Case Report
Guillain–Barré Syndrome Associated with Legionella Infection

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Summary
This is the first report of Guillain–Barré syndrome (GBS) related to Legionella pneumophila infection. A 13-year-old boy presented with acute dysphagia and dyspnea. He lived in a rural area and had a history of drinking potable deep-hole water. The patient was intubated because of increased respiratory distress. A positive direct fluoresein antigen test confirmed L. pneumophila infection in BAL. One week after the first admission, acute weakness was noticed including the lower extremities and was more prominent in the distal than the proximal portions. GBS was considered as the initial diagnosis. Tests for all causes known to trigger GBS were negative. Specific serology for L. pneumophila IgG was positive. He was treated with intravenous immunoglobulins and discharged with minor weakness and difficulty in walking in the second month. On the basis of this case, L. pneumophila should be included in the etiologic spectrum of GBS.

Key words: Guillain–Barré syndrome, Legionella pneumophila, weakness.

Introduction
Guillain–Barré syndrome (GBS) is an acquired, immune-mediated polyradiculoneuropathy causing dysfunction, segmental demyelination, and/or axonal degeneration in peripheral nerves, spinal sensory and motor nerve roots, and, occasionally, in cranial nerves [1]. There is a strong association between GBS and preceding acute infectious illness [2]. Common pathogens include cytomegalovirus, Epstein–Barr virus, Mycoplasma pneumoniae and Campylobacter jejuni [1,3,4]. Less frequently, other viruses are involved, such as parainfluenza, influenza, mumps, herpes virus 6 and varicella zoster [5,6]. Etiology is unknown in many cases. GBS has not been reported previously in association with Legionella infection.

Case report
A 13-year-old male presented with acute dysphagia and progressive dyspnea. He had recently received antibiotic therapy that lasted for 5 days for an upper respiratory tract infection. The patient’s detailed history indicated that he lived in a rural area and used contaminated deep-hole water. A physical examination revealed the patient was agitated, had perioral cyanosis, and had difficulty in swallowing. Other findings were normal.

Complete blood analysis and serum electrolytes were all within the normal limits. A chest X-ray showed left lung hyperinflation, deviated trachea, heart to the right and decreased right lung volume associated with consolidations (Fig. 1). Foreign body aspiration was considered, and bronchoscopy was planned based on this finding. The patient was intubated due to increased respiratory distress. No pathological image was detected concomitant with bronchoscopy and laryngoscopy, but there was a positive result for L. pneumophila IgM. Additionally, direct fluoresein antigen in bronchoalveolar lavage (BAL) was positive for L. pneumophila (Fig. 2). The patient was extubated in 2 days of hospitalization and claritromycin therapy was given for 7 days. The patient discharged without any symptoms. One week later the patient referred to the emergency service with acute neuromuscular weakness.

Acute neuromuscular weakness was confined to the lower extremities and was more prominent in the distal than proximal portions. No pathological results were detected in the blood count, biochemical analysis including serum creatin kinase and liver and renal function. The urine drug screen was found to be negative. Cranial nerves normal but sensation of vibration, pain and touch examination were impaired below the trunk. On the basis of these findings,
we suspected GBS caused by *L. pneumophilia* infection. Serology for *Mycoplasma*, influenza, parainfluenza, adenovirus, cytomegalovirus, rubella, measles, mumps and *C. pneumonia* were negative. Stool cultures for *Campylobacter*, *Salmonella* and *Shigella* were found to be negative. Additionally, *L. pneumophilia* IgG was positive. Craniospinal MR with gadolinium was normal. In nerve conduction studies, reduction in amplitude and the presence of conduction block was observed. Decreased motor conduction velocity in median nerves (17.8 m/sn) was detected and no response in peroneal nerves was observed. This findings were detected by polyneuropathy. Albuminocytological dissociation was detected by cerebrospinal fluid analysis (protein = 181 mg/dl, glucose = 70 mg/dl, and no cells). The patient was given 400 mg/kg/day IVIG therapy for 5 days. The patient did not need mechanical support again. In the second month, he was discharged from the hospital with minor weakness and difficulty in walking.

**Discussion**

There are many causes of acute neuromuscular weakness in children, including a history of drug exposure, episodic abdominal crises, psychiatric illness, seizures, trauma, skin rash, antecedent infection and vaccination [7]. Typically, GBS is characterized by ascending paralysis with lower motor neuron weakness and hyporeflexia [1]. The various forms of GBS are defined by their clinical manifestations and variable involvement of motor and sensory axons of peripheral nerves and the autonomic system [8]. Because there is no single or serological marker for GBS, diagnosis is based on clinical, laboratory and neurophysiological findings [9].

In our case, we detected antecedent infection associated with *L. pneumophilia* and the patient was discharged after therapy without any symptoms. One week later he referred with acute neuromuscular weakness to the emergency service. The clinical course, raised cerebrospinal fluid protein and nerve conduction test confirmed the diagnosis as GBS.

Axonal degeneration in GBS can be caused by an autoimmune cross-reaction directed toward gangliosides of peripheral nerves expressed on the motor axolemma or toward epitopes of Schwann cells or myelin [10]. *Legionella pneumophilia* and the previously reported agents in the etiology may trigger this immune reaction.

*Legionella pneumophilia* is rare infection in children and is usually asymptomatic or mild and unrecognized. Severe disease has occurred in children with malignant neoplasms, severe combined immune deficiency, organ transplantation, end-stage renal disease, underlying pulmonary disease, immunosuppression with corticosteroids, and as a nosocomial infection in newborn infants [11]. Transmission has been reported from contaminated potable water systems, whirlpool spa humidifiers and evaporative condensers [11,12]. Our patient had a history of use of potable deep-hole water, positive serology result for *L. pneumophilia* IgM, positive direct fluorescein antigen in BAL, and a strongly positive result for *L. pneumophilia* IgG at follow-up, all of which confirmed the diagnosis of GBS caused by *L. pneumophilia* infection. In conclusion *L. pneumophilia* should be considered as an infectious agent in GBS, especially in people who consume potable water.

**References**

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