Introduction

Guillain-Barré syndrome (GBS) is an autoimmune inflammatory peripheral neuropathy disease, which results from an aberrant organ specific immune response that affects primarily the myelin sheath of the proximal portion of peripheral nerves. In approximately 60-70% of patients, GBS is triggered by a preceding acute disease by 1-3 weeks before, *Campylobacter jejuni* infection is the most frequent antecedent event (32%); followed by cytomegalovirus (13%); Epstein-Barr virus (10%), and other viral infections such as hepatitis A, B, and C; influenza; and human immunodeficiency virus (HIV).[1-4] Other minor precipitating factors include surgery, immunizations, and pregnancy. The diagnosis of GBS is based on a combination of clinical and laboratory features. We report a case of fulminant GBS that developed myocarditis during the course of illness.

Case Report

A 57-year-old woman presented to the emergency department, complaining of bilateral lower extremities weakness followed by bilateral upper extremities weakness of 2 days duration. There were no prodromal illnesses and no history of recent animal bite or substance abuse and her past medical history were not significant. On initial examination, her heart rate was 96/min, her blood pressure was 140/90 mmHg, and she was afebrile. Neurological examination revealed fully intact mental status, 1/5 power in both lower extremities and 3/5 power in both upper extremities with absent deep tendon reflexes in all four extremities. Sensation was intact and cranial nerves; fundoscopy was normal. The laboratory tests—including hemoglobin, total leukocyte count, erythrocyte sedimentation rate, liver function tests, blood urea nitrogen, and creatinine—were all within normal limits, except for mild hypokalemia (3.2 mEq/L). HIV, hepatitis panel, and antinuclear antibody profile were negative.

A few hours later, muscle power of the upper extremities was reduced to 1/5 and she developed bilateral lower motor neuron facial nerve palsy (left > right), respiratory failure, and autonomic instability; requiring intubation and ventilatory support. Cerebrospinal fluid (CSF) examination showed normal proteins and no cells. Nerve conduction studies could not be performed. The patient was treated with a total of 24 g of intravenous immunoglobulin over a 5-day period, in the medical intensive care unit.

During this time, her weakness remained static with signs of autonomic dysfunction. On the 6th hospital day, the patient developed bronchospasm with fall in oxygen saturation and sinus tachycardia, with ST-segment depression and T-wave inversions in leads V1-V6. X-ray chest taken showed bilateral fluffy shadows suggestive of pulmonary edema. Cardiac enzymes demonstrated an elevated serum creatine phosphokinase at 1,966 IU/L and creatine kinase—MB fractions at 117 IU/L. The peaked troponin T level was 0.78 ng/mL. A transthoracic echocardiography showed moderate left ventricular systolic function without regional wall motion abnormality. This presentation was consistent with acute myocarditis. The patient was started on diuretic and angiotensin-converting enzyme inhibitor.

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On the 7th hospital day, the patient suffered a cardiac arrest. She was resuscitated successfully, which included ventilation with 100% oxygen, cardiac massage, and ionotropic drugs. Subsequently, the patient developed hypoxic ischemic encephalopathy after repeated cardiac arrest due to autonomic instability, and she expired on 20th hospital day.

**Discussion**

The main features of GBS are progressive bilateral and relatively symmetric weakness of the limbs with or without involvement of respiratory muscles or cranial nerve innervated muscles. Weakness can develop acutely or subacutely and reaches a plateau, with subsequent spontaneous resolution of paralysis. The pace of progression to respiratory failure from the onset can be dramatic, leading to quadriaparesis and the need for intubation over 24–48 h. Facial nerve palsy is the most common form of cranial nerve involvement in GBS, occurring in at least 70% of patients. Bulbar and ocoulomotor nerves are less often affected, except in patients with the antiGQ1b antibody syndromes.

The diagnosis is usually straightforward with typical clinical features, electrophysiological examination and albuminocytological dissociation. In atypical patients, a clearly increased CSF cell count raises the possibility of another illness, such as a leptomeningeal malignancy, Lyme disease, West Nile virus infection, HIV-related GBS, or poliomyelitis; particularly in developing countries. In our patient, typical clinical features, normal proteins, no cells in CSF examination, rapid progression of weakness to quadriparesis, and respiratory failure strongly suggested the diagnosis of fulminant GBS.

Due to nonavailability of portable electromyographer, electrophysiological examination was not possible in our case due to ventilatory support and hemodynamic instability. Nerve conduction studies help to confirm the presence, pattern, and severity of neuropathy. These studies are essential for research, given specific criteria for categorizing the diagnosis; but nerve conduction studies are not obligatory for the recently proposed Brighton criteria for diagnosis, which were developed for use in resource poor environments.

The striking feature of this case was the development of pulmonary edema due to myocarditis on the 6th day of hospital admission. Myocarditis is most often due to infection by common viruses, such as parvovirus B19, less commonly nonviral pathogens such as *Borrelia burgdorferi* or *Trypanosoma cruzi*, or as a hypersensitivity response to drugs. Few cases of GBS complicated by myocarditis have been reported in the medical literature. Two cases in children and four cases were seen in adults. The clinical expression of myocarditis ranges from the asymptomatic state associated with limited and focal inflammation to fulminant fatal congestive heart failure due to diffuse myocarditis associated with transient ST and T wave abnormalities, atrial and ventricular arrhythmias, heart failure, and death. The precise causal mechanism for myocarditis was unclear in most of the cases. It is believed that the same inflammatory or autoimmune response to an infectious process could be responsible for both peripheral nerve and myocardial damage.

In conclusion, this report highlights a case of fatal fulminant GBS complicated with myocarditis during the course of illness.

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