Clinical Evaluation and Treatment of Transverse Myelitis

Case Presentation

A 38-year-old right-handed white woman with no significant past medical history presents to her internist’s office with a 3-day history of numbness in her lower extremities. She reports being well until 3 days prior, when she noted tingling and numbness in her feet bilaterally. Over the next 72 hours, these sensations ascended to involve her calves and thighs and now are up to her waistline. Since the morning, she has noted that she is dragging her right leg and has difficulty emptying her bladder. She denies any headache, vision changes, or changes in her upper extremities.

She denies recent travel, infections, or vaccinations. She denies recent trauma. She has never had an event like this before and considers herself to be “very healthy.” She has no significant past medical history but had an appendectomy at age 9. She is married currently, with one daughter. She is a nonsmoker and consumes occasional alcohol. She denies any history of neurologic disease. She denies recreational drug use. A complete 10-topic review of systems was performed, and there were no pertinent positives other than what was noted above.

On exam, her blood pressure was 132/78, pulse was 84, and she was afebrile, with an oral temperature of 98.6°F Fahrenheit. She was saturating 100% on room air. She was anxious but not in distress. Head, ears, eyes, nose, and throat exams were normal. Heart sounds were regular, with no murmurs, rubs, or gallops. Chest was clear to auscultation bilaterally. Her lower abdomen was slightly tender over her pelvis, and she felt full in the midline.

On neurologic exam, she was awake and alert with normal speech and language. She was oriented to situation and place, including her current city and state. She knew the current president of the United States. Her immediate, recent, and past memory was excellent. Pupils were equal in size, round, and reactive. No afferent pupillary defect was noted. Funduscopic exam was normal. Extraocular movements were full. There was no nystagmus and no internuclear ophthalmoplegia. Face sensation was normal. Face was symmetric. Hearing was intact. Tongue and uvula were midline. Sternocleidomastoid strength was 5/5 bilaterally. The upper extremities had normal tone and 5/5 strength throughout. There was no drift. The lower extremities were notable for 4/5 strength at the right iliopsoas, 4-5 at the right quadriceps and hamstrings, and 3/5 strength at the right-ankle dorsiflexion. The left lower extremity had 5/5 strength throughout except for the iliopsoas, which was 4+/5. Sensory exam was notable for normal proprioception and vibration throughout. There was decreased temperature and pinprick in her legs bilaterally, with a T10 sensory level noted. Deep tendon reflexes were 2s at the biceps, triceps, and brachioradialis. They were absent at the patellas and ankles. Toes were mute.
Finger-to-nose is intact, but the patient cannot complete heel-to-shin. Her gait is notable for circumduction of the right leg and a right-foot drop.

The patient is sent to the hospital for admission and testing. At admission, a Foley catheter is placed and one liter of clear urine drained from her bladder. An MRI of the brain and spine is obtained, and you personally view the films. You interpret the brain MRI as normal and note that the thoracic MRI showed increased signal at the T9 region. CBC and comprehensive metabolic panel are normal and a lumbar puncture is performed. The patient is diagnosed with transverse myelitis (TM). You explain to the patient that TM can be idiopathic or secondary to a variety of conditions. You also explain that treatment of TM includes steroids and sometimes plasma exchange. The patient has opted for high-dose methylprednisolone for 5 days, and you prescribe this dose.

Questions

1. Routine testing for patients with TM may include:
   A. MRI of the brain
   B. Aquaporin-4 antibodies
   C. Oligoclonal band (OCB) testing with CSF
   D. MRI of the spine
   E. All of the above

   **The correct answer is E.** Testing for aquaporin-4 antibodies should be considered useful to determine the cause of TM in patients presenting with clinical features of acute complete TM (ACTM) (Level B). CSF examination for cells and OCBs may be considered useful to determine the cause of the TM syndrome (Level C). Brain MRI characteristics consistent with those of MS may be considered useful to predict conversion to multiple sclerosis (MS) after a first episode of partial TM (Level C). Longer spinal lesions extending over more than 3 vertebral segments may be considered useful in determining neuromyelitis optica vs MS (Level C).

2. Acute treatment for TM includes steroids in most cases. Options for alternative and adjunctive therapies are numerous. Which of the following therapies has the best class of evidence to support its use in TM?
   A. Plasma exchange
   B. Intravenous immunoglobulin
   C. Mitoxantrone
   D. Cyclophosphamide
   E. Rituximab

   **The correct answer is A.** Only plasma exchange has Class II evidence supporting its use in acute fulminant CNS demyelinating diseases.
Diagnosis Coding

This report documents the diagnosis “transverse myelitis” (TM). Because this diagnosis statement does not say the TM is “acute,” the appropriate ICD-9-CM code is the “default” code for TM:

323.82 Encephalitis, myelitis, encephalomyelitis. Other causes of myelitis, Transverse myelitis, NOS

Had the diagnosis been “acute transverse myelitis,” the ICD-9-CM code would be:

341.20 Other demyelinating diseases of the CNS, Acute (transverse)

myelitis NOS

Once testing has been done and a cause of the TM is found, a more specific ICD-9-CM code should be used. Available ICD-9-CM codes for the variations of TM are:

323.02 Myelitis in viral diseases classified elsewhere

Excludes: myelitis (in):
   Herpes simplex (054.74)
   Herpes zoster (053.14)
   Poliomyelitis (045.0-045.9)
   Rubella (056.01)
   Other viral diseases of the central nervous system (049.8-049.9)

Use the codes in parentheses for myelitis associated with the diseases listed in the excludes notes

323.1 Encephalitis, myelitis, and encephalomyelitis in rickettsial diseases classified elsewhere

Code also underlying disease (080-083.9)

323.2 Encephalitis, myelitis, and encephalomyelitis in protozoal diseases classified elsewhere

Code also underlying disease, as:
   Malaria (084.0-084.9)
   Trypanosomiasis (086.0-086.9)

323.41 Other encephalitis, myelitis, and encephalomyelitis due to other infections classified elsewhere

Excludes: encephalitis (in):
   Meningococcal (036.1)
   Syphilis:
      NOS (094.81)
      Congenital (090.41)
   Toxoplasmosis (130.0)
   Tuberculosis (013.6)
Meningoencephalitis due to free-living ameba [Naegleria] (136.29)

323.42 Other myelitis due to other infections classified elsewhere

323.52 Myelitis following immunization procedure
Myelitis postimmunization or postvaccinal
Use E code to identify vaccine

323.63 Postinfectious myelitis
Code first underlying disease

323.72 Toxic myelitis
Code first underlying cause

323.82 Other causes of myelitis
Transverse myelitis NOS

341.0 Neuromyelitis optica

The following codes may not be used for any of the conditions described in the codes above:

341.20 Acute (transverse) myelitis NOS

341.21 Acute (transverse) myelitis in conditions classified elsewhere
Code first underlying condition

341.22 Idiopathic transverse myelitis

ICD-9-CM codes will be used until October 1, 2013, when ICD-10-CM codes will be mandatory for reporting diagnosis codes on claims. Coding for TM is very different in ICD-10-CM. Unlike that of ICD-9-CM, the “default” code for TM is:

G37.3 Acute transverse myelitis in demyelinating disease of the central nervous system
Code first underlying disease such as:
    human immunodeficiency virus [HIV] disease (B20)
    poliovirus (A80.-)
Acute transverse myelitis NOS
Acute transverse myelopathy
Excludes1: multiple sclerosis (G35)
    Neuromyelitis optica [Devic] (G36.0)
Other ICD-10-CM codes related to TM are:

G36.0  Neuromyelitis optica

G37.4  Subacute necrotizing myelitis of the central nervous system

A17.82  Tuberculous meningoencephalitis
     Tuberculous myelitis

A52.14  Late syphilitic encephalitis
B00.82  Herpes simplex myelitis
B01.12  Varicella myelitis
B02.24  Post-herpetic myelitis
G05.4  Myelitis in diseases classified elsewhere

Excludes1:
  adenoviral encephalitis, myelitis and encephalomyelitis (A85.1)
  congenital toxoplasmosis encephalitis, myelitis and
  encephalomyelitis (P37.1)
  cytomegaloviral encephalitis, myelitis and encephalomyelitis
  (B25.8)
  encephalitis, myelitis and encephalomyelitis (in) measles (B05.0)
  encephalitis, myelitis and encephalomyelitis (in) systemic lupus
  erythematosus (M32.19)
  enteroviral encephalitis, myelitis and encephalomyelitis (A85.0)
  eosinophilic meningoencephalitis (B83.2)
  herpesviral [herpes simplex] encephalitis, myelitis and
  encephalomyelitis (B00.4)
  listerial encephalitis, myelitis and encephalomyelitis (A32.12)
  meningococcal encephalitis, myelitis and encephalomyelitis
  (A39.81)
  mumps encephalitis, myelitis and encephalomyelitis (B26.2)
  postchickenpox encephalitis, myelitis and encephalomyelitis
  (B01.1-)
  rubella encephalitis, myelitis and encephalomyelitis (B06.01)
  toxoplasmosis encephalitis, myelitis and encephalomyelitis
  (B58.2)
  zoster encephalitis, myelitis and encephalomyelitis (B02.0)

Use the codes in parentheses for TM associated with diseases listed in the
excludes notes

G04.89  Other myelitis
     This is also the code for postvaccinal, postinfectious, and postimmunization TM

G92  Toxic encephalopathy
     Use this code for toxic TM
**Evaluation and Management Coding**

The neurologist sees the patient in his office and admits the patient to the hospital on the same day. The neurologist can render only one evaluation and management (E&M) code for this day, and it would be an initial hospital care day encounter, code 99223. This is based on a comprehensive history and physical and high-complexity medical decision making.

**Procedure Coding**

The patient had a lumbar puncture. The correct code for this procedure is:

62270  Spinal puncture, lumbar, diagnostic

CPT © 2011 American Medical Association. All rights reserved. CPT is a registered trademark of the American Medical Association.


The AAN develops these clinical case examples as educational tools for neurologists and other health care practitioners. You may download and retain a single copy for your personal use. Please contact guidelines@aan.com to learn about options for sharing this content beyond your personal use.

**Disclaimer**

This statement is provided as an educational service of the American Academy of Neurology. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved. The views expressed here are those of the authors and do not represent those of the National Institutes of Health or any other part of the US Government. No official support or endorsement by the National Institutes of Health is intended or should be inferred.

© 2011 American Academy of Neurology