Progressive Weakness in a Previously Healthy 4-year-old Boy

Kristen Pontiff, MD,* Day Breen, MD,† Pamela McMahon, PhD, MPH,* Cristina Zeretzke-Bien, MD,‡ Philip Zachariah, MD,§ Christopher Woodward, DO*x

*Pediatric Residency Program, Our Lady of the Lake Regional Medical Center, Baton Rouge, LA
†Neurodevelopmental Disabilities, Pediatric Neurology, Baylor College of Medicine/Texas Children’s Hospital, Houston, TX
‡Department of Emergency Medicine, University of Florida, Gainesville, FL
§Emergency Medicine Residency Program–Baton Rouge, Louisiana State University Health Sciences Center, Baton Rouge, LA

EDITOR’S NOTE

Ahh, summertime! Some of us look forward to vacations, perhaps reading on a beach or in a quiet forest. We like summer as we share stories with friends, make new memories, and broaden horizons. Of course, you can take Pediatrics in Review along on your summer vacations! You can enjoy the “stories” of Index of Suspicion cases, and you can make new memories as you learn of new diagnoses and treatments. You can broaden the scope of your care of future patients. You might even see a patient later this summer who has one of the diagnoses featured in this month’s issue. Enjoy!

Philip R. Fischer, MD
Associate Editor, Index of Suspicion

PRESENTATION

A previously healthy 4-year-old boy is carried into the emergency department with progressive weakness affecting all 4 extremities for a 24-hour duration. For the past 2 days, the boy has been more fatigued and complaining of right knee and lower back pain. The parents report no history of trauma. He has a normal appetite, without nausea, vomiting, or recent gastrointestinal illness. He has no dysuria, polyuria or incontinence, chest pain, or cough.

On examination he is afebrile, with normal vital signs. He is mildly distressed, without toxic appearance. There are no obvious rashes, abrasions, ecchymoses, or other findings on exposed skin areas. He is alert and oriented to person, place, and time. On cranial nerve examination his pupils are reactive bilaterally, and extraocular movements are intact. However, slowed smooth pursuit and horizontal nystagmus are observed. Bilateral facial droop is present, along with 4/5 strength of the sternocleidomastoid muscles. He is experiencing diffuse hypotonia, with no spontaneous movement of the bilateral lower extremities. He has 2/5 strength in the distal upper extremities bilaterally. Deep tendon reflexes are absent. He has intact sensorium throughout. Examination of the joints reveals
normal passive range of motion. All other physical examination findings are normal.

DISCUSSION

After a thorough examination of the skin, an engorged tick was detected on the scalp and was properly and fully removed. Findings on laboratory evaluation were normal, including a complete blood cell count, C-reactive protein level, complete metabolic panel, and erythrocyte sedimentation rate. The boy was admitted to the hospital. Within 8 hours of tick removal he was sitting unsupported, and by 12 hours he was ambulating without assistance.

Differential Diagnosis

Acute-onset paralysis in children has a broad differential diagnosis, and it is important to establish whether the paralysis is due to an upper motor neuron versus a lower motor neuron lesion. Given the boy’s neurologic examination findings, with absent reflexes, the paralysis is likely due to a condition affecting lower motor neurons. Some conditions to be considered are Guillain-Barre syndrome and transverse myelitis. One can distinguish among these conditions by performing an adequate neurologic examination.

Guillain-Barre syndrome, in particular, affects the peripheral nerves by demyelination. The classic presentation is a viral illness with an upper respiratory tract infection or gastrointestinal illness 1 to 2 weeks earlier, followed by a symmetrical ascending motor weakness. Lower extremities are affected more than upper extremities. Areflexia and paresthesia are also present.

Transverse myelitis is a neurologic condition that presents at a distinct motor or sensory level. Most patients presenting with transverse myelitis will have physical examination findings that point to a specific area of the spinal cord. This helps to differentiate Guillain-Barre syndrome and tick paralysis, which affects the nervous system more globally.

The Condition

Tick paralysis is caused by 2 major tick vectors in the United States: the dog tick and the wood tick. The female tick attaches onto its host and injects a neurotoxin that is believed to block acetylcholine release. In the United States, children and girls are more frequently affected than adults and boys, respectively. It is thought that girls are disproportionately affected because the tick remains unnoticed on the scalp due to their long hair. Its incidence is highest during mating season (February to August). Mortality of those affected by tick paralysis ranges from 6% to 13%. Seventy-nine percent of these deaths occur in children younger than 16 years. Respiratory failure is the primary cause of mortality because of failure to recognize that the paralysis was caused by the tick bite and, therefore, delay in removal of the tick to prevent respiratory muscle paralysis.

Most affected children have nonspecific symptoms, including a 4- to 7-day prodrome of restlessness and irritability, followed by ascending paralysis. Bulbar lesions can be apparent on physical examination and may cause dysphagia and slurred speech. Local skin reactions have also been noted and are described as morbilliform rash. These rashes are different than the centripetal rash seen with Rocky Mountain spotted fever. Although symptoms of many parasite infections continue long after tick removal, tick paralysis symptoms resolve rapidly after removal of the tick.

Diagnosis

Characteristic signs and symptoms coupled with response to removal of the offending tick are enough to establish the diagnosis, and further evaluation is not necessary.

Management

Removal of the tick is the most important aspect of tick paralysis. Proper removal of the tick, including the head, which is usually lodged under the skin, will result in complete resolution of symptoms over a 24- to 48-hour period. A few case studies have reported weakness lasting up to 1 week.

To achieve proper removal of the engorged female tick, one must grasp the tick at the base of the head and remove. This can be accomplished with special tweezers that do not put pressure on the body of the tick. The most safe and efficient way to remove the tick is with tick keys, which slide over the body and, when pulled slowly sideways, removes the entire tick.

Education on how to properly remove ticks is vital, especially for those who live in areas where tickborne illnesses are prevalent. It is imperative to relay that burning and use of petroleum jelly products, 2 commonly used methods, are ineffective in tick removal. In addition, it has been reported that practitioners who removed ticks without gloves have developed transient paralysis.

Progression to respiratory compromise and failure will occur without removal of the offending tick. Patients who experience respiratory failure may need aggressive intervention, including intubation and ventilator support until the toxin effects subside.

To date, there is no evidence of other tickborne illnesses in patients with tick paralysis, and for this reason additional evaluation for tickborne illnesses is not recommended.
Lessons for the Clinician

- Tick paralysis is a relatively rare but easily diagnosed condition in children with acute-onset ascending paralysis.
- Children presenting with ascending paralysis should be carefully examined from head to toe, with care taken to fully examine hair-bearing areas, areas between toes, and in and around the ears.

- Signs of lower motor neuron paralysis, intact sensation, and resolution of paralysis on removal of the tick establish the diagnosis of tick paralysis.
- Proper removal of the tick is the mainstay of treatment.

Suggested Readings for this article are at http://pedsinreview.aappublications.org/content/40/6/302.
Suggested Readings


### Case 1: Progressive Weakness in a Previously Healthy 4-year-old Boy

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