

Guillain Barre Syndrome

Three Case Reports

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Summary: Three cases of Guillain Barre Syndrome (GBS) seen in Ipoh Specialist Centre between May 1987 and January 1988 are described. The clinical manifestations of this condition are briefly discussed.

Introduction

Guillain Barre Syndrome (GBS), was originally described by Guillain et. al in 1916 following two case reports. In 1936 Guillain himself described more cases in a further report¹ and since then this condition has been widely published. Several different terms have been used to describe this illness and these includes acute infective polyneuritis, acute febrile polyneuritis of Osler and acute idiopathic polyneuritis. This condition is uncommon and its incidence is about 1.7 per 100,000 population.

Case One

A 28-year old Chinese female who was seven months pregnant (G_1/P_0) was admitted into Ipoh Specialist Centre on the night of 12th May 1987 with a history of difficulty in breathing at rest for about four weeks which was associated with a choking sensation whenever she swallowed liquids and weakness affecting both thighs.

These symptoms were progressive in nature and over the past three days she had become very breathless at rest and in respiratory distress. On admission, she was breathless at rest with a respiratory rate of 50 per minute, pulse rate 100/minute and blood pressure 100/70 mmHg. She had absent gag reflex and there was no palatal movement on phonation. Proximal muscle weakness of both lower limbs with grade four motor power of both thigh muscles were present with no evidence of muscle tenderness. All lower limb reflexes were absent but upper limb reflexes were intact. Heart sounds were normal and lung fields were clear bilaterally. Electrocardiogram showed sinus tachycardia with a heart rate of 100/minute. Chest X Ray showed no abnormality. Haemoglobin was 12.4 gram%, blood urea and serum electrolytes were within normal limits. Several hours after admission she became more dyspneic and was transferred into the ICU whereby she was put on artificial ventilation.

She became very ill in ICU with drop in blood pressure and urine output not responding to dobutamine infusion, deterioration of conscious level and the

next morning she expired. Lumbar puncture done after her demise showed a raised protein level of 180 mg% (Normal 20–40) with normal sugar and chloride level and there was no increased cells in the cerebrospinal fluid.

Case Two

A 33-year old Chinese female was admitted on 31st December 1987 into Ipoh Specialist Centre with a complain of weakness affecting both lower limbs which started acutely one week ago. She also had weakness affecting the right upper limb with paraesthesia affecting the right hand and front of chest. The weakness of lower limbs involved mainly the proximal thigh muscles and she had difficulty in climbing stairs and difficulty getting up from the squatting position. Two weeks prior to onset of leg weakness she had a febrile illness with diarrhoea which subsided after several days. On admission she had grade four weakness affecting both thighs and she was unable to get up from the squatting position and was unable to walk unaided. Motor power in both upper limbs were normal. All the upper limb reflexes and lower limb reflexes were absent even with reinforcement. No muscle tenderness was present. Sensations were intact. Cranial nerves and coordination were intact.

Haemoglobin was 11.8 gram%, Wbc 4,500 (N57%, L29%, E13%), ESR 23 mm/hour, urea: 5.0 mmol/litre, sodium 138 mmol/litre, potassium 4.4 mmol/litre, chloride 100 mmol/litre, CPK 40u/l. LE cells, Rheumatoid factor and Antinuclear factor were all negative. A tensilon test done in the ward showed a negative response. Lumbar puncture confirmed Guillain Barre Syndrome; CSF protein was 150 mg%, glucose and chloride were normal and cell count was two per high power field. She was started on oral Prednisolone 60 mg daily for two weeks and then gradually tailed off and discontinued over the next two weeks. Eight weeks after onset of illness, the proximal lower limb weakness was still present and there was generalised areflexia. She was given physiotherapy to strengthen the lower limb weakness. No respiratory involvement was noted throughout this period.

Case Three

A 56-year old Chinese woman was admitted on 27th January 1988 with a history of urinary retention of two days duration. She had been previously well until on the night of 19th January 1988 when she developed fever with chills, rigors, aching pains over both shoulders and cramping abdominal pains. Over the next few days she became bedridden due to weakness of both lower limbs and was unable to walk. On admission she was febrile. The abdomen was soft with no signs of acute abdomen. Both lower limbs were weak and she could not walk unaided, neither could she get up from a squatting position. Motor power in both lower limbs were grade three proximally and distally. Upper limbs motor power were normal. All reflexes in upper limbs and lower limbs were absent even with reinforcement and both plantars were flexors. Romberg's sign was negative. Sensory testing for all modalities of sensations were intact.

Haemoglobin was 12.5 gram%, white cell count 13,700 (N76%, L19%, M4%), blood urea and serum electrolytes were normal. CSF analysis, protein 61 mg% (N20–40), sugar 40 mg%, cells one per high power field. Diagnosis of Guillain Barre Syndrome was made and she was started on Prednisolone 15 mg qid. Fever

and abdominal pain subsided and weakness of legs also improved gradually. Prednisolone was maintained at 60 mg daily for one week and then gradually tailed off over four weeks. When seen on 19/2/88 exactly one month after the onset of her illness, all reflexes were still absent but motor power was grade five in both lower limbs proximally and distally and she is fully ambulant.

Discussion

Guillain Barre Syndrome (GBS) is believed to be due to an allergic response of the central nervous system to a previous viral infection. Sometimes there is no preceding viral infection prior to onset of peripheral neuropathy as in the first patient and sometimes a history of prior inoculation may be obtained but there is usually a history of respiratory or gastrointestinal infection 1–2 weeks preceding the onset of paralysis as seen in the second and third patient. The neuropathy is usually predominantly motor but slight sensory involvement may be present as seen in the second case who had paraesthesia but no sensory loss in the right hand and front of chest. The neuropathy affects the peripheral nerve as well as the cranial nerves. The cranial nerves involved are mainly the ocular nerves viz II, III, IV and VI nerve leading to ophthalmoplegia (Miller Fisher Syndrome),² or nerves of the medulla viz IX, X, XI and XII leading to bulbar palsy as seen in the first patient. Bilateral VII nerve palsy is common and VIII nerve palsy may manifest in deafness.

Paralysis of the intercostal nerves leads to respiratory paralysis which may prove fatal as in the first patient. Tachycardia, severe hypotension and urinary retention, as seen in the first and third patient may be due to involvement of the automatic nervous system.³ The diagnostic finding in this condition among other criteria is a raised protein content in the cerebrospinal fluid without an increase in cells (albumino-cytologic dissociation) but the severity of the illness does not appear to correlate with the protein levels.⁴

Whereas most forms of peripheral neuropathy causes distal weakness of lower limbs, in Guillain Barre Syndrome the weakness usually involved the proximal limb muscles as in the first and second case but weakness can also affect both the proximal and distal muscle groups as in the third case. In those cases where there is no involvement of the intercostal nerves the prognosis is usually good but improvement is slow and complete recovery may take three to six months. Many cases of this syndrome appears to recover more rapidly after treatment with steroids especially the relapsing variety.

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