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The Many Faces of Guillain-Barré Syndrome

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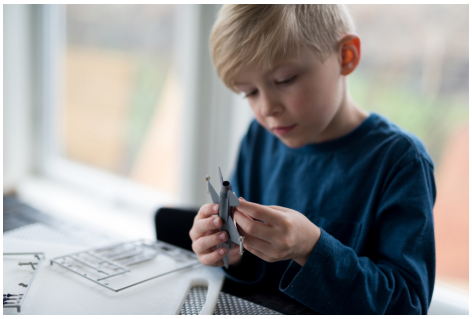

Most of us learned the basic clinical manifestations of Guillain-Barré syndrome in medical school and likely have encountered at least a few children with the condition during residency or in practice. Four January 2018 Pediatrics in Review articles focus on this immune-mediated peripheral neuropathy, emphasizing some of its less common clinical patterns that are apt to be less familiar to most physicians.

In their In Brief on the topic, Chung and Deimling (1) summarize the classic manifestations of Guillain-Barré syndrome. This is an acute-onset, immunemediated peripheral neuropathy that likely is triggered by an infection or another immune stimulus. Guillain-Barré syndrome classically presents as an acute-onset, rapidly progressive, flaccid weakness starting in the legs, associated with diminished or absent tendon reflexes. The clinical severity of Guillain-Barré syndrome varies, but at its worst, it can lead to respiratory insufficiency and death. The cerebrospinal fluid (CSF) typically exhibits an elevated protein level but not an excess of white blood cells, although this pattern is not always present initially, and occasionally individuals have a mildly elevated white blood cell count. (1)

In Index of Suspicion Case 3, Bassal and Lupo (2) present an 11-year-old boy with a 2-day history of gait unsteadiness and diplopia. His examination revealed initially diminished then absent tendon reflexes, dysmetria, and partial dysfunction of cranial nerves III and VI. His extremity muscles were not weak. Results of imaging and laboratory investigations were normal except for elevation of antiganglioside anti-GQ1b immunoglobulin G antibodies.

The differential diagnosis of acute ataxia is extensive. (3) This boy's diagnosis is Miller Fisher syndrome. Generally considered to be a milder variant of Guillain-Barré syndrome, Miller Fisher syndrome features a triad of areflexia, ataxia, and cranial neuropathy. (4) Most individuals with Miller Fisher syndrome recover completely, with or without therapy. Nonetheless, prompt recognition of Miller Fisher syndrome may prevent unnecessary diagnostic tests, and occasionally patients progress to the riskier Guillain-Barré syndrome.

In Index of Suspicion Case 1, Shah et al (5) describe a 5-year-old boy with right leg pain and difficulty walking for 5 days. His right leg was weak and tender to palpation. Tendon reflexes were absent in the right leg. Left leg strength was normal, but even on the left his reflexes were diminished. This presentation might suggest a localized joint problem, but hip magnetic resonance imaging (MRI) findings were normal. Weakness in 1 limb would once have suggested poliomyelitis, which often presents in this manner. More recently, asymmetrical flaccid paralysis due to acute myelitis has been described. (6)

This boy too proved to have a variant of Guillain-Barré syndrome. His CSF revealed increased protein and immunoglobulin G levels. An MRI of the thoracic and lumbar spine documented asymmetrical contrast enhancement of ventral and dorsal nerve roots, and a nerve conduction velocity test confirmed axonal polyneuropathy in both legs. Back or leg pain is common in children with Guillain-Barré syndrome. The weakness of Guillain-Barré syndrome is typically symmetrical, but as this child illustrates, it isn't always.

In Index of Suspicion Case 2, Bashir and Aarons (7) describe a 2-year-old girl who refused to walk for 3 weeks. Based on the tests and treatments initially recommended, her symptoms at first were attributed to musculoskeletal disease. When examined, she had no tendon reflexes in the legs and could not dorsiflex the right foot. Movement of her legs seemed to elicit pain. Eventually an MRI demonstrated abnormal spinal nerve roots, and a lumbar puncture confirmed an elevated CSF protein level in the absence of inflammatory cells.

Persistent, painful dysfunction without complete paralysis has a broad differential diagnosis that includes several conditions that are far more common than Guillain-Barré syndrome. The symptoms of toxic synovitis usually improve with the use of nonsteroidal anti-inflammatory agents and heat. Septic arthritis, diskitis, and osteomyelitis were eliminated because of her lack of fever and normal laboratory test results. Postinfectious myositis could have explained her signs and symptoms. A spinal cord lesion, such as a tumor or epidural empyema, must be considered in this setting.

It is important to recognize not only classic Guillain-Barré syndrome but also its less obvious patterns. (8) Children often improve spontaneously or in response to intravenous immunoglobulin therapy or plasmapheresis, and as is often the case, earlier treatment is generally more effective. There are potentially life-threatening complications of Guillain-Barré syndrome, including respiratory failure and autonomic dysfunction with systemic hypertension or cardiac arrhythmias. These 4 recent articles together provide an excellent overview of both the classic and the atypical manifestations of the syndrome.

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