Severe Autonomic Dysfunction with Cardiac Arrest as the Presenting Feature of Guillain Barre Syndrome

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Abstract: Guillain Barre Syndrome (GBS) typically presents as ascending paralysis. Atypical features are frequently observed. Dysautonomia occurs in majority of the patients with GBS, developing symptoms such as tachycardia (most common), urinary retention, hypertension alternating with hypotension, orthostatic hypotension, bradycardia, other arrhythmias, ileus, and loss of sweating. Severe autonomic dysfunction is important to recognize since this is occasionally associated with sudden death. Here we report a case of GBS in a young female who presented to the emergency department with quadriaparesis and cardiac arrest.

Keywords: Autonomic dysfunction, Cardiac arrest, Guillain Barre Syndrome

INTRODUCTION

Guillain–Barré syndrome (GBS) is a potentially life-threatening postinfectious demyelination that is characterized by rapidly progressive, symmetrical weakness of the extremities [1]. It is an acute polyneuropathy with a variable degree of weakness reaching its maximal severity within 4 weeks [2]. The exact cause is unknown, but often preceded by an infectious illness such as a respiratory infection or the stomach flu [3]. About 25% of patients are reported to develop respiratory insufficiency and many of them exhibit autonomic dysfunction [1].

Several diagnostic criteria for Guillain–Barré syndrome have been proposed. Accurate diagnostic criteria are essential for patient care and research, including clinical trials and vaccine safety studies [4]. Diagnosis is usually clinical, but lumbar puncture and electrophysiological studies can help to substantiate the diagnosis [1]. Molecular mimicry of pathogen-borne antigens leading to generation of cross reactive antibodies which target gangliosides is a part of the pathogenesis of GBS. By the nature of the antecedent infection and specificity of such antibodies the subtype and severity of the syndrome can be partly determined [1]. Both intravenous immunoglobulin (IVIG) and plasma exchange (PE) are found to be effective in GBS, while steroids alone are ineffective [2].

CASE REPORT

A 32 yrs female homemaker with no comorbid diseases was brought to the emergency department in gasping state and she had sudden cardiac arrest. CPR initiated, intubated, reverted and connected to ventilator support. Patient had no pallor, clubbing, icterus, cyanosis or pedal edema. Pulse: 110/min, B.P: 100/70mmHg on Dobutamine infusion 5mcg/kg/min. Cardiovascular, respiratory and abdomen examination was normal. Neurological examination revealed GCS of 2T/15 and all reflexes were absent. Doll’s eye negative, bilateral plantar reflex was absent.

We took a detailed history (as informed by her mother and husband). She was apparently normal 6 months back, had intermittent left ear pain, increased over one month with purulent ear discharge and low grade fever. She went to a local hospital, took injection and oral medication fever subsided. Previous to the present episode patient developed generalized discomfort following which she developed severe pain abdomen. She went to a local hospital and treated as OP. After a few hours her condition worsened, abdomen pain increased and developed pain in hips and legs. She was unable to walk or move lower limbs. With support patient was shifted to car. On the way to hospital she was unable to move all 4 limbs and developed breathlessness. She was brought to our hospital in gasping state and further to which she had cardiac arrest.

A provisional diagnosis of atypical Guillain-Barre syndrome (ascending paralysis with cardiac arrest) was made. She was planned for plasmapheresis after the initial lab values (Table 1), CSF analysis and echocardiogram. Patient gradually started improving with each cycle of plasmapheresis (Five cycles given).
DISCUSSION

Autonomic involvement in GBS occurs frequently and occasionally can be severe. It takes the form of either excessive or inadequate activity of the sympathetic and/or parasympathetic systems. Paroxysmal episodes of excessive autonomic activity signify a poor prognosis. Autonomic dysfunction was closely related to the sudden death [5]. The neuropathy may involve visceral afferent fibers subserving the autonomic nervous system (ANS), parasympathetic efferent fibers, sympathetic efferent fibers, or a combination of these territories. Acute autonomic dysfunction can overtake respiratory failure and thromboembolism as an important cause of death in patients with this disorder [6].

There are important practical considerations in the management of GBS patients with autonomic dysfunction [6]. Prompt diagnosis and early aggressive management with plasmapheresis or immunoglobulin is the key to recovery. In the present case report plasmapheresis was started with high clinical suspicion after the initial labs, CSF analysis and 2D Echo. Interestingly this patient had a dramatic recovery with no significant residual deficit on follow up.

Strictly following the lab criteria for diagnosis of GBS may at times be detrimental to the recovery of the patient. In this patient CSF analysis revealed 20 cells/cumm, no xanthochromia, glucose 61 gm%, protein 55 gm% and no growth in culture.

CONCLUSION

The present case report highlights the atypical presentation of GBS with severe autonomic dysfunction and cardiac arrest. Early recognition and treatment brings a good neurological recovery.

REFERENCES


