BMJ Case Reports

Unusual presentation of more common disease/injury

Unilateral facial palsy in Guillain-Barre syndrome (GBS): a rare occurrence

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Summary

Guillain-Barre syndrome (GBS) is a postinfectious, autoimmune disorder which, apart from limb weakness, is characterised by cranial nerve involvement. Bilateral facial nerve palsy is the most common pattern of cranial nerve involvement in GBS. However, unilateral facial palsy, although uncommon, can be seen in GBS. We report a rare case of unilateral facial palsy in GBS and importance of electrophysiological tests including blink study in such cases has been emphasised.

BACKGROUND

Guillain-Barre syndrome (GBS) is a group of autoimmune disorders which is characterised by acute onset immune-mediated poly-radiculoneuropathies clinically manifested as acute, rapidly progressive weakness of more than one limb and areflexia.[1] One of the characteristic feature of this entity is facial nerve involvement. Facial paresis in GBS is mostly bilateral but rarely it can be unilateral.[2] We report a rare case of an elderly man who presented with unilateral facial paresis and quadriparesis who was later diagnosed to have GBS. Related literature review and importance of electrophysiological studies in such patients is discussed.

CASE PRESENTATION

A 65-year-old man presented to us with history of weakness in both lower limbs since 5 days. Patient had difficulty in getting up from squatting position. He needed support of one person while getting up. But he was able to stand and walk without support. Patient also complained minimal proximal weakness in upper limbs. Patient gave history of facial asymmetry since 5 days such that there was deviation of angle of mouth towards left side, drooling of saliva from right angle of mouth and there was inability to close his right eye (figure 1A). There was history of paresthesias in both feet and hands. There was no history suggestive of bulbar involvement or any sensory complaint. Patient had history of fever with sore throat 10 days before onset of weakness lasting for 4 days. There was no history of any skin rash or tick bite in the past or any promiscuous sexual behaviour. On examination, his vital parameters including pulse, blood pressure (BP 120/70 mm Hg) were normal. His single breath count was 32. Cranial nerve examination revealed right-sided lower motor neuron (LMN)-type facial palsy. Rest of the cranial nerve examination was normal. Motor system revealed hypotonia in all four limbs. Power was grade 4/5 in upper limbs and in lower limbs, power was grade 4 with predominantly proximal weakness. Deep tendon jerks were absent in lower limbs and hypoactive in upper limbs. Sensory system and cerebellar examination was normal. Other system examination revealed no abnormality.

INVESTIGATIONS

Patient's routine investigations including haemogram, blood sugar and chest x-ray were normal. Brain imaging (MRI) did not reveal any abnormality. Work for possible primary malignancy did not reveal anything. His cerebrospinal fluid examination revealed albumino-cytological dissociation (total cells=5/mm^3, all lymphocytes, protein=100.7 mg/dl and normal sugar). Serological tests for HIV and Lyme's disease (ELISA) were negative. Nerve conduction study (NCS) was performed (table 1). It revealed absence of bilateral median and ulnar sensory nerve action potentials (SNAPs) with bilateral normal SNAPs in sural nerve. Motor NCS showed prolongation of distal latencies of motor nerves with decreased amplitude. The motor conduction velocity was normal in upper limbs, but it was slightly reduced in lower limbs. F-wave

Figure 1 (A) Patient photograph of the face revealed unilateral right-sided facial palsy. (B) At follow-up, patient photograph showed partial facial recovery.

Table 1 Nerve conduction study findings including facial motor study

Nerve	CMAP distal latency (ms)	CMAP amplitude distal/ proximal (mV)	MCV (m/s)	Sensory onset latency (ms)	SNAP Amplitude (mV)	F-wave latency (ms)
Median						
Right	8.25	2.2/2.1	53.7	NR	NR	NR
Left	6.45	2.9/2.9	49.4	NR	NR	NR
Ulnar						
Right	4.30	4.3/3.9	52.6	NR	NR	NR
Left	4.55	4.0/3.4	48.5	NR	NR	NR
Tibial						
Right	6.45	2.1/1.6	32.0	–	–	NR
Left	7.95	1.4/1.1	30.8	–	–	NR
CPN						
Right	10.60	2.0/1.5	31.1	–	–	NR
Left	7.00	2.0/1.4	37.3	–	–	NR
Sural						
Right	–	–	–	2.10	47.6	–
Left	–	–	–	2.15	46.5	–
Facial-nasalis						
Right	5.20	1.0	–	–	–	–
Left	3.70	2.0	–	–	–	–
Facial-orbicularis oculi						
Right	5.65	1.4	–	–	–	–
Left	3.45	2.4	–	–	–	–
Blink reflex latency (ms)						
Right-R1	14.65	Right-R2 ipsilateral	51.15	Right-R2 contralateral	55.55	
Left-R1	13.80	Left-R2 ipsilateral	47.06	Left-R2 contralatetral	51.55	

CMAP, compound muscle action potential; CPN, common peroneal nerve; MCV, motor conduction velocity; NR, not recordable; SNAP, sensory nerve action potential.

response was absent in both upper and lower limbs. Thus, nerve conduction study revealed demyelinating polyneuropathy with secondary axonal degeneration. Facial motor NCS done in nasalis and orbicularis oculi was done which suggested demyelinating type of right facial neuropathy as shown in (table 1). Latency was slightly prolonged on left side also while testing for orbicularis oculi. Blink reflex revealed prolonged R1 latencies on both sides, more on right as compared to left side (figure 2).

TREATMENT

As the patient was ambulatory and the weakness had become static by the time patient presented to us, patient was managed symptomatically.

OUTCOME AND FOLLOW-UP

Patient improved over next 4 weeks so that power in all limbs was almost normal when patient came for follow-up after 1 month. Facial paralysis was also recovered partially (figure 1B).

DISCUSSION

GBS is an acute, acquired, autoimmune polyneuropathy which can involve spinal nerve roots, peripheral nerves and, many a time, cranial nerves also.[3] It is clinically characterised by areflexia, rapidly progressive, usually symmetrical, ascending weakness of extremities, trunk and neck muscles.[1] Since the first scientific description of GBS nearly a century ago (1916), number of its variants and clinical subtypes have been described.[4] Clinically evident cranial nerve palsies are commonly seen in GBS, almost in 50% of such patients. Among them, facial nerve is most commonly affected. Facial nerve paralysis in GBS is usually bilateral, but rarely, clinically evident unilateral facial paralysis may be encountered in GBS.[2 3] Most of the patients with GBS start recovering within 4 weeks and treatment with plasmapheresis or intravenous immunoglobulin may hasten recovery in these patients.[5]

As aforementioned, facial palsy in GBS is usually bilateral, but, it maybe asymmetrical and uncommonly unilateral involvement may be seen.[2] The facial nerve palsy in GBS is secondary to direct attack of antibodies either causing demyelination or axonal degeneration depending on the type of antibody involved. However, hypertension which is commonly seen in GBS due to autonomic disturbances, may also contribute to facial paralysis.[2] Although exact mechanism of hypertension causing facial palsy is poorly understood, it has been proposed that it could be because of edema or haemorrhage within the facial canal causing neural compression. This is especially true in paediatric population.[6] In a review of 35 children with severe hypertension, Lloyd *et al*[6] found that LMN-type facial palsy occurred in seven patients (20%).

There have been very few instances in the literature where unilateral facial palsy in GBS has been reported. Smith *et al*[2] reported two children of GBS with hypertension who developed unilateral facial palsy. In this case report, although it was concluded that facial palsy was a consequence of GBS, combined effect of antibody-mediated facial involvement and hypertension was not ruled out. Fulbright *et al*[5] reported a middle-aged man with GBS who had multiple cranial nerve enhancements on MRI. This patient also had unilateral facial paralysis. Sakakibara *et al*[7] reported a 49-year-old woman with axonal GBS who developed unilateral facial, hypoglossal and phrenic palsies. Kamihiro *et al*[8] recently reported a 2-year-old boy with acute motor-sensory axonal type of GBS who had unilateral facial palsy.

Our patient of GBS had presented with predominantly motor quadriparesis with right-sided almost complete facial palsy. Facial palsy in GBS usually follows the limb weakness.[9] However, in our patient, onset of facial weakness was simultaneous with that of limb weakness. Also, it was not associated with hypertension. Nerve conduction study of facial nerve revealed significantly prolonged latencies with normal amplitude on right side suggesting demyelinating pathology of right facial nerve. In some cases of GBS, facial palsy may be unilateral in onset but sequentially it becomes bilateral during the course of illness. Although there was evidence of some degree of demyelination (in the form of slightly prolonged latency) on left side in our patient, it was not clinically evident even at the time of follow-up at 4 weeks when right-sided facial palsy and limb weakness has recovered partially. This signifies the role of electrophysiological studies such as facial nerve conduction study and blink reflex in GBS to detect subclinical facial nerve involvement, if, clinically, patient has unilateral facial paralysis which may masquerade the diagnosis of GBS. We did not give any specific treatment for GBS either immunoglobulin or

BMJ Case Reports 2012; doi:10.1136/bcr-2012-007077

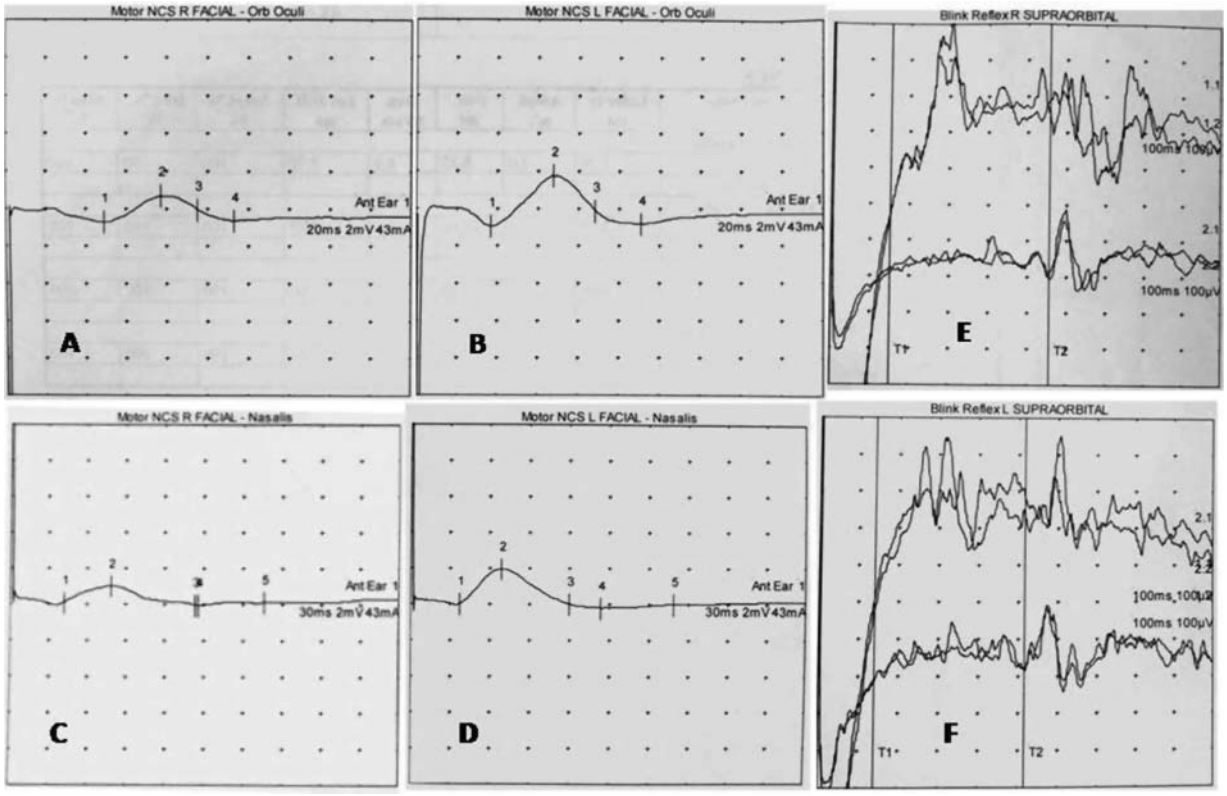

Figure 2 Facial nerve motor and blink studies (A and B, right and left facial—orbicularis oculi; C and D, right and left facial—nasalis; E and F, right and left blink reflexes).

plasmapheresis because treatment may not be necessary if patient remains ambulatory in second week of illness.[10] It also avoids exposing the patient to side effects of these therapies. Other differential diagnoses which need to be considered in this patient include sarcoidosis, Lyme's disease, HIV infection, paraneoplastic infiltration and chronic inflammatory demyelinating polyneuropathy (CIDP). We did not consider CIDP in this patient as patient had history of illness of only 5 days. Work up for rest of the aforementioned causes did not reveal any abnormality.

From this case report, we intend to highlight the fact that unilateral facial paralysis may be a feature of GBS, albeit a rare thing. Electrophysiological test for facial nerve including blink reflex become important part of evaluation of such patients to demonstrate subclinical involvement of facial nerve on opposite side.

Learning points

- Clinically evident cranial nerve palsies are seen in nearly half of the patients with Guillain-Barre syndrome (GBS).
- Facial nerve, usually bilateral, is the most common cranial nerve affected in GBS.
- Unilateral facial palsy, albeit rare, can be seen in GBS.
- Electrophysiological tests, including blink reflexes, assume significant importance in such patient to demonstrate subclinical affection of facial nerve on the other side in such patients.

Competing interests None.

Patient consent Obtained.

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