Case Report

iMedPub Journals
www.imedpub.com

Journal of Neurology and Neuroscience
ISSN 2171-6625

2022
Vol.13 No.2:412

# Descending GBS - An Atypical Presentation

**Rosalia Shivute***

Department of Internal Medicine, University of the Witwatersrand, Johannesburg, South Africa

***Corresponding author:** Rosalia Shivute

rosaliashivute@gmail.com

Department of Internal Medicine, University of the Witwatersrand, Johannesburg, South Africa.

**Tel:** +0768265426

**Citation:** Shivute R (2022) Descending GBS - An Atypical Presentation. J Neurol Neurosci Vol.13 No.2:412.

## Abstract

Guillain-Barre syndrome and its variants form a continuous spectrum of overlapping syndromes. A 15-year-old caucasian male patient previously well presented with a week history of body weak-ness, vomiting and neck pain, (pre Covid pandemic). Further history noted that he suffered multiple infantile apnoeic attacks with mild cognitive impairment. He disclosed that he had an upper respiratory infection 3 weeks prior, treated with fluroquinolones by a general practitioner. On examination he was noted to be febrile. Positive finding included terminal neck stiffness and a sinus tachycardia. All other systems and vitals were normal. Initial CTB was normal with an unremarkable lumbar puncture. Day 5 of admission the patient developed bulbar symptoms; a repeat LP was done where albuminocytogical dissociation was noted. Higher mental function normal, bilateral facial nerve palsy, facial diplegia with decrease palatal movement, an absent gag reflex. It was noted that the patient had significant proximal weakness of upper and lower limbs. Deep tendon reflexes were all decreased with no sensory fallout and no cerebellar signs. Diagnosis of a rare regional variant of Guillan Barre Syndrome (GBS) was clinically made. Immunoglobulin was commenced. The patient responded rapidly to treatment.

**Keywords:** GBS; Variant GBS; Atypical GBS; Bifacial weakness; Facial diplegia; Bulbar GBS; Oropharyngeal; Limb GBS

**Received:** 09 February 2022; Manuscript No. ipjnn-22-12491; **Editor assigned:** 11 February 2022; PreQC No. P-12491; **Reviewed:** 28 February 2022; QC No. Q-12491; **Revised:** 02 March 2022, Manuscript No. R-12491; **Published:** 09 March 2022

## Introduction

A 15-year-old previously well caucasian male patient at Klerksdorp/Tshepong complex, North West province South Africa- presented with a week history of generalized body weakness, vomiting and neck pain, (pre Covid pandemic). Further history noted that he suffered multiple infantile apnoeic attacks with cognitive impairment rendering him unable to attend traditional educational facilities. He is currently employed at local butchery. The patient disclosed that he had an upper respiratory infection 3 weeks prior; treated with fluroquinolones a general practitioner. There was no significant family or social history of note [1].

On examination he was noted to be febrile. General examination was deemed normal. Neurological examination higher functioning and cranial nerves were normal, gait was normal his power, sensation and reflexes were normal. Positive finding were that he had terminal neck stiffness and a sinus tachycardia. All other systems and vitals were normal. CTB and Chest X-Ray were normal [2].

He had a Lumbar Puncture (LP) and a differential diagnosis of possible partially treated bacterial meningitis or of viral meningitis was entertained. Noted on his bloods was a normal white cell count, C reactive protein 0 and a negative Human Immunodeficiency Virus (HIV) (**Tables 1 and 2**).

He was given ceftriaxone, antipyretics with analgesics (Paracetamol/Tramadol), metoclopramide and IV fluids (Ringers Lactate 2 L/daily). Day 2 of admission patient complained of peri-oral numbness, dizziness, neck pain, tingling of feet and legs. Vitamin B12, syphilis screen and anti-nuclear antibodies were requested. Day 5 of admission the patient developed nasal speech, dysphagia, dysarthria, reflexes depressed 1+ globally, power 3/5 proximal, normal tone, bladder/bowel intact/ sensation intact, chest clear, abdomen distended- a repeat LP was done where albuminocytogical dissociation was noted [3-5]. Higher mental function normal, bilateral facial nerve palsy, facial diplegia with decrease palatal movement, an absent gag reflex. It was noted that the patient had significant proximal weakness of upper and lower limbs. Deep tendon reflexes were all decreased with no sensory fallout and no cerebellar signs. The outreach hospital complex did not have access to nerve conduction studies nor antibody testing capabilities [6,7].

 

**Table 1** CSF in day 1 and day 5.

| CSF | Day 1 | Day 5 |
|---|---|---|
| Appearance | Clear | clear |
| Polymorphs | 0 | 0 |
| Lymphocytes | 2 | 6 |
| Red blood cell | 156 | 10 |
| Protein | 0.5 | 1.17 |
| Glucose | 3.6 | 3.3 |
| India Ink | Negative | Negative |
| Cryptococcus Antigen | Negative | Negative |
| VDRL ADA | NEG<br>0.0 | 0.3 |

**Table 2** White cell count, Haemoglobin, Mean corpuscular volume, Platelets, C- reactive protein, Vitamin B12, HIV, TPHA/RPR and ANA in day 1, day 3 and day 7.

| Components | Day 1 | Day 3 | Day 7 |
|---|---|---|---|
| White cell count × 10^9/ | 9.49 | 11.49 | 8.26 |
| Haemoglobin (g/dL) | 16.7 | 15.3 | 16.3 |
| Mean Corpuscular Volume | 85 | 88 | 87 |
| Platelets | 416 | 410 | 374 |
| C- reactive protein | 0 | - | 7 |
| Vitamin B12 | - | 485 | - |
| HIV | - | NEG | - |
| TPHA/RPR | - | NEG | - |
| ANA | - | NEG | - |

**Table 3** AIDP (Acute inflammatory demyelinating polyneuropathy).

| AIDP (Acute Inflammatory Demyelinating Polyneuropathy) | |
|---|---|
| Atypical form of AIDP | Asymmetric |
| | Pure Motor form |
| | Prominent Sensory loss |
| | Preserved reflexes |
| Regional presentation of AIDP | Pharyngo-cervical-brachial |
| | Paraparetic |
| | Facial diplegia with paresthesia |
| Pure sensory | - |
| Pure Autonomic form | - |
| Miller Fischer syndrome | - |
| Axonal forms | AMAN (Acute motor axonal neuropathy) |
| | AMSAN (Acute motor and sensory axonal neuropathy) |

Diagnosis of a rare regional variant of Guillan Barre Syndrome (GBS) was clinically made presenting as facial diplegia and palatal palsy involving symmetrical cranial nerve involvement of VII, IX, X with a symmetrical motor polyneuropathy. Immunoglobulin was commenced at 400 mg/kg over 5 days [8]. The patient responded rapidly to treatment with improvement of his facial palsy and proximal weakness. Day 18 The patient was discharged walking unaided, with slight left sided facial paresis to be followed up at the medical outpatient and physiotherapy departments. Day 62 The patient's symptoms had fully resolved and the patient returned to work [9].

## Discussion and Conclusion

Guillan Barre Syndrome: first described by Charles Guillain,Jean Alexandre Barré,André Strohl in France in 1916 as a form of areflexic paralysis which exhibited on spinal fluid a normal cell count, with an abnormal increase in protein seen in soldiers during the World War 1.GBS is syndrome of heterogeneous conditions with several variant forms. They are acute immune-mediated polyneuropathies presenting as an acute monophasic paralysing illness usually provoked by a preceding infection.

Diagnosis is mostly clinical aided by nerve conductive velocity studies: illustrating motor conduction block, slowing of motor and sensory nerve conduction, temporal dispersion, and prolonged distal latencies. Cerebral spinal fluid can also exhibit -albuminocytologic dissociation, which is present in 50 to 66 percent of patients in the first week after the onset of symptoms and ≥ 75 percent of patients in the third week.

Antibodies against Gangliosides GD1a/GD1b/GT1b are associated with severe GBS. IgG1 and anti-Campylobacter antibodies are associated with poor prognosis. Immunoglobulins IgG1 and IgG3 against Haemophilus influenza lipo-oligosaccharide antibodies are associated with better outcome (**Table 3**).

60-70% GBS occurs 1-3 weeks after respiratory or gastrointestinal infections [9]. 30% cases in North Africa, Europe and Australia preceded by *Campylobacter Jejuni*. Other infections include: CMV, EBV, Mycoplasma, Haemophilus Influenza. Other causes: Vaccines, Influenza, hepatitis, rabies, tetanus, lymphoma, HIV seroconversion, Systemic Lupus Erythematosus surgery, thrombolysis. Recently Sars-COV2, associated COVID vaccines (esp.; adenovirus vector vaccine) and Zika virus.

**Treatment is based on three cornerstones:**

Monitoring, supportive and critical care: most important being vital monitoring, thromboembolic prophylaxis, nutrition,

Immunotherapy and Immunotherapy immunoglobulin is given 400 mg/kg/5 days. Plasmapheresis 40-50 ml/kg: 4-7 exchanges. IVIG as effective PE at least in first 2/52 of motor symptoms. Plasmapheresis reduced mechanical ventilation by 25% and increases full recovery in 1 year by 55-65% but did not change overall mortality. Plasmapheresis however should not be given four weeks after onset of symptoms as its efficacy is reduced Rehabilitation.

## Conclusion

Among severely affected patients 25% need ICU. Indications include poor neck flexion of neck, rapidly progressing symptoms vital capacity of 15 ml/kg or less or/and hypercarbia/hypoxia. Poor prognostic features include: bulbar dysfunction, autonomic dysfunction, severe weakness, rapid onset <3 days, age >60 years, reduced median compound muscle action potential amplitude, pulmonary infections, hypokalemia. Majority of patients will completely recover or left with only minor deficits (distal numbness/foot drop). Ten percent will suffer disabling weakness, imbalance or sensory loss. Three to eight percent will die despite Intensive Care Unit interventions. Main causes of death include: sepsis particularly from- pneumonias, beds sores, acute respiratory distress syndrome and pulmonary embolism.

## References

1 Yuki N, Hartung HP (2012) Guillain-Barré syndrome. N Engl J Med 366:2294-2304.

2 Sejvar JJ, Baughman AL, Wise M, Morgan OW (2011) Population incidence of Guillain-Barré syndrome: A systematic review and meta-analysis. Neuroepidemiol 36: 123-133.

3 Ropper AH (1992) The Guillain-Barré syndrome. N Engl J Med 326: 1130-1136.

4 Vriesendorp FJ, Shefne JM, Targoff IN, Dashe JF (2016) Clinical features and diagnosis of Guillain-Barré syndrome in adults.

5 Willisons HJ, Jacobs BC, Van Doorn PA (2011) Guillain-Barré syndrome. J Peripher Nerv System 2005: 1094-1112.

6 Kuijf ML, Godschalk PC, Gilbert M, Endtz HP, Tio-Gillen AP, et al. (2007) Origin of ganglioside complex antibodies in Guillain-Barré syndrome. J Neuroimmunol 188: 69-73.

7 Longmore M, Wilkinson I, Parkes WE, Foulkes A, Mafi A (2014) Oxford handbook of clinical medicine.

8 Levin KH (2004) Variants and mimics of Guillain Barré Syndrome. Neurol 10: 61-74.

9 Hadden RD, Karch H, Hartung HP, Zielasek J, Weissbrich B, et al. (2001) Preceding infections, immune factors, and outcome in Guillain-Barré syndrome. Neurol 56: 758-765.