Transverse myelitis, a diagnosis that may be made in the emergency department (ED) by emergency physicians, can be difficult to diagnose because of its variable signs and symptoms and its poorly understood pathogenesis. In this article, we recount 2 cases of transverse myelitis to demonstrate its presentation, diagnosis, and management in the ED. [Ann Emerg Med. 2005;46:256-259.]

INTRODUCTION

Transverse myelitis is an acute or subacute inflammatory disorder of the spinal cord. Though quite variable, as demonstrated in the cases included, transverse myelitis often presents with focal neck or back pain, followed by dermatomal paresthesias, sensory loss, paraplegic symmetric motor weakness, sphincter disturbance, and urinary retention. Depending on which portion of the cord is involved, motor, sensory, and autonomic symptoms may predominate.1 These symptoms can evolve over hours or several days and quite often can have atypical presentations.

There are many possible causes of transverse myelitis, and often the individual cause can be obscure. Postinfectious transverse myelitis follows a recent infection or vaccination. Although Epstein Barr2 and cytomegalic3 viruses are most common, practically all human viruses have been associated with transverse myelitis, including human T-cell lymphotrophic viruses.4 Mycoplasma5,6 is the only known bacterial trigger. Some acute infections such as schistosomiasis and Lyme disease can directly cause transverse myelitis. Systemic diseases such as multiple sclerosis,7 systemic lupus erythematosus,8 or cancer9 can be the cause. Cord ischemia from aortic dissection is another diagnostic consideration. If no cause is found, transverse myelitis is said to be idiopathic.

We present 2 cases of transverse myelitis in patients who presented to the emergency department (ED) to familiarize emergency physicians with this condition.

CASE REPORT

Case 1

A 20-year-old man with no significant past medical history presented to the ED with lower extremity weakness. One week before, he had developed sore throat and fatigue without fever, headache, neck stiffness, visual changes, joint pain, or diarrhea. During the ensuing week, he was evaluated twice by other physicians; according to the patient, test results for streptococcal pharyngitis and mononucleosis had been negative, although further information was unavailable. On the evening before his final presentation, the patient noticed difficulty walking, particularly while climbing stairs. He awoke approximately 4 to 5 hours later with burning dysesthesias in both legs. On attempting to get out of bed, he fell to the floor, unable to support his weight. The patient was then transported to the ED, at which time he was unable to move his legs or urinate.

On physical examination, he was afibrile (vital signs of 98.4°F, 132/84 mm Hg, 90 bpm, 19 breaths/min, 100%RA) with a supple neck. His cardiovascular examination result was normal, and he had good distal pulses. His back was unremarkable, with no tenderness or skin findings. His mental status, cranial nerves, and upper extremities were normal. However, both lower extremities were paralyzed, atonic, and areflexic. Sensory testing revealed loss of light touch, temperature, and pinprick below T-8. Rectal sphincter tone was decreased.

Normal respiratory mechanics (negative inspiratory force and vital capacity) were documented before spinal cord magnetic resonance imaging (MRI). MRI showed abnormal enhancement and swelling of the thoracic cord (see Figure).

A lumbar puncture revealed a normal opening pressure (170 mm H2O), pleocytosis (WBC count 247 per cubic millimeter, with 39% polymorphonnuclear cells and 51% lymphocytes), a normal glucose level (45 mg/dL), and an elevated cerebrospinal fluid total protein level (279 mg/dL). The cerebrospinal fluid did not show oligoclonal banding. Blood testing for Borrelia burgdorferi, HIV (antibody levels), herpes simplex (polymerase chain reaction), bacteria and fungi (blood cultures), and antinuclear antibody titers were all negative.

We administered intravenous steroids, as well as empiric ceftriaxone and ganciclovir, and admitted him to the ICU. He was discharged on hospital day 4 to a neurologic rehabilitation center with persistent paraplegia and a T-6 sensory level. He
continued to receive high-dose oral steroids for this acute transverse myelitis. At a 4-month follow-up evaluation, a patchy sensory loss of the bilateral lower extremities remained, and the patient exhibited a spastic paraparesis; however, he is able to ambulate with assistance.

Case 2

A 38-year-old female marathon runner presented in August with 2 weeks of gradual onset, progressively worsening, sharp, midthoracic, diffuse, right paraspinal back pain. She also reported “tight” right upper abdominal muscles and low-grade fevers but denied pleuritic chest pain, bowel or bladder incontinence, nausea, vomiting, or leg weakness. One month previously, she recalled a painless, nonpruritic right midscapular rash; a physician had prescribed cephalaxin and obtained antibody titres to Borrelia burgdorferi (which were negative according to the patient).

On examination, the patient had moderate to severe back pain and was lying flat in bed. Vital signs were 98.6° F, 58 bpm, 142/89 mmHg, 12 breaths/min, 99%RA. There was no midline spine tenderness. Straight leg testing results were negative, and a range of motion of the back was painful but within normal limits. Full neurologic examination results were normal except for a 1-cm band of decreased sensation to light touch along the right T5-6 front distribution just barely crossing the midline. There was a 4-cm irregularly shaped flat purple blanching rash without vesicles in the right subscapular region (the site of her original rash). The patient’s pain was relieved with morphine and diazepam. An ECG demonstrated sinus bradycardia without evidence of ischemia or right-sided heart strain. Plain radiographs of the chest and spine and a computed tomographic examination with angiography of the chest were normal. However, because of her pain severity and her small band of decreased abdominal sensation, we obtained thoracic and lumbar spine MRIs with the suspicion of cord myelitis or epidural abscess from possible Lyme or zoster. Both MRI results were normal. After this negative and thorough evaluation, we diagnosed her with musculoskeletal strain and discharged her from the ED with ibuprofen and diazepam prescriptions.

One week later, she returned with excruciating thoracic and lower back pain that kept her awake at night. She was unable to walk comfortably despite compliance with the prescribed medications. She described her worsening band of abdominal “tightness” as “silicone or Novocain wrapped around me.” Her examination was unchanged except for an expanded area of decreased sensation to pinprick, light touch, and temperature, now from T4 to T12 on both front and back, although more prominent on the front and on the right. Also, she had decreased vibratory sense of bilateral toes, but position, gait, and strength were preserved. Lumbar puncture revealed clear cerebrospinal fluid, with 541 leukocytes per cubic millimeter (90% lymphocytes), normal glucose levels (51 mg/dL), and elevated cerebrospinal fluid protein (171 mg/dL). Her erythrocyte sedimentation rate was normal (5 mm/hr). Repeated thoracic and lumbar MRI results showed patchy longitudinal enhancement of the cord from T4 to T8 levels. She was admitted to the neurology service with a diagnosis of transverse myelitis. During her admission, further testing revealed the etiology of her transverse myelitis to be Lyme disease. Her cerebrospinal fluid was positive for immunoglobulin M and immunoglobulin G antibody to B burgdorferi, and her serum Lyme antibody titer was positive by enzyme-linked immunosorbent assay (but negative by Western blot). She was treated with intravenous ceftriaxone during admission and as an outpatient for a planned course of 3 weeks.

DISCUSSION

Transverse myelitis is a diagnosis that may be made in the ED, although varied presentations are common. Some patients may have a classic presentation (patient 1) with rapidly evolving back pain, most often in the thoracic area, and lower-extremity weakness. Others may present with abrupt segmental back or radicular pain, followed by ascending paresthesias and weakness, similar to that with Guillain-Barré syndrome. Urinary and fecal retention or incontinence is common, as is ataxia and leg
Transverse Myelitis

Hammerstedt, Edlow & Cusick

weakness. As in patient 2, it is also possible for transverse myelitis to present as a slowly progressive back pain with prominent sensory deficit and no other associated symptoms. Symptoms may evolve over hours, days, or weeks.

Transverse myelitis should be included in the differential diagnosis of any patient with back pain and weakness. The variable presentation of transverse myelitis may mimic other emergencies, which should be pursued appropriately by the clinician on an individual case basis. In rapidly progressive cases, one must consider causes such as aortic dissection and other vascular pathology that may cause cord infarction. When symptoms evolve over days or weeks, it is important to exclude cord compression from tumor, abscess, hematoma, or central disc herniation. Patients with slow onset symptoms may have to be admitted to a neurology service for multiple specialist consultations to rule out mimics such as multiple sclerosis, Guillain-Barré syndrome, tick paralysis, botulism, or poliomyelitis because these may be difficult to exclude in the ED.

MRI best establishes the diagnosis of transverse myelitis by demonstrating high-intensity signals on T2-weighted images extending longitudinally along the cord. The number of segments involved may be as few as one, and occasionally, the entire cord is involved. It is possible to differentiate between multiple sclerosis and transverse myelitis by MRI, as transverse myelitis usually affects the central cord and multiple sclerosis affects the peripheral cord and involves brain abnormalities. Also, as in patient 2, magnetic resonance abnormalities may lag behind symptoms. Other tests that may be helpful to elucidate the cause of the transverse myelitis include basic chemistries and CBC counts, antinuclear antibody, erythrocyte sedimentation rate, rapid plasmin reagin, Lyme titers, titers for viruses such as West Nile, polio, hepatitis, Epstein Barr virus, cytomegalovirus, HIV, and mycoplasma antibody and bacterial cultures. Lumbar puncture most commonly demonstrates lymphocytosis and elevated protein, but checking the opening pressure, glucose, immunoglobulins, and protein electrophoresis is warranted to narrow the differential diagnosis. Cultures and polymerase chain reaction sometimes make a specific identification. When the diagnosis of transverse myelitis is strongly suspected in the ED, an admission to neurologic specialists for completion of the evaluation is necessary to specify the cause of the transverse myelitis and obtain further MRI imaging, serial examinations, and culture results.

The most common therapy for transverse myelitis is supportive. Immunoglobulin infusion and corticosteroids have been mainstay therapy in the past; however, one recent study has questioned their utility. Some authors advocate that corticosteroids may only be helpful if acute swelling of the cord is suspected, whereas others strongly argue that early steroid treatment speeds recovery time and morbidity. Because of the dichotomy of academic opinion, until more clinical trials allow for a consensus for standard of steroids in the treatment of transverse myelitis, the addition of steroids should be used at the clinical discretion of the clinician while taking into account the risk-benefit analysis of each patient. When a treatable infection is diagnosed, in particular schistosomiasis or Lyme disease, disease-specific antimicrobials may be useful. Consider Lyme disease in the appropriate setting, keeping in mind that patients presenting with erythema migrans will often be seronegative early in the course and that cephalexin is not an appropriate antibiotic choice. A therapeutic alternative for severe transverse myelitis may be plasma exchange therapy to remove presumptive factors that may contribute to autoimmune-mediated inflammation, and in cases of HTLV or mycoplasma associated transverse myelitis. Recovery depends on the amount of cord necrosis; independent ambulation occurs in 50% of pediatric and 35% of adult cases, and improvement may continue beyond 6 months.

Transverse myelitis is an often obscure disorder that may be diagnosed in the ED in certain clinical scenarios. The emergency physician should include transverse myelitis into the differential diagnosis of back pain, especially when associated with neurological deficits, and obtain an early MRI, both to rule out compression and rule in transverse myelitis. Although the presentation of transverse myelitis, as demonstrated above, can be variable, early diagnosis and treatment can minimize its accompanying morbidity and mortality, the cornerstone of the emergency medicine practice.

Supervising editor: William G. Barsan, MD

Funding and support: The authors report this study did not receive any outside funding or support.

Publication dates: Received for publication February 28, 2005. Revision received April 29, 2005. Accepted for publication May 2, 2005. Available online July 1, 2005.

Reprints not available from the authors.

Address for correspondence: Heather S. Hammerstedt, MD, One Deaconess Road- West CC2, Department of Emergency Medicine, Beth Israel Deaconess Medical Center, Boston MA 02215; 617-754-2323, fax 617-754-2350; E-mail hhammers@bidmc.harvard.edu.

REFERENCES