Bangladesh Crit Care J September 2017; 5 (2): 135-138

Case Report

A case of a recurrent GBS

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Abstract

Background: *Guillain-Barré syndrome is an acquired polyradiculo-neuropathy, often preceded by an antecedent event. It is a monophasic disease but a recurrence rate of 1–6 % is documented in a subset group of patients.*

Case presentation: *Thirty-five-years-old female with past history of near complete recovery following Guillain-Barré syndrome 17 years back presented with acute, ascending symmetrical flaccid quadriparasis extending to bulbar muscles and respiratory compromise needing mechanical ventilation. Nerve conduction study revealed AMAN variant of Guillain-Barré syndrome. Cerebrospinal fluid analysis done after 1 weeks during recurrent episode revealed albuminocytologic dissociation. She was treated with intravenous immunoglobulin resulting in a remarkable recovery.*

Conclusion: *Recurrence of Guillain-Barré syndrome can occur in a subset of patients with Guillain-Barré syndrome even after many years of asymptomatic period. Most patients with recurrent GBS respond favourably to treatment with plasmapheresis or IVIG.*

Keywords : *Recurrent Guillain-Barré syndrome (GBS), Acute motor axonal neuropathy(AMAN), Asymptomatic period.*

Background:

Guillain-Barré syndrome is an acquired polyradiculo-neuropathy, often preceded by an antecedent event[1,2,3]. It is a monophasic disease but a recurrence rate of 1–6 % is documented in a subset group of patients following an asymptomatic period of few months to years (4 months – 10 years)[3,4,5,6]. Guillain-Barré syndrome (GBS) is a heterogeneous group of disorders due to an immune-mediated inflammation and demyelination of the peripheral nervous system, following an antecedent illness in two thirds of the patients, commonly an infection[1,2,3]. It is a medical emergency which usually presents with acute onset, rapidly progressive symmetrical ascending flaccid paralysis of the limbs with accompanying absent or diminished deep tendon reflexes. It is often associated with sensory symptoms, cranial nerve involvement, less commonly autonomic dysfunction and respiratory compromise.

It is suggested by Kuitwaard et al. that there is a subset of patients with GBS who are susceptible for recurrence.Recurrent GBS (RGBS) is characterized by 2 or more attacks of acute inflammatory demyelinating neuropathy with an onset to peak time of 4 weeks or less, and having complete or near complete recovery[3,5]. Compared to non-recurrent GBS group recurrence is characterized by younger age (mean age, 34.2 vs. 46.9 years; 44% were <30 years vs. 22% of the non-recurrent group), milder course of disease and having Miller-Fisher variant of GBS during their first attack. The mean interval between attacks was 7 years (range, 2 months to 37 years)[7].

Literature revealed that the patients with recurrence had similar but more severe symptoms and signs in subsequent episodes while having similar or different antecedent event[3,6]. It is important to distinguish recurrent GBS from GBS with treatment related fluctuations (GBS-TRF) and chronic inflammatory demyelinating polyradiculo-neuropathy (CIDP) as the treatment regimens are different. Cerebrospinal fluid (CSF) shows albuminocytologic dissociation in 82–90% of the patients with GBS after 10–14 days from onset of the illness[8]. Electrophysiological studies and CSF analysis are taken to aid clinical diagnosis of GBS but normal CSF profile can be found in 10 % of GBS patients throughout the disease. Therefore normal values cannot rule out GBS[9].

We present a case of RGBS presenting after 17 years, adding to the limited number of cases with a long asymptomatic interval. Such reported cases from South-Asia are rare.

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Case Presentation:

Mrs Jannatul, 35 years old young lady presented with generalized weakness of four limbs for 2 days with a previous history of loose stool,On next day of admission her condition deteriorates & she has been shifted to ICU due to heaviness in chest with tingling sensation of face. After admission to ICU,patient developed respiratory distress & patient was intubated.Patient was diagnosed as a case of GBS on 2000 and was aided with mechanical ventilation at that time, She Improved with the course of treatment except some residual weakness in right lower limb. Since then she was on her previous health status until 2 days back when she developed loose motion and weakness. On examination upon arrival at ICU she was found conscious and oriented, pulse 80 bpm, BP was recorded 110/70 mm of Hg, afebrile and not dyspneic. On neurological examination higher mental functions were intact. Cranial Nerves including fundoscopy was normal, Bulk of muscles were normal, power was 1/5 in both upper limbs. and 0/5 in both lower limbs. All modalities of sensations were intact. All reflexes were absent and plantar was bilaterally absent. Gait couldn't be evaluated due to muscle weakness, cough reflex were poor. She was intubated on 5th day of illness needed mechanical ventilation and successfully extubated on 23rd day of her illness.

Investigations:

Unfortunately no investigation papers of patients 1st episodes of illness was found except the discharge paper.

CSF study of this episode:

Cerebrospinal fluid profile	Day 07
CSF Glucose (mmol/L)	4.2
Proteins (g/dL)	
(normal: 15 – 40 g/dL)	68
White blood cells/HPF	
(normal: 0 – 5 cells/HPF)	Nill
Neutrophils %	-
Lymphocytes %	-
Red blood cells/HPF	-
Random blood glucose tested at the time of lumbar puncture (mmol/L)	5.7

Sensory Nerve conduction Study:

Nerve	Region	Distal latency (ms)	Conduction velocity (m/s)	Amplitude (uV)
Median (Right)	Wrist	2.24	62.5	44.90 uV
Ulnar (right)	Wrist	1.88	63.8	47.6 uV
Sural(Left)	Wrist	2.18	55	20.50 uV

Motor Nerve conduction Study:

Nerve	Region	Distal latency (ms)	Conduction velocity (m/s)	Amplitude (mV)
Median (Right)	Palm	1.04	19.1 m/s	3.880 mV
	Wrist	4.70	53.3 m/s	2.680 mV
	Cub. fossa	9.20		2.230 mV
Ulnar (Right)	Wrist	4.20	27.7	1.170
	Below Elbow	11.42	22.4	940.0
	Above Elbow	14.10		450.0
Peroneal(left)	Ankle	-	0	-
	Fib Neck	-	0	-
	Popliteal fossa	-	-	-
Tibial (left)	Ankle	7.58	41.8 m/s	990.0 uV
	Pop fossa	16.90		610.0 uV

Discussion:

GBS is an acute, immune mediated inflammatory polyradiculo-neuropathy involving the peripheral nervous system. Onset is preceded by an antecedent event in two thirds of the patients, usually an upper respiratory tract infection or a diarrheal illness[1,2,3], where the causative agent is assumed to trigger an immune response against the gangliosides and glycolipids distributed along the myelin sheaths and peripheral nervous system. This results in marked inflammation of the peripheral nerves, resulting in demyelination and defective impulse propagation. It is a heterogeneous group of disorders which involves motor, sensory and autonomic nervous systems to varying degrees depending on the sub type; [1] Acute inflammatory demyelinating polyneuropathy, [2] Acute motor axonal neuropathy, [3] Acute motor sensory axonal neuropathy, [4] Miller Fisher syndrome, [5] Acute pan-autonomic neuropathy and [6] Pure sensory GBS.

GBS is a monophasic illness, with an annual incidence rate of 1.2–3 per 100 000 population[10]. Yet, recurrence of GBS is observed in 1–6 % of patients, where it is defined as 2 or more attacks of acute inflammatory demyelinating neuropathy with an onset to peak time of 4 weeks or less having complete or near complete recovery [3,4,5,6].

We do not know why a particular trigger will cause one person to develop GBS, while many others in the same situation have no neurological symptoms. Certainly there is an individual susceptibility to developing GBS. This was supported by a study by Dr. K. Geleijns and colleagues, published in 2004. The authors described 12 families of susceptible individuals in which at least two family members developed GBS over the preceding 15 years. The underlying host factors causing susceptibility for these families and for individuals with single-episode and recurring GBS, remain to be discovered. Patients who have residual weakness after GBS may experience excessive fatigue, or has weakness of muscles that were especially affected by the initial GBS. These muscles do not have normal reserve endurance under stress. Weakness may appear after vigorous or prolonged exercise, insufficient sleep, or an unrelated medical illness. This new weakness may be mistaken for a second attack of GBS. Also, decades after recovery from GBS patients may notice slowly increasing weakness in a muscle or limb that was weak during the GBS episode. The most comprehensive study, reported by Dr. R. H. Kennedy and colleagues at Mayo Clinic in 1978, retrospectively followed 40 GBS patients for up to 42 years after the first attack. In that study only one individual had a second episode of GBS, which occurred four years after the initial one[11].

The time lag between two episodes of GBS was 4 months to 10 years in a study done by Das et al. and a mean of 7 years with a range from 2 months to 37 years was described by Kuitwaard et al.[3,5]. Patients tend to get similar clinical presentations and shorter intervals in between subsequent episodes of GBS[3]. Results of the study by Kuitwaard also found that RGBS patients were younger, with milder disease and had Miller-Fisher variant of GBS at the initial episode. Patients with above characteristics on initial presentation of GBS are more prone for recurrences[3]. They also identified that there are similar presentations but more severe clinical deficit and residual effects with each recurrence[3,6]. Yet, there is limited literature addressing why only a certain subset of patients with GBS get recurrences of the disease. The indexed case, although young at presentation, patient had a more alarming disease initially with poor neck muscle power and limb power and did not have Miller-Fisher variant. This shows a deviation from the classically identified features favoring a recurrence of GBS. The time gap between the episodes was 17 years. During the episode of recurrence, she had rapid development of more severe disabling illness involving respiratory compromise needing mechanical ventilation. This episode had AMAN variant of GBS in electrophysiological study with similar initial presentations.

The RGBS patients with similar presentations during the subsequent episodes had different antecedent infections and this may point towards immunogenic and host factors as major determinants of the disease[3,5,6]. Yet, exact mechanism by which similar clinical manifestations occur during recurrence is not established. Our patient also had a GI tract infection preceding the recurrence of GBS.

It is important to distinguish RGBS from two clinical entities; (1) GBS with treatment related fluctuations (GBS-TRF), (2) Chronic inflammatory demyelinating polyneuropathy (CIDP). Since our patient had a long asymptomatic period, GBS-TRF is less likely but CIDP comes as a differential diagnosis. CIDP is suspected when progression of weakness lasts more than 8 weeks followed by a chronic course but it can be of steadily progressive, relapsing remitting or monophasic. The treatment differs as CIDP can be treated with either immunoglobulin or immunosuppressive therapy with a subsequent maintenance immunosuppressive drug treatment whereas GBS and GBS-TRF do not show a response to immunosuppressant therapy but has good response to immunoglobulin or plasmaparesis. GBS is a more likely diagnosis in our patient as there was a rapid onset of symptoms, subsequent complete or near complete recovery, high incidence of an antecedent illness, high CSF protein levels one week after the onset of a recurrence.

Conclusion: In conclusion, GBS rarely recurs. Only about 1–6 % of all former GBS patients will have a second episode.

Multiple recurrences are highly unlikely. Most patients with recurrent GBS respond favourably to treatment with plasmapheresis or IVIG. Former GBS patients who develop a new weakness or sensory symptoms should consult with their primary physician and neurologist before concluding that the cause is a new episode of GBS. Further research is needed to understand what makes some individuals susceptible to GBS in its various forms, and why a very small fraction of those who contract GBS do so more than once.

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