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Guillain–Barré Syndrome with Absent Brainstem Reflexes: A Case Report

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Abstract

A 41-year-old man was admitted to an intensive care unit following respiratory arrest. One day prior to admission, he had complained of nausea and pain involving the lower limbs. On the night of admission, he developed diplopia, dysphagia and rapidly progressive quadriparesis. He developed respiratory failure requiring mechanical lung ventilation 24 hours later. On the fifth day of his hospital stay, the patient became comatose with absent brainstem reflexes and appeared to be brain dead. The cerebrospinal fluid showed albuminocytological dissociation. The electroencephalogram revealed an alpha rhythmic activity. The electrophysiological evaluation revealed an inexcitability of all nerves. Guillain–Barré syndrome was suspected. With supportive treatment, the patient had a remarkable recovery and now is able to independently conduct his daily activities.

Keywords: Guillain–Barré syndrome, autonomic neuropathy, axonopathy, demyelination, brain death, inexcitable nerves.

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Introduction

Guillain–Barré syndrome (GBS) is an important cause of acute neuromuscular paralysis. Molecular mimicry and a cross-reactive immune response play a crucial part in its pathogenesis, at least in those cases with a previous *Campylobacter jejuni* infection and with antibodies to gangliosides. The type of previous infection and patient-related host factors seem to determine the form and severity of the disease. The diagnosis of GBS is based on a combination of clinical and laboratory features. It is typically a monophasic, sub-acute, symmetrical and predominantly motor neuropathy. In rare cases, GBS can present acute quadriparesis and cranial nerve involvement.

We report the observation of a patient who presented a state mimicking cerebral death. In fact, the patient's efferent nerves were completely dysfunctional and he suffered from fulminant GBS with inexcitable peripheral nerves.

Case Report

A 41-year-old man was admitted to the intensive care unit (ICU) following respiratory arrest. He had no previous or concurrent illnesses and was not taking any kind of medication.

There was no history of recent trauma or infection. One day prior to admission, he had complained of lower limb muscular pain and exhaustion. Twelve hours later, he developed diplopia and emesis.

CBC/Biochemistry	Value	Normal range	Unit
Leucocytes	10.9	4.5–11.0	$10^3/\mu l$
Erythrocytes	4.31	4.5–6.5	$10^6/\mu l$
Haemoglobin	13.4	13.0–18.0	g/dl
Platelets	347.0	150.0–450.0	$10^3/\mu l$
Urea	43	8–53	mg/dl
Creatinine	0.79	0.7–1.2	mg/dl
Sodium	136	136–145	mEq/l
Potassium	3.9	3.5–5.10	mEq/l
Chloride	98	98–111	mEq/l
Calcium	8.6	8.4–10.2	mg/dl
Creatine kinase	49.0	<171.0	U/l
AST	30	10–50	U/l
ALT	48	17–63	U/l
C-reactive protein	5.5	<6.10	mg/l
CSF examination	**Value**	**Reference**	**Unit**
Appearance	Clear		
Cells	<1	0–5	cells
Glucose	67.4	40.0–70.0	mg/dl
Protein	92.0	15.0–45.0	mg/dl
Serologies: blood/CSF			
TORCH	Negative		
Respiratory virus	Negative		
Enterovirus	Negative		
EBV	Negative		
VDRL	Negative		
Borrelia	Negative		
Mycoplasma pneumonia	Negative		
HIV 1, 2	Negative		
Hepatitis A, B, C	Negative		
H1N1 virus	Negative		
Campylobacter jejuni	Negative		
Antiganglioside antibodies			
GM1,GM1b, GD1a, GAlNAc-GD1a	Negative		
GQ1b, GD3, GT1a	Negative		
Imagiology exams			
CT scan	Normal		
MRI	Normal		

Table 1 – Laboratory Tests

At admission, he had sixth left cranial nerve paralysis, hypophonia, dysphagia and sensory ataxia. Twelve hours after admission, he was unresponsive and developed gasping respiration. He required immediate intubation and mechanical ventilation due to respiratory arrest. No sedative drugs had been administered.

On examination, he was apyrexial, his heart rate was 85 beats/min and arterial blood pressure was 140/80 mmHg. His pupils were 5 mm wide and did not react to light. There was no voluntary ocular, facial, tongue or pharyngeal movement. The limbs were flaccid and immobile. Motor power was grade 0 (Medical Research Council grade) in all four limbs and deep tendon reflexes were absent. On the fifth day of his hospital stay, the patient became comatose with no cephalic or peripheral response to pain and no corneal and gag reflexes. Vestibulo-ocular and oculo-cephalic reflexes were absent. Tests indicated the absence of all brainstem reflexes and he appeared to be brain dead. Initial and

complementary laboratory tests performed, as shown in *Table 1*, were all normal or negative.

An examination of cerebral fluid showed it to have normal pressure, be clear and colourless, and have a protein concentration of 92 mg/dl (normal <45 mg/dl). It contained 1 mononuclear cell/mm3 and 67.9 mg/dl of glucose (blood glucose was 87 mmol/l). Due to this albuminocytological dissociation, a diagnosis of GBS was suspected.

CT scan and brain magnetic resonance imaging were normal. An electroencephalogram (EEG) carried out on the sixth day of hospitalization revealed posterior alpha activity. Nerve conduction studies and needle electromyography (EMG) were performed on the eighth hospital day. All the motor and sensory nerves were unexcitable.

Day 1 (admission)	▪ Left VI CN palsy ▪ Dysphonia ▪ Dysphagia ▪ Sensory ataxia
Day 1 (12 hours later)	▪ Acute respiratory failure
Day 2	▪ Arreflexic tetraplegia ▪ Arterial hypertension difficult to control
Day 5	▪ Fixed dilated pupils ▪ Absent vestibulo-ocular and oculo-cephalic reflexes ▪ Loss of corneal and gag reflex
Day 18	▪ Opens eyes at request
Month 2	▪ Normal eye movements ▪ Bilateral facial palsy (+right side) ▪ Swallows ▪ Absent gag reflex ▪ Bilateral shoulder movements ▪ Tetraparesis ▪ Absent deep tendon reflexes
Month 3	▪ Spontaneous ventilation
Month 9	▪ Gait with bilateral support ▪ Hand amyotrophy ▪ Distal tetraparesis ▪ Absent proprioception
Today	▪ Bilateral facial palsy (inconspicuous) ▪ Gait with one side support and with no support on even ground ▪ Hand amyotrophy ▪ Distal tetraparesis ▪ Absent distal proprioception ▪ Stocking-and-glove sensory loss ▪ Distal loss of light touch ▪ Dysaesthesias ▪ Absent deep tendon reflexes ▪ Physically independent for routine activities

Table 2 – Neurologic evolution

The patient was first treated (first 5 days) with IV immunoglobulin, 35 g/day, with no neurological response. Immediately after, he was treated with six plasma exchanges and repeated IV immunoglobulin (5 more days).

The repeated neurological examinations during the first month are shown in *Table 2*.

A tracheotomy was performed 2 days after admission and weaning from mechanical ventilation was started 2 months later. Three months later, spontaneous ventilation was possible without oxygen. He was able to communicate with staff and relatives. Nowadays, our patient is able to walk unaided on even ground. His hands are still amyotrophic, and distal proprioception and deep tendon reflexes are still absent. He has stocking-and-glove sensory loss, distal loss of light touch and dysaesthesias. He is physically independent for routine activities.

Discussion

Fulminant GBS mimicking brain death is a rare occurrence, with about 20 cases reported in the literature[1-4]. As with milder forms, there is a slight male predominance, with peak presentation in the fifth decade of life, often with a history of a recent minor respiratory or gastrointestinal illness.

Our patient was a 41-year-old man with no history of recent trauma or infection. The most relevant feature of this case was the initial clinical presentation. He rapidly progressed to fulminant GBS with complete efferent nerve dysfunction resulting in flaccid quadriplegia, total areflexia, absent brainstem reflexes and respiratory paralysis. Hypothermia, metabolic derangements and exposure to drugs or toxins were ruled out. Nevertheless, our patient did not meet the criteria for brain death declaration[5], as there was no consistent aetiology, which is an inescapable requirement. Absent response with a peripheral nerve stimulator as well as a normal imaging study of the brain and a normal EEG prompted a search for a peripheral cause. Presenting history and rapidly progressive areflexic paralysis, as well as the 'albumin-cytological dissociation' in the cerebrospinal fluid studies, suggested the diagnosis of GBS.

When confronted with a patient in a 'comatose state', the diagnosis of GBS does not seem apparent. Pupillary abnormalities have rarely been described[4] and our patient had total ophthalmoplegia, not mentioned in the criteria for the diagnosis of GBS. Miller-Fisher syndrome, a rare variant of GBS that typically presents with the classic triad of ataxia, areflexia and ophthalmoplegia, should always be suspected in such presentations[1]. Fulminant GBS has a poor recovery rate with permanent disabling weakness[2]. The different therapeutic methods are specified in only some cases. It is therefore difficult to establish treatment guidelines for these types of patients. In addition, in a recent study of patients with GBS, including those with unexcitable nerves, the outcomes in response to plasma exchange or infusion of gamma globulin, or a combination of both treatments, did not differ[3]. Most patients had a prolonged ICU stay and at the time of discharge they were physically dependent for routine activities. Death occurred in some cases caused mainly by cardiac arrest related to dysautonomia[4]. Our patient recovered progressively and presently he can carry out routine activities independently. GBS with absent

brainstem reflexes is an important variant of GBS to consider, because it is potentially easy to make a misdiagnosis of brain death with the inherent consequences. This case illustrates the importance of electrophysiological tests and laboratory and imaging studies in patients with suspected brain death where the cause is not clearly determined.

Learning points

- GBS can mimic brain death.
- GBS should be considered as a possible diagnosis in a comatose patient.
- It is fundamental to perform electrophysiological tests, laboratory and imaging studies in patients with suspected brain death where a cause is not clearly determined.
- Unlike most reported cases, fulminant Guillain–Barré syndrome treated with plasma exchange followed by IV immunoglobulin can be associated with an excellent recovery.

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