



A rare case of fulminant GBS mimicking brain death with clinical recovery

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Abstract: A 28-year-old male patient admitted with an acute onset areflexic quadriparesis requiring mechanical ventilation within 24 h of admission. The clinical findings of absent motor response and loss of brainstem reflexes mimicked a “brain death” pattern. MRI brain and spine were normal. Nerve conduction studies were suggestive of early acute inflammatory demyelinating polyneuropathy and cerebrospinal fluid (CSF) showed albumino-cytological dissociation. Electroencephalographic (EEG) showed 5–6 Hz generalized slowing. He was treated with 2 cycles of intravenous immunoglobulins (0.4 gm/kg) for 5 days starting on day 1 and 19. Extensive work up was done to rule out other causes of acute flaccid areflexic quadriparesis with multiple cranial nerve involvement. Five sessions of plasma exchange (50 mL/kg) were carried out between day 45 to 55. He showed gradual recovery after being in “locked in” state for almost 55 days. At 9 months post on-set the patient has regained bulbar function and has MRC grade 3/5 power in all four limbs. Other published cases with a similar admission pattern were reviewed. The prognosis is usually very poor as most of the patients died or remained severely disabled.

Keywords: Quadriparesis; Guillain-Barré syndrome (GBS); brainstem reflex; mechanical ventilation; motor recovery

Received: 05 July 2017; Accepted: 24 July 2017; Published: 29 August 2017.

doi: 10.21037/jecm.2017.08.02

View this article at: <http://dx.doi.org/10.21037/jecm.2017.08.02>

Introduction

In 1916, Guillain, Barré and Strohl described the main clinical features of Guillain-Barré syndrome (GBS): motor weakness, areflexia, minor paresthesias and cerebrospinal fluid (CSF) albumino-cytological dissociation. The frequent finding of motor conduction blocks and reduced conduction velocities provide confirmation of widespread demyelination. Common subtypes of GBS are acute inflammatory demyelinating polyradiculoneuropathy, acute motor axonal neuropathy, acute motor-sensory axonal neuropathy and Miller Fisher syndrome.

Fulminant cases of GBS mimicking brain death have been reported in which a rapid clinical deterioration occurs over hours to days (1,2). This clinical presentation is very rare, disease diagnosis can be challenging and patient mortality rate is high.

Case presentation

A 28-year-old male patient presented to emergency department with complaints of acute onset diplopia, difficulty in walking and inability to hold objects in his hands since waking on the day of admission. There was history of acute diarrhoeal illness 1 week prior to the onset of symptoms. His neurological exam was consistent with acute flaccid quadriparesis with complete areflexia in all four extremities. There was marked restriction of extraocular muscle movement in all primary directions of gaze. He had bifacial weakness, dysarthria and nasal intonation of voice. He followed commands briskly in spite of limited movements of limbs, eyes and facial musculature. On day 2 of ICU stay, his neurological exam became consistent with clinical brain death as he developed comatose appearance, bilaterally fixed and dilated pupils, absence of corneal reflex

Table 1 Comparison of CSF analysis

CSF analysis	Day 1	Day 10
Color	Colorless	Colorless
Appearance	Clear	Clear
Glucose (mg/dL)	65	112
Protein (mg/dL)	44.6	57.7
Cells	0	5
Clot/cobweb	absent	absent
RBC	0	0

CSF, cerebrospinal fluid; RBC, red blood cells.

and complete loss of extraocular and facial movements. He required mechanical ventilation within 24 h of admission. His MRI brain and spine did not show any parenchymal lesion. Nerve conduction studies showed decreased H amplitude bilaterally with conduction block in left ulnar and right radial nerve suggestive of early acute inflammatory demyelinating polyneuropathy. CSF study on admission showed no cells with protein 44.5 mg/dL. He was started on intravenous immunoglobulins 0.4 gm/kg for 5 days starting from day 1. Repeat nerve conduction studies on day 7 showed inexcitable peripheral nerves. Repeat CSF study showed 5 cells with protein 57.7 mg/dL (*Table 1*). Electroencephalographic (EEG) study demonstrated intact cortical function with patient in persistently awake state. ANA and vasculitis panel, antibody to Acetylcholine receptor, urine for porphyrins and toxicology screen were negative. ELISA for HIV, CSF VDRL and Lyme disease serology were negative. Antiganglioside antibodies were negative. The patient developed autonomic dysfunction in the form of labile blood pressure and “tachy-brady syndrome”. He was treated with IV pulse Methylprednisolone in view of complete absence of response to IVIG. Tracheostomy and PEG tube insertion were planned in view of persistent neurological deficit. On day 18 of admission the patient did not show any change in neurological status. His serum IgG levels were 1,782 [791–1,643]. Second cycle of intravenous immunoglobulins (0.4 gm/kg) for 5 days was administered starting day 19.

The patient remained clinically unresponsive for a period of 55 days during which he was provided DVT and bed sore prophylaxis, physiotherapy for prevention of contractures and muscle wasting and appropriate antibiotics

for nosocomial infections. Plasma exchange (50 mL/kg) was carried out on 5 days from day 45 to 55 of hospital stay. The patient showed extraocular muscles and facial muscle movement from day 55 onwards. He showed progressive recovery over the period of next 45 days and was gradually weaned off mechanical ventilator support. He was discharged after 134 days of hospital stay (*Table 2*). At discharge, he was able to open and close eyes, move head to question /commands and able to mouth words. He had bladder and bowel control. At 9 months post onset of symptoms the patient is able to speak and swallow normally. MRC grading for muscle power at 9 months is as below.

Discussion

GBS is the most common cause of acute flaccid paralysis worldwide with incidence of 1.8 per 100,000. There is slight male predominance with male: female ratio 1.5:1 (3). Dysautonomic features consisting of arrhythmias, hypertension or hypotension can occasionally turn fatal and are associated with poor clinical outcome (4). Mortality rate of 3 to 8 percent arises from complications like sepsis, acute respiratory distress syndrome (ARDS), pulmonary embolism or in rare cases, unexplained cardiac arrest, related to dysautonomia (3). Sixty-five percent of patients are left with persistent minor deficits, 20% with serious disability and only 20% with no residual problems (3). Fulminant GBS with absent brainstem reflexes is a rare occurrence and is usually associated with high mortality. The diagnosis of fulminant GBS is based on rapidly progressive areflexic quadriparesis and early need for mechanical ventilation.

There is presence of albuminocytologic dissociation on CSF analysis with Nerve Conduction study suggestive of demyelination or primary axonal neuropathy (1). This condition has a poor recovery rate with high morbidity and mortality (5). Outcomes of this subset of patients have been described by Vargas *et al.* in the largest case series of 13 patients in 2000. Of the 3 deaths, 2 died from cardiac arrest due to dysautonomia and 1 died due to large anterior wall MI. Only 2 patients recovered with minor deficits and rest were severely disabled (5). Bernard *et al.* described a patient with fulminant GBS mimicking brain death who died from nosocomial infection after 158 days without any motor recovery (6). Nithyashree *et al.*

Table 2 Chronology of clinical events

Day 1: diagnosis of GBS, conscious, flaccid quadriplegia, restricted extraocular movements
Day 2: coma, bilaterally fixed and dilated pupils and loss of extraocular movements
Day 7–10: inexcitable nerves on NCV with CSF protein 57.7 mg/dL
Day19: second cycle of IV immunoglobulins
Day 45: plasma exchange initiated
Day 55: eyelid and extraocular muscle movement
Day 134: extraocular movements, word mouthing, self-voiding, shoulder elevation and neck movements
9 months: as described in <i>Tables 3 and 4</i>

GBS, Guillain-Barré syndrome; NCV, nerve conduction velocity.

Table 3 Upper limb MRC grading

Movement	Right	Left
Shoulder abduction	4/5	4/5
Shoulder adduction	4/5	4/5
Elbow flexion	3/5	3/5
Elbow extension	3/5	3/5
Wrist flexion	3/5	3/5
Wrist extension	3/5	3/5

MRC, Medical Research Council.

Table 4 Lower limb MRC grading

Movement	Right	Left
Hip flexion	3/5	3/5
Hip extension	3/5	3/5
Knee flexion	3/5	3/5
Knee extension	3/5	3/5
Ankle dorsiflexion	2/5	2/5
Ankle plantar flexion	2/5	2/5

described age above 60 years, prodromal diarrhoea, rapid involvement of the respiratory muscles requiring ventilatory support within 24 h of the illness, severe muscle weakness with grade 0 power in all four limbs, presence of cardiovascular autonomic symptoms and electrophysiology showing inexcitable peripheral nerves as factors indicating poor outcome in fulminant GBS (7). Our patient had all the above-mentioned features except

that he was young. In general, the therapeutic response following IVIg is good in those with demyelinating neuropathy and unsatisfactory with axonal features (1). Few studies have shown that in those who are not responding to the first course of IVIg, a second course may show good response even in those with axonal changes (7,8). Our patient had received two cycles of intravenous immunoglobulins 2 weeks apart but remained unresponsive. The decision to treat with IVIg against Plasma exchange was taken considering severe dysautonomia though review of literature suggested equal efficacy of both modalities. However, when the patient did not show any response after two cycles of IVIg, Plasma exchange was done from day 45 to 55 of hospital stay. He responded well to the comprehensive care that included immunotherapy, rehabilitation and management of nosocomial infections. There has been progressive recovery of bulbar weakness and power in both upper and lower limbs. A review of literature for characteristics in cases of fulminant GBS with early requirement of mechanical ventilation can be summarized as below. All cases had requirement of mechanical ventilation within 48 h of admission. Absence of brainstem reflexes with coma is common to all cases. Neuroimaging was normal with nerve conduction study suggestive of severe demyelinating/axonal neuropathy (*Table 5*).

Conclusions

Fulminant GBS has a rapidly deteriorating clinical course associated with high mortality. Patients treated with multiple course of immunotherapeutic agents and persistent

Table 5 Review of literature

Cases	Age/sex	H/O diarrhoeal illness	Clinical features	Complication	Treatment	Outcome
Case 1	57/F	No	Ascending areflexic quadriparesis	Cardiac arrest	Gamma globulin	Death
Case 2	47/M	Yes	Acute onset diplopia and dysphonia with paresthesia of all 4 limbs	Multiple episodes of nosocomial infection	Gamma globulin and plasma exchange	Residual motor deficit
Case 3	32/M	Yes	Acute onset ascending areflexic quadriparesis with bilateral facial weakness	Dysautonomia	Gamma globulin (3 times)	Residual weakness in both lower limbs
Case 4	44/M	Yes	Acute onset flaccid quadriparesis with respiratory distress	Dysautonomia and cardiac arrest	Gamma globulin	Death
Case 5	65/M	No	Acute onset quadriparesis following LRTI with H influenzae	–	Gamma globulin and plasma exchange	Residual quadriparesis with dyscognitive features

H/O, history of; LRTI, lower respiratory tract infection.

supportive care can have good outcome.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript.

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doi: 10.21037/jecm.2017.08.02

Cite this article as: Trivedi T, Reddi R, Agarwal P. A rare case of fulminant GBS mimicking brain death with clinical recovery. *J Emerg Crit Care Med* 2017;1:20.