Guillain-Barré Syndrome: A Case Study

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Abstract

Introduction: Dallas Cowboys football player, Travis Frederick was diagnosed with Guillain-Barré Syndrome (GBS) after experiencing stingers in his neck during training camp. This disease caused Frederick to sit out for the 2018 season because he developed weakness in his extremities.

Case Presentation: A 48-year-old female presented to the neurology office a five-day history of muscle pain, progressive weakness of the lower limbs, and difficulty in walking. She also developed neck flexor weakness and was brought to the hospital.

Management and Outcome: During the neurological examination, the patient was conscious and alert, but restless and anxious. Her muscle tone was decreased. The CSF analysis suggested that the patient had cyto albumino dissociation. In addition, the nerve conduction study (NCS) showed a slow conduction velocity. Such results are indicative of GBS, so the patient was given an IVIG treatment of 2mg/kg spread out over five doses. By the twelfth day, the patient was discharged from the hospital.

Discussion: This case demonstrates a classical presentation of Guillain-Barré Syndrome (GBS). Often times, an antecedent infection can cause this autoimmune disorder to develop; and consequently the myelin sheaths of the peripheral nervous system are destroyed.

Introduction

In August of 2018, Travis Frederick, a football player who plays the position of center for the Dallas Cowboys, started experiencing stingers in his neck during training camp. After extensive testing by doctors, he was diagnosed with Guillain-Barré Syndrome (GBS). This rare neurological condition prevented him from playing football for the entire 2018 season (Stevenson, 2018).

Guillain-Barré Syndrome (GBS) causes patients to develop weakness in their extremities, and thus physical activity becomes exceptionally difficult. However, Frederick's onset of GBS was caught within two weeks, and since the prognosis for full recovery is excellent if diagnosed early, he is expected to return next season.

Guillain-Barré Syndrome can impact virtually anyone, and this condition will be explored more in depth by analyzing the case of a different patient suffering from the same condition.

Case Presentation

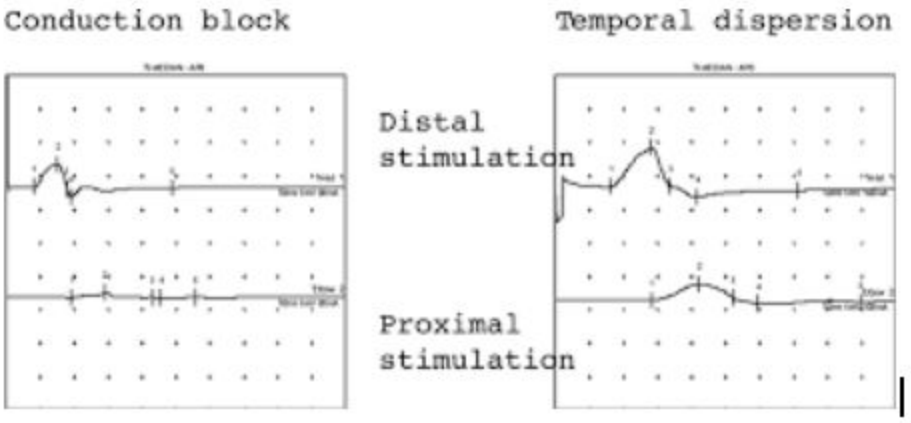
This is a case report over a 48-year-old female who presented to the neurology office a five-day history of muscle pain, progressive weakness of the lower limbs, and difficulty in walking. This difficulty in ambulatory function eventually led to an inability to walk, and and she began to require significant support. By the third day, the weakness had progressed gradually to the upper limbs, with difficulty in lifting her arms. She also developed neck flexor weakness and was brought to the hospital. There was neither an associated bulbar nor a respiratory or autonomic disturbance. Two weeks prior to the onset of symptoms, she had an episode of gastroenteritis that lasted for four days.

Management and Outcome

When examined, the patient was quite anxious, but she was not pale, febrile, icteric, cyanosed, or dehydrated. In addition, respiratory and cardiovascular examinations showed no abnormalities. During the neurological examination, the patient was conscious and alert, but restless and anxious. All her cranial nerves were intact, and there was no sign of meningism. She had flaccid quadriparesis with grade 2 power in both of the lower limbs and grade 3 power in the upper limbs. The patient's muscle tone was also decreased; hyporeflexia of the deep tendons and plantar reflexes were flexor on both sides. The sensory system was normal, and no bladder or bowel dysfunctions were occurring. All of these observations made it clear that the patient had GBS.

The serum urea was 5.0 mmol/l, creatinine was 80 μmol/L, sodium was 132 mmol/l, potassium was 3.5 mmol/l, chloride was 99 mmol/l and bicarbonate was 25 mmol/l. Haemoglobin concentration was 10 g/dl, white cell count was 6.0 × 10^9/l, and urinalysis was normal. The erythrocyte sedimentation rate was 75 mm/h. Venereal disease, hepatitis B surface antigen, hepatitis C virus, and human immunodeficiency virus were also negative.

The CSF analysis showed a white blood cell count of 4 and protein count of 118; such results suggested a cyto albumino dissociation. In addition, the Anti-Ganglioside antibody (Gq1b) was checked and it showed a high positive value. A nerve conduction study (NCS) was performed, and it showed a slow conduction velocity of 26 m/s, which is suggestive of a demyelinating lesion. A sample nerve conduction study is shown below.



Afterwards, in-house physiotherapists were called upon. They reviewed the patient, and she was started on physiotherapy. Physiotherapy interventions include strength, endurance, and gait training with graduated increases in mobility, maintenance of posture, alignment, and joint function. Psychologists provided counselling and support to allay anxiety, fear, and depression.

The patient's symptoms and signs were closely observed and followed up. She was given an IVIG treatment of 2mg /kg spread out over 5 doses. She tolerated the treatment well .

There was no drop in breath count and even during her hospital stay, such a change was not observed. Improvement in muscle weakness were noted. The patient was discharged on the twelfth day of the illness. At the time of discharge, the power was 5/5 in her upper limbs, 4/5 in her lower limbs, and there was no neck flexor weakness.

Discussion

Guillain-Barré Syndrome (GBS) is an acute-onset, monophasic, autoimmune disorder that affects the peripheral nervous system by damaging the axons and myelin sheath of this part of the neural network. Overall, the nervous system is divided into three main components: the central nervous system, which is responsible for communicating signals between the brain and the spinal cord, the peripheral nervous system, which communicates signals to different body parts, and the autonomic nervous system, which is responsible for conducting signals that are not consciously directed. Because of the nature of GBS, individuals who are affected by this condition often experience symmetrical, progressive ascending weakness and/or sensory loss in their extremities, and more serious conditions can even affect the autonomic nervous system, causing patients to lose their ability to breathe and regulate blood pressure. In most cases, the symptoms of GBS reach their nadir, or peak, by the second or third week (National Institute of Neurological Disorders and Stroke, 2018).

GBS can impact virtually anyone regardless of age, gender, race, or family history, and it occurs most often after an antecedent infection or surgery. Culprit pathogens include viruses such as *Epstein-Barr* (mono virus), cytomegalovirus, and bacteria such as *Campylobacter Jejuni*, which cause a diarrheal disease. The flu vaccination has also been reported to cause this condition (Mazidi et al, 2013).

Although there is no exact explanation for why antibodies begin attacking the peripheral nerves, many medical professionals have generally developed a consensus around the Molecular Mimicry Theory. In order to explain this theory in greater detail, some key concepts about the immune system must be established. To begin, B-cells produce antibodies, or infection-fighting agents. Antibodies attach themselves to the epitope of an antigen, or foreign object, in order mark them for destruction by the cytotoxic T-cells. Multiple experiments, including a study by Fujinami et al, have shown how viruses tend to have epitopes that are similar to and react with the host protein; this can lead to the production of monoclonal, or asexually cloned, antibodies that react with these same proteins in uninfected cells. In terms of GBS, an infection can lead to the production of one antibody that attacks uninfected neurons, and as the B-cell creates antibodies, more of these faulty antibodies are manufactured.

However, B-cells are not the only part of the immune system that the Molecular Mimicry Theory applies to; this theory can also be explained in terms of T-cells. Often times, viral peptides can activate T-cells to attack a certain protein, thus resulting in the destruction of uninfected cells. Although the reaction of a cross-reactive immune response is quite common, it must occur at an epitope that is directly affected by the infection in order for the autoimmune disorder to form.

GBS progresses quickly, and thus doctors often find it difficult to diagnose this disease. However, the most important signs to look out for are muscle weakness and symmetrical loss of

coordination that starts distally from the feet and/or hands and ascends proximally to the shoulders and/or hips. Abnormal sensations such as paresthesia and formications, and an abnormal heart rate are also indicators of GBS.

Such observations can be made from a physical examination, and if there is reasonable doubt that this condition is present, then confirmatory tests such as an electromyography (EMG), nerve conduction study (NCS), lumbar puncture, and serological testing can be performed.

The nerve conduction study (NCS) begins with an electrical stimulation of the peripheral nerves in the arms and legs, and this allows for the recording of several neurophysiological markers such as conduction velocity and amplitude. In GBS, the myelin sheaths are more specifically affected, so the conduction velocity of the peripheral nerves is markedly slow. The EMG, which is a test consisting of a needle inserted into a muscle, may or may not be abnormal in GBS. However, if it is abnormal, the EMG would show evidence of acute neurogenic changes.

The lumbar puncture is done by inserting a needle into the L3 or L4 lumbar spine interspace and collecting cerebrospinal fluid (CSF). This is then analyzed for several markers. The most important marker in GBS is the cell and protein count. In GBS, the white cell count is normal, but the protein count is significantly elevated, leading to a term called cyto albumino dissociation. (Soliven, 2008). This is virtually diagnostic of GBS. However, before making a solid conclusion, it is prudent to check for other inflammatory syndromes such as HIV, Sjogrens, Lyme disease, Sarcoidosis, and Lupus.

Treating GBS is particularly difficult because there is no known cure; however, different treatments can be given to patients in order to reduce the severity of the symptoms and shorten recovery time. Overall, there are two main treatments for this syndrome: plasmapheresis (PE) and intravenous immunoglobulin therapy (IVIg). Both of these treatments are quite effective as long as they are started within two weeks of the onset of symptoms. During plasmapheresis, blood is filtered in order to remove the harmful antibodies that are damaging the axons. In immunoglobulin therapy (IVIg), injections of immunoglobulins, a natural protein that attacks infections, is given to patients. This treatment essentially dilutes the number of harmful antibodies with healthy antibodies, so the probability of the autoimmune mechanism occurring is much lower (Jolles et al, 2005).

In addition to acute care, rehabilitative care can also be given to patients who have lost their ability to move and perform daily activities. Through physical therapy, muscle contracture can be avoided and patients can regain full control of their bodies.

Although GBS progresses rapidly, more than 70% of patients are able to recover if proper acute and rehabilitative care is started at an appropriate time. The recovery period varies from patient

to patient, but overall the process is slow and may be incomplete; in fact, 15% of patients become wheelchair or walker-dependent (Mercer, 2017).

Recently, the controversy of vaccines has brought GBS to more prominence; due to the Molecular Mimicry Theory, it is possible that vaccines could also cause this condition. As a result, critics of vaccines often use the GBS as evidence to support their opinions. However, a recent study conducted by Nicola Principi proves that "Less than 1 case of GBS per million immunized persons might occur for these vaccines." Hence, the odds of this condition forming due to vaccines are fairly low.

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