Ir Med J; April 2024; Vol 117; No. 4; P950
25th April, 2024

Guillain-Barré Syndrome In Immunocompetent Patient

T.M. Chia[1,2], N. Zaffar[1], N. Onwasigwe[1], D. Nagi[1]

1. Department of Medicine, Portiuncula University Hospital, Co. Galway, Ireland.
2. School of Medicine, University of Galway, College Rd., Co. Galway, Ireland.

Abstract

Presentation
We present a case of 53-year-old man who present with 6 days history of lower limb weakness and impaired light touch perception on feet.

Diagnosis
Nerve conduction study revealed severe demyelinating polyneuropathy with conduction block in all the motor nerves. The sural sensory action potential is attenuated but still present and radial sensory nerve action potential is normal. Electrophysiological findings confirm a progressive demyelinating polyneuropathy.

Treatment
He was initially treated with potassium infusion and after confirmation of findings on electromyography and nerve conduction study, he was started on intravenous immunoglobulin treatment.

Discussion
This is a unique case in that GBS affected a previously healthy man. Remarkably, our patient regained the ability to walk after administration of intravenous immunoglobulin.

Introduction

Guillain-Barré syndrome (GBS) and its variants are a rare but serious post-infectious, immune-mediated neuropathies[1]. In this report, we present the case of a 53-year-old immunocompetent man who presented with 6 days history of acute onset bilateral lower limb weakness, hyporeflexia, and absent light touch perception.

Official Journal of the Irish Medical Organisation

Ir Med J; April 2024; Vol 117; No. 4; P950
25th April, 2024

Case Report

A 53-year-old man with background history of diverticulosis, coeliac disease, encephalomalacia, anxiety and alcohol abuse, present with experiencing weakness in his legs for 6 days and this has steadily deteriorated to the point where he is not ambulant and need assistance to mobilize. He reported feeling weak using his hand and noted some paresthesia in his fingertips on both hands. On examination, there is moderate weakness of finger abduction and extension. Both supinator and biceps reflexes are hypoactive. He was unable to dorsiflex his ankle bilaterally and the lower limb reflexes are absent. The examination also showed impaired light touch perception to the mid feet and there is impaired vibration perception to the malleoli bilaterally.

Initial laboratory testing showed hypokalemia of potassium level 3.3 mmol/L (reference range 3.4 - 5.1 mmol/L) which we corrected with normal saline infusion and oral supplementation. Despite the correction of electrolyte derangements, his paresis persisted, necessitating further workup. CT brain was unremarkable and MRI brain was consistent with encephalomalacia. MRI cervical show degenerative spondylosis throughout the entire spine however there is no neural impingement. At this point differential diagnosis of GBS, vitamin B12 and folate deficiency was considered keeping in with his history of alcohol abuse. Electromyography (EMG) and nerve conduction studies (NCS) were conducted (Figure 1). It showed a very severe demyelinating polyneuropathy with conduction block in all the motor nerves. The electrophysiological findings confirm a progressive demyelinating polyneuropathy. Patient's lumbar puncture also show classic pattern of albumin cytologic dissociation which the spinal fluid shows a normal amount of white blood cells and an elevated CSF protein level. He was started on intravenous immune globulin (IVIG) and transferred under care of neurology department.

Sensory Nerve Conduction Studies

Nerve	Peak Lat	Amp	CV	Area	Dur
	ms	uV	m/s	ms*uV	ms
Radialis Sensory Right					
Forearm-Thumb	3.10	17.2	--	12.0	1.50
Suralis Sensory Right					
Calf-Ankle	4.68	5.3	31.9	7.3	2.6

-- indicates response absent

Motor Nerve Conduction Studies

Nerve	Lat	Amp	CV	Amp	Area	F-M Lat
	ms	mV	m/s	%	ms*mV	ms
Medianus Motor Right						
Wrist- APB |APB	4.40	3.1				
Pos. 2-Wrist | APB	13.1	1.10	29.3	-64.5	8.1	
Peroneus Motor Right						
Ankle – EDB | EDB	46.9	--			--	
Tibialis Motor Right						
Ankle – Abd hal | Abd Hal	9.17	1.43			3.4	
Stim 2-Ankle | Abd hal	24.6	0.60	24.6	-58.0	3.0	
Ulnaris Motor Right						
Wrist – ADM | ADM	3.34	3.9			13.3	
Bl. elbow-Wrist | ADM	13.6	1.28	22.4	-67.2	7.4	
Ab. Elbow-Bl.elbow | ADM	16.5	1.18	79.3	-7.8	5.3	

-- indicates response absent

Figure 1: EMG / NCS report

Discussion

A total number of 488 cases with GBS were reported in the last 20-year period (1992 to 2012) in Ireland. The incident rate varied from 0.3 per 100,000 in early 1990s to a maximum of 1.3 in 2010. Among all, acute inflammatory demyelinating polyradiculoneuropathy (AIDP) was the most common subtype, follow by Miller-Fisher syndrome (MFS), acute motor-sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN)[2].

Most GBS cases are reported to have a preceding history of infectious condition, either gastrointestinal or respiratory. Campylobacter jejuni and Mycoplasma pneumoniae are common microorganisms known to trigger GBS while Epstein-Barr virus and Sars-Cov-2 virus also reported in GBS patients. The pathophysiology consists of cross-react of antibodies and inflammatory cells with peripheral nerve and roots, which result in demyelination or axonal damage[3,4]. Intravenous immunoglobulin (IVIG) or plasmapheresis are two treatment options currently considered the standard care in GBS patients. IVIG and plasma exchange have been shown to be equally effective and IVIG are usually preferred as it is cost effective and readily accessible[5,6].

One of the most common complication of GBS is respiratory failure. The Erasmus Guillain–Barré Syndrome Respiratory Insufficiency Score (EGRIS) is often used to estimates the risk of respiratory insufficiency and need for mechanical ventilation within first week from hospital admission. Score of >4 will warrants the needs for an ICU admission[7]. Most patients show good recovery especially in the first year after disease onset. The modified Erasmus GBS outcome score (mEGOS) prognostic tool was developed to calculate the probability of regaining walking ability in GBS patients[8].

In conclusion, we present the case of a healthy, immunocompetent adult man with GBS. This case highlights the importance of recognizing GBS in patient present with acute flaccid paralysis at early stage. A higher index of suspicion is required for GBS as early recognition and treatment improve the clinical outcome.

Declaration of Conflict of Interest:
None declared.

Corresponding author:
Tiong Ming Chia,
Department of Internal Medicine,
Portiuncula University Hospital,
Co Galway,
Ireland.
E-Mail: tiongming.chia@hse.ie

References:

1. Nguyen TP, Taylor RS. Guillain Barre Syndrome. Nih.gov. StatPearls Publishing; 2023.
2. Lefter S, Hardiman O, Ryan AM. A population-based epidemiologic study of adult neuromuscular disease in the Republic of Ireland. Neurology. 2017;88(3):304–13.
3. Piña-Rosales E, Gonzalez-Salido J, Barron-Cervantes N M, et al. (February 17, 2024) Guillain-Barré Syndrome in an Immunocompromised Patient: A Case Report. Cureus 16(2): e54380.
4. Zaka A, Dehkordi O, Weir R, et al. (November 30, 2023) A Case of Guillain-Barré Syndrome With Multiple Causative Factors in a Young Male. Cureus 15(11): e49745.
5. Van der Meché FG, Schmitz PI. A randomized trial comparing intravenous immune globulin and plasma exchange in Guillain-Barré syndrome. Dutch Guillain-Barré Study Group. N Engl J Med. 1992 Apr 23;326(17):1123-9.
6. Shang P, Feng J, Wu W, Zhang HL: Intensive care and treatment of severe Guillain-Barré syndrome. Front Pharmacol. 2021, 12:608130. 10.3389/fphar.2021.608130

7. Walgaard C, Lingsma HF, Ruts L, et al. Prediction of respiratory insufficiency in Guillain-Barre syndrome. Ann Neurol 2010;67:781–787.
8. Leonhard SE, Mandarakas MR, Gondim FA, et al.: Diagnosis and management of Guillain-Barré syndrome in ten steps. Nat Rev Neurol. 2019, 15:671-83. 10.1038/s41582-019-0250-9