been described although they are thought to occur only in pure motor GBS in contrast to areflexia which occurs in sensory-motor forms of GBS.⁶ The case presented here appears to be an exception to this rule. Susuki et al has previously reported two cases of facial diplegia with brisk reflexes as a GBS variant.⁷

Moreover, in our case the NCS showed absent H reflexes and F waves, findings typical of early GBS.⁸ A significant diagnostic quandary pertains to his frequent engagement with sexual workers in unprotected sex. Although it has been 4-5 months since his last unprotected sexual encounter and the preliminary retroviral screen was negative, it is possible that the presentation of GBS portends HIV seroconversion in this

patient.^{9, 10} B-FNP is a rare but recognised manifestation of HIV seroconversion illness. In general, GBS occurs in less immunosuppressed HIV-infected patients (CD4 count $>300/\mu\text{L}$), and is rare in the setting of profound immune compromise (CD4 count $<50/\mu\text{L}$). His utter unease at our probing questions coupled with a less-than-forthright account of the exact timing of his sexual encounters probably led to his eventual at-own-risk discharge on day 5 and subsequent loss to follow up.

It is unclear however if GBS patients with isolated facial diplegia actually warrant treatment. The unpredictability of the early clinical course of GBS makes it difficult to judge which patient will deteriorate. Unpredictable and rapid (within hours) deterioration in respiratory capacity and progression of muscle weakness can occur in up to 30% of patients with GBS, thereby necessitating mechanical ventilation. In addition, autonomic dysfunction may be severe enough to merit ICU monitoring. However treating all "mild" cases may risk exposing them to the potential side effects of IVIG and plasmapheresis. Treatment may be unnecessary in patients who remain ambulatory during the second week of illness.¹¹ Therefore, observation into the second week is advisable to ascertain that the disease does not progress or relapse.

In conclusion, although GBS presenting as B-FNP has been reported before, GBS variant with intact nerve stretch reflexes and presenting as unilateral oculomotor nerve and B-FNP is exceedingly rare. We should withhold immunotherapy in patients with the isolated facial diplegia variant of GBS or "mild" GBS who are still able to walk until the eighth day or so before committing them to IVIG or plasmapheresis.

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