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Guillain-Bare Syndrome in a Patient with HIV Managed with Plasma Exchange: A Case Report from Zambia

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Abstract

A 23-year-old female presented with acute flaccid paralysis following an episode of acute diarrhoeal disease. The patient and family declined a lumbar puncture for cerebrospinal fluid sampling. They did consent to electrodiagnostic studies, which revealed absence of F-waves in all motor nerves examined. This supported a diagnosis of Guillain-Barré syndrome with a certainty of level 2 according to the Brighton criteria. A Human Immunodeficiency Virus (HIV)-test done after admission was positive. Because of the unavailability intravenous immunoglobulins, Plasma Exchange (PLEX) treatment was initiated. After the first session of PLEX her condition deteriorated, and she required ventilatory support, for which she was transferred to the intensive care unit. She had continued sessions of PLEX and physiotherapy while in the intensive care unit. Under these treatments the need for ventilatory support gradually decreased, and the muscle strength improved. The patient was discharged 21 days after admission, and initiated on HIV-treatment as an outpatient. The patient regained full neurological function, and she currently remains virologically suppressed and has had no further neurological complaints. In summary, we report the case of a young HIV-positive female with Guillain-Barré syndrome, who was successfully treated with PLEX. This case highlights the diagnostic and management challenges of Guillain-Barré syndrome in our resource-limited setting.

Keywords: Guillain-Barré syndrome; HIV; Plasmapheresis; Plasma exchange; Lumbar puncture

Introduction

Guillain-Barré Syndrome (GBS) is the most common form of acute paralytic neuropathy [1]. There are several identifiable variants of GBS, which have distinct clinical and pathological characteristics [1]. GBS can be a life-threatening illness, and 20-30% of patients develop respiratory failure and require ventilation at an Intensive Care Unit (ICU) [2]. The estimated incidence of GBS is 0.8-1.9 per 100 000 per year, but these data are mostly from Europe and North America [2]. GBS is slightly more prevalent in men than in women with a relative risk of 1.78 and the incidence of GBS increases with age for both sexes [2]. GBS often follows a bacterial infection of the respiratory or gastrointestinal tract such as *Campylobacter jejuni* and *Mycoplasma pneumonia*. However, viral infections such as Cytomegalovirus (CMV), Epstein-Barr Virus (EBV), Zika virus, influenza A virus and Human Immunodeficiency Virus (HIV) have also been associated with GBS [3]. The infection can induce an autoimmune response in the host that targets peripheral nerves and spinal nerve roots [3]. In the setting of HIV infection GBS has been reported in the context of acute infection (seroconversion), or in severely immunocompromised patients with Immune-Reconstitution Inflammation Syndrome (IRIS) [3-5].

Co-infection with other common viruses or bacteria can also trigger GBS in HIV infected individuals [3]. In Zambia and other African countries, a substantial proportion of GBS patients are HIV-positive [6-9]. Cerebrospinal Fluid (CSF) studies with the typical protein cellular dissociation is what has been the diagnostic tool of choice in our setting as electrodiagnostic facilities are a rarity [10]. The diagnosis of GBS can be made with the help of the Brighton criteria, which takes into account the possibility of suboptimal data for making the diagnosis (Table 1) [11].

Diagnostic criteria	Level of diagnostic certainty			
	1	2	3	4
Bilateral and flaccid weakness of limbs	+	+	+	+/-
Decreased or absent deep tendon reflexes in weak limbs	+	+	+	+/-
Monophasic course and time-phase between nadir 12h to 28 days	+	+	+	+/-
CSF cell count <50 µl/min	+	+[a]	-	+/-
CSF concentration > normal value	+	+/[a]-	-	+/-
NCS findings consistent with one of the subtypes of GBS	+	+/-	-	+/-
Absence of alternative diagnosis for weakness	+	+	+	+

[a]If CSF is not collected or results not available, nerve electrophysiology results must be consistent with the diagnosis of Guillain-Barré syndrome. Level 1 is the highest level of diagnostic certainty; level 4 is the lowest level of diagnostic certainty.

Table 1: Brighton criteria as adapted from Fokke et al. [11]: + present; − absent; +/− present or absent; CSF = cerebrospinal fluid; NCS = nerve conduction studies.

Both Intravenous Immunoglobulins (IVIG) and Plasma Exchange (PLEX) can be used in the treatment of acute GBS, and seem to be equally effective [12,13]. The former is the preferred treatment in many European and North American centres, because of wide availability, minor side effects and convenience of administration [1]. However, it is expensive, and not widely available in developing countries. Plasma exchange is less expensive, and may be more readily available in developing countries. One of the key factors for favourable outcomes in GBS is early initiation of treatment, preferably within two weeks of onset of disease [14]. There is no evidence to support the use of steroids in GBS and this may even be detrimental in the setting of HIV-infection [15]. Here, we present an HIV-positive patient with GBS who was treated with PLEX. The case illustrates diagnostic and therapeutic challenges in a sub-Saharan African setting.

Case Report

A 23-year-old female presented to our services with history of non-bloody diarrhoea one week before presentation that lasted a day and was associated with fever. This was followed by weakness in both feet that was of sudden onset four days before presenting to hospital. This weakness quickly ascended causing inability to walk and shortly after, progressed to involve the upper limbs. She also noticed difficulty in swallowing and would choke when eating food with episodes of nasal regurgitation. At presentation, she could not walk and complained of weakness in both upper and lower limbs. She denied any visual disturbances and had no symptoms of meningeal irritation. Other than an unknown HIV status, she had an unremarkable past medical history and denied being aware of any exposure to toxins. On initial examination, she was alert and oriented with no abnormality in extraocular muscles or pupils. She had nasal speech and showed right facial weakness. Her tongue protrusion was in the midline and she was able to shrug her shoulders symmetrically. Her muscles were of normal bulk in both upper and lower limbs, with no fasciculations or atrophy [16,17]. The muscle tone was generally reduced and she was noted to be areflexic in all four limbs. Muscle strength was scored using the Medical Research Council (MRC) Scale for muscle strength out of 5. By this scale she had a score of 3/5 for shoulder abduction on both sides and a score of 4-/5 and 4/5 for elbow flexion on the right and left side respectively. She scored 0/5 for hip flexion, knee extension, ankle dorsiflexion and planter flexion on both sides.

Sensation was present throughout the body with some hyperalgesia noted in the lower extremities. Vibration sense was intact throughout. Her baseline haematological and biochemistry profile was essentially normal aside an elevated liver transaminase level (Table 2).

	Day 0	Day 11	1 ½ year	2 years	3 years
White blood cell count ($x10^9$/l)	6.82	6.82	6.87	8.79	-
Haemoglobin (g/dl)	15.5	13.2	15.5	13.7	-
Platelet count ($x10^9$/l)	287	99	95	326	-
ALT (U/l)	73	48	-	18.4	-
AST (U/l)		-	-	20.6	-
Potassium (mmols/l)	3.8	3.1	-	-	-
Sodium (mmols/l)	141	142	-	-	-

Creatinine (µmol/l)	57	32	-	53.6	-
Urea (mmols/l)		3.2		-	-
RPR	NR	-	-	NR	-
HBsAg	NR	-	-	NR	-
CD4 count (cells /µl)/ (%)	229/24	-	893(30)		-
Viral load (copies/ml)	-	-		420	

Table 2: Laboratory test results from bloods over the course of two years. HBsAg = Hepatitis B surface antigen; NR = non-reactive; RPR = rapid plasma reagin

An HIV antibody test done on the second day of admission was positive. CSF studies were not done as the patient declined to give consent for a lumbar puncture, after having deliberated with her family. With a strong clinical suspicion of GBS she was moved to the ICU and nerve conduction studies and electromyography were done on the fourth day of admission. The findings of the nerve conduction studies were consistent with GBS, giving her a score of 2 according to the Brighton criteria (Table 1). Treatment with PLEX was initiated on day 5 of admission with follow up sessions on day 6, 8, 11 and 13. Twenty-eight units of Fresh Frozen Plasma (FFP) were used throughout the course of the PLEX treatment. In addition, she received five physiotherapy sessions. After the first session of PLEX, the patient had a stronger voice and improvement in movement was noted all extremities. However, on the second day of PLEX she went into respiratory distress and her oxygen saturation dropped. She improved on Continuous Positive Airway Pressure (CPAP) via closed circuit, but later needed to be intubated and ventilated. Four days after intubation she self-extubated, but she was reintubated the same day as she was unable to maintain her airway. She was kept on the ventilator for the next 8 days, during which there were several unsuccessful attempts to wean her off the ventilator. Her hospital course was further complicated by electrolyte imbalances (Table 2), autonomic dysfunction, and aspiration pneumonia. She gradually regained full strength in the upper limbs by the 12th day of admission, a day before her last session of PLEX. Twenty-one days after admission she was discharged with strength of 3/5 in lower limbs. Ten days later, she was seen in the outpatient clinic and was noted to be fully ambulant. She was initiated on HIV-treatment with a standard combination regimen of tenofovir disoproxil fumarate, emtricitabine, and efavirenz, a month after her HIV diagnosis. She had full immune reconstitution on this regimen and remains virologically suppressed (Table 2). She has had follow up review visits to the outpatient clinic over the course of three years and has retained full neurological function with no new neurological complaints.

Discussion

In this article, we present the case of a young female that presented with flaccid paralysis after an episode of diarrhoeal disease. During admission she was found to be HIV-positive. It was unclear when our patient acquired the HIV infection, as this was her first test in two years, and her first admission to our hospital. Therefore, it is also not certain if the GBS was related to acute HIV infection or to the preceding diarrheal illness that the patient reported. She presented with the acute inflammatory demyelinating polyneuropathy (AIDP) variant of GBS, which is the commonest in HIV infected patients [18]. As the patient declined a lumbar puncture for CSF studies, the diagnosis of GBS was made based on clinical grounds and the results of nerve conduction studies which were consistent with GBS.

There was normal sensory and motor nerve conduction, except for absence of F-waves in all motor nerves studied. This pattern is frequently seen during acute phase of acute inflammatory demyelinating polyneuropathy. She eventually started treatment with PLEX and she made a full recovery. The Brighton criteria is a clinically validated tool [11] that is useful for a diagnosis of GBS as many other conditions may present with acute flaccid paralysis. In resource-limited settings the diagnostic certainty may be limited by a lack of CSF studies and electrodiagnostic studies, hence a level 3 of certainty on the scale may be acceptable. Even though our lab is well able to perform CSF studies, it is not uncommon for patients and families to withhold consent for lumbar puncture [19]. According to a recent survey conducted at our hospital's adult medical wing in February 2017, up to 90% of patients requiring a lumbar puncture denied consent for the procedure. At present, a significant number of patients seeking services at this tertiary institution require a lumbar puncture procedure for various conditions especially in the context of HIV/AIDS. The high rate of decline complicates patient care, and the cost implications in terms of life and finances (though not quantified by studies) are bound to be large. This is particularly so in conditions such as GBS, which require early diagnosis and treatment for improved prognosis.

Physiotherapy, IVIG and PLEX are used in the treatment of GBS. A study in Zambia showed that only a fraction of patients with GBS are referred for physiotherapy [7] while there is no data on outcomes of these therapies in GBS patients in our setting. The first documented reports of GBS in Zambia [10] were of three supportively managed patients with HIV with variable outcomes. With a functioning renal unit, the use of PLEX for GBS is now feasible at our hospital. Intravenous immunoglobulins remain an expensive alternative that is not readily available for all patients. The outcomes for IVIG and PLEX are comparable, but in resource limited settings such as ours PLEX may be the preferred treatment as it is cheaper [20,21]. In our patient we used FFP, but PLEX may

be modified to be more cost-effective with the use of hydroxyethyl starch [22], though the evidence to support this is insufficient [23]. For optimal response, PLEX should be initiated within two weeks of disease onset, and 4-5 sessions should be given over two weeks. There were several challenges in our setting in the timely treatment of our patient. These included a delay in diagnosis due to time lost on awaiting consent for lumbar puncture, and getting electrodiagnostic studies done. Additionally, FFP was provided based on availability from our blood bank which proved challenging on days when the blood bank was running low. Despite these challenges we were still able to provide treatment in accordance with the recommended 5 sessions in 2 weeks.

In summary, we report the case of a young female with Guillain-Barré syndrome, who was diagnosed with HIV during her admission. She developed respiratory failure requiring intubation, but was successfully treated with PLEX. She has fully recovered, and is maintained on HIV-treatment without any neurological sequelae. This case highlights the diagnostic and management challenges of Guillain-Barré syndrome in our resource-limited setting.

Consent

Was obtained from the patient before writing up this report.

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Conflicts of Interest

None declared.

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