Guillain-Barré Syndrome

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Mr G.B. presented to casualty with bilateral weakness in distal lower limbs which was progressive. He had a history of gastrointestinal illness with severe diarrhoea. On examination he had decreased deep tendon reflexes (ankle jerk) and an unsteady gait. A lumbar puncture was done and the findings were unremarkable. An EMG was also done and confirmed Guillain-Barré Syndrome. He was treated with intravenous immunoglobulin and physiotherapy and he is currently still in hospital recovering well.

Fact file on Guillain-Barré Syndrome:
Guillain-Barré Syndrome (GBS) is a neurological disorder characterised by weakness, most prominently in the distal extremities at first and which progresses proximally. Absent or decreased myotactic reflexes are also an important feature of this syndrome (Walling & Dickson, 2013). It was first described by Guillain, Barré and Strohl, back in 1916 (Guillain et al, 1916). GBS is a rare condition with incidence estimated at 1.1/100,000/ year – 1.8/100,000/year. The incidence also increases with age; over 50 years incidence is between 1.7/100,000/year – 3.3/100,000/year (McGrogan et al, 2009). Sejvar et al also noticed that males have a higher tendency to develop GBS; the ratio is 3:2 (Sejvar et al, 2013). Some complications of this disorder include breathing difficulties due to weakness or paralysis of the respiratory muscles, blood clots and pressure sores due to immobility, cardiac arrhythmias, dysphagia and also relapse (Alshekhlee et al, 2008).

Some criteria that must be present for GBS diagnosis are: bilateral symptoms, reduced myotactic reflexes and weakness with decreased or absent reflexes. Moreover, there can be autonomic and cranial nerve involvement and some sensory disturbances. Cerebrospinal fluids obtained through lumbar puncture usually reveal a high protein content in the fluid but normal amount of white blood cells (Walling & Dickson, 2013).

GBS have 5 subtypes. The most common is acute inflammatory demyelinating polyradiculoneuropathy (AIDP) with demyelination of peripheral nerves causing a symmetrical weakness, hyporeflexia or areflexia. Acute motor axonal neuropathy (AMAN) is another subtype characterised by antibodies targeting different gangliosides such as GM1 and GD1a. This subtype only presents with motor symptoms unlike another subtype called acute motor-sensory axonal neuropathy, having both motor and sensory involvement. Miller Fisher syndrome and acute autonomic neuropathy are the rare subtypes of GBS (Walling & Dickson, 2013).

A study done by Hadden et al, discovered that some infections, particularly caused be Campylobacter jejuni, Cytomegalovirus and Epstein-Barr virus, could be the cause of some developments of GBS (Hadden et al, 2001). In fact the majority of patient present with a history of gastrointestinal symptoms, fever, a cough, sore throat etc. This link is explained by the fact that GBS is an inflammatory neuropathy.

C.jejuni was found to express antigens similar to gangliosides. So due to molecular mimicry and a cross-reactivity, antibodies start targeting the infectious agent as well as the gangliosides. In AIDP, the antibodies target the myelin whereas in AMAN the nodes of Ranvier are damaged (van Doorn et al, 2008). Some have also suggested that following certain vaccinations such as influenza, tetanus and hepatitis, one can also have a higher risk of developing GBS but this is very rare and the benefits outweigh the risks (De Wals, 2012). Recently an upsurge of GBS was linked to the Zika virus epidemic (Wahid et al, 2016).

Treatment of patients with GBS includes plasma exchange to eliminate circulating immune complexes or intravenous immune globulin therapy. Most patients recover however about 3% still die regardless of therapy (Walling & Dickson, 2013).

Case report on Guillain-Barré Syndrome

Presenting complaint:
A 32 year old man presented to casualty with decreased power in both his distal lower limbs after abruptly falling while walking on the pavement. Mr G.B has also decreased sensation on both lower limbs and the weakness is progressing proximally.

History of presenting complaint:
4 weeks previous to the incident he experienced diarrhoea, fever, chills and sweating. The diarrhoea was sometimes with mucus but without blood and brown in colour. He says he was having diarrhoea about 20 times a day. He also complained of dull hypogastric pain before an onset of diarrhoea. The pain did not radiate to anywhere else and stopped when Mr G.B did not have any more episodes of diarrhoea. He did not experience any vomiting. He noticed about 7kg weight loss in the past month with a loss for appetite. 1 week before the incident he noticed his hand cramping and also diminished movement in his legs. He also mentioned that he hobbled a bit before the collapse and complained of different sensation on both his knees. Mr G.B did not have any shortness of breath nor swallowing difficulties. He did not have vision problems, headaches or any other pain.

Previous medical/surgical history:
Mr G.B. had a fracture in his hand and was treated with a metal plate. He does not suffer from diabetes, hypertension, liver or kidney problems, high cholesterol

Drug history:
Mr G.B. does not take any drugs and he does not have any drug or food allergies.

Family history:
His father suffers from Type 1 Diabetes and raised blood pressure which is controlled.

Social history:
Mr G.B. consumes 2 pints of beer a day. He does not smoke nor consumes recreational drugs. He lives with his girlfriend in an apartment. He works as an operations manager in finance.

Systemic enquiry:
Nil to note

Physical examinations and Preliminary investigations:

Examination/Investigation	Result
Blood pressure	109/71 mmHg
Heart rate	84 beats per minute
Respiratory rate	18 breaths per minute
SpO2	100% on RA
Temperature	36.8 °C
Chest	Clear R=L
Cardiovascular	+S1 +S2 +0
Abdomen	SNT
Neurological exam	UL power 5/5 bilaterally LL power GC 4.5 bilaterally, UJ 2/5, AJ 2/5 otherwise power 5/5 bilaterally. Sensation in UL and LL bilaterally intact Plantars downgoing CN grossly intact Decreased ankle DTR jerk and normal knee DTR jerk bilaterally. Gait very unsteady
Chest X-Ray	Lungs are clear. No pneumothorax or pleural effusions. The heart is not enlarged
ECG	NAD
CT brain	The brain, ventricles and sulci are normal. The skull is normal. There is a mucosal polyp within right maxillary sinus
Pulmonary function tests	Unremarkable
MRSA Screen	No MRSA isolated
Blood analysis	Haemoglobin: 15.2 g/dL Na+: 140 mmol/l K+: 4.9 mmol/l Cl-: 99.7 mmol/l Glucose: 5.21 mmol/L

Table 1: examinations and investigations carried out when Mr G.B. was admitted to casualty.

Differential diagnosis:

- Guillain-Barré Syndrome
- Chronic inflammatory demyelinating polyneuropathy

Diagnostic investigations:

- Requested investigation: Lumbar Puncture
- Justification for procedure: To obtain CSF composition
- Result and conclusion: No cells or bacteria seen. The findings are unremarkable
- Requested investigation: Nerve conduction studies and Electromyography
- Justification for procedure: To confirm neuropathy and identify the extent, severity and whether the syndrome is demyelinating or axonal.
- Result and conclusion: This study shows a predominantly demyelinating patchy motor polyneuropathy. The findings in keeping with GBS in the appropriate clinical setting

Diagnosis:

The symptoms were suggestive of GBS. Mr G.B. had distal weakness in his lower limbs with progression to proximal muscle. He also had decreased ankle jerk reflexes. This agreed with the GBS diagnosis. On investigation the cerebrospinal fluid did not have any elevated protein which was not in keeping with GBS, although his white blood cell count was normal. Diagnosis of GBS was finalised with EMG showing a demyelinating patchy motor polyneuropathy.

Management:

- Immunoglobulin therapy: Intravenous immunoglobulin 0.4g/kg body weight per day were given for 5 days
- Physiotherapy: the patient started getting out of bed with assistance and started regaining power in his lower limb muscles. Mr. G.B. is not experiencing any disability now however he has some mild symptoms (1 in the Modified Rankin Scale). He goes about his normal activities without any help.

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