

Autonomic Neuropathy as Presenting Feature of Guillain-Barré Syndrome

Dr Ganesh Bavikatte
MBBS, MD
(Medicine), MRCP
(UK), MRCP (London)

Specialist Registrar,
9 Averhill,
Worsley, Manchester M28 1ZN.
Tel: 0044 7917123577
Email: ganeshbavikatte@
ukdoctor.org

Dr Ali Hassoon
MB ChB, FRCP
(London)

Consultant Neuro rehabilitation,
Salford Royal NHS Foundation
Trust, Manchester, UK
Email: Ali.Hassoon@srft.nhs.uk

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Guillain-Barré syndrome (GBS) is an acute inflammatory polyradiculoneuropathy that may lead to autonomic dysfunction. Autonomic neuropathy can manifest as cardiovascular, sudomotor, gastrointestinal and other systems involving both sympathetic and parasympathetic fibres. The degree of dysautonomia is highly variable from being mild with presentation found only by specialist tests to profound illness. Although autonomic neuropathy is an important complication of GBS, only few cases in the literature report autonomic dysfunction as the presenting feature of GBS.¹ There is very little information available regarding its impact on disability and long term prognosis.^{2,3,4}

Case report

A 20-year-old Afro Caribbean healthy male presented with a brief loss of consciousness at home on standing. He had fever, chills and cough one week prior to admission, which was treated with oral antibiotics. He started to have dizziness with recurrent collapses on standing three days prior

to admission. On examination he was a febrile, chest was clear. Neurological examination showed normal tone and power in all four limbs, areflexia, down going plantars, diminished sensation of oral cavity, face along with glove and stocking type of anaesthesia over limbs. His Blood Pressure (BP) was 125/60mmhg (lying), 66/30 (sitting). The patient's cerebrospinal Fluid examination shown albumino-cytological dissociation and nerve conduction studies confirmed abnormal sensory and motor conduction in upper and lower limbs suggestive of axonal degeneration (Table 1). His blood tests were largely normal (negative GQ1b, normal cortisol levels) except for positive mycoplasma titres (suggestive of atypical pneumonia). Imaging of brain and spinal cord were normal. Electrocardiogram (ECG) tests show depressed ST segment. The diagnosis of GBS with prominent dysautonomia probably secondary to atypical pneumonia was made.

He was treated for GBS with Intravenous Immunoglobulin and postural hypotension managed with thrombo-embolus deterrent (TED)

Table 1: Nerve conduction test

Motor Nerves						
Nerves	Latency (ms)		Amp(mv)		CV (m/s)	
Right median Wrist-APB Elbow-Wrist	4.2		10.6		51.8	
	9.8		9.6			
Right ulnar Wrist-ADM A Below-Wrist	3.8		2.6		46.5	
	10.9		2.3			
Tibial Ankle-AbH Knee-Ankle	Right 5.6 18.8	Left 6.4 21.4	Right 2.1 1.4	Left 2.0 1.5	Right 37.9	Left 33.3
Peroneal Ankle-EDB Pop Foss-Ankle	Right 6.4 19.8	Left 5.6 19.3	Right 0.0 0.0	Left 0.0 0.1	Right 38.8	Left 37.2
Sensory Nerves						
Nerves	Latency (ms)			Amplitude (uV)		
Right Radial Forearm-Hand	-			-		
Right superficial per Shin-Ankle	-			-		
Right Sural Calf-Ankle	-			-		
Right Digits to wrist Med II-Wrist Ulnar V-Wrist	-			-		

Table 2: Blood pressure

Admission	Lying	30 degrees	45 degrees	90 degrees	Standing
June 2009	125/60	–	–	60/30	Unable
August 2009 Reclining chair	115/68	112/68	108/69	55/36	Difficult to record
July 2010	103/53, Pulse- 67/min	103/53 Pulse- 67/min	87/44 Pulse-58/min	71/42 Pulse-51/min	Unable to tolerate more than 1 minute

stockings, fludrocortisone and midodrine. In spite of that he continued to suffer with severe autonomic dysfunction (postural drop in BP when sitting more than 30 degrees upright with relative bradycardia, loose bowels, excessive sweating). Due to his poor sensation in face and oral cavity he had poor oromotor control of saliva and food. He needed the support of PEG (percutaneous enteroscopic gastrostomy) feeding to maintain adequate nutrition and regular medication intake. He started to make slow progress (loose bowels settled, weight started to improve) but postural drop of blood pressure continued to persist even one year after presentation.

One year of multidisciplinary rehabilitation reduced his disability and he became able to take care of his personal care, change his feed, transfer and self propel a wheel chair. He continued to walk with a high stepping gait under supervision, a maximum distance of 100 meters. However he continued to have severe sensory deficits in lower legs and drop in his BP on standing, which significantly limited his mobility.

Discussion

Guillain Barré Syndrome was first described in 1916. It is sometimes called Landry's paralysis, after the French physician who first described a variant of it in 1859. Autonomic dysfunction is well known feature of GBS since 1892, when Osler described "paralysis of heart" as a cause of death in GBS. Acute panautonomic neuropathy is the rare variant of GBS. It is associated with a high mortality rate, due to cardiovascular involvement, and associated dysrhythmias. The most common symptoms at onset are related to orthostatic intolerance, as well as gastrointestinal and sudomotor dysfunction. Autonomic dysfunction can be seen in up to two thirds of patients with GBS, but dysautonomia as presenting feature is very rare.⁵ Clinical features vary, and cardiovascular involvement appears to be the commonest manifestation. Wide fluctuations in pulse and blood pressure have been found in fatal cases of dysautonomia. Litchenfeld⁶ found evidence of postural hypotension in 43% of patients and episodic hypotension in 57%, whereas most other studies identified postural hypotension in 19-35% of patients but rarely found as presenting feature.³

Clinical features of autonomic dysfunction are due to excessive sympathetic outflow and hypertension; sympathetic hypofunction and hypotension; parasympathetic dysfunction; arrhythmias and bladder or bowel dysfunction. Features usually manifest 2- 21 days following onset of illness with mean of seven

days following onset of motor weakness. Excessive sympathetic outflow results in hypertension with severe systolic fluctuations, if not treated adequately may complicate by subarachnoid haemorrhages, pulmonary oedema, and even sudden death. Sympathetic hypofunction resulting in postural hypotension can be seen up to 43% and episodic hypotension in 57% of patients with GBS.¹⁰ Causes of postural hypotension may be due to denervation super sensitivity of alpha adrenergic receptors and prolonged recumbence and loss of muscle tone as late feature. Sympathetic hypofunction can be found out by provocative testing (Abnormal response to sustained handgrip, cold immersion, response to atropine) or by direct evidence (anhidrosis). Parasympathetic dysfunction manifests as either excessive or decreased vagal tone. Excessive vagal tone causes bradycardia, asystole (sometimes needing pacemaker) which may worsen following tracheal suctioning or Valsalva manoeuvres. While decreased vagal tone causes sinus tachycardia, noticed in up to 37%. Other arrhythmias noted include atrial tachycardia (fibrillation, flutter), ventricular tachycardia, ST segment (elevation, depression), T wave (flat, inverted), QT prolongation, axis deviation, conduction blocks. Bladder and bowel dysfunction includes urinary retention, overflow incontinence, impaired bladder sensation, constipation, gastroparesis, diarrhoea, sexual dysfunction. Pupillary abnormalities are also commonly observed.

The impact of autonomic dysfunction on rehabilitation process and prognosis of such patients are not well understood. Many studies looked in to motor or sensory recovery following rehabilitation in GBS,¹¹ but not many reported evidences available regarding outcome following autonomic dysfunction secondary to GBS. Evidences suggest motor and sensory impairment were still detectable in more than 50% of patients with GBS even after two years.¹¹

Conclusions

Few cases in the literature have reported autonomic dysfunction as the presenting feature of GBS, such as in this case. In a previously asymptomatic patient, acute onset of autonomic dysfunction should alert the physician to the possibility of an acute polyneuropathy, such as GBS. This case demonstrates dysautonomia even without severe motor weakness can cause significant long term disability and can pose complex rehabilitation challenges. We also noted that the residual impairment is significant and likely persistent. ♦

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