

Guillain-Barré Syndrome associated with Typhoid Fever. A case study in the Fiji Islands

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Abstract

Guillain-Barré Syndrome is a very rare neurological complication of typhoid. We report a young girl with blood culture proven typhoid septicaemia that developed this very rare neurological complication of the disease. Following treatment with intravenous antibiotics she improved but developed the complications during the third week of her illness while admitted in hospital. To our knowledge this neurological complication of typhoid has never been reported in Fiji. (PHD 2011; Vol. 16(2): p85-88).

Key words Typhoid Fever, Guillain-Barré Syndrome, Neurological manifestations, Fiji Islands

Introduction

Typhoid fever is caused by Salmonella group of organisms. It has a high prevalence in tropical countries such as Asia and Africa. Fiji experienced a Typhoid outbreak in 2010 and clinicians were faced with different variations in the clinical presentations of the disease. Fever, anorexia and headache are common symptoms of the disease. Abdominal pains, diarrhea, constipation and myalgia are other known symptoms of typhoid. The typical symptoms may not be seen in all patients and the disease may manifest itself in an atypical form-of-which neuropsychiatric manifestations constitute an important presentation. This case report is to highlight a rare neurological complication of typhoid fever that clinicians need to be aware of.

Case Report

A 17 year old girl was admitted to the Lautoka Hospital Surgical Unit, with "acute abdomen". Two weeks prior to admission she complained of fever, loose bowel motion, epigastric pain and vomiting. Her sister was admitted at the area Sub Divisional Hospital for similar problems.

On examination, she was lethargic, dehydrated and mildly pale. Her blood pressure was 100/70mmHg, heart rate of 98 beats/minute, in sinus rhythm and temperature of 39°C. Heart sounds were dual with no murmur. Lung fields were clear on auscultation. Abdomen was soft with generalized tenderness and voluntary guarding. There was no rebound tenderness and bowel sounds were hyperactive. The spleen and liver were not palpable. There was no other mass noted. Her extremities were normal. Neurological examination was normal as well.

Laboratory examination showed Haemoglobin was 10.7g/dl, MCV 86FL, PCV 31%, Platelets count 12,000/ul, White cell count 3600/ul. Urea and creatinine were normal. Sodium 133mmol/l and potassium 2.7mmol/l. Total bilirubin 12umol/l, ALP 131U/L, AST 300U/L, ALT 73U/L. Total protein 73g/l and albumin 30g/L.

A provisional diagnosis of Salmonella was made based on her clinical presentation, laboratory results and prevalence of disease in her area of residence. She was administered with intravenous Ceftriaxone

administered with intravenous Ceftriaxone and Cloxacillin immediately. On the fifth day her blood culture result confirmed *Salmonella typhi* which was sensitive to Ampicillin, Cephalothin, Ceftriaxone, Ciprofloxacin, Amikacin and Meropenem. Ceftriaxone was continued for the duration of two weeks.

The patient developed bilateral lower limbs weakness on the eighth day of admission deteriorating to involve her upper limbs and face. A day later she had experienced difficulty in breathing and the powers in the lower limbs were graded as 1/5 and upper limbs were 2/5. She was areflexic but her sensations to touch and pin prick was intact. She didn't have neck stiffness. Her extra ocular eye movements were normal. Chest examination revealed clear lung fields. The other systems were essentially normal. A diagnosis of Guillain-Barré Syndrome was made. She was transferred to the intensive care unit. A lumbar puncture was performed which showed slightly turbid and xanthochromic cerebrospinal fluid (CSF). The total CSF protein was 8048 mg/dl, red cell was 12/mm³ and there was no white cell seen, CSF sugar was 3.5mmol, Random Blood Sugar was 5.5mmol/l, Direct smear and cultures were negative.

The patient was electively ventilated as her respiratory status had deteriorated (reduced vital capacity). A tracheostomy tube was inserted after eight days on the ventilator and a trial of intravenous steroids commenced. Plasma exchange or immunoglobulin therapy is not available in Fiji.

She required 20 days of tracheostomy and Continuous Positive Airway Pressure (CPAP) support before she was able to breathe spontaneously on the tracheostomy tube alone. On the 43rd day her right upper limb power improved to 4/5 while the right was 3/5. Her lower limb power was 2/5.

The patient was transferred to the general ward after thirty eight days in the intensive

care unit. On the eleventh week, she was able to stand and walk with minimal support. She was referred to the ENT surgeon for further management of her tracheostomy tube and seen at the outpatient's clinic. Her total length of stay in hospital was 89 days of which 38 days were spent in intensive care and 51 days in the general ward.

Discussion

Guillain-Barré Syndrome is an acute or subacute condition associated with onset of distal paraesthesia, muscle pain and weakness with a tendency for the symptoms and signs to spread proximally and with increased spinal fluid protein and a normal cell count. Guillain-Barre' Syndrome as a complication of Typhoid Fever was not known to us prior to this interesting clinical case.

Common complications of typhoid fever presented in clinical practice include acute confusion, haematological complications ranging from anaemia, thrombocytopenia and leucopenia, often in combination rather than isolated abnormality.

Pancytopenia is a common laboratory finding in our clinical experience with Typhoid in Fiji. Intestinal perforation is a known complication. This interesting case made us more aware of the neuropsychiatric complications of typhoid fever. One of the earliest case describing Guillain-Barré Syndrome as a complication of typhoid fever was reported in 1969 in Sri Lanka occurring in a 27 year old farmer.¹ In 1977 a similar report of a case was described in a 15 year old girl. She recovered uneventfully from her neurological illness ten weeks from the onset of her symptoms.² A 10 year girl with similar presentation of Guillain-Barré Syndrome as a complication of typhoid fever was reported in a Paediatric department in India as well.³

A Nigerian study reported confusional state or delirium as the commonest neuropsychiatric manifestation of typhoid fever.⁴ It

accounted for 57 % of the total neurological manifestations in a series of 959 patients. In the same study, seven patients had symmetrical sensorimotor neuropathy but without the usual CSF abnormal findings seen in Guillain-Barré Syndrome.⁴ Five patients (0.5%) were initially diagnosed to have schizophrenia. Other rare manifestations and neuropsychiatric complications observed in this series were convulsions, focal neurological deficits such as deafness, hemiplegia and infranuclear facial palsy.⁴

In a study of 791 cases of typhoid fever in India over a six year period, 665 (84%) developed neuropsychiatric complications.⁵ Acute confusional state was the commonest (73%), followed by myelitis (6%), cerebellitis (1%), parkinsonism (1%), acute psychosis (0.6%), meningo-encephalitis (0.5%), encephalitis (0.25%).⁵ This same study noted sensory motor polyneuropathy occurring in 0.12% of patients.⁵ Gazanfar et al observed that sensory motor polyneuropathy developed two weeks after onset of illness and gradually improved within four weeks of treatment.⁵ The authors also reported typhoid cerebellitis took 6-24 weeks to resolve completely in their study group.⁵

Other reports of cerebellar complications of typhoid noted a shorter period of recovery of 1-2 weeks.⁶ The major cerebellar symptom was gait ataxia. Similar neurological manifestations were observed in a study in Western Rajasthan, India.⁷ Neurological complications of the disease were seen in 63 of their patients. Typhoid delirium state was observed in 42.8%.⁷ Commonly encountered specific neurological disorders in the same study were encephalitis, psychiatric manifestations, cerebellar ataxia and meningitis. Most of the neurological complications of typhoid described by Lakhota et al were seen during the course of the illness or during recovery period.⁷ Some occurred during convalescent such as neuropathy and psychosis. Others occurred well after recovery such as motor neuron disease.

The patient in this case report developed neurological complications on the eighth day of admission which was three weeks from the onset of her illness. The mechanisms for the neurological manifestations of typhoid are not well understood?. Hyperpyrexia, electrolyte disturbances, neurotoxins, vasculitis, autoimmune process is thought to be the mechanism.⁴ The prognosis of neurological deficits in typhoid fever is usually good.^{7,8} The recovery in most cases is slow and complete but in some cases deficits may persist. Our patient showed gradual improvement in her muscle strength. Her return to normal walking and mobilization took about three months.

The management of this patient was challenging as it was difficult to differentiate between weakness due to Typhoid septicaemia and the symptom of Guillain-Barre'. The delay in diagnosis was attributed to a musculoskeletal problem and her hypokalemic state. This was corrected, however her paralysis remained. A neurological examination and diagnosis was initially overlooked as the weakness was thought to be musculoskeletal. Additionally as highlighted by this case, we were not aware of the association between typhoid and Guillain-Barré Syndrome. The diagnosis was confirmed by clinical presentation and the elevated cerebrospinal fluid protein content with normal cells. Nerve conduction studies are not available in Fiji. The treatment of Guillain-Barré Syndrome is early high dose intravenous Immune globulin (IVIg) or plasmapheresis⁸ which is not available at our hospital. Intravenous steroids were used instead although steroid has not been found to be effective.⁸ Steroid was tapered off and stopped after two months. The patient was managed by ENT specialists after discharged and has recovered fully.

As clinicians in a country where typhoid fever is increasingly encountered, we need to be aware of the complications of the disease. Guillain-Barré Syndrome is easily missed if the index of suspicion is low. In this case,

complications occurred over a weekend and the team realized that a new problem was happening about 48 hours later. Failure to recognize this complication can result in death from respiratory failure.

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Ward rounds with Dr. Lisi Tikoduadua

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