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SPINAL CORD DISORDERS

ARTICLE 1: SPINAL CORD ANATOMY AND LOCALIZATION

Todd A. Hardy, PhD, MBBS, FRACP. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):12–29.

ABSTRACT

PURPOSE OF REVIEW:

This article focuses on clinically relevant teaching points in spinal anatomy and localizing the lesion in myelopathy.

RECENT FINDINGS:

The principles underlying spinal cord lesion localization are well established, but improvements in MRI and the discovery of pathologic antibodies associated with causes of transverse myelitis distinct from multiple sclerosis, such as aquaporin-4 IgG and myelin oligodendrocyte glycoprotein IgG, have assisted in diagnosis.

SUMMARY:

The spinal cord has a highly organized neuroanatomy of ascending and descending tracts that convey sensory, motor, and autonomic information. Using integration of clues from the patient's history and neurologic examination, the effective clinician can distinguish spinal cord from peripheral nerve or brain pathology, often determine the level and parts of the spinal cord affected by a lesion, and focus on a likely diagnosis. The advent of MRI of the spine has revolutionized investigation of spinal cord disorders, but an important place for strong clinical acumen still exists in assessing the patient with a myelopathy.

KEY POINTS

- Lumbar puncture is typically performed at the L3-L4 or L4-L5 level.
- The butterfly-shaped area of the spinal cord in cross section is known as the central gray matter.
- The descending motor pathways in the cord before the anterior horns are called upper motor neurons, and those of the anterior horns and somatic motor nerves are called lower motor neurons.
- The monosynaptic spinal reflex is caused by activation of peripheral stretch receptors transmitting an impulse along primary sensory afferents that synapse directly on alpha motor neurons, causing a final efferent motor response.
- The artery of Adamkiewicz is the large radiculomedullary artery that supplies the anterior spinal artery between T9 and T12 in most individuals.

- The somatotopic organization of the spinal cord allows determination of the approximate or, in some cases, precise level of a spinal cord lesion.
- The Uhthoff phenomenon commonly occurs when patients with multiple sclerosis experience an exacerbation of symptoms with an increase in body temperature.
- Spinal injury can lead to either a flaccid or spastic bladder, with a range of symptoms, including urinary frequency, urgency, incontinence, and urinary retention due to a lack of coordination between the detrusor muscle of the bladder and the urinary sphincter.
- The patient's previous medical history and the temporal onset of neurologic symptoms can be used to narrow the differential diagnosis of a spinal cord lesion.
- The likelihood of upper motor neuron pathology is increased if hyperreflexia occurs accompanied by other upper motor neuron signs, such as an extensor plantar response or pyramidal weakness, or both.
- The C4 dermatome abuts the T2 dermatome on the chest.
- A truncal sensory level is defined as the highest dermatomal area of normal sensation to pinprick and temperature on the trunk.
- A lesion affecting the spinothalamic tract of the right hemicord only will cause impaired temperature and pinprick sensation on the left trunk and lower limb two to three vertebral levels below the level of the cord lesion because the spinothalamic tracts ascend as they decussate.
- The term *paresis* is used to denote weakness, whereas *plegia* is used to denote absence of any voluntary movement.
- Spinal shock occurs when hyperacute or acute injury (particularly trauma) to the spinal cord results in flaccid areflexia below the level of the lesion.
- Neurogenic shock occurs due to acute pathology above the level of T6, which leads to loss of sympathetic tone below the lesion causing hypotension and unopposed vagal activity leading to bradycardia.
- Autonomic dysreflexia occurs when patients have injury above the T6 level, leading to an exaggerated sympathetic nervous system response to sensory stimuli below the level of the lesion (eg, bladder filling).
- A central intraaxial spinal cord lesion often causes sensory symptoms and signs in the upper limbs and trunk before the lower limbs and sacral regions (called a suspended sensory level). This is because the lower extremity spinothalamic tracts run more laterally than those of the upper extremities and so take longer to be affected by an expanding central cord lesion.
- A partial transverse myelitis refers to spinal cord inflammation in which symptoms and signs occur that are attributable to only a portion of the spinal cord in cross section rather than involving the entire transverse diameter. A complete transverse myelitis is attributable to spinal cord inflammation involving its entire cross section.
- Patients with a longitudinally extensive transverse myelitis should be tested for aquaporin-4 IgG and myelin oligodendrocyte glycoprotein IgG.

ARTICLE 2: VASCULAR MYELOPATHIES

Nicholas L. Zalewski, MD. Continuum (Minneap Minn). February 2021;
27 (1 Spinal Cord Disorders):30–61.

ABSTRACT

PURPOSE OF REVIEW:

Neurologists should be able to identify clinical and neuroimaging features that distinguish vascular disorders from other causes of myelopathy.

RECENT FINDINGS:

Although certain clinical features suggest a vascular etiology in acute and chronic myelopathy settings, accurate MRI interpretation within the clinical context is key. Recent studies have

shown vascular myelopathies are frequently misdiagnosed as transverse myelitis, and recognition of this diagnostic pitfall is important. Many different vascular mechanisms can cause myelopathy; this article provides a comprehensive review that simplifies disease categories into arterial ischemia, venous congestion/ischemia, hematomyelia, and extraparenchymal hemorrhage.

SUMMARY:

It is important to recognize and manage vascular disorders of the spinal cord as significant causes of acute, subacute, and progressive myelopathy.

KEY POINTS

- Two large retrospective studies recently showed that patients initially diagnosed with idiopathic transverse myelitis frequently had alternative myelopathy diagnoses, with vascular etiologies among the most common.
- Vascular disorders of the spinal cord have important time-to-treatment considerations as delays in diagnosis can be associated with worse outcomes, highlighting the importance of considering vascular causes early.
- The vascular anatomy of the spinal cord consists of a single anterior spinal artery and paired posterior spinal arteries that run along the length of the spinal cord.
- Open or endovascular thoracic aortic aneurysm repair is the most common procedure associated with spinal cord infarction, representing approximately 50% of periprocedural spinal cord infarction cases. Spinal cord infarction has also been associated with other aortic surgeries and an array of other procedures (eg, cardiac surgery, spinal decompression, epidural injection, angiography, nerve block, embolization, other vascular surgery, and thoracic surgery).
- During spinal cord ischemia, the goal of treatment is to increase spinal cord perfusion pressure through collaterals by lowering pressure within the spinal canal via CSF drainage or mean arterial blood pressure augmentation.
- Two large studies highlighted the frequent misdiagnosis of spinal cord infarction as "transverse myelitis" in approximately 15% of referred cases.
- Although an older patient population with vascular risk factors is common in spinal cord infarction, mechanisms affecting younger patients also occur (eg, fibrocartilaginous embolism, vertebral dissection), highlighting that spinal cord infarction can occur at any age.
- From the earliest days, it has been recognized that spinal cord infarction frequently results in acute deficits localized to an anterior spinal artery territory (bilateral corticospinal tract, lower motor neuron at lesion level, and pain/temperature loss) or, less frequently, a posterior spinal artery territory (dorsal column dysfunction); these deficits may distinguish spinal cord infarction from other myelopathies.
- Severe acute pain (back, chest, neck, limb) at or before onset is another helpful feature that is reported in approximately 70% of patients with spinal cord infarction but is atypical acutely in myelitis.
- Once spontaneous spinal cord infarction is suspected, it is important to understand the typical MRI appearance in acute, subacute, and chronic settings.
- A low threshold for MRI of the entire spine should exist, unless the localization is clear (eg, cervical spinal cord in quadriplegia). Diffusion-weighted imaging/apparent diffusion coefficient should be performed, but the sensitivity is incomplete and sometimes takes days to evolve. In the initial hours of symptoms, imaging is likely normal or equivocal.
- It is reasonable to discuss risks and benefits of IV recombinant tissue plasminogen activator within the first 4.5 hours after onset if suspicion of spinal cord infarction is high and the patient understands the limited evidence.
- Atherosclerosis, arterial dissection, and fibrocartilaginous embolism are the most common presumed mechanisms of spontaneous spinal cord infarction.
- Outcomes after spontaneous spinal cord infarction are variable. Despite severe deficits, approximately 50% of patients ultimately ambulate without a gait aid.

- The incidence of spinal dural arteriovenous fistula is 5 to 10 cases per million per year. An older population is typical (40 to 80 years), with male predominance (80%), and patients frequently report previous back surgeries or trauma that may contribute to the development of a fistula.
- The clinical presentation of spinal dural arteriovenous fistula typically includes a gradually progressive thoracic myelopathy with leg weakness/numbness, bowel/bladder dysfunction, symptoms frequently referable to the conus/roots, and episodic worsening with exertion/Valsalva. The strongest clinical clue is dramatic worsening of deficits with activities elevating venous pressure, such as exertion, Valsalva, or lumbar puncture.
- Inappropriate corticosteroid use for suspected alternative diagnoses can lead to clinical worsening in spinal dural arteriovenous fistulas from exacerbation of venous hypertension and thus should be avoided.
- Spinal cord T2-hyperintense signal is present in approximately 95% of cases of spinal dural arteriovenous fistula, is often longitudinally extensive (three or more vertebral body segments), and frequently extends to the conus (90% of cases).
- Flow voids (engorged perimedullary veins) are seen on the dorsal more than the ventral surface of the cord in approximately 80% of cases of spinal dural arteriovenous fistula.
- Gadolinium enhancement of the spinal cord is common in spinal dural arteriovenous fistula, and clinicians should not be misled to suspect an inflammatory or neoplastic etiology when features are suspicious for spinal dural arteriovenous fistula.
- Although digital subtraction angiography is the gold standard to identify a fistula, a noninvasive spinal magnetic resonance angiogram can initially be considered to potentially help localize the fistula before the formal angiography.
- Treatment options for spinal dural arteriovenous fistula include embolization of the fistula via digital subtraction angiography or surgical disconnection of the draining vein. Improvement or stabilization is expected after treatment.
- Intraparenchymal spinal cord hemorrhage (hematomyelia) is very rare. Trauma is the most common cause, followed by intramedullary spinal cord cavernous malformations and arteriovenous malformations; many additional causes have also been reported.
- A well-defined lobulated masslike lesion within the parenchyma of the spinal cord with heterogeneous T1- and T2-weighted signal intensity surrounded by a well-defined dark T2-hypointense rim (hemosiderin deposition) with classic popcorn appearance is typical of an intramedullary spinal cord cavernous malformation, which can be best appreciated with gradient recalled echo and susceptibility-weighted imaging.
- Management of intramedullary spinal cord cavernous malformation is focused on avoiding further deterioration with recurrent hemorrhage; observation is typically recommended for asymptomatic cavernous malformations or those with minimal symptoms.
- Although spinal dural arteriovenous fistula (type I) comprises 70% of spinal arteriovenous shunts, 30% of cases are secondary to types II through V: intramedullary glomus arteriovenous malformation (type II), intramedullary juvenile arteriovenous malformation (type III), perimedullary arteriovenous fistula (type IV), and extradural arteriovenous fistula (type V).
- The predominant clinical presentation of spinal arteriovenous malformation is acute myelopathy secondary to hematomyelia.
- Spinal epidural hematoma is most commonly encountered in trauma or after surgery, epidural catheterization, or lumbar puncture.
- Treatment of symptomatic spinal epidural hematoma is emergent surgery, with improved outcomes when performed within 12 hours of symptom onset.

ARTICLE 3: MYELITIS AND OTHER AUTOIMMUNE MYELOPATHIES

Sebastian Lopez Chiriboga, MD; Eoin P. Flanagan, MBBCh. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):62–92.

ABSTRACT

PURPOSE OF REVIEW:

This article provides an update on the clinical diagnosis and management of immune-mediated myelopathies, including the relevance of imaging, ancillary testing with an emphasis on autoantibody biomarkers, recognition of myelitis mimics, and therapeutic approach.

RECENT FINDINGS:

The imaging characterization of immune-mediated myelopathies and the discovery of neural autoantibodies have been crucial in improving our ability to accurately diagnose myelitis. The identification of autoantibodies directed against specific central nervous system targets has led to major improvements in our understanding of the mechanisms underlying inflammation in myelitis. It has also allowed distinction of these myelopathy etiologies from noninflammatory etiologies of myelopathy and from multiple sclerosis and provided insight into their risk of recurrence, treatment response, and long-term clinical outcomes. Prompt recognition and appropriate testing in the setting of acute and subacute myelopathies is critical as timely administration of immunotherapy can help improve symptoms and prevent permanent neurologic disability. A patient should not be classified as having "idiopathic transverse myelitis" without a comprehensive evaluation for a more specific etiology. Achieving the correct diagnosis and learning to recognize noninflammatory myelitis mimics is crucial as they have therapeutic and prognostic implications.

SUMMARY:

Identifying the clinical and radiographic features of immune-mediated myelitis and recognizing mimics and pitfalls will help clinicians treat confirmed autoimmune myelitis appropriately.

KEY POINTS

- The differential diagnosis of immune-mediated myelopathies is broad and includes noninflammatory myelopathies from compressive, vascular, neoplastic, metabolic, nutritional, infectious, toxic, and inherited causes.
- The length of the T2-hyperintense lesion seen on sagittal spinal cord imaging is a very useful discriminator between multiple sclerosis (less than three vertebral segments) and aquaporin-4 (AQP4) IgG–seropositive neuromyelitis optica spectrum disorder (NMOSD) (three or more vertebral segments).
- Idiopathic transverse myelitis should be considered a diagnosis of exclusion, with a comprehensive evaluation for both inflammatory and noninflammatory etiologies before assigning that diagnosis.
- Significant advances in the field of autoimmune neurology, including the discovery of neural autoantibodies, have assisted in identifying a specific cause for patients previously classified as having idiopathic transverse myelitis.
- Better radiographic characterization of immune-mediated myelopathies and their mimics has improved our ability to diagnose patients with myelopathies of uncertain etiology.
- The time from onset to maximal neurologic deficit is the most important feature to determine when evaluating a myelopathy as it helps narrow the differential diagnosis.

- The time to nadir in myelopathy can be classified as hyperacute (<12 hours), acute/subacute (1 to 21 days), or chronic progressive (progression beyond 21 days).
- In spinal cord infarction, the rapid onset of severe deficits reaching nadir within a few hours (up to 12 hours) is typical and occurs in approximately 80% of patients.
- Most patients with idiopathic or disease-associated transverse myelitis reach nadir in 1 to 21 days.
- The natural evolution of attacks of myelitis associated with central nervous system inflammatory demyelinating diseases is subacute development reaching nadir within 21 days with potential plateau, followed by subsequent improvement that may be sped up by intervening with treatment.
- AQP4-IgG–seropositive NMOSD can coexist with systemic autoimmune disorders, including systemic lupus erythematosus, Sjögren syndrome, and antiphospholipid syndrome. Testing for AQP4-IgG in such cases is prudent as a seropositive result confirms a coexisting autoimmune neurologic disorder rather than a neurologic manifestation of a systemic connective tissue disease.
- Typical findings of myelitis on neurologic examination include a sensory level across the trunk and an upper motor neuron pattern of weakness, hyperreflexia, spasticity, and extensor plantar responses, but these may take time to develop.
- Detailed evaluation of the gadolinium enhancement pattern on MRI is critical in the evaluation of myelopathies as it can provide clues to determine specific etiologies.
- A lesion extending less than three vertebral segments (a short lesion) is most suggestive of multiple sclerosis, and multiple peripheral short T2 hyperintensities within the spinal cord with or without typical brain lesions is strongly suggestive of multiple sclerosis.
- Multiple sclerosis lesions are usually wedge-shaped on axial images and involve the periphery of the cord in either the lateral or dorsal columns.
- Short lesions are less common with AQP4-IgG–seropositive NMOSD, occurring in about 15% of patients.
- An isolated longitudinally extensive T2-hyperintense lesion that extends over three or more vertebral segments is typical of AQP4-IgG–seropositive NMOSD and supports this diagnosis over MS.
- Similar to AQP4-IgG–seropositive NMOSD, MOG-IgG–associated disorder myelitis is frequently associated with longitudinally extensive transverse myelitis, although often multifocal cord lesions are seen rather than the solitary lesion typical of AQP4-IgG–seropositive NMOSD.
- Patients with MOG-IgG–associated disorder often have conus involvement.
- Linear dorsal subpial enhancement extending inward from the posterior aspect of the cord and spanning over multiple vertebral segments is seen in approximately 60% of cases of spinal cord sarcoidosis. When this dorsal subpial enhancement is accompanied by central canal enhancement, an axial trident sign can be seen, which is very suggestive of spinal cord sarcoidosis.
- Obtaining an MRI of the brain is standard in the evaluation of autoimmune myelopathy, and the features of the lesions detected can help suggest the underlying diagnosis.
- AQP4-IgG and MOG-IgG are two important antibody biomarkers of transverse myelitis that should be tested in patients with transverse myelitis in whom the clinical and paraclinical findings are not suggestive of MS.
- For AQP4-IgG and MOG-IgG, serum yields the optimal sensitivity (more so than CSF) and cell-based assays are the most reliable.
- Care is needed with low-positive MOG-IgG results as false positives can occur, particularly when ordered in low-probability situations, and the positive MOG-IgG test result should not replace clinical judgment.
- With AQP4-IgG cell-based assays, false positives are extremely rare, although with older-generation techniques, false positives at low titer can be found.
- Several neural autoantibodies are associated with paraneoplastic myelopathies, and the most commonly encountered are amphiphysin and collapsin response mediator protein-5 (CRMP-5)/anti-CV2.
- The majority of autoimmune/inflammatory myelitis episodes will be accompanied by an elevated CSF white blood cell count, and its absence should at least raise consideration of alternative etiologies (eg, vascular myelopathies).
- Understanding the differential diagnosis of autoimmune/inflammatory myelopathy is crucial as many diagnostic pitfalls can lead the clinician to the wrong diagnosis.

- For most autoimmune/inflammatory myelopathies, after reasonable exclusion of alternative etiologies such as extrinsic compression, spinal cord infarction, and infections, expert consensus recommends prompt administration of high-dose IV corticosteroids with 1 g IV methylprednisolone once daily for 5 days.
- In patients with myelitis as a manifestation of central nervous system inflammatory demyelinating disease with severe neurologic deficits despite steroids, plasma exchange should be considered.

ARTICLE 4: INFECTIOUS MYELOPATHIES

Michel Toledano, MD. Continuum (Minneap Minn). February 2021;
27 (1 Spinal Cord Disorders):93–120.

ABSTRACT

PURPOSE OF REVIEW:

This article reviews infectious etiologies of spinal cord dysfunction, emphasizing the importance of recognizing common clinicoradiographic syndromes and interpreting them in the context of exposure risk and individual host susceptibilities.

RECENT FINDINGS:

This article discusses the shifting spectrum of neurologic infectious diseases, the growing population of patients who are immunocompromised, and the emergence of effective antiretroviral therapies. In addition, it discusses new molecular and serologic tests that have the potential to enhance our ability to rapidly and accurately diagnose infectious diseases of the spine.

SUMMARY:

When evaluating patients with suspected infectious myelopathies, it is imperative to narrow the range of pathogens under consideration. The geography, seasonality, and clinicoradiographic presentation and immunocompetence status of the patient define the range of potential pathogens and should guide testing and initial management.

KEY POINTS

- Infections can result in spine pathology through direct invasion of neural structures or by immune-mediated mechanisms triggered by systemic infection in the absence of neuroinvasion.
- Although considerable overlap exists, recognizing common clinicoradiographic syndromes is critical when generating a differential diagnosis for infectious myelopathies.
- The sensitivity of CSF varicella-zoster virus polymerase chain reaction starts decreasing steadily the further away from symptom onset. A low serum to CSF IgG ratio demonstrating intrathecal production of antibodies is more sensitive.
- Varicella-zoster virus myelitis can occur in the absence of a characteristic herpes zoster rash.
- The myelitis associated with *Mycoplasma pneumoniae* is likely caused by parainfectious or postinfectious immune-mediated mechanisms.
- Meningomyelitis is the most common spinal cord manifestation of syphilis.
- Treponemal tests remain positive for life following infection. Negative treponemal tests essentially rule out a diagnosis of syphilis.
- Elsberg syndrome is characterized by subacute onset of sacral myeloradiculitis and is commonly associated with herpes simplex virus type 2.
- Meningoradiculitis is the most common spinal manifestation of *Borrelia burgdorferi*.
- Neuroschistosomiasis can present as an insidious lumbosacral myeloradiculitis.

- Human T-cell lymphotropic virus type 1–associated myelopathy presents with slowly progressive proximal greater than distal spastic paraparesis and early urinary retention.
- Human immunodeficiency virus–associated vacuolar myelopathy occurs most commonly in advanced infection, but the pathophysiology does not seem to be caused by viral cord infection or inflammation.
- Although tabes dorsalis was common in the preantibiotic era, it is only rarely seen in contemporary practice.
- The clinical presentation of poliomyelitis is usually monoparesis with reflex loss.
- Despite the strong epidemiologic link with enterovirus D68, the etiology of epidemic acute flaccid myelitis remains elusive.
- Viremia is short-lived with most flaviviruses, and polymerase chain reaction is insensitive. Blood or CSF IgM in the acute setting establishes the diagnosis.
- *Aspergillus* can present with spinal cord ischemia and hemorrhage.
- Ampicillin should be initiated empirically in cryptogenic spinal cord abscess for *Listeria* coverage.
- Tuberculous spondylitis (Pott disease) is the most common spinal manifestation of tuberculosis.
- Fever is present in less than 50% of patients with pyogenic spondylodiskitis or epidural abscess.
- Fungal and mycobacterial infection can cause adhesive arachnoiditis, resulting in spinal block and myelopathy with or without syringomyelia.
- Unlike intracerebral disease, which predominantly involves the brain parenchyma, most spinal neurocysticercosis occurs in the subarachnoid space, resulting in compressive myelopathy.

ARTICLE 5: NEOPLASTIC MYELOPATHIES

Amy A. Pruitt, MD, FAAN. Continuum (Minneap Minn). February 2021;
27 (1 Spinal Cord Disorders):121–142.

ABSTRACT

PURPOSE OF REVIEW:

This article reviews the current classification system of primary spinal cord tumors and explores evolving diagnostic and therapeutic strategies for both primary tumors and metastatic tumors to various compartments of the spinal cord.

RECENT FINDINGS:

The 2016 World Health Organization classification system allows for more precise prognostication of and therapy for spinal cord tumors and has identified new entities, such as the diffuse midline glioma, H3 K27M mutant. Whole-exome sequencing reveals that the genetic background of primary glial spinal cord neoplasms differs from that of their intracranial histologic counterparts in ways that can potentially influence therapy. Targeted and immune checkpoint therapies have improved survival for patients with melanoma and lung cancer and have simultaneously produced novel complications by enhancing radiation toxicity in some cases and by facilitating the emergence of novel autoimmune and paraneoplastic syndromes involving the spinal cord, such as neuromyelitis optica spectrum disorder and syndromes associated with anti-Hu and collapsin response mediator protein-5 (CRMP-5) antibodies. These conditions must be distinguished from tumor or infection. Epidural spinal cord compression treatment paradigms have changed with the advent of robotic surgery and advances in radiation therapy.

SUMMARY:

Neoplastic myelopathies subsume a wide spectrum of pathologies. Neoplastic cord involvement may be primary or secondary and may be approached diagnostically by the particular spinal cord compartment localization. Primary spinal cord tumors account for only 2% to 4% of primary

central nervous system tumors, ranging from low-grade glial neoplasms to malignant tumors. Metastatic malignancy to the epidural or leptomeningeal spaces is more common than primary cord tumors. Differential diagnoses arising in the course of evaluation for cord tumors include myelopathies related to radiation or chemotherapy and paraneoplastic syndromes, all of which are sources of significant morbidity. Knowledge of genetic syndromes and the biologic behavior of diverse histologies together with selective application of surgery, radiation, and targeted therapies can facilitate diagnosis, minimize surgical morbidity, and prolong quality of life.

KEY POINTS

- Although spinal cord tumors represent only 2% to 4% of all primary central nervous system tumors, they cause significant morbidity and are often confused clinically and radiographically with non-neoplastic processes.
- Neoplastic myelopathies are classified by the compartment affected as intramedullary, intradural-extramedullary, and extradural tumors. Differential diagnostic considerations and workup are dictated by the particular neuroanatomic compartment involved.
- Overall, metastatic tumors, which are usually in the epidural space, account for many more cases of adult spinal cord tumors than do primary spinal tumors, whereas in children (in whom primary tumors are more common), the intramedullary compartment is the most common tumor site.
- Ependymomas are the most common primary intramedullary spinal tumors in adults, but the most common primary spinal tumors overall in adults are meningiomas.
- Lung, breast, prostate, thyroid, and renal cell cancers represent the majority of spinal metastatic tumors, the vast majority of which are extradural. Leptomeningeal dissemination is seen most frequently with adenocarcinoma of the breast and lung, non-Hodgkin lymphoma, melanoma, and gastrointestinal tumors.
- Ependymomas are the most common intramedullary primary spinal cord tumor in all age groups.
- Back, radicular, or central pain, often asymmetric and without motor involvement, is the most common symptom preceding the diagnosis of intramedullary neoplasm.
- Spinal glial tumors show no association between increasing grade of malignancy and patient age at diagnosis.
- Cellular ependymomas may be World Health Organization (WHO) grade II or grade III and arise from the intraspinal canal, usually in the cervical and thoracic regions; myxopapillary ependymoma, a WHO grade I tumor, is most frequently seen in the conus medullaris arising from the filum terminale, where they comprise 90% of tumors.
- Ependymomas are often well-demarcated isointense lesions that enhance with gadolinium.
- Gross total resection of astrocytomas is unlikely, but ependymomas, which are often encapsulated, are more amenable to total resection.
- Recognized for the first time in the 2016 WHO classification of tumors is the WHO grade IV diffuse midline glioma, H3 K27M mutant, previously called diffuse intrinsic pontine glioma.
- Most H3 K27M-mutant diffuse midline gliomas occur in the thalamus and brainstem, but brainstem cases can extend to the spinal cord and show a propensity for intramedullary drop metastases and leptomeningeal dissemination.
- Pilocytic astrocytomas of the spinal cord account for about 11% of pediatric spinal cord tumors. These often are associated with neurofibromatosis type 1. Most are well circumscribed and WHO grade I.
- Hemangioblastoma, a WHO grade I tumor, is rare except in von Hippel-Lindau syndrome, an autosomal dominant disorder characterized by chromosome 3p deletion; von Hippel-Lindau syndrome accounts for up to 30% of cases of hemangioblastoma.
- Meningiomas are the most common primary spinal cord neoplasm in adults, accounting for one-fourth of all primary spinal cord tumors. The majority of meningiomas are WHO grade I slow-growing tumors. Genetic predisposition (neurofibromatosis type 2) and prior radiation are risk factors.
- Radiosurgery is used for incomplete resection or recurrence of spinal meningioma, and protons are gaining a larger role in the treatment of spinal meningioma. No role for chemotherapy has been established, but intracranial meningiomas have been reported to respond to everolimus, sunitinib, and bevacizumab.

- Schwannomas are benign nerve sheath tumors, the majority of which are WHO grade I. They represent nearly 30% of spinal root tumors, and multiple schwannomas can be found in patients with neurofibromatosis type 2 or schwannomatosis.
- Extradural metastases are most likely to occur from lung, breast, prostate, thyroid, and renal cancers, whereas leptomeningeal dissemination of solid tumors is most commonly seen from breast and lung cancers, melanoma, non–Hodgkin lymphoma, and gastrointestinal tumors.
- Treatment of epidural cord compression is palliative, with the principle goals of pain relief, preservation of neurologic function, maintenance of spinal stability, and improvement in quality of life while avoiding the toxic consequences of radiation and chemotherapy.
- A thin rim of peripheral enhancement and a flame-shaped appearance in the region of enhancement at the superior and inferior margins should suggest a non–central nervous system metastatic intramedullary tumor rather than one of primary spinal cord origin.
- Compared with patients with nonparaneoplastic neuromyelitis optica spectrum disorder (NMOSD), patients with paraneoplastic NMOSD are older at symptom onset and more frequently male. Thus, older patients presenting with NMOSD, particularly if male, should be investigated for neoplasia.
- In the context of immune checkpoint inhibitor treatment, paraneoplastic antibody–associated spinal cord syndromes have emerged. Reported antibodies include anti-Hu/ANNA-1, CRMP-5/anti-CV2, and aquaporin-4 IgG.

ARTICLE 6: METABOLIC AND TOXIC MYELOPATHIES

Natalie Elizabeth Parks, MD. Continuum (Minneap Minn). February 2021;
27 (1 Spinal Cord Disorders):143–162.

ABSTRACT

PURPOSE OF REVIEW:

This article describes the clinical presentation, relevant diagnostic investigations, and treatment of metabolic and toxic myelopathies.

RECENT FINDINGS:

Metabolic myelopathies, including those due to deficiency of vitamin B_{12}, folate, copper, or vitamin E, are preventable and typically respond to supplementation. In metabolic myelopathy, early recognition and treatment are important to reduce morbidity, particularly due to subacute combined degeneration of the spinal cord. Toxic myelopathies, including those due to medical interventions (eg, methotrexate, radiation), dietary toxins (eg, lathyrism, konzo), and drugs of abuse (eg, heroin), typically result in permanent neurologic deficits. Toxic myelopathy due to hepatic dysfunction may be reversible if patients receive early intervention, whereas nitrous oxide myelopathy responds to vitamin B_{12} replacement and cessation of exposure. In toxic myelopathy, it is best to avoid the provoking factor when possible or attempt to mitigate risk by identifying risk factors for developing myelopathy.

SUMMARY:

Metabolic and toxic myelopathies are important causes of morbidity that require a high index of suspicion for diagnosis.

KEY POINTS

- Vitamin B_{12} deficiency is common among older adults.

- Subacute combined degeneration of the spinal cord presents with posterior column dysfunction (reduced vibration/proprioception) along with variable severity of lateral column dysfunction (upper motor neuron signs).
- Vitamin B_{12} deficiency may be present despite serum cobalamin within the normal range, although plasma methylmalonic acid or plasma homocysteine, or both, may be elevated.
- The treatment for subacute combined degeneration of the spinal cord due to vitamin B_{12} deficiency is IM or subcutaneous cyanocobalamin 1000 mcg/d for 5 days followed by 1000 mcg once per month.
- Vitamin B_{12} replacement should be given indefinitely following subacute combined degeneration of the spinal cord due to vitamin B_{12} deficiency.
- Nitrous oxide causes inactivation of vitamin B_{12}, which may result in subacute combined degeneration of the spinal cord.
- Folate deficiency is uncommon since the introduction of national fortification programs aimed at improving folate levels among reproductive-age women to reduce neural tube defects in their offspring.
- Serum folate level reflects recent folate intake, whereas red blood cell folate level reflects intake over approximately the past 3 months.
- Copper deficiency is an underrecognized cause of subacute combined degeneration of the spinal cord.
- Copper deficiency may be caused by bariatric surgery, celiac disease, or excessive zinc intake as a supplement or in denture cream.
- Serum copper and serum ceruloplasmin levels are typically low in copper deficiency.
- Vitamin E deficiency that results in spinocerebellar ataxia is an increased risk among those with impaired fat absorption from disorders such as cystic fibrosis and rare genetic conditions, including ataxia with vitamin E deficiency and abetalipoproteinemia.
- Grass peas (*Lathyrus sativus*) contain a neurotoxin that may result in neurolathyrism manifesting with acute-onset spastic paraparesis.
- Bitter cassava contains cyanogens that may cause konzo, manifesting with spastic paraparesis, due to cyanide toxicity.
- Subacute combined degeneration of the spinal cord may occur with intrathecal methotrexate, which is a folate antagonist.
- Tumor necrosis factor-α inhibitors and immune checkpoint inhibitors are associated with transverse myelitis.
- Subacute myelo-optico-neuropathy was caused by clioquinol, a metal chelator that may cause copper deficiency.
- Reintroduction of heroin following a period of abstinence may cause acute-onset complete myelopathy.
- Radiation myelopathy is a delayed effect of radiation occurring 6 to 24 months after radiation exposure.
- Hepatic myelopathy occurs in chronic liver disease with portosystemic shunting.
- Decompression myelopathy occurs within 1 hour of diving and is treated with hyperbaric oxygen therapy, typically with good recovery.

ARTICLE 7: SPONDYLOTIC AND OTHER STRUCTURAL MYELOPATHIES

Shamik Bhattacharyya, MD, MS. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):163–184.

ABSTRACT

PURPOSE OF REVIEW:

This article highlights both common structural causes of myelopathy, such as spondylotic disease, and infrequent but treatable causes, such as syringomyelia, spinal cord herniation, arachnoid cyst, arachnoid band and web, epidural lipomatosis, Hirayama disease, and arachnoiditis.

RECENT FINDINGS:

Neuroimaging improvements and availability have uncovered many structural abnormalities in the spines and spinal cords of patients who were asymptomatic or minimally symptomatic. Recent published clinical series have improved our knowledge of the natural history of structural abnormalities and the risks of intervention versus conservative management.

SUMMARY:

Myelopathy from a suspected structural cause is a common reason for neurologic consultation. Correlation between the history, examination, and imaging are especially important to determine whether intervention is necessary or conservative management is the best option.

KEY POINTS

- Cervical spondylotic myelopathy is caused by degenerative disease of the cervical spine resulting in narrowing of the spinal canal.
- Cervical spondylotic myelopathy is overdiagnosed in some patients (symptoms misattributed to imaging findings) and missed in others (mild symptoms that are not investigated).
- Congenital narrowing of the spinal canal is a frequent risk factor for the development of cervical spondylotic myelopathy.
- Patients with Klippel-Feil syndrome clinically have decreased neck mobility, a low posterior hairline, and a short neck; imaging shows fusion of multiple cervical vertebral bodies.
- Cervical spondylotic myelopathy is likely caused by a combination of canal narrowing, stretch of the spinal cord over the stenotic region, and microvascular ischemia.
- Cervical spondylotic myelopathy can have acute, subacute, and chronic presentations.
- Acute cord injury from extension in patients with cervical spondylotic myelopathy causes central cord syndrome in which patients have urinary retention and greater weakness in their arms than in their legs.
- Chronic cervical spondylotic myelopathy causes initial symptoms of progressive gait disorder.
- Lack of neck pain does not exclude cervical spondylotic myelopathy.
- Bladder and bowel sphincter dysfunction are atypical in chronic progressive cervical spondylotic myelopathy.
- The Babinski sign is not very sensitive for cervical spondylotic myelopathy and may be absent in early disease. The Hoffman sign may be positive more often.
- MRI of the cervical spine without contrast is the preferred study to evaluate for cervical degenerative disease.
- X-ray of the cervical spine is useful to evaluate instrumentation and for dynamic instability of bony structures with flexion and extension of the neck.
- The majority of older adults will have degenerative changes of the cervical spine on MRI.
- Categorization as moderate or severe stenosis of the cervical spine based on the degree of CSF obliteration has modest correlation with clinical symptoms.
- Clinical myelopathy from cervical spinal stenosis can occur without any T2 cord signal changes.
- The presence or absence of cord signal hyperintensity does not correlate with outcome after surgery.
- T2 hyperintensity in the spinal cord from cervical spondylotic myelopathy may have a snake-eye appearance, with areas of hyperintensity in the anterior horns bilaterally.
- The natural history of untreated cervical spondylotic myelopathy is unclear and has considerable variability.
- Minor trauma is an unusual precipitant of acute myelopathy in patients with asymptomatic severe cervical spine stenosis.
- Patients with untreated cervical spondylotic myelopathy often have a stepwise course, with periods of stability and then episodes of acute deterioration. Some patients relentlessly progress, whereas others can remain stable for years.
- Conservative therapy for cervical spondylotic myelopathy generally involves physical therapy and gentle cervical spine range-of-motion exercises.
- Cervical spinal stenosis can be decompressed via either an anterior or a posterior approach. No clear consensus exists on which approach is superior.

- C5 radiculopathy can be a postoperative complication of cervical spine surgery.
- Upper extremity strength recovers best following surgery for cervical spondylotic myelopathy, whereas recovery of leg strength and sensory dysfunction are less complete.
- An enlarged central canal is often incidentally found on imaging and is generally not pathogenic.
- Chiari type I malformation can be clinically silent and associated with syringomyelia.
- Syrinx formation from spinal cord injury can be delayed by many years.
- Free CSF flow impairment in the spinal subarachnoid space is a common theme among the different predisposing causes of syringomyelia.
- A small midline syrinx causes interruption of crossing spinothalamic tracts and a capelike distribution of numbness to pain and temperature.
- The natural history of syringomyelia is unpredictable. Many patients remain asymptomatic, whereas others can progress with time.
- Idiopathic spinal cord herniation is characterized by a defect through which the spinal cord is displaced, typically in the ventral dura and generally presenting with progressive myelopathy.
- Spinal arachnoid cysts are intradural-extramedullary cysts in the subarachnoid space that can cause myelopathy by compression of the spinal cord.
- Spinal arachnoid webs are intradural bands of arachnoid tissue that usually attach to the dorsal surface of the spinal cord.
- Spinal arachnoid webs may not be seen directly on MRI but rather inferred from change in caliber of the spinal cord with dorsal cord indentation.
- Spinal epidural lipomatosis refers to accumulation of fat in the epidural space that can be asymptomatic or cause symptoms from compression of nerve roots or the spinal cord.
- Hirayama disease is characterized by the insidious onset of weakness and atrophy of the hand and forearm, predominantly in young males in their teens or twenties without other cranial or pyramidal signs.
- In Hirayama disease, when imaged with the neck extended and flexed, the diameter of the dural sac decreases during flexion with corresponding stenosis and pressure on the spinal cord without movement in the bony elements.
- Spinal adhesive arachnoiditis refers to progressive fibrosis of the arachnoid membrane with injury to the nerve roots, tethering of the spinal cord, and disruption of free flow of CSF.
- Arachnoiditis often presents after a time delay from the initial spinal injury ranging from weeks to years.

ARTICLE 8: HEREDITARY MYELOPATHIES

John K. Fink, MD. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):185–204.

ABSTRACT

PURPOSE OF REVIEW:

This article guides clinicians in the clinical recognition and differential diagnosis of hereditary myelopathies.

RECENT FINDINGS:

Rather than a disease, a disease process, or relating to specific cellular vulnerability, the term *hereditary myelopathy* refers to diverse inherited disorders in which major aspects of the clinical syndrome reflect disturbance of elements within the spinal cord (specifically, the dorsal columns and dorsal root ganglia, corticospinal tracts, and anterior horn cells). It is important to note that the clinical features of almost all hereditary myelopathies reflect not only disturbance of elements within the spinal cord but also disturbance of extraspinal structures (particularly,

but not limited to, peripheral nerves and the cerebellum) and that these extraspinal clinical features can be very helpful in recognizing specific myelopathy syndromes. The value of classifying disorders as inherited myelopathies lies primarily in facilitating their clinical recognition and differential diagnosis. It is useful to recognize that many hereditary myelopathies conform to one of four clinical paradigms: (1) spinocerebellar ataxia, (2) motor neuron disorder, (3) leukodystrophy, or (4) distal motor-sensory axonopathy predominantly affecting the central nervous system. Although they are myelopathies, spinal dysraphisms such as spina bifida and myelomeningocele are not included in this context because they are not usually due to single-gene mutation and have low hereditability.

SUMMARY:

This article illustrates clinical paradigms of hereditary myelopathy with clinical examples emphasizing the spectrum, clinical recognition, and differential diagnosis of hereditary myelopathies.

KEY POINTS

- In addition to symptoms arising from disturbance within the spinal cord, neurologic involvement in nearly all hereditary myelopathies includes structures outside the spinal cord.
- Many hereditary myelopathic syndromes can be recognized as one of four clinical paradigms: (1) spinocerebellar ataxia, (2) motor neuron disorder, (3) leukodystrophy, or (4) central nervous system–predominant distal motor-sensory axonopathy.
- Spinocerebellar degenerations (eg, Friedreich ataxia, spinocerebellar ataxia type 3, Bassen-Kornzweig syndrome, and vitamin E deficiency) are recognized by a combination of progressive cerebellar ataxia, often accompanied by peripheral neuropathy; dorsal column (or dorsal root ganglia) impairment (which may cause sensory ataxia); and variable corticospinal tract involvement.
- Spinocerebellar ataxia type 3 is caused by a trinucleotide repeat (CAG) expansion that, like other polyglutamine expansions, is thought to be pathogenic through protein misfolding.
- The vast majority of patients with Friedreich ataxia are homozygous for expanded trinucleotide repeat in the *FXN* gene, which encodes a mitochondrial protein. Rarely, individuals will have an expanded trinucleotide repeat in one *FXN* allele and a point mutation in the other *FXN* allele.
- In primary lateral sclerosis, there is either no evidence of lower motor neuron involvement, or, at most, minimal evidence of chronic denervation is seen on EMG late in the disease. At the other extreme, spinal muscular atrophy is characterized by muscular weakness and atrophy due to anterior horn cell degeneration with preservation of corticospinal tracts.
- Demyelinating peripheral neuropathy, which may accompany childhood-onset leukodystrophies (eg, Krabbe disease and metachromatic leukodystrophy), may be absent in the rare adolescent- and adult-onset forms of these disorders.
- Childhood-onset adrenoleukodystrophy and adolescent- and adult-onset adrenomyeloneuropathy are X-linked disorders in which *ABCD1* gene mutation leads to impaired peroxisomal beta-oxidation and accumulation of very long chain fatty acids systemically.
- Adrenoleukodystrophy/adrenomyeloneuropathy phenotypes include rapidly progressive childhood, adolescent, and adult cerebral forms; slowly progressive myelopathic forms (characterized by slowly progressive spastic paraparesis and peripheral neuropathy, often with complete sparing of the brain); and isolated adrenal insufficiency.
- Clinical distinction of leukodystrophies from axonopathies is based on the presence of additional neurologic findings, particularly cognitive impairment, optic neuropathy, deafness, and sensory disturbance.
- Sensory impairment in uncomplicated motor-sensory axonopathies (eg, uncomplicated hereditary spastic paraplegia) typically results in mild dorsal column impairment affecting longer fibers and manifests as impaired vibration perception in the toes with preservation of other sensory modalities.

- *ATL1*/atlastin gene mutation is the most common cause of childhood-onset autosomal dominant hereditary spastic paraplegia. *ATL1* hereditary spastic paraplegia usually causes nonprogressive infantile-onset spastic gait and resembles spastic diplegic cerebral palsy.
- Central nervous system–predominant distal motor-sensory axonopathy (eg, uncomplicated hereditary spastic paraplegia) can be considered analogous to Charcot-Marie-Tooth disease type 2, in which axonopathy affects predominantly the distal ends of long motor and sensory fibers in the peripheral nervous system.
- *SPAST* mutations are the most common cause of autosomal dominant hereditary spastic paraplegia.

ARTICLE 9: DISORDERS OF THE CAUDA EQUINA

Samantha LoRusso, MD. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):205–224.

ABSTRACT

PURPOSE OF REVIEW:

Cauda equina dysfunction (often referred to as *cauda equina syndrome*) is caused by a diverse group of disorders that affect the lumbosacral nerve roots. It is important to recognize dysfunction of the cauda equina quickly to minimize diagnostic delay and lasting neurologic symptoms. This article describes cauda equina anatomy and the clinical features, differential diagnosis, and management of cauda equina disorders.

RECENT FINDINGS:

The diagnosis of disorders of the cauda equina continues to be a challenge. If a compressive etiology is seen, urgent neurosurgical intervention is recommended. However, many people with clinical features of cauda equina dysfunction will have negative diagnostic studies. If the MRI is negative, it is important to understand the diagnostic evaluation and differential diagnosis so that less common etiologies are not missed.

SUMMARY:

Cauda equina dysfunction most often occurs due to lumbosacral disk herniation. Nondiskogenic causes include vascular, infectious, inflammatory, traumatic, and neoplastic etiologies. Urgent evaluation and surgical intervention are recommended in most cases of compressive cauda equina syndrome. Other types of treatment may also be indicated depending on the etiology.

KEY POINTS

- Cauda equina syndrome results from dysfunction of lumbosacral nerve roots leading to symptoms of urinary retention and incontinence, constipation, bowel incontinence, sexual dysfunction, sensory changes (particularly saddle anesthesia), back pain, and lower extremity weakness.
- Examination findings that suggest cauda equina dysfunction include reduced or absent reflexes in the lower extremities, loss of perineal or lower extremity sensation, reduced rectal tone, and lower extremity flaccid weakness.
- The sensory changes in cauda equina syndrome can be unilateral or bilateral, with the most common areas of involvement being the posterior thighs, buttocks, and perineum.
- No single symptom or sign has been found to have consistently high sensitivity and specificity in diagnosing MRI-positive cauda equina syndrome.
- If any question exists regarding the localization to the cauda equina based on history and examination, then imaging of the entire neuraxis (brain and spinal cord) should be considered.

- Neurosurgery should be consulted immediately in a case of suspected cauda equina dysfunction due to a compressive lesion.
- The degree of neurologic dysfunction before surgery is the most consistent prognostic factor in cauda equina syndrome.
- Disk herniations are the most common cause of cauda equina dysfunction, occurring the majority of the time at the L4-L5 or L5-S1 levels.
- Constitutional symptoms, such as fevers, night sweats, and weight loss, should lead to consideration of an infectious etiology of cauda equina dysfunction in the appropriate clinical setting.
- Elsberg syndrome likely accounts for about 10% of patients with a clinical presentation of cauda equina syndrome and myelitis.
- Myxopapillary ependymomas are the most common primary tumor to affect the cauda equina.
- Sarcoidosis is likely the most common inflammatory disorder that can present with cauda equina dysfunction.
- Trauma, especially from motor vehicle accidents, falls, and gunshot wounds, is a potential cause of cauda equina syndrome, often because of a low lumbar or transverse sacral fracture.
- Although not technically a disorder of the cauda equina, pudendal neuropathy can closely mimic cauda equina syndrome since it originates from the S2 through S4 nerve roots and innervates the perineum.

ARTICLE 10: NEUROIMAGING OF SPINAL CORD AND CAUDA EQUINA DISORDERS

Felix E. Diehn, MD; Karl N. Krecke, MD, FACR. Continuum (Minneap Minn). February 2021; 27 (1 Spinal Cord Disorders):225–263.

ABSTRACT

PURPOSE OF REVIEW:

This article reviews the neuroimaging of disorders of the spinal cord and cauda equina, with a focus on MRI. An anatomic approach is used; diseases of the extradural, intradural-extramedullary, and intramedullary (parenchymal) compartments are considered, and both neoplastic and non-neoplastic conditions are covered. Differentiating imaging features are highlighted.

RECENT FINDINGS:

Although T2-hyperintense signal abnormality of the spinal cord can have myriad etiologies, neuroimaging can provide specific diagnoses or considerably narrow the differential diagnosis in many cases. Intradural-extramedullary lesions compressing the spinal cord have a limited differential diagnosis and are usually benign; meningiomas and schwannomas are most common. Extradural lesions can often be specifically diagnosed. Disk herniations are the most commonly encountered mass of the epidural space. Cervical spondylotic myelopathy can cause a characteristic pattern of enhancement, which may be mistaken for an intrinsic myelopathy. A do-not-miss diagnosis of the extradural compartment is idiopathic spinal cord herniation, the appearance of which can overlap with arachnoid cysts and webs. Regarding intrinsic causes of myelopathy, the lesions of multiple sclerosis are characteristically short segment but can be confluent when multiple. Postcontrast MRI can be particularly helpful, including when attempting to differentiate the long-segment myelopathy of neurosarcoidosis and aquaporin-4 (AQP4)-IgG–seropositive neuromyelitis optica spectrum disorder (NMOSD) and when characterizing spinal cord tumors such as primary neoplasms and metastases. Spinal dural arteriovenous fistula is another do-not-miss diagnosis, with characteristic MRI features both precontrast and postcontrast. Tract-specific white matter involvement can be a clue for

diseases such as subacute combined degeneration, paraneoplastic myelopathy, and radiation myelitis, whereas gray matter–specific involvement can suggest conditions such as cord infarct, viral myelitis, or myelin oligodendrocyte glycoprotein (MOG)-IgG associated disorder.

SUMMARY:

Knowledge of the neuroimaging findings of the many causes of spinal cord and cauda equina dysfunction is critical for both neurologists and neuroradiologists. A structured approach to lesion compartmental location and imaging feature characterization is recommended.

KEY POINTS

- IV administration of gadolinium with postcontrast T1-weighted imaging performed in at least the sagittal plane, if not also the axial plane, is recommended for complete evaluation of suspected intrinsic myelopathy with blood–spinal cord barrier breakdown.
- Inclusion of diffusion-weighted imaging in the MRI protocol is suggested for any hyperacute or acute myelopathy, particularly to help assess for infarct.
- The central canal can be physiologically prominent and thereby evident as a normal variant on MRI; this is thin, usually only a few millimeters in diameter, and should not be confused with a syrinx.
- The normal gray matter is slightly more hyperintense than the white matter, which may simulate abnormal T2 hyperintensity anteriorly and centrally on sagittal T2-weighted images.
- The most common mass in the epidural space is a disk herniation. Most disk herniations are located in the ventral/ventrolateral epidural space and remain in anatomic continuity with their parent disk, a key clue to the diagnosis.
- A highly prevalent finding for dorsal disk herniations is that the abnormal epidural soft tissue still typically maintains continuity with the parent disk in the ventrolateral epidural space, wrapping around the thecal sac dorsally.
- The heterogeneous internal signal characteristics of synovial cysts are wide-ranging and include T1 and T2 hypointensity or hyperintensity or a combination.
- MRI of extradural abscesses may show associated adjacent inflammatory changes, including in the paraspinal soft tissues and bones, or frank findings of spondylodiskitis.
- On CT, ossification of the posterior longitudinal ligament is readily identifiable as flowing ossification along the expected course of the posterior longitudinal ligament in and along the midline at the ventral aspect of the spinal canal.
- After gadolinium administration, T1-weighted images in cervical spondylotic myelopathy often demonstrate a characteristic narrow transverse (pancakelike) band of cord enhancement at or slightly caudal to the focus of spinal stenosis.
- Findings that suggest a relatively high grade and symptomatic lumbar stenosis include redundancy/tortuosity of the cauda equina nerve roots and the sedimentation sign.
- If Hirayama disease is suspected clinically or based on neutral position MRI, flexion MRI should be performed as it can demonstrate findings to better advantage and increase diagnostic confidence.
- For enhancing intradural-extramedullary masses, the two most likely possibilities are meningiomas and nerve root sheath tumors (schwannomas and neurofibromas).
- Characteristic, although not entirely specific, imaging findings suggestive of spinal meningioma include dural tail(s) of contrast enhancement and avid and homogeneous enhancement of the lesion, which may be relatively T2 hypointense because of cellularity or calcification.
- When an isolated, incidental, small enhancing nodule of the cauda equina nerve roots is encountered, the most likely consideration is a nerve sheath tumor.
- Myxopapillary ependymomas are usually relatively large, oval or sausage shaped, well circumscribed, T2 hyperintense, and avidly enhancing.
- Varied appearances of arachnoiditis include nerve roots that are irregularly clumped, clumped into a mass of neural tissue centrally, or dispersed to the margins of the dura (the empty thecal sac sign).

- The most common location for an arachnoid cyst is dorsal to the thoracic cord; although these lesions tend to be well circumscribed, their wall is often imperceptible, especially on MRI.
- Typically, arachnoid webs cause mass effect on and flattening of the dorsal cord, with a characteristic but not pathognomonic morphology termed the scalpel sign that is best seen sagittally on either T2-weighted images or CT myelography.
- On MRI or CT myelography, the characteristic findings of idiopathic spinal cord herniation include ventral displacement of a short segment of cord that is focally distorted/kinked, with the subarachnoid space being lost ventrally and expanded dorsally.
- Neoplasms tend to expand the spinal cord, to have a mass or masslike appearance, and to enhance. The presence of associated adjacent cord cysts or hemorrhage typically suggests a neoplastic process.
- Internal or adjacent heterogeneity, including related to the presence of cystic change (polar cyst) or hemorrhage (T2-hypointense hemosiderin cap sign) at the margins of the lesion, is more commonly encountered with ependymoma than with astrocytoma.
- Although astrocytomas usually enhance, they may not; when they do enhance, it may be fairly mild in amount/intensity or ill-defined.
- Hemangioblastomas have a propensity to be eccentrically located and abut the surface of the spinal cord, with an enhancing pial/subpial nodule especially dorsally. The neoplasms may present as cystic or partially cystic lesions with nodular enhancement.
- Two highly specific enhancement characteristics of intramedullary spinal cord metastases are reasonably prevalent in these lesions and not commonly seen in primary cord tumors: the rim and flame signs.
- Characteristically, multiple sclerosis lesions are short segment (fewer than two segments craniocaudally), asymmetrically and eccentrically located (not involving the entire cross-sectional area), and affect white or white plus gray matter. Common cross-sectional locations include the lateral and dorsal aspects of the cord.
- Three characteristic features of aquaporin-4-IgG–seropositive neuromyelitis optica spectrum disorder that may be present are involvement of the cervicomedullary junction, T1-hypointense components of the lesion, and foci on axial images that are at least as T2 hyperintense as CSF (bright spotty lesions).
- The enhancement pattern in aquaporin-4-IgG–seropositive neuromyelitis optica spectrum disorder is often patchy and heterogeneous, and ring or partial ring enhancement can strongly suggest the diagnosis over neurosarcoidosis.
- The classic finding of neurosarcoidosis of the cord is a long-segment myelopathy, with characteristic enhancement at the dorsal subpial/pial aspect of the cord.
- Selective viral involvement of the anterior horn cells can result in acute flaccid paralysis. An example is the 2014 outbreak related to enterovirus D68, which typically resulted in long-segment T2 hyperintensity of the central gray matter, particularly in the cervical cord.
- When present, the spinal cord T2 hyperintensity of paraneoplastic myelopathy is often long segment, symmetric, and tract specific, such as confined to the lateral or dorsal columns.
- MRI is relatively insensitive for subacute combined degeneration but, when positive, demonstrates T2 hyperintensity of the dorsal columns with an inverted V or inverted rabbit ears morphology on axial images, especially in the cervical and upper thoracic cord.
- A key ancillary imaging clue for radiation-induced myelopathy is associated vertebral body marrow T1 hyperintensity (fatty marrow replacement) encompassing the radiation port.
- Similar to arterial infarcts in the brain, restricted diffusion (with high signal on diffusion-weighted imaging and low signal on apparent diffusion coefficient images) can be seen in acute spinal cord infarcts.
- On T2-weighted images of an anterior spinal artery infarct, preferential involvement of the gray matter manifests as an H-shaped or butterfly-shaped appearance, or as an owl-eyes or snake-eyes sign.
- Because of the shared blood supply of the spinal cord and vertebral bodies, vertebral body infarcts may be observed, usually 1 to 2 weeks after the initial presentation of spinal cord infarct.
- Enhancement of the ventral cauda equina roots may be evident in the subacute phase of spinal cord infarct.

- **The classic finding of a spinal dural arteriovenous fistula is increased posterior pial serpentine vascularity, typically best seen on T2-weighted sagittal images as prominent flow voids within the dilated veins along the dorsal cord surface.**
- **Adjacent hemorrhage in the spinal cord extending craniocaudally away from a cavernous malformation is relatively prevalent; a 2020 retrospective series demonstrated that this finding is more common than some classic features of these lesions, such as popcorn morphology and T2-hypointense rim.**