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A 55-Year-Old Woman With an Abrupt Onset of Weakness

A 55-year-old woman presented to the emergency department with the complaint of neck and shoulder pain. This pain was accompanied with acute bilateral lower-extremity weakness that occurred during the same time. She stated that the neck and shoulder pain had been ongoing for the last 2 days and was worsening. She noted earlier in the day while walking to the bathroom that she had a sudden worsening of her symptoms. She started to collapse to the ground, but her husband assisted her, preventing any traumatic fall. After this episode, she developed numbness and paresthesias in all 4 of her extremities. Her husband carried her downstairs and called 911. The patient described the shoulder and neck pain as an 8 of 10 in intensity. It was sharp and stabbing in nature and radiated down her right arm distally. Her symptoms did not improve, and any type of activity seemed to make her symptoms worse. Her past medical history was significant for a diagnosis of optic neuritis, and she was legally blind. Her primary care physician had recently started evaluating her for multiple sclerosis. A referral to a neurologist was currently pending. No other significant relevant history was obtained at the time of the initial evaluation.

On her physical examination, the patient had an initial blood pressure of 133/89, pulse of 81, an oral temperature of 99.5°F, and pulse oximetry of 100% on room air. She appeared well nourished and appeared her stated age. She was in no apparent distress. There were no acute physical findings other than altered muscle strength. She had decreased muscle function at the biceps, deltoids, and wrist with muscle strength at only 3 of 5. This reflects that she was only able to move those muscle groups against gravity but not against resistance. She had muscle strength at 5 of 5 bilaterally in the lower extremities. Her deep tendon reflexes were 2 of 4 without clonus bilaterally in the upper and lower extremities. She had flexor plantar responses (downgoing) bilaterally. The patient's symptoms prompted concern for possible spinal cord inflammation and injury. The sending facility did not have magnetic resonance imaging (MRI) equipment. Because of the significant alteration of strength, a local flight team was activated to transfer this patient to a neurologic specialty center. There was an initial concern for the possibility of worsening loss of muscle in the thorax leading to respiratory impairment.

Upon arrival at the sending hospital, the flight crew prepared the patient for transfer. The neurosurgeon at the receiving facility requested that no steroid medications be administered. They asked that the patient's neurologic status be monitored during transport. A neurologic examination was performed before departure and during transport. The patient remained awake and alert. Her vital signs remained stable. Her neurologic examination revealed weakness at 3 of 5 in her upper extremities before being loaded into the aircraft.

During the flight, the patient had difficulty moving her extremities against gravity. The flight crew relayed this information to the receiving facility. The patient was very anxious about the flight, and she was treated with midazolam 2 mg intravenously. She developed light-headedness and nausea. This was treated with ondansetron with resolution. Throughout the remainder of the flight, her vital signs remained stable.

Upon arrival to the receiving tertiary facility, the patient was immediately evaluated by the emergency department physician. Her neurologic examination was found to be consistent with the examination performed before departure from the sending facility. She had muscle strength at 3 of 5 in the bilateral deltoid, biceps, and wrist. The remainder of her neurologic examination was identical to that documented before her transfer. Her light-headedness and nausea had subsided. The neurosurgeon evaluated this patient in the emergency department. A head computed tomographic scan of the brain was completed because of her near syncopal episode. This did not show evidence of acute intracranial bleeding or ischemic stroke. An electrocardiogram was completed, which showed a normal sinus rhythm without evidence of ischemic changes. Her laboratory values were evaluated and were only significant for a mildly elevated white blood cell (WBC) count of 12.83 and mild anemia with a hemoglobin level of 10.3. Her urinalysis revealed a bacterial urinary tract infection. She was treated with ceftriaxone intravenously. Her remaining laboratory values were unremarkable.

Because of the acute onset of her symptoms and neurologic findings, MRI of the brain and cervical spine were obtained. No acute intracranial abnormality was visualized. However, the MRI of the cervical spine showed increased T2 signal hyperintensities of the spinal cord consistent with myelitis at the levels of C1/2 through C7. Bulging discs with mild stenosis at C5/6 and C6/7 were also observed (Figs. 1 and 2).

The patient's MRI showed T2 signal hyperintensities associated with mild areas of enhancement at the upper cord. These findings are consistent with myelitis of C1/2 to C7. There were also bulging discs with mild stenosis at C5/6 and C6/7. The patient was diagnosed with transverse myelitis and started on high-dose methylprednisolone.

Acute transverse myelitis (ATM) is a broad clinical syndrome that is characterized by acute or subacute spinal cord

dysfunction below the level of a lesion in the spinal cord. This manifests as motor dysfunction, a sensory level, and autonomic dysfunction.[1,2] ATM is an inflammatory disease process of the spinal cord that can be the result of an underlying disease or idiopathic in nature. Use of the term "transverse" implies involvement of the whole cross-sectional area of the spinal cord (acute complete transverse myelitis [ACTM]), but the definition has been expanded to include neurologic dysfunction of varying degrees and at 1 or more levels (acute partial transverse myelitis [APTM]).[1,3] Manifestations of dysfunction must be attributed to the spinal cord and are typically bilateral but they can be asymmetric.[1,2]

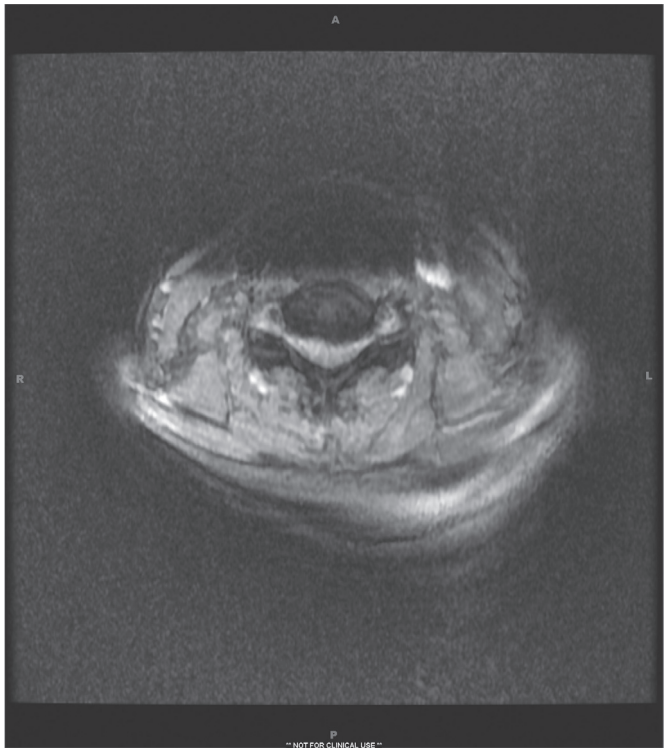
Acutely, muscle tone and deep tendon reflexes may be diminished or even absent, but over time, muscle spasticity, hyperreflexia, and extensor (upgoing) response to plantar reflex testing become evident.[1] By the time of maximal debility, half of the individuals have lost all movement in their legs, virtually all have bladder dysfunction, and the majority have paresthesias numbness or bandlike dysesthesias.[2,4-9] Autonomic dysfunction is almost always present. It may manifest as difficulty voiding, urinary urgency, bowel or bladder incontinence, sexual dysfunction, incomplete evacuation, constipation, or even cardiovascular and thermoregulatory dysfunction.[1,2,10]

There are multiple identified diseases and syndromes that can cause subsequent findings similar to those in ATM and transverse myelitis (TM)-like syndromes. These include parainfectious myelitis, myelitis caused by systemic disease (systemic lupus erythematosus, Sjögren syndrome, antiphospholipid syndrome, Behcet disease, and other mixed connective tissue diseases).[1,3] Additionally, ATM syndromes can occur as a result of treatments for other ailments including delayed radiation myelopathy and as a result of receiving chemotherapeutic agents. Multiple other etiologies exist; however, there are many patients to whom a specific etiology cannot be attributed.[1,2,11]

Multiple sclerosis (MS), neuromyelitis optica (NMO), and acute disseminated encephalomyelitis are acquired demyelinating disorders that can cause TM.[1] TM in individuals with MS typically presents as partial myelitis with manifestations of a sensory nature and evolve slowly, over a period of 1 to 3 weeks or longer.[1] The lesions are usually asymmetrically placed, less than 2 segments in length, and often have a predilection for the cervicothoracic region of the spinal cord.[1,2,12] Patients with ATM and a history of clinically significant optic neuritis meet the criteria for NMO. They are more likely to present with ACTM, have recurrent and progressive disease, and have less complete recovery.[2,13] Spinal cord lesions related to NMO tend to be centrally located cervical lesions and be longitudinally extensive, meaning they cover more than 3 vertebral segments. Additionally, TM associated with MS and NMO often have brain lesions on MRI.[2,12] Idiopathic TM more commonly has lesions on the thoracic spinal cord.[3,11]

Parainfectious etiologies associated with TM mainly include viruses (especially hepatitis) and bacteria (*Mycoplasma pneumonea*, *Campylobacter jejuni*, and several others), but it can occasionally be caused by fungal and parasitic infections (pinworm).[1,14] The infections precede the neurologic manifestations, and the antecedent infection usually resolves before manifestations of TM begin.[1] It is thought that the bacterial and viral infections cause postinfectious, immune-mediated, inflammatory damage. Fungal and parasitic agents may cause ATM by direct pathogenic effects.[1,14]

Figure 1. MRI of the Cervical Spine



The Transverse Myelitis Consortium Working Group proposed criteria for diagnosing idiopathic ATM in 2002. They require bilateral sensory, motor, or autonomic dysfunctions attributable to the spinal cord with a clearly defined sensory level. Compressor etiologies must be excluded, and evidence of spinal cord inflammation must be proven by cerebrospinal fluid (CSF) pleocytosis, elevated immunoglobulin (Ig) G index, or gadolinium enhancement on MRI. Symptoms should also progress to a nadir between 4 hours and 21 days. In addition to these criteria, exclusion criteria for idiopathic TM include a history of radiation to the spine within the last 10 years, deficits clearly attributed to anterior spinal artery thrombosis, arteriovenous malformation on the surface of the spinal cord, serologic or clinical evidence of connective tissue disease, central nervous system manifestations of infection, brain MRI suggestive of MS, or clinically apparent optic neuritis.[2]

Because ATM is a rare neurologic condition with only 1 to 4 new cases per million people per year,[2-4] it requires a high degree of suspicion to diagnose. There are few helpful demographic features because there is no geographic variation in