Original article:

Guillain-Barre Syndrome with Severe Rheumatic Heart Disease: A case report

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ABSTRACT

Guillain–Barre syndrome (GBS) is the commonest acute inflammatory demyelinating polyradiculoneuropathy (AIDP) that is autoimmune in nature. It is characterized by mainly motor weakness but sometimes involves sensory and autonomic nerves. Weakness usually begins in the lower extremities and progressively involves the trunk, upper limbs and face. We report a fifty-one year old female patient who presented with complaints of weakness in both upper and lower extremities of 5 days duration diagnosed to be due to GBS. She coincidentally had conservatively managed severe rheumatic aortic stenosis which proved to be a therapeutic challenge in this scenario?

Key words: Acute or sub acute inflammatory demyelinating polyradiculoneuropathy, Guillain–Barre syndrome, Rheumatic Heart Disease.

INTRODUCTION

Guillain-Barre syndrome (GBS) was first reported by Landry in 1859 and later detailed by Guillain, Barré and Strohl, in 1916. The disease has become well-known internationally under the name -Guillain Barré Syndrome.[1] Guillain-Barre Syndrome (GBS) or acute idiopathic polyradiculoneuropathy is a disorder of immune mediated etiology, involving the peripheral nervous system. It is characterized by rapidly progressive muscle weakness. The immune response depends on antigen factors (specificity of lipo-oligosaccharide) and on host factors (immune status). The presence of antibodies leads to activation of T cells and complements, leading to a cascade of inflammation and demyelination. The demyelination decreases the velocity of nerve conduction and slows the impulse transmission along the axons. Clinical features include progressive, symmetrical ascending muscle weakness of limbs, areflexia with or without sensory and autonomic abnormalities.[2]

CASE REPORT:

A fifty-one year old female , who was a known case of rheumatic heart disease – severe aortic stenosis on conservative treatment since 2011, presented with complaints of progressive symmetrical weakness ,initially of both lower limbs followed by upper limbs of 5 days duration. Patient had an episode of fever one week prior to the date of admission. Fever was associated with chills and body pain, lasted for one day and subsided with medication. Patient noticed weakness of bilateral lower limbs in the form of difficulty in getting up from sitting and squatting positions followed by difficulty in holding objects. She had no history of dysphagia, nasal regurgitation, urinary or fecal incontinence.

On admission, her EGRIS (The Erasmus GBS Respiratory Insufficiency Score) score was 3 and EGOS (The Erasmus GBS Outcome Score) was 3.She was conscious and oriented to time, place and person. She had a

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respiratory rate of 20/min and her single breath count was 16. Her pulse was 70 beats per minute, regular, low volume and blood pressure was 120/66 mmHg.

Her higher mental functions were normal. Cranial nerves examination was unremarkable .Motor examination revealed normal bulk with hypotonia in both upper limbs and lower limbs. The power of the upper limbs was Medical research council (MRC) grade 3 at both shoulder and elbow and was MRC grade 2 at both wrists. She had small muscle weakness of both hands. The lower limb power was MRC grade 4 in both lower extremities both proximally and distally. She had generalized areflexia with mute plantar response. Her sensory examination was normal.

Historically she had significantly impaired effort tolerance corresponding to NYHA class - 3 - this could not be tested clinically as the patient could not walk independently. The patient had a Grade 4 ejection systolic murmur in the aortic area, radiating to all other areas but her chest was clear on auscultation.

She had no hypokalemia or other metabolic abnormalities and her CPK (Creatine phosphokinase) was normal.

The motor nerve conduction studies conducted showed essentially normal distal latencies, markedly reduced amplitudes with mild slowing of conduction velocities of bilateral tibial and peroneal nerves with absent F waves in these nerves bilaterally. The median and ulnar motor studies showed reduced amplitudes with mild slowing of conduction velocities in these nerves with the left median and ulnar nerves showing possible conduction blocks in the arm segments and prolonged distal latency in the left median nerve. F wave latencies were prolonged in both median and ulnar nerves bilaterally. The sensory conductions were essentially normal. All these findings were suggestive of a demyelinating motor neuropathy with significant axonal involvement, especially in the lower limbs, afflicting both the upper and lower limbs.[1,2]

Given the clinical presentation and findings of the patient in the absence of other reversible metabolic diseases, and the above nerve conduction findings a diagnosis of GBS was made.Her echocardiography showed features of Rheumatic heart disease, aortic valve disease, severe calcific aortic stenosis (AVA – 0.6 CM2), moderate AR, mild mitral stenosis, dilated LA with concentric LVH and trace pericardial effusion with a LVEF of 60%.

Plasma exchange (PLEX) was initiated along with supportive treatment. IV Immunoglobulin's was not considered in this patient owing to her cardiac status and hyper viscous nature of the solution. Low volume PLEX (1litre/day) was planned with a total of 7 cycles. In view of her cardiac status the cardiologist advised monitoring her volume status by insertion of a Swan Ganz catheter. However it could not be done as the patient did not give consent. The patient was carefully monitored in an intensive care unit during the procedure of plasma exchange as she had hypotension during first few cycles.

There was much improvement in her power (power of upper limbs improved to MRC grade 4 at bilateral shoulder, elbow and wrists).

She was discharged after 7 sessions' of plasma exchange and neurorehabilitation. Two week after discharge the patient came for follow up to the neurology outpatient and was ambulatory with one person support.

DISCUSSION:

GBS is an acute inflammatory demyelinating neuropathy. Multifocal demyelination with inflammation results in conduction block; severe forms show secondary axonal degeneration. The disease is characterized by progressive motor weakness of limbs with areflexia. Bladder involvement is rare in GBS and even when is seen is mild and hardly lasts longer than 1 week. Preceding antecedent infections, mostly viral, are seen in half of the cases. Variants of GBS include acute motor axonal neuropathy (AMAN), acute motor and sensory axonal

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neuropathy (AMSAN) and Miller Fisher syndrome. AMAN is distinguished from AIDP by its involvement of exclusively motor nerves and an electrophysiological pattern indicating axonal involvement.[3]

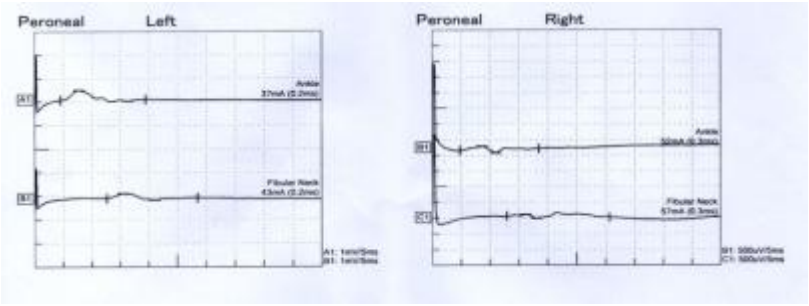
Patient was also a known case of has rheumatic heart disease with valvular involvement which proved to be a therapeutic challenge in the management of this case.

CONCLUSION:

GBS with severe rheumatic heart disease is one of the most challenging scenarios to clinicians with variable range of presentation and clinical course severity, because of the cardio vascular risks involved during the treatment of GBS. Repeated cycles of low volume plasma exchange with appropriate interval between the cycles are suggested to lower the cardio vascular risks and mortality. However, since there are no other documented similar cases scenarios available, further studies will be prudent to establish a therapeutic protocol in such patients.

Motor Nerve Conduction Study

Site	Lat. (ms)	Dur. (ms)	Amp.	Area	Stim. (mA)	Segment	Dist. (mm)	Intst. (ms)	NCV (m/s)
Median	Left								
Wrist	4.7	12.4	8.0mV	25.8mV	35.0	Wrist-Elbow	220	4.7	46.8
Elbow	9.4	14.7	3.5mV	12.5mV	99.0				
Median	Right								
Wrist	3.5	11.4	9.2mV	30.0mV	45.0	Wrist-Elbow	220	4.3	51.2
Elbow	7.8	13.1	6.0mV	21.5mV	43.0				
Ulnar	Left								
Wrist	2.8	15.2	5.4mV	22.4mV	36.0	Wrist-Elbow	240	5.0	48.0
Elbow	7.8	15.0	2.8mV	10.8mV	50.0				
Ulnar	Right								
Wrist	3.2	12.0	7.8mV	26.5mV	51.0	Wrist-Elbow	250	5.5	45.9
Elbow	8.7	11.0	3.7mV	11.2mV	52.0				
Peroneal	Left								
Ankle	4.6	14.9	8.5mV	1.7mV/m	37.0	Ankle-Fibular Neck	300	8.1	37.3
Fibular Neck	12.7	15.9	8.2mV	1.0mV/m	43.0				
Peroneal	Right								
Ankle	4.0	13.7	208.6uV	538.7uV	52.0	Ankle-Fibular Neck	300	8.2	36.6
Fibular Neck	13.0	17.8	107.6uV	801.0uV	57.0				
Tibial	Left								
Ankle	6.8	11.0	2.0mV	4.0mV/m	70.0	Ankle-Popliteal Fossa	380	10.8	35.2
Popliteal Foss	17.8	13.3	1.4mV	4.1mV/m	76.0				
Tibial	Right								
Ankle	6.6	12.9	2.2mV	5.0mV/m	74.0	Ankle-Popliteal Fossa	370	9.7	38.3
Popliteal Foss	16.3	18.8	1.8mV	5.5mV/m	99.0				



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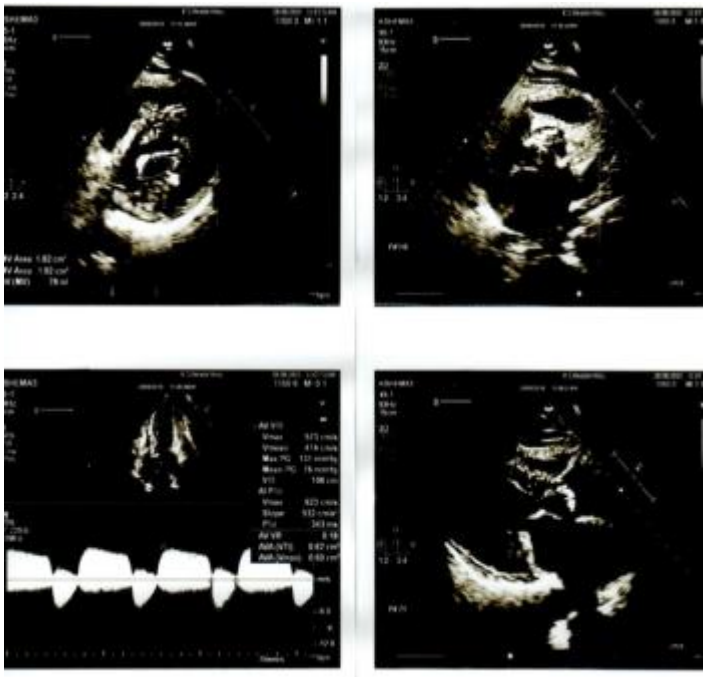

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