PERSPECTIVE

Spinal cord disorders encompass a wide range of pathologic entities and affect all age groups. Some spinal cord disorders may have catastrophic outcomes if they are not recognized early in the clinical course. The ultimate neurologic outcome with many of these disorders may depend on expeditious recognition in the emergency department (ED), with appropriate initial investigations, neuroimaging, management, and consultation for definitive therapy. Early presentations of spinal cord syndromes are frequently subtle, and diagnosis may be difficult if not impossible in the early stages of disease. As with many disease processes affecting the nervous system, correct diagnosis and appropriate management require knowledge of the anatomic organization of the spinal cord and skill in taking the history and in performing the neurologic examination.

This chapter generally is concerned with nontraumatic processes affecting the spinal cord and its vascular supply as well as processes compressing the spinal cord. Spinal cord trauma is discussed in Chapter 43.

PRINCIPLES OF DISEASE

Anatomy

In adults, the spinal cord is approximately 40 cm long and extends from the foramen magnum, where it is continuous with the medulla oblongata, to the body of the first or second lumbar vertebra. Like the brain, the spinal cord is covered by three meningeal layers: the inner pial layer, the arachnoid, and the outer dural layer. At its lower end, the spinal cord tapers into the conus medullaris, where several segmental levels are represented in a small area. The lumbar and sacral nerve roots form the cauda equina as they descend caudally in the thecal sac before exiting the spinal canal at the respective foramina. The non-neural filum terminale runs from the tip of the conus and inserts into the dura at the level of the second sacral vertebra.

Two symmetrical enlargements of the spinal cord contain the segments that innervate the limbs. The **cervical enlargement** (cord level C5 to T1) gives rise to the brachial plexus and subsequently to the peripheral nerves of the upper extremity. The **lumbar enlargement** (L2 to S3) gives rise to the lumbosacral plexus and peripheral nerves of the lower extremity. The space surrounding the spinal cord within the spinal canal is reduced in the area of the enlargements, potentially leaving the cord more vulnerable to compression in these regions. At each segmental level, anterior (ventral) and posterior (dorsal) roots arise from rootlets along the anterolateral and posterolateral surfaces of the cord. At each level, the anterior root conveys the outflow of the motor neurons in the anterior horn of the spinal cord, and the posterior root contains sensory neurons and fibers that convey sensory inflow.

The arterial supply of the spinal cord is derived primarily from two sources. The single anterior spinal artery arises from the paired vertebral arteries. This anterior spinal artery runs the entire length of the cord in the midline anterior median sulcus and supplies roughly the anterior two thirds of the spinal cord. Blood supply to the posterior third of the spinal cord is derived from the smaller paired posterior spinal arteries. The anterior and the posterior spinal arteries receive segmental contributions from radicular arteries, the largest being the radicular artery of Adamkiewicz, which typically originates from the aorta between T8 and L4. The venous drainage of the cord largely parallels the arterial supply.

The internal anatomy of the spinal cord is divided into central gray matter, which contains cell bodies and their processes, and surrounding white matter, where the ascending and descending myelinated fiber tracts are located. These fiber tracts are organized into discrete bundles; the ascending tracts convey sensory information, and the descending tracts convey the efferent motor impulses and visceral innervation.

For clinical purposes, neuroanatomy of the spinal cord may be greatly simplified, as depicted in Figure 106-1. Major ascending sensory tracts are represented on the right side of the figure, with motor tracts on the left side. The posterior columns carry afferent ascending proprioceptive and vibratory information on the ipsilateral side of the cord to the area stimulated; decussation of these fibers occurs in the medulla so that contralateral cortical representation is consistent. The lateral spinothalamic tract conveys afferent information about pain and temperature in a portion of the lateral column of white matter. (Tracts are named with the point of origin first; the spinothalamic tract, for example, arises in the spinal cord and travels to the thalamus.) The tract is laminated so that sacral fibers are represented most laterally. Crossing of fibers from this tract occurs near the level of entry of the spinal nerve; a cord lesion affecting only one lateral spinothalamic tract results in decreased or absent pain and temperature perception below the level of injury on the contralateral side of the body.

For clinical purposes, the major descending motor tract is represented in the lateral corticospinal tract (which, as the name implies, originates in the cortex and descends to the spinal cord). This tract also is anatomically organized, with efferent motor axons to the cerebral area located medially and the sacral efferent axons located laterally. Decussation of this descending tract occurs in the medulla. The cell bodies of the lower motor neurons (anterior horn cells) are in the ventral (anterior) portion of the gray matter of the spinal cord.
usually carries a better prognosis than a complete lesion. 1

voluntary toe movement suggest a partial cord lesion, which
tone or voluntary rectal sphincter contraction, and even slight
persistent perineal sensation (sacral sparing), reflex rectal sphinc-
Any evidence of preserved cord function below the level of injury
more than 24 hours, functional recovery does not occur in 99%.3,4

patients with acute complete transverse syndromes that persist for
hemorrhage, and entities causing extrinsic compression. In
Other causes of acute complete cord syndrome include infarction,

CORD SYNDROMES

CLASSIFICATION OF SPINAL CORD SYNDROMES

The anatomic organization of the spinal cord lends itself to a cor-
responding anatomic-pathophysiologic classification of cord dys-
function. Any of the different anatomic syndromes may be the
final clinical picture of a variety of clinical processes. The syn-
dromes frequently exist in partial or incomplete forms, adding to
the diagnostic difficulty.

Complete (Transverse) Spinal Cord Syndrome

Complete spinal cord lesions may be manifested as either acute or
subacute pathologic processes. A complete spinal cord lesion is
defined as a total loss of sensory, autonomic, and voluntary motor
innervation distal to the spinal cord level of injury. Reflex responses
mediated at the spinal level, such as muscle stretch (deep tendon)
reflexes, may persist, although they also may be absent or abnor-
mal. Autonomic dysfunction may be manifested acutely with
hypotension (neurogenic shock) or priapism. The most common
cause of the complete transverse cord syndrome is trauma, although this anatomic syndrome is nonspecific as to etiology.1,2

Other causes of acute complete cord syndrome include infarction,
hemorrhage, and entities causing extrinsic compression. In
patients with acute complete transverse syndromes that persist for
more than 24 hours, functional recovery does not occur in 99%.3,4

Any evidence of preserved cord function below the level of injury
denotes a partial rather than a complete lesion. Signs such as
persistent perineal sensation (sacral sparing), reflex rectal sphinc-
ter tone or voluntary rectal sphincter contraction, and even slight
voluntary toe movement suggest a partial cord lesion, which
usually carries a better prognosis than a complete lesion.1

Spinal shock refers to the loss of muscle tone and reflexes with
complete cord syndrome during the acute phase of injury. The
intensity of the spinal shock increases with affected spinal cord
level.1 Spinal shock typically lasts less than 24 hours but has been
reported occasionally to last days to weeks.5,6 A marker of spinal
shock is loss of the bulbocavernosus reflex, which is a normal
cord-mediated reflex that may be preserved in complete cord
lesions. The bulbocavernosus reflex involves involuntary reflex
contraction of the anal sphincter in response to a squeeze of

Brown-Séquard Syndrome

Brown-Séquard syndrome, first described in 1846 by the one physi-
cian for whom it is named,7 is the result of an anatomic or
functional hemisection of the spinal cord. Usually associated with
penetrating injuries,8 Brown-Séquard syndrome also may be seen
with compressive or intrinsic lesions. The syndrome has been
reported in association with spinal cord tumors, spinal epidural
hematoma, vascular malformations, cervical spondylosis, degen-
erative disk disease, herpes zoster myelitis, and radiation injury
and as a complication of spinal instrumentation.9,10 The syn-
drome in its pure form is characterized by ipsilateral loss of motor
function and proprioception or vibration, with contralateral loss
of pain and temperature sensation below the spinal cord level of
injury. Because fibers associated with the lateral spinothalamic
tract ascend or descend one or two spinal cord segments before
crossing to the contralateral side, ipsilateral anesthesia (pain and
temperature modalities) may be noted one or two segments above
the lesion, although this observation is variable. Most patients
with Brown-Séquard syndrome incur only partial sensory and
motor impairment, and the classic pattern is not seen.9,10,11

Brown-Séquard syndrome carries the best prognosis of any of the
incomplete spinal cord syndromes. Fully 80 to 90% of patients
with Brown-Séquard syndrome regain bowel and bladder func-
Central cord syndrome

Variable position and vibration sense loss
Contralateral pain and temperature sensation loss
Motor loss ipsilateral to cord lesion
Variable

Brown-Séquard syndrome

Loss of pin and touch sensation
Vibration, position sense preserved
Motor loss or weakness below cord level
Variable

Anterior cord syndrome

Loss of sensation below level of cord injury
Loss of voluntary motor function below cord level
Sphincter control lost

Transverse cord syndrome—complete

Conus medullaris syndrome

Saddle anesthesia may be present, or sensory loss may range from patchy to complete transverse pattern
Weakness may be of upper motor neuron type
Sphincter control impaired

Cauda equina syndrome

Saddle anesthesia may be present, or sensory loss may range from patchy to complete transverse pattern
Weakness may be of lower motor neuron type
Sphincter control impaired

Table 106-1 Spinal Cord Syndromes

<table>
<thead>
<tr>
<th>SYNDROME</th>
<th>SENSORY</th>
<th>MOTOR</th>
<th>SPHINCTER INVOLVEMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central cord syndrome</td>
<td>Variable</td>
<td>Upper extremity weakness, distal &gt; proximal</td>
<td>Variable</td>
</tr>
<tr>
<td>Brown-Séquard syndrome</td>
<td>Ipsilateral position and vibration sense loss</td>
<td>Contralateral pain and temperature sensation loss</td>
<td>Motor loss ipsilateral to cord lesion</td>
</tr>
<tr>
<td>Anterior cord syndrome</td>
<td>Loss of pin and touch sensation</td>
<td>Motor loss or weakness below cord level</td>
<td>Variable</td>
</tr>
<tr>
<td>Transverse cord syndrome—complete</td>
<td>Loss of sensation below level of cord injury</td>
<td>Loss of voluntary motor function below cord level</td>
<td>Sphincter control lost</td>
</tr>
<tr>
<td>Conus medullaris syndrome</td>
<td>Saddle anesthesia may be present, or sensory loss may range from patchy to complete transverse pattern</td>
<td>Weakness may be of upper motor neuron type</td>
<td>Sphincter control impaired</td>
</tr>
<tr>
<td>Cauda equina syndrome</td>
<td>Saddle anesthesia may be present, or sensory loss may range from patchy to complete transverse pattern</td>
<td>Weakness may be of lower motor neuron type</td>
<td>Sphincter control impaired</td>
</tr>
</tbody>
</table>

Anterior Cord Syndrome

Anterior cord syndrome is characterized by loss of motor function, pinprick, and light touch below the level of the lesion with preservation of posterior column modalities, including some touch, position, or vibratory sensation. Although most reported cases of anterior spinal syndrome follow aortic surgery, the syndrome also may occur after severe hypotension, infection, myocardial infarction, vasospasm from drug reaction, and aortic angiography. The anatomic lesion may be caused by a cervical hyperflexion injury resulting in a cord contusion or by protrusion of bone fragments or herniated cervical disk material into the spinal canal. Rarely, it is produced by laceration or thrombosis of the anterior spinal artery or a major radicular feeding vessel. Patients present with characteristic mixed motor and sensory neurologic findings as noted before. Functional recovery varies; most improvement occurs during the first 24 hours, but little improvement is expected thereafter. Although anterior cord lesions from ischemia usually are incomplete, patients without motor function at 30 days have little or no likelihood of regaining any motor function by 1 year. Overall, only 10 to 20% of patients with this entity regain some muscle function, and even in this group, there is little power or coordination.

Conus Medullaris and Cauda Equina Syndromes

The separation of conus medullaris and cauda equina lesions in clinical practice is difficult because the clinical features of the
disorders overlap. In addition, a combined lesion may occur that masks clear clinical symptoms or signs of either an upper or a lower motor neuron type of injury. The conus medullaris is the terminal end of the spinal cord, located at approximately the L1 level in adults. The conus medullaris syndrome may involve disturbances of urination (usually from a denervated, autonomic bladder that is manifested clinically with overflow incontinence) and sphincter impairment or sexual dysfunction. Sensory involvement may affect the sacral and coccygeal segments, resulting in saddle anesthesia. Pure lesions of the conus medullaris are rare.19 Upper motor neuron signs, such as increased motor tone and abnormal reflexes, may be present, but their absence does not exclude the syndrome. The conus medullaris syndrome can be caused by central disk herniation, neoplasm, trauma, or vascular insufficiency. Because the conus is such a small structure, with lumbar and sacral segments represented in a small area, a lesion usually causes bilateral symptoms. This finding may help distinguish lesions of the conus from lesions of the cauda equina, which often are unilateral.19

The **cauda equina** (Latin for “horse’s tail”) is the name given to the lumbar and sacral nerve roots that continue on within the dural sac caudal to the conus medullaris. Not a true “cord syndrome,” cauda equina syndrome represents dysfunction at the level of nerve roots, but the anatomic clustering of nerve roots within the lumbar dural sac allows injury to several nerve roots to occur simultaneously. The etiologic lesion in the cauda equina syndrome usually is a midline rupture of an intervertebral disk, most commonly at the L4-5 level. Tumors and other compressive masses also may cause the syndrome. As in the conus medullaris syndrome, patients generally present with progressive symptoms of fecal or urinary incontinence, impotence, distal motor weakness, and sensory loss in a saddle distribution. Muscle stretch reflexes also may be reduced. Urinary retention is the most consistent finding, with a sensitivity of 90%.20 Low back pain may or may not be present.

**CLINICAL FEATURES**

**History**

Weakness, sensory abnormalities, and autonomic dysfunction are the cardinal manifestations of spinal cord dysfunction. The tempo and degree of impairment often reflect the disease process. Past medical history is vital because a history of coagulopathy or other systemic processes may be elicited. A history of cancer should suggest the possibility of metastatic disease. Recent trauma raises the possibility of vertebral fracture or disk protrusion.

**Physical Examination**

The physical examination pertinent to spinal cord dysfunction involves testing in three areas: (1) motor function, (2) sensory function, and (3) reflexes. Each component is best tested with the anatomic organization of the spinal cord in mind to help determine the level of the spinal cord dysfunction.

**Motor Function**

Testing of motor function encompasses examination of muscle bulk, tone, and strength. Muscle bulk is easily examined in large motor groups, such as the thigh or calf muscles, the biceps, and the triceps. Inspection of the intrinsic hand muscles also may be helpful for determination of muscle bulk; wasting may be evident as hollowed or recessed regions of the hand. Decreased mass, asymmetry, or fasciculations should be noted. Tone is tested with repeated passive knee, elbow, or wrist flexion, with the examiner feeling for abnormally increased or decreased resistance. Rapid pronation-supination of the forearm is another useful method to assess tone. Increased tone may indicate spasticity or an upper motor neuron lesion, whereas decreased tone corresponds with lower motor neuron, motor endplate, or muscle problems. Finally, motor strength is graded in the upper and the lower extremities. Motor grading for the neurologic examination is relatively straightforward. Note that a tremendous gradient of strength is within the fourth grade of the scale. Scored on a scale of 0 to 5, neuromuscular functioning is graded as follows:

0. No firing of the muscle is present.
1. The muscle fires but is unable to move the intended part.
2. The muscle is able to move the intended part with gravity eliminated.
3. The muscle is able to move the intended part against gravity.
4. The muscle is able to move the intended part, but not at full strength.
5. Full muscle strength is present.

A rectal examination is performed to assess voluntary sphincter contraction, resting tone, and, as described previously, the bulbocavernous reflex. Although it is not commonly thought of as a physical examination maneuver, a post-void residual urine volume may be important for assessment of bladder function. A post-void residual volume of more than 100 to 200 mL in a patient without prior voiding difficulty might suggest bladder dysfunction of neurologic cause.

**Sensory Function**

Sensory testing requires a cooperative patient and an attentive examiner. The spinal cord–related modalities that may be clinically useful include testing for pinprick and light touch (contralateral lateral spinothalamic tract) and proprioception (ipsilateral posterior column). Assessment of the patient’s response to pinprick, light touch, and proprioception in all four extremities is necessary if a neurologic injury is suspected. Testing of sacral dermatomes is an important part of the examination in some patients. As previously noted, sacral sparing is an important finding indicating that spinal cord dysfunction may be incomplete. The sensory fibers from sacral dermatomes are more peripherally located in the ascending fiber bundles; central or partial cord lesions may ablate sensation in the extremities yet allow some perception of sensation in the sacral area.

**Reflexes**

Muscle stretch (deep tendon) reflexes may be tested rapidly at the bedside. Responses are graded on a scale of 0 to +4+, with 2 being normal. Hyperactive reflexes suggest upper motor neuron disease (affecting the neurons or their outflow from the brain or spinal cord), as do sustained clonus and Babinski’s sign. The absence of these reflex changes does not constitute evidence that a myelopathy is not present. In fact, one small series noted a low incidence of extensor plantar responses as well as a lack of hyperreflexia in patients presenting to the ED with acute or progressive cord compression or myelopathies.21 Reflexes also may be diminished or absent when sensation is lost or when spinal shock is present. Diseases of muscles or neuromuscular junctions also may decrease reflexes. In acute cord injury, reflexes may be diminished in the acute phase. The bulbocavernous reflex may be helpful in this assessment.

**DIAGNOSTIC STRATEGIES**

Historical or physical examination findings that suggest spinal cord dysfunction prompt further investigations. The basic strategy is to detect or to exclude extrinsic compressive lesions or other potentially treatable entities. Magnetic resonance imaging (MRI)
has changed the diagnostic approach to patients with suspected spinal cord dysfunction. Plain radiographs and computed tomography (CT) scans may show bone and some soft tissue abnormalities. Conventional radiographs and CT scans are required in patients with trauma or suspected bone involvement by tumor or degenerative processes, but MRI shows many of these abnormalities and defines the spinal cord as well as the soft tissue structures associated with it. Tissue damage patterns within the cord, such as hemorrhage and edema, also may be detected with MRI. CT myelography may be able to answer some of these questions in patients in whom implanted metal precludes MRI but generally does not yield the same level of detail. After imaging studies exclude compressive lesions or other masses affecting the spinal cord, the possibility of inflammatory or demyelinating disorders remains. Lumbar puncture may be useful in diagnosis by revealing an inflammatory cerebrospinal fluid (CSF) formula.

**DIFFERENTIAL CONSIDERATIONS**

The prime principle in management of spinal cord dysfunction is to consider and exclude potentially treatable clinical conditions. The clinical assessment of spinal cord dysfunction is limited to detection of weakness, sensory alterations, sphincter dysfunction, and perhaps reflex abnormalities. There may be pain in the back, depending on the pathologic process, but generally it is not helpful in formulating a list of considerations for the differential diagnosis. Because potential functional loss and impact on quality of life are great, the detection of a process for which some intervention is possible assumes great importance. A likely diagnosis of spinal cord infarction may be entertained, but the pursuit of a treatable process, such as spinal cord compression from an epidural hematoma, should be seriously considered.22 This discovery process may involve specialty consultation or studies that may not be readily available in all settings, such as MRI. As a general rule, liberal use of consultation and imaging is recommended when the possibility of spinal cord dysfunction is considered. The history may suggest a specific cause and will guide the tempo of investigation. The caveat is that spinal cord diseases may mimic many other disease processes, and neither the history nor physical examination may allow diagnosis until appreciable neurologic dysfunction has developed.

The picture of a complete transverse spinal cord syndrome with paraplegia, sensory loss at a clear anatomic level, and sphincter dysfunction cannot be fully simulated by other anatomic disorders. Incomplete or evolving spinal cord syndromes may be imitated by other disease processes. It is always prudent to focus the differential diagnosis on anatomic considerations—the classic “where is the lesion?” approach (Table 106-2). Progressive lower extremity weakness and sensory alteration may represent cord dysfunction but could reflect an intracranial vertex mass with bilateral cortical dysfunction. Ataxia may be a finding in cerebellar disease but also has rarely been reported as an isolated finding with spinal cord compression. Another example is rapidly progressive paralysis in a patient with areflexia and quadriplegia; ascending paralysis (Landry-Guillain-Barré syndrome) at times may mimic an acute cord lesion.

In general, pathologic processes involving the spinal cord may be divided into processes affecting the cord or its blood supply primarily, such as demyelination, infection, or infarction, and processes that compress the cord, most often originating outside the dura (Box 106-1). Myelitis is a comprehensive term for spinal cord inflammation with dysfunction, and the potential causes are legion. The clinical presentation is often similar across the variety of entities that may cause cord compression. The tempo of the process may yield a different clinical picture. In chronic compression, muscle wasting and abnormal reflexes may be present, whereas both of these may be lacking in acute compression. A neurologic deficit in concert with back pain strongly suggests a spinal cord lesion causing compression of neural elements, necessitating prompt investigation to identify a specific cause. Atypical presentations for these lesions are the rule, and additional diagnostic studies should be pursued as appropriate.

![Table 106-2 Clinical Characteristics of Neuromuscular Diseases](image)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Relevant Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myelopathy</td>
<td>Trauma, infection, cancer</td>
</tr>
<tr>
<td>Motor neuron disease (ALS)</td>
<td>Progressive difficulty with swallowing, speaking, walking</td>
</tr>
<tr>
<td>Neuropathy</td>
<td>Recent infection, Ascending weakness</td>
</tr>
<tr>
<td>Neuromuscular junction disease</td>
<td>Food (canned goods), Tick exposure, Easy fatigability</td>
</tr>
<tr>
<td>Myopathy</td>
<td>Thyroid disease, Previous similar episodes</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Strength</th>
<th>DTR</th>
<th>Sensation</th>
<th>Wasting</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal to decreased</td>
<td>Increased</td>
<td>Normal to decreased</td>
<td>No</td>
</tr>
<tr>
<td>Decreased</td>
<td>Increased</td>
<td>Decreased</td>
<td>Yes</td>
</tr>
<tr>
<td>Normal or decreased</td>
<td>Distal &gt; proximal</td>
<td>Decreased</td>
<td>Yes</td>
</tr>
<tr>
<td>Decreased</td>
<td>Distal &gt; proximal</td>
<td>Normal to fatigue</td>
<td>No</td>
</tr>
<tr>
<td>Normal or decreased</td>
<td>Distal &gt; proximal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Decreased</td>
<td>Proximal &gt; distal</td>
<td>Normal</td>
<td>Yes</td>
</tr>
</tbody>
</table>

*ALS, amyotrophic lateral sclerosis; DTR, deep tendon reflex.*
MANAGEMENT

Just as the clinical manifestations of spinal cord dysfunction are nonspecific with respect to etiology, the treatment of many of the disease entities often is nonspecific. Steroid administration has been traditionally recommended as therapy in spinal cord trauma, although currently this use of steroids has been seriously questioned in the medical literature.23-26 Steroids have also been used with many nontraumatic causes of cord compression, despite the lack of rigorous clinical studies supporting this use. Radiation treatment is recommended for cord compression by tumor. Surgical consultation for decompression may be considered, although the indications for surgery and timing of surgery are controversial.

A specific diagnosis is needed to guide therapy. Accordingly, involvement of appropriate consultants and discussion of what may be understudied therapies are suggested.

SPECIFIC DISEASE PROCESSES

As noted earlier, spinal cord disorders may be grouped into lesions resulting from processes intrinsic to the cord or vasculature and lesions causing extrinsic compression. The order of the following discussion roughly corresponds with the organization of Box 106-1.

Intrinsic Cord Lesions

Multiple Sclerosis

Principles of Disease. Demyelination denotes a disease process with the prominent feature of partial or complete loss of the myelin surrounding the axons of the central nervous system. Multiple sclerosis (MS) is the most common example of such a process; spinal cord involvement may dominate the clinical picture.27

Clinical Features. Central nervous system lesions that are “scattered in time and space” are the hallmark of MS, as described in more detail in Chapter 105. The demyelinated segments do not transmit action potentials normally, resulting in a wide variety of spinal cord–related abnormalities, depending on the location and extent of the demyelination. In addition to patchy motor and sensory deficits, patients with MS may complain of bladder dysfunction or tremor or demonstrate evidence of a transverse partial or complete cord syndrome mimicking a compressive spinal lesion.27,28 Spinal cord lesions in MS primarily involve the lateral corticospinal tracts, the posterior columns, and the lateral spinothalamic tracts. Motor system dysfunction is the most frequent manifestation of MS involvement of the spinal cord, usually as a result of lesions in the lateral corticospinal tracts.

The examination of patients with MS often reveals paresis, increased muscle tone, hyperreflexia, clonus, and Babinski’s response. Spinal cord involvement also may result in dysautonomias.

Diagnostic Strategies. Spinal MRI is the diagnostic imaging modality of choice because it can exclude cord compression and show lesions suggestive of MS.29-31 CSF testing for myelin basic protein and oligoclonal bands also is a diagnostic option, but no CSF abnormalities are entirely specific for MS.31,32 Oligoclonal bands in the CSF may aid in the diagnosis, but they are significant only if they are not present in the serum as well.31

Differential Considerations. Considerations in the differential diagnosis include systemic lupus erythematosus, Lyme disease, neurosyphilis, human immunodeficiency virus (HIV) myelopathy, and other disorders.

Management. MS exacerbations may be treated with high-dose methylprednisolone, followed by a tapering dose of prednisone.28 Consultation and referral to a neurologist usually are indicated. Immunosuppressive therapy in patients with the chronic progressive form of the disease has met with variable success.27,28 Because numerous disorders can mimic MS, the definitive diagnosis of this disease is usually not made in the ED.33

Transverse Myelitis

Principles of Disease. Acute transverse myelitis refers to acute or subacute spinal cord dysfunction characterized by paraplegia, a transverse level of sensory impairment, and sphincter disturbance. It describes a heterogeneous group of inflammatory disease processes that can affect the spinal cord by interruption of the ascending or descending pathways in the spinal cord.34 It is relatively rare, with a reported annual incidence of 1 case per 1.3 million population. The presentation may be mimicked by compressive lesions, trauma, infection, or malignant infiltration. The exact pathogenesis is unknown, although it is noted to follow viral infection in approximately 30% of patients and commonly is termed postinfectious myelitis.34,35 Other postulated etiologic categories include infectious, autoimmune, and idiopathic.35-37 No apparent cause of acute transverse myelitis is identified in 30% of the patients.35 Progression of symptoms usually is rapid, with 66% of the cases reaching maximal deficit by 24 hours.36 Symptoms may progress, however, during days to weeks. The thoracic cord region is affected in 60 to 70% of cases;66 the cervical spinal cord is rarely affected.39

Clinical Features. In addition to motor, sensory, and urinary disturbances, patients with acute transverse myelitis may complain of back pain and may have low-grade fever, raising concern for spinal epidural abscess. As with MS, the examination may reveal weakness progressing to paresis, hypertonia, hyperreflexia, clonus, and Babinski’s response. Spinal cord involvement also can result in dysautonomias.

Diagnostic Strategies. Evaluation for acute transverse myelitis is done primarily with emergent MRI to exclude compressive lesions. Results of CSF studies are normal in 40% of the cases, with only mildly elevated protein level or pleocytosis in the remaining 60%.40 The most essential aspect of the evaluation is to eliminate a potentially treatable cause, such as spinal epidural abscess, neuroplasm, or hematoma.

Differential Considerations. Considerations in the differential diagnosis for transverse myelitis include MS, spinal epidural abscess, spinal neoplasm, and hematoma.

Management. Treatment with steroids is of unknown benefit. Anecdotal reports of improvement after steroid administration exist,40,41 but some studies have found no benefit to their use.37 Neurologic consultation is suggested, and hospitalization usually is required.

The clinical course of acute transverse myelitis varies widely, ranging from complete recovery to death from progressive neurologic compromise.37 Maximal improvement usually is obtained within 3 to 6 months.32,43 At 5-year follow-up evaluation of one series of patients with this disease, 30% had a good recovery, 25% had a fair recovery, 30% had a poor outcome, and 15% had died as a result of complications of the disease.43

Spinal Subarachnoid Hemorrhage

Principles of Disease. Intraspinal hemorrhage is rare and may occur in the same anatomic locations as intracranial hemorrhages; epidural, subdural, subarachnoid, and intramedullary hemorrhages are all possible.7 Spinal subarachnoid hemorrhage usually is caused by an arteriovenous malformation.44 Hemorrhage from tumors or cavernous angiomas and spontaneous hemorrhage secondary to anticoagulation therapy also have been reported.45,46 Bleeding may occur exclusively in the subarachnoid space or within the substance of the spinal cord itself.
Clinical Features. Patients present with excruciating back pain, of paroxysmal onset, at the level of the hemorrhage. This pain also may be in a radicular distribution or extend into the flank. Patients may complain of headache and exhibit cervical rigidity if the blood migrates into the intracranial subarachnoid space, simulating an intracranial subarachnoid hemorrhage. Variable neurologic deficits depend on the magnitude and anatomic location of the hemorrhage. These deficits typically include extremity numbness, weakness, and sphincter dysfunction.45,47 Nuchal rigidity or signs of meningeal irritation may also be present.

Diagnostic Strategies. The diagnostic study of choice is MRI. Lumbar puncture also can confirm the presence of blood in the CSF.

Differential Considerations. Considerations in the differential diagnosis include epidural abscess, tumor, transverse myelitis, ischemia from aortic dissection, and anterior spinal artery thrombosis.

Management. Treatment depends on the etiology of the hemorrhage. Neurosurgical referral is obtained for further evaluation and for clot evacuation if compression is present. Angiography may be recommended if arteriovenous malformation is suspected.

Syringomyelia

Principles of Disease. Syringomyelia is the presence of a cavitary lesion within the substance of the spinal cord. A syrinx usually is a chronic progressive lesion, and its location within the cord determines the constellation of neurologic findings on examination.

Clinical Features. Headache and neck pain are the most common complaints, followed by sensory disturbance, gait disorder, and lower cranial nerve dysfunction.48 The classic pattern of sensory deficit involves a loss of pain and temperature sensation in the upper extremities with preservation of proprioception and light touch. This phenomenon is described as a “dissociative anesthesia” because of the discrepant loss of sensory modalities. The sensory deficit often is described as being in a “capelike” distribution over the shoulders and arms. The anatomic basis for the neurologic features of a syrinx is the location near the central canal. Crossing fibers of the lateral spinothalamic tract carrying pain and temperature fibers may be impaired. Crude touch, position, and vibratory sensation typically are unaffected. Sensory fibers from the lower limbs are similarly spared.

The symptoms of syringomyelia develop and progress in accordance with the intracavitary pressure and location of the syrinx. The most common features on physical examination are lower limb hyperreflexia, weakness and wasting in the hands and arms, dissociated sensory loss, and gait disorder. Symptoms may be exacerbated by a sneeze, cough, or Valsalva maneuver.49 Ninety percent of patients with syringomyelia have Arnold-Chiari I malformation (projection of cerebellar tonsils and medulla into the spinal canal).50 Syringomyelia also may result from spinal cord trauma (often months to years later) or compressive tumors, or it may follow meningitis.51 Diagnostic Strategies. Syringomyelia is best seen on MRI. No other study currently in widespread use is equal to MRI in diagnostic ability.

Differential Considerations. Considerations in the differential diagnosis for syrinx include intrinsic spinal tumor and demyelination.

Management. When the diagnosis of syringomyelia is considered, emergent imaging in the ED is not necessary if follow-up evaluation can be arranged because this condition usually is a slowly progressive process. In patients for whom MRI studies are obtained and the diagnosis is made, referral to a neurosurgeon is indicated because symptoms progress in approximately two thirds of patients.52

Idiopathic Spastic Paraparesis

Idiopathic spastic paraparesis is a progressive disorder characterized by progressive weakness and signs of spasticity of the lower extremities. This condition sometimes also is referred to as primary lateral sclerosis, which describes the demyelination pattern in the lateral column of the spinal cord. This disorder typically occurs in older men. A heritable form may sometimes be discovered. It is a diagnosis of exclusion.53-55

Human Immunodeficiency Virus Myelopathy

HIV myelopathy typically occurs in patients with advanced HIV disease. Weakness, gait disturbance, sphincter dysfunction, sensory abnormalities, and signs of spasticity are features of this progressive process. This is a diagnosis of exclusion because disorders such as toxoplasmosis, lymphoma, variella-zoster, and cytomegalovirus infection may produce a similar clinical picture in immunocompromised patients. On pathologic examination, vacuolization of myelin sheaths in the cord may be found. Treatment is directed at the retroviral infection, although there is no proven treatment.56,57

Spinal Cord Infarction

Spinal cord infarction is another diagnosis of exclusion. Aortic dissection, surgery, and global ischemia are the more common causes, although this disorder may occur as a complication of systemic lupus erythematosus or may be cryptogenic. An anterior spinal cord syndrome is the most common clinical picture. Some recovery is possible, although it generally is less complete than in cerebral stroke. The site of clinical dysfunction may be distant from the site of vascular occlusion.58

Extrinsic Cord Lesions

Spinal Epidural Hematoma

Principles of Disease. Spinal epidural hematoma is a relatively rare condition resulting from a variety of etiologic disorders. Its incidence is 0.1 per 100,000 population.59,60 The etiology may be traumatic after lumbar puncture, epidural anesthesia, or spinal surgery.1,5,61 Spinal epidural hematoma is more likely to occur in anticoagulated or thrombocytopenic patients or in patients with liver disease or alcoholism.62 Spontaneous bleeding is rare but may arise from spinal arteriovenous malformation or vertebral hemangioma. Approximately one fourth to one third of all cases are associated with anticoagulation therapy, including low-molecular-weight heparin.63,64

Clinical Features. The patient usually presents with sudden, severe, constant back pain with a radicular component. It may be noted to follow a spreading episode. The pain may be worsened by percussion over the spine and maneuvers that increase intraspinal pressure, such as coughing, sneezing, or straining.65 The pain often causes the patient to seek care before the development of neurologic signs, possibly leading to delays in diagnosis.6 Neurologic deficits follow and may progress during hours to days.60 Anticoagulant use or an intrinsic coagulation abnormality may be present.

The patient usually is in significant distress from the pain. Motor and sensory findings depend entirely on the level and size of the hematoma but can include weakness, paresis, loss of bowel or bladder function, and virtually any sensory deficit.

Diagnostic Strategies. MRI, as with virtually all suspected intrinsic spinal disorders, is the diagnostic study of choice.66

Differential Considerations. Considerations in the differential diagnosis include abscess, epidural neoplasm, acute disk herniation, and spinal subarachnoid hemorrhage.
Management. Recovery without surgery is rare, and surgical consultation for emergent decompressive laminectomy should be considered. The overall mortality rate is approximately 8%.66,67 Functional recovery is related primarily to the length of time the symptoms are present. Recovery after 72 hours of symptoms is rare but has been reported even without surgery.66,67

Spinal Epidural Abscess

Principles of Disease. Spinal epidural abscess is an infectious process usually confined to the adipose tissue of the dorsal epidural space, where there is a rich venous plexus. It is an uncommon disease, with an overall frequency of 0.2 to 1.2 per 10,000 hospital admissions.68,69 Major risk factors include diabetes, injection drug abuse, chronic renal failure, alcoholism, and immunosuppression, although the disease can be seen in patients who have none of these conditions.69-71 Whereas the disease may be manifested in subacute and chronic forms, the acute presentation is seen most frequently in the ED. Thoracic and lumbar sites of infection predominate, with cervical epidural abscess being much less common.72,73 Infection typically extends over four or five spinal vertebral segments.74 The dura mater limits the spread of an epidural infection, making subdural or intraspinal spread uncommon. Hematogenous spread of infection to the epidural space is the most common source (seen in 26-50% of cases),68,69,74 either to the epidural space or to the vertebra with extension to the epidural space. Skin and soft tissue infections are the most frequently reported identified source (in 15%).70,74,75 *Staphylococcus aureus* is the most prevalent organism, being cultured in more than 50% of cases.68,75 Other frequently identified pathogens include aerobic and anaerobic streptococci, *Escherichia coli*, and *Pseudomonas aeruginosa*. Multiple organisms are identified in approximately 10% of cases; no organism is identified in 40%.65,72

Clinical Features. The classic clinical presentation of spinal epidural abscess begins with a backache that progresses to localized back pain often associated with tenderness to percussion. The duration of symptoms is typically a few days but may extend for weeks. Fever, sweats, and rigors are common, being reported in 30 to 75% of patients.68,75 The classic triad of back pain, fever, and progressive neurologic deficits is present in only a few patients, however, and delayed clinical diagnosis is common.68 Radicular symptoms may not be present initially but usually develop as the disease progresses.

Without treatment, myelopathic signs will develop, usually beginning with bowel and bladder disturbance. Weakness ensues, followed by paraplegia or quadriplegia. Approximately 10% of patients with spinal epidural abscess present with altered mental status due to encephalopathy.68,74

Diagnostic Strategies. MRI is the imaging modality of choice and needs to be performed emergently if the diagnosis is entertained. Other diagnostic testing is nonspecific for spinal epidural abscess, but a complete blood count may support the diagnosis because leukocytosis is commonly present, with a typical white blood cell count of 13,000 to 16,000/µL.74 The erythrocyte sedimentation rate, although not specific for epidural abscess, is virtually always elevated with this condition.68,69,74 Plain films usually are normal in appearance unless evidence of osteomyelitis of an adjacent vertebral body is seen. Lumbar puncture is relatively contraindicated with known epidural abscess but is often performed as part of the evaluation for meningitis. CSF findings are consistent with a parameningeal infection, showing elevation of protein and some increase in inflammatory cells.

Differential Considerations. Any compressive spinal lesion, including tumor or hematoma, can mimic spinal epidural abscess.

Management. Urgent surgical consultation for decompression usually is required. Antibiotics effective against the most common pathogens (particularly *S. aureus*) should be started empirically.

One such regimen that covers gram-positive and gram-negative organisms consists of a third-generation cephalosporin plus vancomycin, both given intravenously, plus rifampin given orally.

Outcome is usually related to the speed of diagnosis before the development of myelopathic signs. The disease is fatal in 18 to 23% of cases, and patients with neurologic deficit rarely improve if surgical intervention is delayed more than 12 to 36 hours after onset of paralysis.68,74 Patients operated on before development of neurologic symptoms have an almost universally good outcome.69,70

Diskitis

Principles of Disease. Diskitis is an uncommon primary infection of the nucleus pulposus, with secondary involvement of the cartilaginous endplate and vertebral body. It may occur after surgical procedures or spontaneously, the latter being more common in pediatric patients.76,77 An increased incidence of diskitis has been noted in immunocompromised patients and in patients with systemic infections. Both a chronic disease and a more common acute course have been described.76

Clinical Features. Patients present with moderate to severe pain, localized to the level of involvement and exacerbated by almost any movement of the spine. Radicular symptoms are present in 50 to 90% of cases.76,77 The lumbar spine is the most common site of disease. Elevated temperature is noted in more than 90% of patients.76 Patients experience pain with range of motion. Neurologic deficits are the exception with diskitis.

Diagnostic Strategies. Plain radiographs usually are not helpful for early diagnosis, but destruction of the disk space is highly suggestive if present. The radiographic findings become abnormal after 2 to 4 weeks of disease. In addition to disk space narrowing, plain films may show irregular destruction of the vertebral body endplates. Often there is a latent period (2 to 8 weeks) between the onset of back pain and the development of other clinical symptoms or abnormalities on the physical examination. MRI is the radiographic study of choice because it can not only diagnose diskitis but also rule out paravertebral or epidural abscess. Laboratory studies often show an elevated erythrocyte sedimentation rate, but the white blood cell count usually is normal.76 *S. aureus* is the most common pathogen, but gram-negative, fungal, and tuberculous infections all have been recognized.

Differential Considerations. Considerations in the differential diagnosis include vertebral osteomyelitis, spinal epidural abscess, neoplasm, and hematoma.

Management. With timely diagnosis and treatment, outcome generally is good, and medical treatment with intravenous antibiotics usually is curative. Surgery often is not necessary.76,77

Neoplasm

Principles of Disease. Spinal cord tumors are classified according to their relationship to the dura and spinal cord (extradural, intradural extramedullary, and intradural intramedullary). Spinal cord tumors produce neurologic symptoms by compression, invasion, or destruction of myelinated tracts. The resulting neurologic symptoms are directly related to the growth rate and the location of the tumor. Spinal cord tumors account for 4 to 10% of central nervous system tumors but for only 1% of all cancers. Primary tumors occur with an incidence of 1 per 1 million population.78 Most tumors affecting the spinal cord are metastatic. Approximately 10% of patients with known cancer are diagnosed with a spinal metastasis at some point in the course of their disease, and 5 to 10% of patients ultimately diagnosed with cancer first present with a spinal metastasis.78 Lung cancer, breast cancer, and lymphoma represent more than 50% of the primary malignant neoplasms that subsequently develop spinal metastasis, spreading by both the hematogenous route and direct extension. Most
metastases occur in the thoracic spine, and nearly 20% of patients with tumor spread to the spine will be found to have disease at multiple levels.2,79,80

Clinical Features. In 95% of patients with spinal neoplasm, the initial complaint is pain, either in the back at the level of the tumor or in a radicular distribution. Pain often is characterized as dull, constant, and aching and commonly is said to worsen with recumbency (in contrast with the pain of herniated disk).6 Nighttime pain that is severe is characteristic of spinal neoplasm.81 Any action that increases intraspinal pressure (Valsalva maneuver, sneeze, cough) may be associated with increased pain. Neurologic deficits vary by the location of the lesion. Besides a thorough neurologic examination, a search for possible primary sites should be done on the physical examination.

Diagnostic Strategies. Plain radiographs are usually the initial diagnostic test, and 70 to 85% of patients with spinal column involvement show some abnormality on these films.81 Patients with neurologic abnormalities and suspicious findings on plain films are candidates for emergent MRI or CT myelography. In patients with a known history of neoplasm and new back pain, some authorities recommend foregoing plain films and proceeding directly to MRI because plain films can be misleading or nondiagnostic.82

Differential Considerations. Considerations in the differential diagnosis include any of the compressive lesions (hematoma, infection). Tumor can also mimic intrinsic spinal cord lesions, such as transverse myelopathy and cord infarction.

Management. Acute compressive myelopathy from neoplasm constitutes an oncologic emergency. Immediate treatment is required to preserve function and to prevent deterioration. With onset of paraplegia and incontinence, less than 5% of patients regain ambulatory status.1,83 Of patients who are ambulatory at the time of diagnosis, 60% remain ambulatory.8 High-dose steroids, radiotherapy, and surgery all may be necessary acute interventions, and consultation with neurosurgeons, neurologists, oncologists, and therapeutic radiologists may be required.

KEY CONCEPTS

- Spinal cord disorders may be manifested in subtle fashion and with nonspecific clinical signs and symptoms. In the absence of neurologic abnormalities or complaints, diagnosis of these disorders can be extremely difficult.
- Patients with rapid onset and progression of spinal cord symptoms should receive specialized imaging and consultation in the ED.
- MRI frequently is required for a definitive diagnosis of spinal cord syndromes.
- With compressive lesions of the spinal cord, duration of neurologic dysfunction is directly related to ultimate neurologic outcome. The diagnosis should be made expeditiously and definitive therapy begun as soon as possible.

The references for this chapter can be found online by accessing the accompanying Expert Consult website.