Case Report

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Case report on atypical Guillain Barre Syndrome with bulbar dysfunction and descending paralysis

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ABSTRACT

Guillain-Barrie syndrome (GBS) *is a common cause of acute flaccid, usually ascending paralysis, characterized by symmetrical weakness of the limbs and hyporeflexia or areflexia, which reaches maximum severity within 4 weeks. The motor and sensory axons of the peripheral and autonomic nervous systems may be locally or regionally involved in the atypical presentation group of Guillain-Barré syndrome. We describe the case of a male patient, age 17, who came to our ED with symptoms of bulbar dysfunction and descending arreflexic quadriparesis. A nerve conduction test confirmed the diagnosis of atypical GBS. He was treated in the emergency room with mechanical ventilation support for respiratory failure and airway protection and other fundamental supportive care like analgesia and sedation. He was then admitted to the intensive care unit (ICU) and treated for complications, such as autonomic dysfunction and ventilator-associated pneumonia that arose during his stay at the emergency room. After a three-month stay in the ICU, he was transferred to the medical ward, where he was discharged walking with support and able to feed himself with no swallowing difficulty with instructions on how to comply with ongoing medical management of his dysautonomia and follow-up.*

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1. Introduction

GBS is a common cause of acute flaccid paralysis, characterized by symmetrical weakness of the limbs, and hyporeflexia or areflexia, which reaches a maximum severity within 4 weeks.[1] A molecular mimicry attacks the peripheral nervous system responsible for the pathogenesis of this immune system-mediated polyneuropathy.[2] The classical feature of GBS is an acute ascending weakness accompanied by the absence of deep tendon reflexes. Respiratory failure requiring intensive care also occurs in 20-30% of patients. However, other patients showed more benign and uncommon clinical features, which is usually referred to as atypical GBS. Typical presentations of GBS include acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motoric axonal neuropathy (AMAN), and acute motorsensory axonal neuropathy (AMSAN). Atypical GBS is characterized by local or regional involvement of motor and sensory axons of the peripheral nerves and autonomic nervous system. Atypical presentations include cranial nerve involvement, pharyngo-cervical-brachial, and cranial polyneuritis, as well as other presentations such as acute pandysautonomic and acute sensory neuropathy.[3-6] The clinical manifestations of atypical GBS often overlap with those of other diseases in early onset, thus making diagnosis more difficult. In initial onset, serial EMG and sometimes MRI are required to rule out the differential diagnosis if the clinical presentation is atypical. Currently, intravenous immunoglobulin (IVIG) and plasma exchange are proven effective treatments for GBS. However, despite these treatment options, many patients have a severe disease course, pain, and residual deficits.[7-12]

Here, we report a case of atypical GBS, which imposed diagnostic and treatment challenges that resulted in delayed diagnosis and treatment.

2. Case report

A 17-year-old male patient came to St. Paul Adult ED with a complaint of worsening difficulty in swallowing and communication (speaking) for one-day duration.

The symptoms were progressively worsening over the past three days before his presentation. He had a preceding nausea and malaise. On the last day of his presentation, he started to have an associated body weakness, which initially involved the upper extremities, and over half a day involved the lower extremities.

For the above complaints, he visited the Health Center and private clinics and received unspecified IV medications and IV fluids for the diagnosis of acute tonsillopharyngitis (as the attendant claimed). Also, he was evaluated at ENT for considerations of acute tonsillopharyngitis and at the psychiatry unit for possibilities of underlying psychiatric illness like depression at St. Paul's Hospital Millennium Medical College in Addis Ababa. Both sides evaluated and ruled out any illness concerning the respective departments, and finally, he was sent to adult ED. However, the family didn't take him to the ED. Instead, they took him home against the will of the referring physician as the situation was cumbersome for them, only to bring him back the next day to the ED for worsening of symptoms and new onset agitation. Otherwise, he has no history of fever, cough, chest pain, no history of preceding upper respiratory tract infection or diarrheal illness, no history of similar illness in the vicinity or among family members, no known personal and family history of chronic illness (medical/surgical), no previous history of similar episode, and no history of trauma.

On physical examination, he looked acutely sick, in respiratory distress, agitated and non-communicative, having progressive bradypnea (RR was decreasing from 34 to 28, 24, 16…), desaturating to the level of 80%, hypertensive (150/85), and also progressively became bradycardic (pulse decreased from a baseline of 90bpm to 50 to 55bpm). ECG monitor was displaying sinus rhythm. He had

also coarse crepitation bilaterally in the lower 2/3rd of the chest. He had GCS 12(E4V2M6), pupils midsized reactive bilaterally, power 0/5 in upper extremities and 2/5 in lower extremities, hypotonic and arreflexic in all extremities. Cranial nerves IX, X, and XII were affected. Meningeal signs were negative, and there were no sensory level conditions. No other pertinent finding in other systems examination was found.

For the above history and examination findings, respiratory failure secondary to massive aspiration and possible diaphragmatic paralysis plus acute flaccid(descending) paralysis with bulbar dysfunction secondary to botulism r/o atypical GBS was considered.

Investigations: CBC organ function test and serum electrolyte results were unremarkable initially

VDRL, PICT, and Hepatitis B surface antigen were also negative. Brain CT with C-spine and Nerve conduction test was planned to be done after stabilization. Serum and stool assays for identification of Clostridium botulinum toxin were not available in our setup.

At ED the patient was managed with intubation (intubated with sedation alone) and MV support, sedation and analgesia, and ulcer prophylaxis. After initial stabilization on MV, he was admitted to the ICU with the same assessment after 24 hours of stay.

Mechanical ventilation support, physiotherapy, DVT prophylaxis, and other supportive management continued at the intensive care unit. Low-dose morphine, gabapentin, and as-needed IV labetalol were added for the consideration of dysautonomia (Evidenced by persistently and significantly fluctuating vital signs and diaphoresis). Brain and C-spine CT became unremarkable, and nerve conduction test done (On 2nd week) showed reduced motor nerve amplitude with conduction blocks and delayed all F waves latencies with normal sensory nerve exam suggesting motor dominant demyelinating polyradiculoneuropathy likely acute inflammatory demyelinating polyneuropathy. Therefore, the assessment of atypical GBS strengthened, and four doses of IVIG were given subsequently.

Over his ICU stay, he was also managed for ventilator-associated pneumonia (evidenced by persistent fever, leucocytosis with left shift, and consolidative changes seen on bedside US and Chest x-ray), otitis media, and oral candidiasis.

After two weeks of MV support, as he was showing significant improvement, passing the weaning and spontaneous breathing trials, extubation was tried but failed after 24 hours. For this reason, he was re-intubated, and subsequently, a tracheostomy was done. Then, progressively, he was off mechanical ventilation and put on direct oxygen support via a tracheostomy tube. Other supportive management such as tracheostomy care, gabapentin and morphine (For the dysautonomia), analgesia, chest and musculoskeletal physiotherapy, high protein diet via NGT, ulcer, and DVT prophylaxis continued.

Generally, during his ICU stay he showed progressive improvement with relatively stable vital signs, well-controlled dysautonomia, off mechanical ventilation support, desculating oxygen dose support via tracheostomy tube, and improved muscle power (3/5 in all extremities) from the baseline of zero out of five and two out of five in upper and lower extremities, respectively.

After ninety days of stay at the Medical ICU, his condition improved, and he was discharged with advice on continued basic care, physiotherapy, and medical management of the dysautonomia with a short appointment.

3. Discussion

Landry first described acute ascending weakness in 1859, but Guillain, Barré, and Strohl expanded its

extent and characteristics in 1916, naming the disease Guillain Barré Syndrome.[13] The disease gained international notoriety under the name that remains today. Guillain Barré Syndrome.[14] GBS is a rare disease with a median incidence of 0.81-1.89 per 100,000 person-years, more common in men.[1,15] Worldwide, incidence varies, with low rates in Brazil and high rates in Curaçao and Bangladesh.[15-19]

GBS is an autoimmune disease caused by an infectious disease that results in nerve damage or blockage. The type of infection and antiganglioside antibodies determine the subtype and clinical course of GBS. Campylobacter Jejuni is the most common pathogen causing antecedent infections, which is associated with the AMAN subtype of GBS.

GBS patients often experience sensory symptoms like paraesthesia or numbness, cranial nerve deficits, and pain. About half have cranial nerve deficits, while one-third experience muscle weakness.[20] About 25% develop respiratory insufficiency requiring artificial ventilation.[13,21-23] Autonomic dysfunction (predominantly cardiovascular dysregulation) is present in about two-thirds of patients, although its severity is highly variable, with one-third remaining able to walk.[24-26]

Although most GBS cases show classic arreflexic descending paralysis, appropriate attention should be paid to atypical presentations due to the risk of overlooking atypicals, as seen in our instance. The atypical presentation group of Guillain-Barré syndrome is distinguished by localized or regional involvement of peripheral nerve motor and sensory axons as well as the autonomic nervous system.[27,28] AIDP and axonal forms(AMAN and AMSAN) are classified as the typical ascending GBS. The atypical presentation included prominent cranial nerve involvement, Miller Fisher syndrome, Bickerstaff brainstem encephalitis, pharyngo-cervical-brachial and polyneuritis cranialis, and others, which included acute pandysautonomia and acute sensory neuropathy.[13,29] About 8% of patients with GBS present with paraparesis, which often complicates the diagnosis and requires extensive diagnostic work-up. Definite asymmetrical limb weakness, however, is very uncommon in patients with GBS.[14] Although the 1990 GBS criteria require hyporeflexia or areflexia for the diagnosis of GBS, in one cohort of patients with GBS, 9% had normal tendon reflexes in weak arms, and 2% had normal tendon reflexes in weak legs at presentation.[14] During follow-up, all patients developed hyporeflexia or areflexia in their legs, but in some patients, normal reflexes persisted in the arms.[14] For unknown reasons, a small proportion of patients with GBS, especially those with the AMAN subtype, have well-preserved or even exaggerated tendon reflexes.[30,31] Given the supportive feature of nerve conduction studies and improvement with IVIG, though incomplete, our case is also under the common subtype, AIDP, but with a rare form of presentation. The diagnoses are more clinical using criteria, with nerve conduction study and CSF analysis being part of supportive features.[32,33] Electrodiagnostic tests (nerve conduction studies and electromyography) can help with the diagnosis, prognosis, and follow-up of GBS patients. However, they are not required to diagnose patients with typical presentation. Because there are several entities that might induce weakness and sensory impairments, it is critical to rule out other aetiologies before diagnosing GBS. Poliomyelitis, myasthenia gravis, electrolyte disruption, botulism, acute myopathy, diphtheria, vasculitis, porphyria, tick paralysis, and toxic neuropathy are all possible GBS mimics.[34, 35]

Treatment of GBS includes plasmapheresis or intravenous immunoglobulin (IVIg), as well as respiratory support when needed. Plasmapheresis and IVIg in GBS have been shown to be equal in efficacy; however, the ease of use of IVIg has made this the treatment of choice.[36-42] There was controversy regarding whether a steroid regimen

added benefit to therapy, but current recommendations do not support steroid use.[41, 43] Ventilator support is needed in approximately 25% of GBS cases and in cases with more rapid progression. With treatment, most will have a linear progression of recovery in weeks to months. However, those with a more aggressive onset tend to do more poorly with recovery, and overall, 10-20% are left with a disabling motor deficit.[38]

We report this case as it will remain the best example of how differently a GBS patient can present in the ED. Our patient presented with atypical features, such as features of bulbar dysfunction preceding descending limb paralysis. The progression of the weakness is also fast, involving all the four extremities within twenty-four hours. Because of this unusual presentation, the patient was initially subjected to additional health costs, including receiving medications and other costly treatments for tonsillopharyngitis from private clinics (as claimed by his father), delayed diagnosis, and extensive unnecessary visits to various departments. However, this instance will serve as a useful reminder to emergency departments and other departments to thoroughly evaluate patients before sending them out for another visit. Our case is also a typical patient who needs an electrodiagnostic test (EDT) as soon as possible for diagnostic and prognostication purposes. EDT and other pertinent investigation modalities should better be near by ED. We also recommend that the responsible body make serum and/or stool assays for botulinum toxin as botulism is not uncommon in our setup as there are more case reports than atypical GBS in Ethiopia.[44] However, the immediate management of patients with acute peripheral neuropathy or Acute neuromuscular diseases with bulbar dysfunction and/or respiratory muscle weakness in the ED is almost the same, with due attention always being given to anticipation and early airway protection and respiratory support with intubation and mechanical ventilation. And the other supportive management will continue subsequently.

A 10-year retrospective study was conducted in Ethiopia at Addis Ababa University from September 1992 to September 2001 to assess the clinical profile and outcomes of GBS patients in Ethiopia. The study showed that the major presenting feature was an ascending arreflexic quadriparesis(78.5%), followed by descending arreflexic quadriparesis(12.7%). In 7.4% of patients, the weakness was confined to the lower extremities (paraparetic variant). The commonest electrophysiologic abnormality was demyelinating (55.3%), followed by mixed (25.5%) and axonal (19.1%) ones.[45]

This case is one of the rarest presentations of GBS, and no exactly similar case report has been found in the world so far. However, there are few case reports of atypical GBS (including descending weakness pattern but without features of bulbar dysfunction), including in Ethiopia, as seen in the above study.

4. Conclusion

Even though there are various GBS types, our situation does not fit properly into any of them. Our case had atypical GBS symptoms that included initial onset with bulbar palsy and respiratory muscle weakness, which necessitated early intensive care in the emergency room, such as intubation.

Atypical presentations are notorious for delaying diagnosis and therapy. To avoid this, emergency physicians should be aware of GBS patients' varied clinical presentations. A high index of suspicion and early proactive management, including consultation with the appropriate department in the ED, can save lives.

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Competing interests

No conflicts of interest declared.

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