Nonpoliovirus Poliomyelitis Simulating Guillain-Barré Syndrome

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Background: Paralytic poliomyelitis due to the wild-type poliovirus has been eradicated in the United States because of effective immunization programs. In the post-vaccination era, most cases are caused by other RNA viruses, such as coxsackievirus or echovirus. The condition usually begins with a fever and upper respiratory tract or gastrointestinal tract symptoms that progress to a "paralytic" phase characterized by limb weakness, areflexia, and, occasionally, respiratory failure that superficially resemble Guillain-Barré syndrome.

Objective: To describe 2 patients with nonpoliovirus poliomyelitis and highlight the findings on magnetic resonance imaging of the spinal cord to distinguish these cases from variants of Guillain-Barré syndrome.

Design and Setting: Case series from an academic medical center.

Patients: Following a viral illness, the patients, aged 35 and 50 years, had painless, progressive, asymmetrical weakness in the arms followed by respiratory failure in one patient, and generalized limb weakness in the other patient, reaching a nadir in 1 week. Both patients had fevers but no signs of meningitis at onset. Tendon reflexes were absent or reduced in affected regions. The cerebrospinal fluid findings were as follows: mononuclear leukocyte counts of 100,000 cells/mm³ and 700,000 cells/mm³, respectively, and the protein level was above 10 g/dL in both patients. Compound muscle action potential amplitudes were reduced in some nerves with active denervation in clinically affected muscles, and F-responses were absent but there were no other demyelinating features. Magnetic resonance imaging showed discrete T2-weighted signal changes of the ventral horns of the spinal cord, and one had elevated coxsackievirus titers in the serum. There was little recovery and significant atrophy in weak muscles after 3 years.

Conclusions: The poliomyelitis syndrome still occurs in adults in developed countries. It has superficial similarities to a motor axonal variant of Guillain-Barré syndrome but can be distinguished by clinical, cerebrospinal fluid, and, perhaps specifically, magnetic resonance imaging characteristics.

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PARALYTIC poliomyelitis currently results most often from oral polio vaccination rather than from the wild-type poliovirus, although the latter still plagues developing countries. In the United States, nonvaccine-related cases have been caused by other RNA viruses, such as coxsackievirus, echovirus, and other enteroviruses. The illness usually begins with a fever and upper respiratory tract or gastrointestinal tract symptoms that progress to a "paralytic" phase characterized by flaccid, asymmetrical limb weakness, areflexia, and occasionally, respiratory failure, superficially resembling Guillain-Barré syndrome (GBS). We describe 2 patients with nonpoliovirus poliomyelitis initially diagnosed as having GBS; in addition to fever and cerebrospinal fluid (CSF) pleocytosis, abnormalities on magnetic resonance imaging (MRI) of the spinal cord helped distinguish these cases from the acute motor axonal variant of GBS.

REPORT OF CASES

CASE 1

A 35-year-old man had acute, painless, progressive weakness of the right hand over 1 day. Within 4 days, the entire right arm was virtually paralyzed and the left arm became weak. Several days later, the weakness had progressed to cause bibralchial paralysis and respiratory failure requiring ventilatory assistance. There were no sensory symptoms, and his cranial nerves, sphincter function, and legs were
unaffected. He had a mild frontal headache and diffuse arthralgias during the preceding week, without fever or diarrhea. He had received polio vaccinations as a child, had no contact with recently vaccinated infants or children, and had not traveled outside the country.

There was a low-grade fever (temperature, 37.8°C) but no meningism. His mentation, cranial nerves, and limbs were normal, but mild weakness was present in the neck extensor muscles. The right arm and hand, and left deltoid, infraspinatus, and supraspinatus were paralyzed. There was 3/5 power (Medical Research Council scale) in the left biceps and triceps, and 4+/5 strength in the left hand. Sensation was normal. The triceps tendon reflex was trace and the biceps and brachioradialis reflexes were absent bilaterally. Reflexes in the legs were normal.

The CSF showed a mononuclear leukocyte count of 100 000 cells/mm³ (56% lymphocytes, 44% neutrophils), a protein level of 10.7 g/dL, and normal glucose concentration. Results of the complete blood cell count; liver, kidney, and thyroid function studies; erythrocyte sedimentation rate; and serum rapid plasma reagin test; human immunodeficiency virus and Lyme disease titers, and anti-GM1 antibody titers were normal or negative. Bacterial cultures of the blood, CSF, urine, and sputum were negative for organisms. Results of urine porphyrin screening were normal. The patient was tentatively diagnosed as having a variant of GBS and was treated at a local hospital. There was no immediate improvement in the patient’s condition after treatment with intravenous corticosteroids, plasma exchange, and intravenous immune globulin, and he was referred to our facility.

Electrodiagnostic studies were performed at our institution 6 weeks after presentation. The right median compound muscle action potential amplitude was 0.40 mV (reference range, >4.0 mV); conduction velocity, 58 m/s (reference range, >50 m/s); distal latency, 5.8 milliseconds (reference range, <4.4 milliseconds); and F-response, absent. The ulnar compound muscle action potential amplitude was absent, and the peroneal motor potential was normal. The right median, ulnar, and sural sensory nerve action potentials were normal. Findings from needle electromyographic examination showed widespread active and chronic denervation with absent or greatly reduced recruitment in the muscles of the upper limbs, cervical, and thoracic paraspinal region (Table). An MRI of the cervical spine showed an abnormal signal within the ventral gray matter of the spinal cord (Figure 1).

He was weaned slowly from the mechanical ventilator over 3 weeks. At 30 months, the patient’s right arm remained paralyzed with severe atrophy of the shoulder girdle, forearm, and hand muscles. The left shoulder girdle also remained paralyzed with moderate weakness of other left arm and hand muscles.

**CASE 2**

A 50-year-old man had a 3-day history of urinary retention, abdominal cramping, diarrhea, and chills without fever, followed by leg weakness. Within several days he be-
came lethargic and developed arm weakness. He had received polio vaccinations as a child, had no contact with recently vaccinated infants or children, and was not immunodeficient.

He was febrile (temperature, 38.4°C), drowsy and inattentive, dysarthric, and had a reduced gag reflex; other cranial nerve responses were normal. There was generalized weakness with 4/5 power in the arm and intrinsic hand muscles, 2/5 strength in the hip muscles, bilateral foot drop (0/5 tibialis anterior), generalized areflexia except for +1 right biceps reflex, and normal sensation. Forced vital capacity was 2.1 L (<50% of predicted). He was diagnosed as having acute GBS and referred to our institution for treatment.

Results of routine laboratory study findings were normal except for an erythrocyte sedimentation rate of 100 mm/h. The following serological study findings were also normal or negative: immunofixation, rapid plasma reagin, cold agglutinins, human immunodeficiency virus, hepatitis B and C virus, Epstein-Barr virus, Lyme disease, *Mycoplasma pneumoniae*, *Rickettsia*, herpes simplex virus 1 and 2 titers, and anti-GM1 antibody titers. Stool culture and serum titers for *Campylobacter jejuni* were negative for organisms. Urine porphyrin screening test results were normal. The CSF findings were as follows: a mononuclear leukocyte count of 695,000 cells/mm³ (91% lymphocytes, 9% neutrophils), a protein level of 10.8 g/dL, and a normal glucose level. Bacterial, fungal, and mycobacterium tuberculosis cultures of the CSF were negative for organisms. A second lumbar puncture performed 1 week later showed a mononuclear leukocyte count of 120,000 cells/mm³ (98% lymphocytes, 2% monocytes), a protein level of 26.1 g/dL, and a glucose level of 0.7 g/dL (3.9 mmol/L). Oligoclonal bands were not detected and CSF Lyme titer and rapid plasma reagin results also were negative. Cerebrospinal fluid viral titers, including Epstein-Barr virus; herpes simplex virus 1 and 2; cytomegalovirus; varicella zoster virus; coxsackievirus types B1, B2, B3, B4, B5, and B6; echovirus types 4, 9, 11, and 30; and poliovirus types 1, 2, and 3 were not detected; however, serum viral titers of coxsackievirus type B4 were elevated (1:256), suggesting recent infection. Modified barium-swallow esophagogram demonstrated moderate pharyngeal-phase dysphagia with aspiration of thin liquids.

Four weeks after presentation electrodiagnostic studies showed an ulnar compound muscle action potential amplitude of 2.6 mV (reference range, >6.0 mV) with a normal conduction velocity and distal latency. The ulnar F-response was absent. The tibial compound muscle action potential amplitude was 4.6 mV (reference range, >4.0 mV); conduction velocity, 39 m/s (reference range, >40 m/s), and distal latency, 4.5 milliseconds (reference range, <5.9 milliseconds). The tibial F-response and H-reflex were absent. The ulnar and sural sensory nerve action potentials were normal. Findings from a needle electromyographic examination showed fibrillation potentials, modest reinnervation changes (+1 duration, amplitude, and phases), and reduced recruitment patterns in proximal and distal muscles of the left arm and leg. Magnetic resonance imaging showed slight enlargement of the spinal cord in the cervical and thoracic regions with discrete signal changes within the gray matter (Figure 2).

He was treated with broad-spectrum antibiotic agents and acyclovir and his mental status improved over several days, but the weakness remained unchanged during the hospitalization. There was slow improvement in his condition and after 20 months his arm, hand, and hip girdle muscles were normal but the foot drop was unchanged. Deep tendon reflexes were normal except for absent Achilles reflexes bilaterally. He could walk with leg braces.

**COMMENT**

Our patients had acute, rapidly progressive regional or generalized weakness that followed an infectious pro-
drome and simulated the acute motor axonal neuropathy variant of GBS. Findings from electrodiagnostic studies demonstrated a pure motor axonopathy indistinguishable from abnormalities that have been observed in cases of acute motor axonal neuropathy. Although our patients were initially misdiagnosed as having a motor variant of GBS, the fever, short latent period, and CSF pleocytosis suggested an acute infectious process, and with the MRI findings were characteristic of poliomyelitis.

The prodrome of paralytic poliomyelitis consists of malaise, headache, fever, myalgias and muscle stiffness, and sore throat that last a few days before progressing to the paralytic phase. Our second patient displayed other salient prodromal features such as mental status changes, irritability, and restlessness, reflecting aseptic meningitis. Weakness peaks rapidly over several days in most cases, rarely evolving in a saltatory fashion over 1 week or longer. In contrast to GBS, weakness occurs at the height of the fever and generally does not progress after the fever subsides. An asymmetrical, flaccid paralysis develops in most patients, typically affecting proximal more than distal muscles. The legs are involved more often than the arms and differential involvement of muscles within a limb is common, but complete paralysis of an affected limb is unusual. In contrast, in 1 of our patients the illness began asymmetrically in the arms and rapidly evolved into a symmetrical pattern, and the other patient had a symmetrical distribution of weakness from the outset, similar to another study. Oropharyngeal muscles are affected in 10% to 15% of cases, and mechanical ventilatory failure occurs when the intercostal muscles and diaphragm also are affected, similar to 1 of our patients. Fasciculations may be prominent early in the illness but are usually transient and were not noted in either of our cases. Deep, aching muscle pain, indistinguishable from what occurs in some cases of GBS, is common as weakness progresses. Reflexes are reduced in the clinically affected limbs. Although some patients complain of paresthesias early in the illness, objective sensory loss is not present.

Vaccine-associated poliomyelitis has been linked to poliovirus type 3 and is extraordinarily rare. It generally occurs within 30 days of immunization with a relative risk of infection of 0.02 to 0.04 cases per 1 million doses of oral poliovirus vaccine. However, coxsackieviruses groups A and B, echovirus, and other viruses are the main causes of sporadic, nonpoliovirus poliomyelitis and produce a paralytic syndrome indistinguishable from poliovirus. Schellinger et al recently described 6 patients with a poliomyelitislike illness following a tick bite and presumed infection with flavivirus, causing an unusual form of central European encephalitis. Recognition that a suspected poliovirus infection is due to another virus is invariably retrospective, based on negative acute and convalescent poliovirus titers or culture, or raised titers of the offending virus.

Although poliomyelitis is a common cause of acute flaccid paralysis in many countries around the world, GBS and its variants remain the most common cause of rapidly progressive generalized weakness in the United States and Europe. For example, Busby and Donaghy recently described 4 patients with an acute, bibrachial, motor predominant neuropathy that was considered a regional variant of GBS, like one of our cases and similar to others that have been reported with cervico-pharyngeal-brachial involvement. A pure motor variant of GBS was observed in 18% of cases in one large series, but the figure is closer to 3% in our experience. These cases are characterized by distally predominant weakness, areflexia, lack of sensory symptoms or signs, sparing of cranial nerves, an early clinical nadir, higher than usual rates of raised anti-GM1 or anti–GalNAc-GD1a antibody titers, and preceding C jejuni infection. These cases may be clinically similar to Chinese patients who had the acute
motor axonal neuropathy variant, but demyelinating changes detected by electromyographic studies are more frequent in the former group. In contrast to the other well-described pattern of axonal GBS, acute motor sensory axonal neuropathy, our patients lacked sensory symptoms or findings, electrically inexorable motor and sensory nerves, and acellular CSF characteristic of acute motor sensory axonal neuropathy. In addition to pure motor variants of GBS, rare disorders such as lead poisoning, acute fulminant myasthenia gravis, diphtheritic neuropathy, and tick paralysis are the other major diagnostic considerations in patients with acute flaccid paralysis.

A useful finding in our cases was the striking T2-weighted signal changes observed within the gray matter of the spinal cord on MRI, corroborating a disorder of the anterior horn cell rather than the ventral roots or motor nerves. In addition to the marked CSF pleocytosis, the MRI abnormalities excluded a motor variant of GBS. The appearance of abnormal signal in the gray matter of the spinal cord may not be specific but is characteristic of the anatomical distribution of poliomyelitis, in contrast to nerve root enhancement, a common MRI abnormality in patients with GBS. The changes on MRI in poliomyelitis likely reflect edema, perivascular and intraparenchymal inflammation, gliosis, neuronophagia, or anterior horn cell loss, and identical MRI abnormalities, mostly in children, have been described in the literature. The lack of recovery and substantial residual weakness suggests irreversible damage of the anterior horn cells in our cases, in contrast to the usually favorable recovery of most patients with GBS.

Nonpoliovirus poliomyelitis is a rare cause of acute flaccid paralysis and usually can be differentiated from typical GBS by an active, infectious prodromal phase, inflammatory CSF profile, and lack of sensory features typical GBS by an active, infectious prodromal phase, in contrast to the usually favorable recovery of most patients with GBS.

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