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Child Neurology: Tick paralysis

A diagnosis not to miss

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A 4-year-old girl presented to our tertiary care hospital with a complaint of lower extremity weakness and unsteady gait for 2 days. She was able to pull herself to stand but could not stand unsupported. She had no sensory symptoms or pain. She did not complain of any weakness in her arms, trunk, face, or neck. She had no bowel or bladder incontinence or retention. On presentation to the emergency department, she had minimal antigravity strength of the lower extremities but normal strength elsewhere. In addition, she was areflexic in both lower extremities and had a wide-based, unsteady gait but no appendicular dysmetria or titubation. Sensory examination was normal.

After consultation by the neurology service, MRI of the brain and total spine were completed and a plan was made for subsequent lumbar puncture. Lyme disease antibodies were drawn because of exposure to a wooded area in West Virginia; these were negative. MRI of the spine showed syringomyelia extending from T5 to T8 and an extramedullary, intradural cystic lesion dorsal to the spinal cord from T1 to T4, which was believed to be consistent with an arachnoid cyst. Due to this unexpected finding, the neurosurgical service was consulted, who believed that this cyst and the associated syrinx were the source of her paralysis. The following day, she was taken to the operating room for fenestration. Subsequent to the fenestration, repeat imaging showed resolution of syringomyelia.

The following night, the patient developed increasing respiratory distress, requiring mechanical ventilation. Over the subsequent postoperative period, she failed multiple attempts at extubation. Extensive evaluation including infectious workup, chest x-ray, ultrasonography of the diaphragm, and upper airway endoscopy revealed no reason for her ongoing breathing difficulties. In addition, it was noted that the patient had not been able to move her upper extremities at any point during the day of surgery or in the following days.

The neurology service was consulted again for further evaluation. Seven days after the initial surgery, the patient's neurologic examination revealed flaccid paralysis of all 4 extremities, bifacial weakness, minimal gag reflex, and complete areflexia. She had full extraocular movements and normal pupillary response to light. Bulbar function was difficult to evaluate due to intubation. Repeat MRI of the brain and cervical spine revealed continued resolution of syringomyelia and no new abnormalities. A lumbar puncture showed mild albuminocytologic dissociation with protein of 90, 2 leukocytes, and 1 erythrocyte. IV immunoglobulin (IVIg) therapy was instituted for presumed acute inflammatory demyelinating polyneuropathy.

On postoperative day 8, EMG and nerve conduction studies were completed. Nerve conduction studies revealed low compound motor action potentials in multiple nerves with preserved sensory nerve action potentials. There was no prolonged conduction velocity seen and normal F-wave responses were noted. EMG/needle study revealed increased insertional activity and positive sharp waves. The summation of these results suggested a possible diffuse motor axonal neuropathy or a presynaptic neuromuscular junction disturbance. Moreover, it did not fulfill criteria for a primary demyelinating neuropathy.

Based on EMG results, we performed a thorough evaluation of the patient's skin and scalp. Along the superior retroauricular scalp, a 3-cm engorged tick was found and removed. This tick was identified by an infectious disease specialist as a gravid female *Dermacentor* species tick.

DISCUSSION Tick paralysis (TP) is a rare and easily reversible condition that if missed can lead to significant morbidity and mortality. In one series of children with TP between 1946 and 1906, 6% died.[1] However, in the modern era of respiratory support and intensive care, survival may be higher. TP in the United States is more common in girls younger than 8 years with long hair, presumably due to the ability of the tick to go unnoticed on the scalp.[2,3]

Most cases reported in the literature have been identified in Australia, where the causative species is *Ixodes holyclus*. In North America, most cases reported in the Rocky Mountain region, US Pacific Northwest, and Southwestern Canada are transmitted by *Dermacentor andersoni* species and in the Southeast region are transmitted by *Dermacentor variabilis*.[4]

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These distinctions are relevant due to the differences in clinical presentation produced by the 2 species. Pupillary changes and focal weakness are more common in Australian cases (i.e., *Ixodes* cases). In addition, symptoms tend to remit immediately upon removal of a *Dermacentor* tick, whereas they persist for a day or two after removal of an *Ixodes* tick. Duration of recovery is more prolonged in Australian cases, often lasting days to weeks.[1]

TP is thought to be caused by a neurotoxin produced in the insect's salivary glands. The toxin is thought to decrease presynaptic acetylcholine release at the neuromuscular junction, similar to botulinum toxin. It is possible that variations in the toxin of *Dermacentor* ticks compared to *Ixodes* ticks may account for the variation in clinical features.[1]

The classic clinical presentation of TP is an acute symmetric, ascending flaccid paralysis occurring over hours to days. There can be a prodrome of restlessness, irritability, fatigue, and myalgias, but fever is noticeably absent. Weakness usually begins in the lower extremity, and as the tick continues to feed, the weakness ascends from the legs to the arms and then to the muscles supplied by the cranial nerves, causing dysphagia, dysphonia, and facial weakness. Deep tendon reflexes are diminished or absent.[1,5] A case series reported from Australia noted frequent pupillary involvement and external ophthalmoplegia in 2 of their patients, although this has not been the case in the United States.[6] Respiratory involvement and requirement for mechanical ventilation occur invariably if the tick remains in place, though in some patients the tick may have fallen off, accounting for those patients who recover without assisted ventilation.[1] Atypical presentations have been reported, including lower motor neuron facial nerve palsy, in which ticks were identified in the external auditory canal,[7] and left-sided arm weakness in a brachial plexus distribution, which resolved after an engorged tick was removed from the subclavian fossa.[8]

TP presents as an acute-onset flaccid paralysis of the lower extremities with hyporeflexia or areflexia. Therefore, the differential diagnosis typically includes pathologies of the lower motor neuron or neuromuscular junction. See the table for full differential diagnosis.

Table Differential diagnosis of tick paralysis[1,9]

Guillain-Barré syndrome (acute inflammatory demyelinating polyneuropathy or acute motor axonal neuropathy)	Spinal cord compression	Transverse myelitis
Cerebellar ataxia	Poliomyelitis	Myasthenia gravis
Botulism	Organophosphate ingestion	Lambert-Eaton syndrome
Encephalomyelitis	Periodic paralysis	Diphtheria
Porphyria	Electrolyte imbalance	Heavy metal poisoning

The diagnosis of TP is made by finding the engorged tick on a patient with symptoms that correlate clinically. The importance of a complete skin evaluation including the scalp, external ear canals, groin, and axillae is irrefutable. Neuroimaging studies including CT and MRI are normal, although on closer inspection they may show the embedded tick if located on the scalp. CSF should also be normal.[9] The albuminocytologic dissociation in our patient was believed to be postsurgical. If performed, electrophysiologic tests show a diffuse reduction in the compound muscle action potentials (CMAPs) with preserved sensory nerve action potentials. The low CMAPs are not usually accompanied by any abnormality of neuromuscular transmission with repetitive nerve stimulation testing. Published cases have proved that the function is reversible after removal of the tick.[10]

Despite attention to other tickborne diseases such as Lyme disease and Rocky Mountain spotted fever, TP remains a frequently misdiagnosed entity. A recent meta-analysis reviewed 50 cases of TP in the United States between 1946 and 2006 and revealed that 11 (22%) of these cases were initially misdiagnosed, with mean time to correct diagnosis of 2.16 days. Of these 11 cases, 9 were initially diagnosed as Guillain-Barré syndrome (GBS), 1 as chronic polyneuropathy, and 1 as postinfectious polyneuritis. In this analysis, preparations for invasive IV therapy for GBS were initiated in 4 patients before tick attachment was discovered and 3 patients received IVIg, while 1 case was discovered during the process of placing a central catheter to prepare for plasmapheresis.[4]

The definitive treatment of TP is removal of the offending tick, after which symptoms rapidly resolve. Careful inspection of the rest of the body for additional ticks is mandatory. The tick should be carefully removed by grasping it as closely as possible to the attachment site and using steady traction to avoid leaving the head or mouthparts engaged.[5] During paralysis, standard supportive therapy should be utilized, including mechanical ventilation when necessary for respiratory support.[9] Antitoxin, a hyperimmune dog serum used in veterinary medicine, has been used in severe cases but carries a high risk of adverse reaction.[6]

CASE SUMMARY Within 24 hours after removal of the tick, our patient started to regain some movement in her upper and lower extremities. Within 48 hours, reflexes were elicited in the patella, ankles, biceps, and brachioradialis bilaterally. By 3 days after removal, she was extubated. She had normal facial strength but continued to have some weakness, primarily in the upper extremities, likely due in part to deconditioning. The patient was able to sit independently, feed

herself, and walk with minimal assistance 6 days after tick removal and was discharged home.

AUTHOR CONTRIBUTIONS

Sarah L. Chagnon: corresponding author responsible for case report, literature review, and primary content of the manuscript. Monica Naik: coauthor responsible for revision of the manuscript. Hoda Abdel-Hamid: coauthor responsible for revision of the manuscript and execution and interpretation of EMG and nerve conduction studies.

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