Case Number 1
Guillain-Barré Syndrome (GBS)

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Case summary:

Demographic details:
Mr. JS, male, Gudja
Referred from: GP

A 57-year-old Caucasian gentleman presented with bilateral progressive distal upper limb paraesthesiae, which he described as a feeling of "heaviness" followed by distal lower limb and mild tongue parasthaesia. He complained of dysaesthetic symptoms in his upper limbs with intermittent burning and tingling and autonomic disturbances such as excessive sweating of the face, hands and legs. He later developed epigastric pain that radiated to the chest, which was not related to exercise. According to the patient, symptoms got worse after taking the influenza vaccine. On examination, he had gait disturbance with weaker left lower limb muscles. During his stay in hospital, he also developed slight dysarthria and diplopia, together with urinary retention and constipation. He had had a similar, though much less severe, episode six years previously where he was diagnosed with Guillain-Barré Syndrome based on his clinical features, EMG result and his high protein levels in the CSF (more than 8g/L). He was treated with IVIG and recovered completely. This was his second presentation of neuromuscular weakness and he was referred for immunoglobulin treatment and intensive physiotherapy.

Presenting complaint:

JS presented with progressive bilateral parasthaesia of the upper extremities. It was followed by bilateral lower limb and later, tongue parasthaesia. He had been referred to hospital due to epigastric pain that progressed to chest pain. His chest pain was associated with slight sweating and it did not radiate anywhere. It was exacerbated by inspiration.

History of presenting complaint:

He suffered from a similar episode six years previously where he was diagnosed with GBS. He recovered completely after being treated with intravenous immunoglobulins (IVIG) and had no symptoms until this second presentation.

Past medical and surgical history:

Past medical history:

- Hypertension
- Diabetes
- Hypercholesterolemia
- Gastro-oesophageal reflux disease

Drug history:

Drug	Dosage	Frequency	Type	Reason
Metformin	500mg	TDS	Anti-diabetic	Treatment for Diabetes Mellitus type 2
Perindopril	4mg	Nocte	ACE Inhibitor	Treatment of hypertension
Simvastatin	20mg	Nocte	Lipid lowering agent	Treatment of hypercholesterolaemia
Aspirin	75mg	Once Daily	Analgesic	Ischaemic Heart Disease

Family history:

Father died of a myocardial infarction.
Mother suffers from arthritis.

Social history:

A married public transport driver. He is a non-smoker and drinks socially.

Systemic inquiry:

- General Health: looks well in general
- Cardiovascular System: no abnormalities
- Respiratory System: clear chest
- Gastrointestinal System: constipation, bloated
- Genitourinary System: urinary retention
- Central Nervous System: unstable gait, cannot walk on toes and heels, needs help to stand, impaired sensation in a glove and stocking distribution, reduced power of muscles L>R
- Musculoskeletal System: chest pain especially on inspiration
- Endocrine System: no abnormalities

Current Therapy:

Immunoglobulins via IV route were given for 5 days in order to suppress the acute inflammatory demyelination of the peripheral nervous system.

Discussion of results of general and specific examination:

Neurological examination:

General: The patient did not have any nystagmus or facial asymmetry, but had a slight dysarthria.

Tone: There was reduced tone in both his upper and lower limbs.

Power: Power assessment showed marked weakness in both upper and lower limbs, being more pronounced in the lower limbs, with the left more severe than the right.

Upper limb examination of power:

Muscle	Right Limb	Left Limb
Deltoid	4	3
Triceps	4	3

Lower limb examination of power:

Muscle	Right limb	Left limb
Glutei	3	2
Quadriceps	3	2
Hamstrings	3	2
Iliopsoas	3	2

Reflexes: Reflexes of both upper and lower limbs were reduced.

Coordination: Upper and lower limb coordination examination was normal.

Sensation: He had reduced sensation in both his upper and lower limbs.

Gait: He had mild difficulty raising himself from a sitting position. His gait was unstable and was unable to walk on his toes or heels.

Differential diagnosis:

- GBS
- Mononeuritis multiplex

Diagnostic procedures:

Laboratory exams:

Test: Lumbar Puncture
Justification for test: In order to obtain CSF composition.
Result: Raised protein (889 mg/L) with no cells diagnostic of GBS.
Conclusion: GBS

Instrumental exams:

Test: Nerve Conduction Studies and Electromyography
Justification for test: To show any evidence and distribution of a neuropathy
Result: Findings are consistent with a severe sensori-motor predominantly demyelinating polyneuropathy that is compatible with an acute inflammatory neuropathy
Conclusion: GBS

Therapy:

Drugs:

Drug Name (Generic)	Dosage	Frequency	Type	Reason
IVIG	0.4g/kg	Once daily	Immunoglobulin	Suppression of demyelination

Diagnosis:

This gentleman's symptoms were suggestive of GBS. The typical clinical presentation consists of lower limb symmetrical parasthaesia that ascends to the upper limbs, progressing to weakness. However, this patient's presentation was atypical since he first complained of a "heavy" feeling of his upper extremities which later progressed to parasthaesia. The same later occurred in the lower limbs, thus his presentation had a 'descending' pattern rather than 'ascending'. Together with that, he had tongue parasthaesia which progressed to dysarthria. In view of these signs and symptoms, a lumbar puncture was done to assess the CSF. Elevated protein levels, in the absence of high white blood cell count, was indicative of GBS. EMG is a specific and sensitive investigation for GBS since the results were consistent with demyelination.

Final treatment and follow ups:

The patient was started on IVIG which was continued for five days. However he had minimal immediate benefit and required intensive rehabilitation. He complained of constipation, urinary retention and diplopia. He was transferred to rehab for further physical therapy.

Fact Box 1

Title: Guillain–Barré syndrome

General overview: GBS is an acute inflammatory immune-mediated disorder affecting the peripheral nervous system[1]. GBS typically manifests as sudden distal symmetrical parasthaesia that ascends to the upper limbs and progresses to weakness. A few patients undergo sensory dysfunction especially in the demyelinating forms of GBS[2]. About a third of hospitalised patients are mechanically ventilated due to diaphragmatic, respiratory and oropharyngeal muscle weakness[3].

Long term signs and symptoms: GBS patients may have persistent weakness, areflexia, ataxia and sensory loss. About 7-15% of patients suffer from permanent neurologic sequelae such as bilateral foot-drop, intrinsic hand muscle wasting, ataxia and dysaesthesia. Moreover, they may experience long-term functional impairment and differences in pain intensity[4].

Epidemiology: GBS is made up of different subtypes with variable incidence rates in different countries. In Europe, acute inflammatory demyelinating polyradiculoneuropathy (AIDP) contributes to 90% of the cases. This subtype includes predominantly motor, bilateral facial and pharyngeal disturbances, with occasional abnormal sensory and autonomic manifestations[5].

Risk factors: Usually, a bacterial or viral infection such as Campylobacter jejuni, Mycoplasma pneumonia, CMV, EBV or influenza virus precede the onset of GBS [6, 7]. Surgery has also been shown as a risk factor[8]. Several vaccines have been associated with GBS (mainly past rabies vaccination[9], swine flu (H1N1) influenza vaccine used in 1976-77 and oral polio vaccination[10]), but controversy remains for the influenza vaccines[11]. A recent study, however, found an increased risk of GBS with the seasonal influenza vaccination[12]. Moreover, the risk increases with age[13].

Prognosis: Up to 85% achieve a full functional recovery within 6-18 months [14]. Acute relapse occurs in about 10% after initial improvement after treatment [15]. Some undergo clinical fluctuations during their treatment course. Recurrence of Guillain-Barré syndrome is rare but has been reported in 2-5% of patients [16, 17]. Mortality is rare, 2-12% and this occurs due to GBS complications: acute respiratory distress syndrome, sepsis, pneumonia, venous thromboembolism, cardiac arrhythmias and arrest. The most common cases are due to severe autonomic instability or from the complications of prolonged intubation and paralysis [18-21].

References:

1. Levin KH. Review Variants and mimics of Guillain Barré Syndrome. Neurologist. 2004;10(2):61-74.
2. Gupta SK, Taly AB, Suresh TG et al. Acute idiopathic axonal neuropathy (AIAN): a clinical and electrophysiological observation. Acta Neurol Scand. 1994;89(3):220-4.
3. Lawn ND, Fletcher DD, Henderson RD et al. Anticipating mechanical ventilation in Guillain-Barré syndrome. Arch Neurol. 2001;58(6):893-8.
4. Rudolph T, Larsen JP, Farbu E. The long-term functional status in patients with Guillain-Barré syndrome. Eur J Neurol.2008;15(12):1332-7.
5. Meena AK, Khadilkar SV, Murthy JMK. Treatment guidelines for Guillain–Barré Syndrome. Ann Indian Acad Neurol. 2011;14(Supp1):S73–S81.
6. Jacobs BC, Rothbarth PH, van der Meché FG et al. The spectrum of antecedent infections in Guillain-Barré syndrome: a case-control study. Neurology.1998;51(4):1110-5.
7. Ravi V, Taly AB, Shankar SK et al. Association of Japanese encephalitis virus infection with Guillain-Barré syndrome in endemic areas of south India. Acta Neurol Scand. 1994; 90(1):67-72.
8. Gensicke H, Datta AN, Dill P et al. Increased incidence of Guillain-Barré syndrome after surgery. Eur J Neurol. 2012;19(9):1239-44.
9. Hemachudha T, Griffin DE, Chen WW et al. Immunologic studies of rabies vaccination-induced Guillain-Barré syndrome. Neurology. 1988;38(3):375-8.

10. Haber P, Sejvar J, Mikaeloff Y et al. Vaccines and Guillain-Barré syndrome. Drug Saf. 2009;32(4):309-23.
11. Stowe J, Andrews N, Wise L et al. Investigation of the temporal association of Guillain-Barre syndrome with influenza vaccine and influenzalike illness using the United Kingdom General Practice Research Database. Am J Epidemiol. 2009;169(3):382-8.
12. Dieleman J, Romio S, Johansen K et al. Guillain-Barré syndrome and adjuvanted pandemic influenza A (H1N1) 2009 vaccine: multinational case-control study in Europe. BMJ. 2011;343:d3908
13. McGrogan A, Madle GC, Seaman HE et al. Review The epidemiology of Guillain-Barré syndrome worldwide. A systematic literature review. Neuroepidemiology. 2009; 32(2):150-63.
14. Bersano A, Carpo M, Allaria S et al. Long term disability and social status change after Guillain-Barré syndrome. J Neurol. 2006;253(2):214-8.
15. http://emedicine.medscape.com/article/315632-overview#aw2aab6b2b5 – last viewed 6th Dec 2012.
16. Das A, Kalita J, Misra UK. Recurrent Guillain Barre' syndrome. Electromyogr Clin Neurophysiol. 2004;44(2):95-102.
17. Roper TA, Alani SM. Recurrent Guillain-Barré syndrome: lightning does strike twice. Br J Hosp Med. 1995;53(8):403-7.
18. Zochodne DW. Autonomic involvement in Guillain-Barré syndrome: a review. Muscle Nerve. 1994;17(10):1145-55.
19. Maher J, Rutledge F, Remtulla H et al. Neuromuscular disorders associated with failure to wean from the ventilator. Intensive Care Med. 1995;21(9):737-43
20. Hund EF, Borel CO, Cornblath DR et al. Intensive management and treatment of severe Guillain-Barré syndrome. Crit Care Med. 1993; 21(3):433-46.
21. Teitelbaum JS, Borel CO. Respiratory dysfunction in Guillain-Barré syndrome. Clin Chest Med. 1994; 15(4):705-14.