Cervical–pharyngeal–brachial Variant of Guillain–Barré Syndrome: A Sequel of Leptospirosis

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ABSTRACT

Guillain–Barré syndrome (GBS) is a lower motor neuron disease due to postinfectious immunological reaction leading to demyelination. Usual preceding etiologies are acute viral episodes caused by cytomegalovirus, Epstein–Barr, and Zika infections, among others. Bacterial organisms causing GBS are Mycoplasma pneumonia, Campylobacter jejuni, Haemophilus influenzae, and Shigella. It can also occur postimmunization against rubies, influenza, MMR (measles, mumps, rubella) and conjugated meningococcal vaccine. A rare case of GBS (acute inflammatory demyelinating polyneuropathy) occurring after an episode of leptospirosis infection is presented.

Keywords: Cervical–pharyngeal–brachial variant of Guillain–Barré syndrome, Leptospirosis, Unusual presentation.


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INTRODUCTION

Guillain–Barré syndrome is an autoimmune disorder often considered a postinfectious polyneuropathy involving mainly not only motor but also sensory and sometimes autonomic nerves. It is characterized by progressive symmetrical ascending acute flaccid weakness and mild sensory changes. Progression is often maximal by the end of 4 weeks,1 and then the condition usually plateaus before improving slowly. Particularly, in cases with an abrupt onset, tenderness on palpation and pain in muscles are common in the initial stages. Affected children are irritable. Weakness can progress to inability or refusal to walk and later to flaccid tetraplegia. Maximal severity of weakness is usually reached by 4 weeks. Bulbar involvement occurs in about half of cases, resulting in respiratory insufficiency. Dysphagia and facial weakness are often impending signs of respiratory failure.2 They interfere with eating and increase the risk of aspiration. The facial nerves may be involved. Occasionally, additional infectious precursors of GBS include mononucleosis, Lyme disease, cytomegalovirus, and Haemophilus influenzae [for Miller–Fisher syndrome (MFS)]. There are variants of GBS, namely acute inflammatory demyelinating polyneuropathy, which is the classical variant, acute motor axonal neuropathy, acute motor sensory axonal neuropathy, acute sensory neuropathy, MFS, cervical–pharyngeal–brachial variant (lower cranial nerve palsy mimicking botulism) and Bickerstaff brainstem encephalopathy.3 However, GBS post spirochete illness, leptospirosis is extremely rare. Only few such cases have been reported till date.

CASE REPORT

An 11-year-old female child was brought to the emergency department, Mahatma Gandhi Mission Hospital, Kamothe, Navi Mumbai, India, with chief complaints of fever, yellowish discoloration of urine, and myalgia since 4 days. Child was admitted in pediatric intensive care unit. Soon after admission, the child developed abdominal distension. On neurological examination, child had altered sensorium with preserved reflexes. Examination of abdomen showed liver enlarged 3 cm below costal margin and with smooth surface and rounded margin. Spleen was not palpable. Respiratory and cardiovascular examination was found to be normal.

Complete blood count showed leukocytosis with normal platelets. Liver enzymes were elevated (serum glutamic-pyruvic transaminase – 980; serum glutamic-oxaloacetic transaminase – 4000); serum bilirubin 6.2 (direct 4.2; indirect 2); serum ammonia 100; serum proteins 5.4 (albumin 3.2; globulin 2.2). Child had a deranged coagulation profile (prothrombin time 24.9; international normalized ratio 1.88); Lumbar puncture and serum ceruloplasmin levels were normal. Dark ground illumination was negative. Leptospira immunoglobulin M was positive, which confirmed the diagnosis of leptospirosis. Child was started on restricted intravenous fluids, Tab. Doxycycline and other supportive measures. She started responding well to treatment and was started on nasogastric feeds, which were gradually shifted to full oral feeds.

However, in the second week of illness, the child developed drooling of saliva, pooling of secretions, difficulty in...
speech associated with a nasal twang, difficulty in deglutition along with clumsy movement of hands. On examination, child had a weak gag reflex. Direct laryngoscopy was normal. Lower motor neuron bulbar palsy was noted. Provisional diagnosis of cervical-pharyngeal-brachial variant of GBS was considered at this point.

Repeat lumbar puncture was done. It showed cytoalbuminologic dissociation. Cerebrospinal fluid proteins were elevated to more than twice the upper limit of normal, and glucose levels were normal. Fewer than 10 white blood cells/mm³ were found on smear examination. Bacterial cultures were negative. Nerve conduction study of median and ulnar nerves showed decreased nerve conduction velocity. Child was confirmed to have diagnosis of cervical-pharyngeal-brachial variant of GBS. Injection methyl prednisolone was given for 3 days. Gradually, child showed complete recovery without any residual bulbar weakness.

DISCUSSION

Guillain–Barré syndrome is an autoimmune disorder often considered a postinfectious polyneuropathy involving mainly not only motor but also sensory and sometimes autonomic nerves. This syndrome affects people of all ages and is not hereditary. It is a common cause of acute flaccid paralysis in children. The paralysis usually follows a nonspecific gastrointestinal or respiratory infection by approximately 10 days. The original infection might have caused only gastrointestinal (especially Campylobacter jejuni) or respiratory tract (especially Mycoplasma pneumoniae) symptoms. The syndrome includes several pathological subtypes, of which the most common is a multifocal demyelinating disorder of the peripheral nerves. Evidence from histological examination of peripheral nerve biopsy and postmortem samples suggests that both cell-mediated and humoral mechanisms are involved in the pathogenesis. Immunological studies suggest that at least one-third of patients have antibodies against nerve gangliosides, which in some cases also react with constituents of the liposaccharide of C. jejuni. The GBS has been reported after administration of vaccines against rabies, influenza, measles, mumps, rubella, and following administration of conjugated meningococcal vaccine, particularly serogroup C. Other infectious precursors of GBS are infectious mononucleosis, Lyme disease, cytomegalovirus, and H. influenzae (for MFS) infections.

Leptospirosis is a spirochete infection of great public health importance in the tropics. The source of infection in humans is usually either direct or indirect contact with the urine of infected animals. These bacteria infect humans by entering through abraded skin, mucous membrane, and conjunctivae. Leptospirosis infection is described as anicteric and icteric form. The septicemic phase of anicteric leptospirosis has an abrupt onset with flu-like symptoms of fever, shaking chills, lethargy, severe headache, malaise, nausea, vomiting, and severe debilitating myalgia. The immune phase can follow a symptomatic interlude and is characterized by recurrence of fever and aseptic meningitis which manifest as headache, photophobia, and nuchal rigidity. Complications, such as optic neuritis, uveitis, iridocyclitis, chorioretinitis, and peripheral neuropathy may occur. Nervous system involvement is essentially immune-mediated and gross changes that include leptomeningeal edema, brain and spinal cord congestion, and hemorrhage. Microscopically, perivascular round cell infiltration of small- and medium-sized blood vessels along with patchy demyelination are the prominent features. In icteric leptospirosis (Weil syndrome), initial manifestations are similar to those described for anicteric leptospirosis. The immune phase is characterized by jaundice, renal failure, thrombocytopenia, and in fulminant cases hemorrhage and cardiovascular collapse.

Two case reports of GBS post leptospirosis have been published as per our knowledge. Bal et al described classical variant of GBS postleptospirosis infection. Silva et al described a case of ascending progressive leg weakness with acute pancreatitis postleptospirosis. Our case presented as cervical-pharyngeal-brachial variant of GBS postleptospira infection.

CONCLUSION

A case of cervical-pharyngeal-brachial type of GBS due to leptospirosis in an 11-year-old girl is presented. Leptospirosis is an uncommon cause of GBS as per published reports.

REFERENCES