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CASE REPORT

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A case of Recurrent Guillain-Barre Syndrome observed by the same clinician 12 years apart

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Abstract

Recurrent GBS is a rare neurological condition in which patients develop similar symptoms of motor weakness after different preceding infections and suffer shorter intervals in between subsequent episodes of GBS. However, the majority of RGBS patients undergo full recovery.

KEYWORDS

autoimmune, flaccid paralysis, Guillain-Barre syndrome, immunology, lower motor neuron lesion, neurology

1 | INTRODUCTION

The rate of recurrence of Guillain-Barre Syndrome (GBS), after an initial primary insult, runs at approximately 5% (1). Although there is ample analysis and literature review regarding primary occurrence (2), seldom does a clinician treat the same patient for a recurrence a decade apart.

We present the case of a young man with two separate episodes of ascending weakness—each occurrence an established diagnosis of Guillain-Barre Syndrome 12 years apart. Both times, the level of paralysis extended to the diaphragm and subsequent respiratory failure, requiring intubation and a prolonged period of mechanical ventilation. After the first insult, the patient made a complete recovery and is close to a complete recovery during the second, current episode.

2 | CASE

A 43-year-old right-handed man presented to the emergency room, brought in directly from a hospital in Mexico by air ambulance. The medical records accompanying the patient stated he had suffered an upper respiratory tract infection 3 weeks previously, subsequently developing ascending weakness that eventually required intubation and mechanical ventilation 7 days from the onset of symptoms. Further history of the patient revealed that it was his second occurrence of GBS and that he had experienced a complete recovery after the first insult 12 years previously. The patient's family did not choose a hospital close to the United States-Mexican border because they wanted the patient to be seen by the neurologist who treated him during the first occurrence.

On arrival, initial vital signs were stable. The patient was heavily sedated, and his physical examination established a GCS score of 3 (1/1/1), fixed and dilated pupils, unresponsive deep tendon reflexes, and flaccid extremities in all four limbs. Initial CT scan of the head and stat EEG returned normal. On repeat evaluation, the pupils regained some activity, but remained highly fluctuating in terms of their size and reactivity to light.

After the initial assessment, the patient was promptly admitted to the intensive care unit. Due to the confounding factor of sedation, a formal assessment of consciousness could not take place for 3 days. Once the designated time had passed, the patient displayed an ability to nod in response to questions. The patient was inquired whether he had retained sensation, to which he responded with a nod, thereby officially establishing a "locked-in" state.

Family reported that the patient did not receive any treatment in Mexico, as they could not afford either the IVIG or the plasma exchange. On day 2 of arrival (approximately 10 days after the onset of symptoms), plasma exchange was initiated. Due to the fact that Adventist Health White Memorial only carries medical records up to 8 years, the initial episode of GBS that occurred 12 years previously was not available for review. However, from the clinician's own records, in addition to the family's detailed history, it was determined the patient had the same course on the previous occurrence, albeit unknown whether or not the first episode was triggered by a URI or diarrheal illness. Based on the same notes, EMG and nerve conduction test was done after the first presentation and showed reduced conduction velocities as well as absence F waves and H waves. However, the first insult involved therapy in the form of both plasma exchange as well as IVIG. It was only after a full year of therapy and physical rehabilitation the patient returned to his previous baseline and experienced a full recovery. Since then, the patient led a normal professional and personal life until the recurrence. Of note, he did not receive any vaccinations that could have potentially triggered the recurrence in the previous 12 years.

Laboratories included a positive ganglioside antibody. They were measured prior to and after the plasma exchange. Before exchange, GM1 Antibody IgG was 251, and 1 week after exchange, it was 88. Similarly, GM1b Antibody IgG was 255 before plasma exchange, and 1 week after it was 121. All other ganglioside antibodies were within normal limits. Ganglioside antibody testing was not available commercially at our hospital 12 years ago. Unfortunately, nerve conduction studies were not preformed during the relapse. Otherwise, all chemistry, renal function tests, liver function tests, and coagulation panels were all within normal limits.

After a month long stay in acute unit with 2 rounds of plasma exchange (total of 10 exchange sessions), the patient was transferred to an acute rehabilitation unit with a tracheostomy and a gastric feeding tube in place. On departure from acute rehabilitation, he was able to mouth words, his upper extremity strength was 2/5, and his lower extremity was 1/5. There was no reflex loss, no sensory loss, or no bowel/bladder incontinence.

At the 6-month follow-up, he was able to sit up and swallow. The tracheostomy collar had been discontinued, and his upper extremity strength was 4/5, and lower extremity strength was 3/5.

At the 14-month follow-up, he was able to lift both upper extremities above his shoulders and reach the back of his head. In addition to the above, he was able to stand up on his feet and take a few steps with the help of a walker. The patient also was able to speak normally, and his swallowing function had returned to normal. All in all, signifying a gradual and steady road to recovery.

3 | DISCUSSION

Guillain-Barre Syndrome is defined as an acute, immune-mediated inflammatory ascending polyradiculoneuropathy, which targets the peripheral nervous system. The neurological pathology is triggered by either a URI or diarrheal episode in two-thirds of the affected patients[1] and involves the triggering illness that initiates an immune response against gangliosides and glycolipids of the myelin sheaths of the peripheral nervous system. Going further, according to Das' study in 2004, Recurrent Guillain-Barre Syndrome is a rare entity that only occurs in approximately 1%-6% (11 out of 200 in Das' particular study) of the patients suffering an initial insult of the disease[2] with some patients showing whole recuperation while others showing permanent disability (up to 10%) and residual signs such as food drop.[7]

There have been only a few published cases of RGBS in children aged less than 30 and even fewer in adults over the age of thirty.[2,4-6] According to literature (Kuitwaard's study[3]), those recurrences under the age of 30 usually present with varying symptoms and in Miller-Fisher Syndrome (a variant of GBS with a unique antibody as compared to GBS which involves the occulomotor nerves and the brain stem). However, those with the Miller-Fisher variant were more prone to suffer a recurrence as well as more severe clinical deficits and residual effects with each subsequent recurrence.[3]

The current belief is that genetic or immunological host factors may play an important role in recurrent GBS as those particular develop similar symptoms after different preceding infections and patients suffer similar clinical presentations and shorter intervals in between subsequent episodes of GBS.[3] In our case, the interval between episodes of GBS was 12 years, a significant number longer compared to the mean of previous studies in which the lag time between two episodes was 4 months to 10 years in a study conducted by Das et al, and a mean of 7 years with a range from 2 months to 37 years (an outlier) done by Kuitwaard et al.[2,3]

However, despite the longer interval between episodes, our case had identical symptoms, mirroring the initial insult with very similar recovery trajectories, similar to most of the reported cases of RGBS. Wijdicks et al[7] in 1990 had reported five patients having RGBS in a period 4, 10, 15, 17, and 36 years apart, all of whom showed a total recuperation. Kawada et al[8] also reported a case of RGBS five years and seven years after an initial insult showing clinical signs and outcomes identical to the initial episode. Therefore, it is safe to conclude that non-Miller-Fisher variants have a better prognosis and similar roads to recovery as demonstrated by our study in addition to the above.

To summarize, for our particular case, it is important to monitor the indexed patient for another episode of RGBS, the interval of a potential recurrence with worse symptoms, and

the possibility of developing the Miller-Fisher variant of the disease.

AUTHOR CONTRIBUTION

Antonio Liu is P. Z. Kamran Imam is first author working under Dr. Liu's support.

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CONFLICT OF INTEREST

None declared.

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