Spondylotic and Other Structural Myelopathies

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REVIEW ARTICLE

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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.**

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INTRODUCTION

Structural causes of myelopathy are important to diagnose early and accurately because symptoms can often be improved or at least arrested with timely intervention. Despite the widespread availability of imaging, structural etiologies are the leading source of nontraumatic spinal cord injury.[1] On the other hand, most practicing neurologists know of patients who have undergone spinal surgeries with little benefit or were worse off after the surgery. Accurate estimates of "unnecessary surgeries" are hard to come by, but a single-center study showed about 17% of patients recommended to undergo spinal surgeries were unlikely to benefit from them.[2] The neurologist has a critical role in assessing the cause of neurologic symptoms, prognosis, and risks and benefits of intervention in structural causes of myelopathy.

CERVICAL SPONDYLOTIC MYELOPATHY

Cervical spondylotic myelopathy is defined broadly as spinal cord dysfunction caused by degenerative disease of the cervical spine with narrowing of the vertebral canal through which the spinal cord traverses. Because the mechanisms of narrowing of the vertebral canal and injury to the spinal cord are usually multifactorial, involving bony, soft tissue, and vascular structures, some prefer to refer to cervical spondylotic myelopathy more generally as degenerative

cervical myelopathy. Accurate estimates of the incidence and prevalence of cervical spondylotic myelopathy are unavailable. This paucity of data for a common disease results from lack of accurate coding, lack of definite correlation between imaging findings and symptoms, lack of accuracy in presurgical diagnoses (not all who received surgery for cervical spondylotic myelopathy likely had the disease), and lack of recognition of cervical spondylotic myelopathy in patients with mild symptoms. With these limitations, an estimate of the prevalence of cervical spondylotic myelopathy in North America is 605 per 1 million.[3]

Pathogenesis

As with many other slowly progressive diseases, the final clinical development of cervical spondylotic myelopathy is frequently preceded by years of subclinical changes. Susceptibility factors for clinical disease are congenital spinal stenosis, osteoarthritis, ossification of the ligamentum flavum and posterior longitudinal ligament, inflammatory arthritides such as rheumatoid arthritis or ankylosing spondylitis, Down syndrome, and Klippel-Feil syndrome. In congenital stenosis of the cervical spine, patients developmentally have a narrow cervical spinal canal and are predisposed to get cervical spondylotic myelopathy from relatively minor spinal degenerative disease. Based on large series of cadaveric measurements, an anterior-posterior cervical spinal canal measurement of 13 mm or less may define congenital narrowing of the cervical canal (FIGURE 7-1).[4] Others have countered that absolute measurement of the spinal canal width is less important than the relative fraction of space occupied by the spinal cord compared to the width of the canal itself. This parameter is known as the spinal cord occupancy ratio, and values greater than 70% may more accurately capture the concept of a congenitally narrow canal.[5] Among different inflammatory arthritis syndromes, rheumatoid arthritis frequently affects the cervical spine; MRI of the cervical spine shows evidence of degenerative disease in more than 70% of patients with chronic rheumatoid arthritis.[6] The cause of this tropism is unclear but may have to do with the exclusively synovial joints in the upper cervical spine.

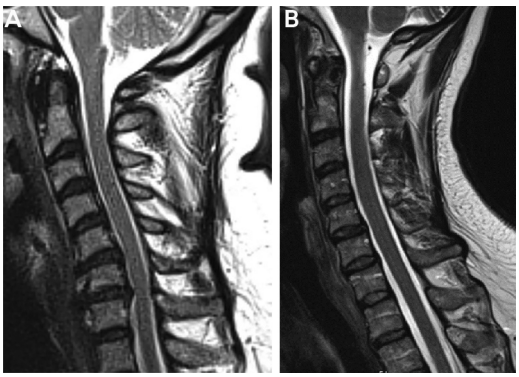

FIGURE 7-1
Congenital spinal stenosis. *A*, Sagittal T2-weighted MRI of the cervical spine showing congenital stenosis. The anterior-posterior distance of the thecal sac averages about 10 mm, putting the patient at risk for spondylotic myelopathy from mild degenerative disease. *B*, For comparison, sagittal T2-weighted MRI of the cervical spine of a patient without congenital spinal stenosis.

Klippel-Feil syndrome is characterized by congenital fusion of multiple cervical vertebral bodies, resulting in spinal instability and propensity for injury. Clinically, patients with Klippel-Feil syndrome have decreased neck mobility, a low posterior hairline, and a short neck. Most patients with Klippel-Feil syndrome do not have all these features, and no single unifying genetic basis for

this syndrome has been identified (the syndrome is seen as part of other discrete genetic disorders).

Among nonbony susceptibility factors, ossification of the posterior longitudinal ligament demonstrates the importance of ligamentous structures in cervical spondylotic myelopathy (FIGURE 7-2). This disorder was traditionally thought to be more common in Japan, presumably from genetic influence, but 2016 data show that more than 20% of patients with cervical spondylotic myelopathy in the United States have posterior longitudinal ligament ossification as well.[7] Many patients have questions about the risk of cervical spondylotic myelopathy from various occupations. Anecdotal reports indicate that workers experiencing high levels of cervical spine stress, such as those in construction who use jackhammers, may be at higher risk. However, definitive data are lacking. A small study of grape-growing farmworkers with extension strain concluded that they have a higher risk of degenerative spine disorder.[8]

In spondylotic cervical myelopathy, although narrowing of the spinal canal is always present, the narrowing itself is not enough to cause disease. Clinical symptoms result from a combination of static stenosis, dynamic stress of movement, and microvascular ischemia. In most cases, the first stage of spondylotic disease is thought to be degenerative disk disease driven by desiccation of the inner layer of the intervertebral disk (nucleus pulposus). Disk degeneration results in increased stress on the bony and ligamentous elements of the cervical spine, which is hypothesized to lead to disk fragment herniation, ligament hypertrophy and ossification, misalignment of vertebral bodies, osteophyte growth, and uncovertebral joint hypertrophy.[3] In most patients, canal stenosis is caused by a combination of these disease processes.

Static stenosis alone, however, may not cause myelopathy. This was seen in a large-scale cervical spine MRI study of more than 1200 asymptomatic subjects. About 5% had significant spinal cord compression, and 2.3% even had cord signal changes.[9] The importance of other factors was also demonstrated in an animal study of 14 dogs, in which the spinal canal was experimentally narrowed by about 29%. In 12 of the 14 dogs, the onset of clinical myelopathy was delayed an average of 7 months, after which the animals developed progressive signs.[10] A proposed mechanism of delayed injury is microcirculatory ischemia caused by extension of the spinal cord over a stenotic region. This may explain the predilection for injury of the anterior horns of the spinal cord, which are supplied by the anterior spinal artery and are near the end arterial zone. Other mechanisms include neuroinflammatory injury induced by chronic microglia/macrophage activation, endothelial cell

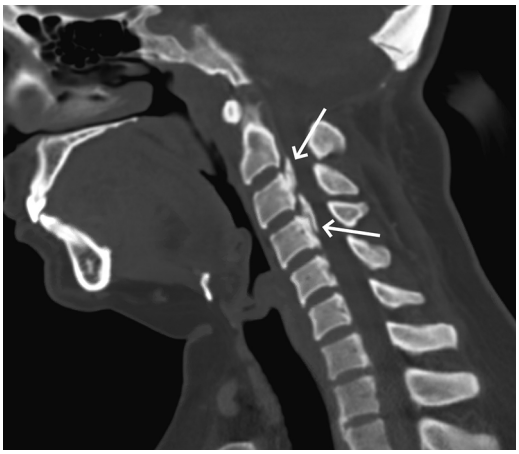

FIGURE 7-2
Sagittal CT of the cervical spine showing ossification of the posterior longitudinal ligament, causing spinal stenosis at the C2-C3 and C3-C4 levels (*arrows*).

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.**

dysfunction, and blood–spinal cord barrier disruption.[3] The neurobiology of injury is likely multifactorial, which may explain why significant variability exists in clinical course, response to therapy, and correlation between imaging and symptoms.

Clinical Symptoms and Signs

Cervical spondylotic myelopathy can become symptomatic acutely or in a more chronic fashion. Acute symptoms often follow trauma to the cervical spine in the background of prior cervical spinal stenosis. Hyperextension injuries, such as from a car accident, and falls with head injury are frequent triggers for acute presentations in older patients. In younger patients, high-impact accidents with fracture of the cervical spine and compression of the spinal cord are a more common cause. Acute injury of the spinal cord generally causes a central cord syndrome. In central cord syndrome, patients have more motor weakness in their arms than in their legs and urinary retention.[11] Anatomically, the corticospinal tract projections that control the arms are located more internally compared to the projections to the legs; hence, in central cord syndrome, the arms are affected more significantly. A variable degree of sensory loss is seen below the level of the lesion. Other myelopathic examination findings, such as spasticity, hyperreflexia, or upgoing toes, may not be present acutely; these findings may develop over the space of days to weeks.

A chronic presentation of cervical spondylotic myelopathy is more common. In contrast to acute central cord syndrome, gait disorder is one of the earliest symptoms in chronic disease, usually described as a stiff gait with a feeling of clumsiness and uncertainty. In brief office examinations of very early cases, gait abnormalities may not be clearly visible. Having the patient walk for an extended length, such as down the hallway, may elicit subtle stiffness in walking. As the disease progresses, more symptoms are seen, such as decreased dexterity in the hands, especially with fine movements such as writing, typing, or opening jars. Paresthesia in the fingertips is common as well. It is important to note that neck pain does not correlate with the severity of spinal cord compression. In a clinical series of cervical spondylotic myelopathy, axial neck pain was only present in about one-third of patients.[12] Axial neck pain is generated by disease of the musculoskeletal structures, such as the paraspinal muscles, ligaments, and bones, rather than nerve root or spinal cord compression. Another important clinical note is that bowel/bladder dysfunction, although typical in central cord syndrome, is infrequent in slowly progressive cervical spondylotic myelopathy. Bladder dysfunction is likely present in less than 5% of patients.[12] From an anatomic viewpoint, the autonomic fibers are present in the central areas of the spinal cord and are likely spared from slow extrinsic compression. Overactive bladder is a frequent symptom in patients with cervical spondylotic myelopathy, but its relationship with cord compression is unclear because of lack of consistent improvement after treatment.[13]

On neurologic examination, the usual findings are signs of myelopathy, including weakness in the intrinsic muscles of the hands, spasticity in the legs, and increased muscle stretch reflexes. If concurrent nerve root compression is present, findings of cervical radiculopathy might be present as well, causing mixed lower and upper motor neuron findings. An example is the inverted brachioradialis also known as the inverted supinator reflex first described by Joseph Babinski.[14] This finding refers to percussion of the brachioradialis muscle

causing flexion of the fingers (exaggerated reflex at the C8 level) without flexion of the elbow (absent reflex at the C5-C6 level).

A clear sensory level is usually not seen as is found in other causes of acute myelopathy, but decreased sense of vibration and joint position sense in the legs are common. In more severe cases, weakness in an upper motor neuron pattern can be found, especially in the flexor muscles in the legs. Among the different signs of corticospinal injury in patients with mild myelopathy, the Hoffman sign (involuntary flexion of the thumb after flicking the fingernail of the middle finger downward) may be more sensitive than the Babinski sign. In a clinical series of 225 surgically treated patients with cervical spondylotic myelopathy, the Hoffman sign was present before surgery in 46% of those with mild myelopathy, whereas the Babinski sign was present in only 10%.[15] In those with more severe myelopathy, the Hoffman and Babinski signs were both present in more than 80%.

Imaging

Generally, clinical suspicion for cervical spondylotic myelopathy is followed by imaging. In neurologic clinical practice, the opposite also occurs when patients are referred for concerning imaging and the goal is to evaluate the clinical significance. MRI is the preferred imaging modality for evaluation of the spinal cord because it can show the parenchyma of the spinal cord, boundaries of the cord and CSF, and soft tissue structures. In patients for whom MRI is contraindicated, a CT myelogram can also accurately show the contours of the spinal cord and degree of cord compression. However, cord signal abnormalities cannot be seen by myelography. X-ray of the cervical spine is useful, especially after prior instrumentation, to assess for displacement of the hardware (FIGURE 7-3). Another use of x-ray imaging is in flexion/extension images. In this procedure, an x-ray image is first taken with the neck flexed, and then the image is repeated with the neck extended. If significant change in the alignment of the bones is seen, dynamic instability is present; static images obtained in a flat position may underestimate the degree of dynamic stenosis. In some centers, flexion and extension views of the cervical spine can also be obtained by MRI and show ligamentous and bony instability better than x-ray.[16]

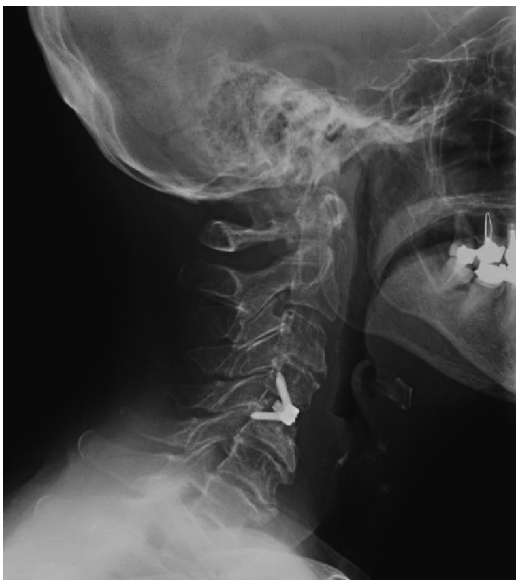

FIGURE 7-3
Sagittal x-ray of the cervical spine showing C4-C5 anterior cervical diskectomy with intervertebral disk graft placement and fusion. X-ray imaging can show hardware without significant artifact.

When examining an MRI of a patient with suspected cervical spondylotic myelopathy, the most important parameters are the degree of stenosis, the nature of bony or soft tissue changes, and cord signal intensity. Having no cervical spinal stenosis essentially excludes the

KEY POINTS

- **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.**

KEY POINTS

- 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.

possibility of cervical spondylotic myelopathy. However, as discussed before, even severe cervical spinal stenosis can exist on imaging without corresponding clinical symptoms. Degenerative changes in the cervical spine of any kind are extremely common on imaging. More than 80% of adults over the age of 60 who are asymptomatic have degenerative disk changes.[17]

How to accurately assess the clinical significance of stenosis on imaging is a work in progress. At first pass, a broad categorization is to classify areas of spinal canal narrowing as having no stenosis or some narrowing. The degree of narrowing is usually classified further as moderate or severe based on T2-weighted images (FIGURE 7-4). In moderate stenosis, some CSF is still present around the contours of the spinal cord, whereas in severe stenosis, the CSF rim is completely effaced. This categorization into moderate or severe stenosis unfortunately does not translate well into clinical symptoms. Some patients with moderate stenosis are symptomatic, whereas others with severe stenosis do not have any symptoms of myelopathy. Research to improve interpretation of MRI of the cervical spine to correlate better with clinical symptoms is ongoing. For example, the cross-sectional area of the spinal cord at the area of stenosis may correlate better with symptoms than qualitative interpretation of the degree of obliteration of the subarachnoid space.[18] Some of these techniques will likely enter clinical practice in the coming years.

After examination of the musculoskeletal structures and vertebral canal, the spinal cord is generally examined by T2-weighted sequences to look for cord signal changes. Absence of cord signal changes does not exclude the presence of myelopathy. Summarizing multiple series, the sensitivity of cord signal change for the presence of cervical spondylotic myelopathy is in the range of 58% to 85%.[18] Hence, patients may have symptoms and benefit from intervention without necessarily having cord signal changes, especially patients with mild myelopathy (CASE 7-1). In fact, in clinical series of imaging before and after surgery, the presence of any cord signal hyperintensity does not correlate strongly with outcome either positively or negatively.[19]

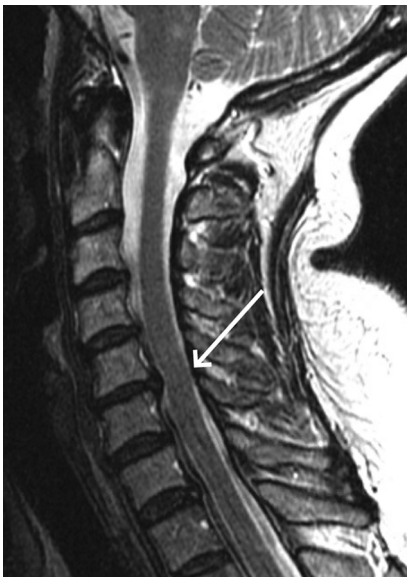

FIGURE 7-4
Sagittal T2-weighted MRI showing moderate cervical spine stenosis at C5-C6 (*arrow*). A thin rim of CSF is still visible posteriorly at this level.

Even in patients with cord signal hyperintensity, varying underlying pathology is present, ranging from edema and demyelination to necrosis and gliosis. If very bright cord signal T2 changes (same intensity as the CSF) are seen in combination with T1 hypointensity, then underlying gliosis that may not be reversible is likely present.[20] Fluffy, mildly hyperintense T2 changes may reflect edema that may improve with treatment; however, this remains to be prospectively validated in large clinical series. When cord signal changes occur from cervical spondylotic myelopathy, they can have variable morphology. However, especially

with chronic compression, a snake-eye pattern (in which there are circumscribed areas of high T2 signal intensity in the anterior cord) can evolve (FIGURE 7-6). The location of the T2 signal hyperintensity may not occur at the region of greatest stenosis but rather just adjacent to it. Finally, although IV gadolinium contrast is generally not used to evaluate for cervical spondylotic myelopathy, enhancement may be seen on postcontrast T1-weighted images. A specific pattern of enhancement called the pancakelike pattern has been described in patients with spondylotic myelopathy. It refers to a flattened appearance of enhancement different from the typical rostrocaudal enhancement throughout a lesion that accompanies transverse myelitis and has the following characteristics:

CASE 7-1

A 54-year-old man presented for evaluation of long-standing neck pain and new paresthesia in his fingertips. His primary care doctor obtained an MRI of his cervical spine, which led to referral for neurosurgical consultation and initial recommendation for spinal surgery. He sought a second opinion from a neurologist.

On examination, he had minimal weakness in left knee flexion, hyperreflexia in his left leg, and downgoing plantar responses. He had restricted range of motion of his neck to lateral movement bilaterally. MRI of his cervical spine (FIGURE 7-5) showed severe stenosis of the cervical spinal cord at C3-C4 by a right-sided disk protrusion.

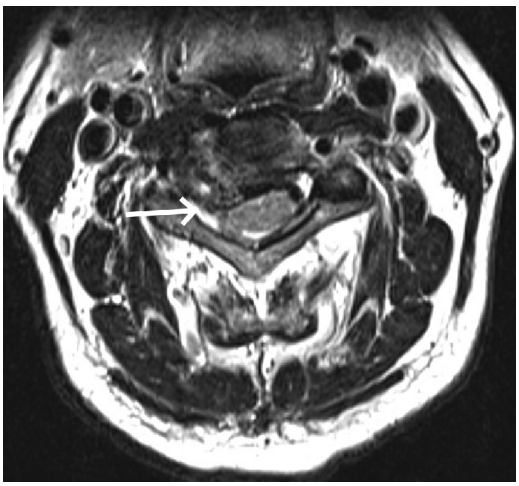

FIGURE 7-5
Imaging of the patient in CASE 7-1. Axial T2-weighted MRI shows a right-sided disk protrusion with flattening of the right side of the cervical cord (*arrow*) and rotation of the spinal cord.

COMMENT

The optimal management strategy in this case is unclear. The patient had signs of mild myelopathy from cervical spondylotic disease that could stabilize or progress with time. After discussing different options, he elected to continue conservative management with physical therapy and sequential neurologic examinations. Five years later, he continued to be stable with minimal myelopathic signs. Another observation is that although the patient had more significant right-sided spinal cord compression, he was symptomatic on the left side of the body. This mismatch between the side of the compression and side with more symptoms is a known phenomenon and does not exculpate the compressive lesion.

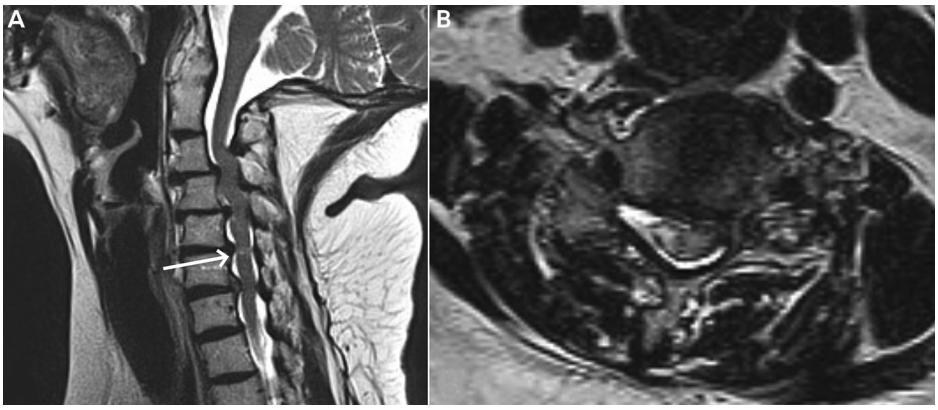

FIGURE 7-6

Severe multilevel degenerative disease. *A*, Sagittal T2-weighted MRI shows severe multilevel degenerative disease of the cervical spine with abnormal focal T2 hyperintensity of the cord at C5-C6 just caudal to the level of the greatest stenosis at C5-C6 (*arrow*). *B*, Axial T2-weighted MRI at C5-C6 (level of *arrow* in *panel A*) shows bilateral anterior cord T2 hyperintensity that is more hyperintense on the right compared to the left.

(1) a transverse band of enhancement with width of enhancement that is greater than or equal to the height, (2) location at or just below the site of maximal stenosis and at the center of the T2 hyperintensity, and (3) circumferential enhancement of white matter on axial sequences sparing the gray matter (FIGURE 7-7[21]).[22] The enhancement resolves slowly and can persist for many months even after surgical decompression. Clinically, apart from the diagnostic utility of the imaging sign, it is important for neurologists to be aware of the possibility of enhancement with cervical spondylotic myelopathy. The author is personally aware of patients who have undergone multiple investigations for myelitis and neoplasms because of "atypical" finding on contrast-enhanced MRI.

Since conventional MRI does not accurately capture the underlying neuropathology and corresponding prognosis with intervention, multiple new imaging techniques are being developed to look for injury to the spinal cord in cervical spondylotic myelopathy. Some of the more promising techniques include diffusion tensor imaging, magnetization transfer, and magnetic resonance spectroscopy. Most of these techniques, which were initially developed for brain imaging, show artifacts introduced by spinal imaging. It is likely that in the coming 5 to 10 years, these techniques will become better refined and enter routine clinical practice for better evaluation of the spinal cord in cervical spondylotic myelopathy.

Treatment

How best to treat cervical spondylotic myelopathy is controversial, and significant regional and practitioner variation exists. This variability originates from a lack of understanding of the natural history of untreated disease and lack of robust trials comparing conservative and interventional methods. To begin with, patients with severe cervical spinal stenosis on imaging without myelopathic symptoms likely do not all need surgery. The natural history of cervical spondylotic myelopathy was evaluated in a cohort of 199 patients with asymptomatic severe cervical spinal stenosis followed for a median of 44 months. Although about 22% developed symptoms of myelopathy during follow-up, no

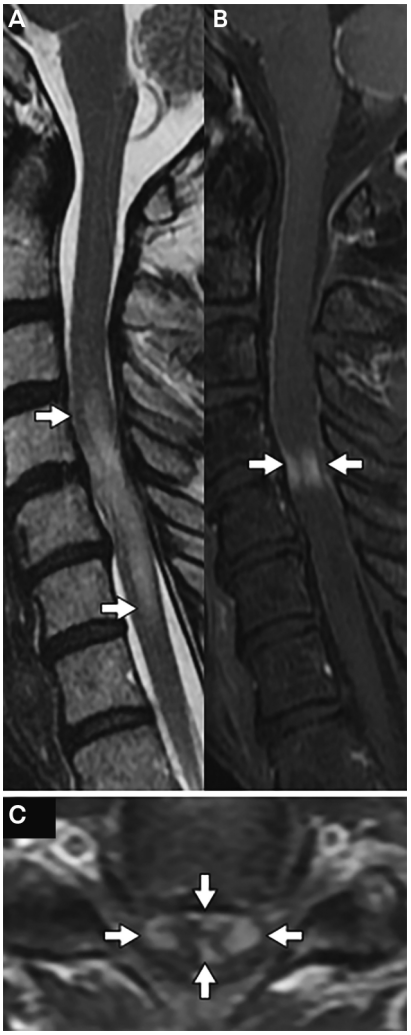

FIGURE 7-7

Pancakelike enhancement in cervical spondylotic myelopathy. Sagittal T2-weighted cervical spine MRI shows a longitudinally extensive T2-hyperintense lesion in the cervical cord (*A*, *arrows*) with moderate to severe stenosis at the C4-C5 level. The presence of a flat pancakelike enhancement in which the width is equal to the height on sagittal postcontrast T1-weighted MRI (*B*, *arrows*) involving the white matter and sparing the gray matter on axial sequences (*C*, *arrows*) is highly characteristic of the enhancement associated with cervical spondylotic myelopathy.

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association was seen between minor traumatic events and the onset of myelopathy.[23] This is in contrast to the often-heard advice that the goal of early surgery is to prevent the development of severe myelopathy from relatively minor trauma (such as hitting one's head on a doorframe). For those who progressed to clinical myelopathy within a year, predictive factors were the presence of clinical cervical radiculopathy and abnormal evoked potentials (somatosensory or motor); MRI factors, such as cord hyperintensity or cord cross-sectional area, were not predictive of early conversion to myelopathy.

Most experts would agree that surgery aimed at decompressing the cervical spinal canal is indicated for patients with myelopathy with significant disability (CASE 7-2). What to do with symptoms of mild myelopathy (such as findings of hyperreflexia in the legs and a mild sense of instability) is far less clear. Even in the earliest clinical series, it was observed that not all patients relentlessly progress to disability and that the untreated clinical course often had a stepwise progression with phases of deterioration followed by long periods of stability.[24] Clinical trial evidence for the benefit of conservative management versus surgical intervention is principally limited by small numbers. One trial randomly assigned 68 patients with mild or moderate cervical spondylotic myelopathy to either conservative or surgical treatment and followed them for 3 years.[25] At last follow-up, patients treated conservatively did no worse on scores of myelopathy than those treated surgically. In fact, on the timed walking test, patients treated conservatively walked 10 meters faster than those treated surgically after 3 years. In both groups, about one-third of patients had T2 hyperintensity on initial cervical spinal cord MRI. Another smaller trial randomly assigned 49 patients with mild or moderate cervical spondylotic myelopathy to surgical or conservative

KEY POINTS

- **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.**

treatment and followed them for 2 years.[26] In this trial also, no clear differences were seen in myelopathy rating scores or in electrophysiologic measures between the two groups after 2 years.

These small randomized trials contrast with usual clinical practice at many centers. Prospective clinical series demonstrate neurologic and pain improvement with surgery, although any surgical intervention necessarily has a large placebo effect.[27,28] More patients likely have surgery for mild myelopathy than benefit in the long term. On the other hand, cervical spondylotic myelopathy continues to be a major cause of morbidity and neurologic injury. The clinical trials are small and underpowered, and the clinical scales used in the trials may not be sensitive to small improvements in spinal cord function. In clinical practice, the author of this article generally recommends conservative therapy for signs or symptoms of mild myelopathy present for many months or years (CASE 7-2) and recommends intervention for significant disability or progressive neurologic injury in sequential clinical examinations.

CASE 7-2

A 50-year-old man with prior spinal cord trauma and residual mild left-sided weakness experienced increasing difficulty walking and loss of dexterity with fine finger movements. On examination, he had moderate weakness in the flexor muscles of his legs, spasticity in his left arm and both legs, and diminished sensation to vibration in both legs.

MRI of his cervical spine showed disk protrusion at the C6-C7 level causing severe stenosis of the cervical spine in addition to prior cord trauma at C4-C5 (FIGURE 7-8A). Surgery was recommended, but the patient did not follow up. He returned 3 years later and was walker dependent with persistent spasticity in the legs. Repeat MRI showed resolution of the disk protrusion at C6-C7 (FIGURE 7-8B).

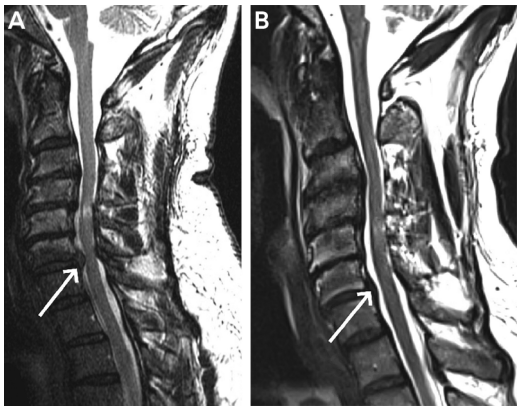

FIGURE 7-8
Imaging of the patient in CASE 7-2. *A*, Sagittal T2-weighted MRI shows an area of T2 cord hyperintensity at C4-C5 and disk protrusion causing severe canal stenosis at C6-C7 (*arrow*). *B*, Repeat sagittal T2-weighted MRI obtained 3 years later shows resolution of the prior disk protrusion at C6-C7 (*arrow*) without surgery.

COMMENT

This case illustrates that if the spinal cord is compressed for long enough, irreversible injury to the spinal cord can occur. In this case, the disk reabsorbed back without any intervention, but the patient's neurologic signs did not improve. His disability may have been preventable if he had gotten surgical decompression.

Even in cases in which conservative management is chosen, no clear consensus exists on what is meant by this aside from no surgery. In some centers, patients undergo physical therapy to improve strength and balance in combination with gentle neck stretching exercises to improve any coexistent neck pain. All patients should avoid activities that cause severe flexion/extension neck movements that may cause further injury to the spinal cord and can provoke an acute central cord syndrome. None of these interventions have individually been tested in clinical trials, and which are most beneficial is unclear.[29]

SURGICAL APPROACHES. The concept that surgery could potentially improve spondylotic myelopathy originated in the 1920s when Byron Stookey of Bellevue Hospital described decompression of the cervical canal and improvement in neurologic function in patients with ventral herniations of what he called "cervical chondromas."[30] Since then, many different surgical techniques have been developed to address cervical spinal cord compression from spondylotic disease. However, in the surgical community, no clear consensus exists on the best operative technique. Hence, an identical structural pathology could be addressed differently based on the surgeon and the center. Generally, surgery for cervical spondylotic myelopathy can be divided into anterior and posterior approaches. In the anterior approach, the compressing disk is generally removed, often in combination with placement of a graft in the intervertebral space for fusion. Instrumentation with plating of the vertebra anteriorly is frequently used to improve rates of fusion. Innovations in hardware, including plates with dynamic load sharing, plates with smaller screws, and low-profile plates, have decreased rates of fusion failure. Biodegradable plates are in the process of development and may enter clinical use in coming years. The alternative decompression approach is a posterior one, in which the goal is to provide more space in the spinal canal via laminectomy or laminectomy with fusion in cases of dynamic instability. Similar to innovations in anterior instrumentation, the hardware used for posterior instrumentation has also changed, with improved axial load sharing and better sagittal balance with goals of decreasing stress on the adjacent spine and decreasing rates of hardware failure.

Whether one approach is better than the other is a matter of controversy and surgeon preference. Overall, for multiple levels, the posterior approach is likely easier, whereas for a single level of compression from a large ventral disk, the anterior approach may be preferred. Primary surgical complications include wound infection, bleeding, displacement of hardware, and failure of fusion. Some more specific neurologic complications include C5 radiculopathy, even in surgeries that do not directly involve the C5 nerve root. The cause of this complication is unclear. Injury to the recurrent laryngeal nerve can occur, especially in anterior approaches, causing hoarseness of voice.

Following surgery, some improvement in upper extremity function occurs in about 60% to 80% of patients based on unblinded clinical series.[28,31] Recovery of lower extremity dysfunction and sensory dysfunction are usually less robust compared to arm strength recovery. Neurologic recovery continues to occur over the first few months but typically plateaus at around 6 months. A risk of spondylosis occurring at adjacent levels exists (called adjacent segment disease), especially after fusion surgeries of the spine. This risk is calculated to be around 2% per year, and patients often need careful monitoring for recurrent symptoms following surgery.[28]

KEY POINTS

- 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.

SYRINGOMYELIA

Syringomyelia refers to the presence of a syrinx, a fluid-filled, glial-lined cavity, within the parenchyma of the spinal cord. This entity must be distinguished from an enlarged central canal (hydromyelia), which is an ependymal-lined enlargement of the normally obliterated central canal. This is an important distinction that is sometimes not recognized and can lead to inappropriate diagnosis and therapies. The central canal is a circular midline structure located at the junction of the ventral one-third and dorsal two-thirds of the spinal cord. The central canal is an embryologic remnant that progressively becomes obliterated during adulthood. However, not uncommonly, some portions of the central canal can remain visible on imaging and are often found incidentally (FIGURE 7-9). In an MRI spine series of 794 patients, the central canal was enlarged in 12, corresponding to a rate of 1.5%.[32] In none of the patients was the central canal enlargement pathogenic or related to the clinical syndrome leading to the imaging study. When enlargement of the central canal is visible, the contours are generally smooth and without nodularity. Most instances of incidental enlarged central canal are in the thoracic spinal cord, although cervical spinal cord enlarged central canals are also seen.[32]

By contrast, syringomyelia can occur in the cervical or thoracic segments and has variable appearance. The prevalence of syringomyelia is estimated to be 3.3 per 100,000 to 8.5 per 100,000, with variation in different ethnic groups.[33] Epidemiologically, syringomyelia can be primary or associated with other predisposing conditions. The most common predisposing condition is Chiari type I malformation, which can be clinically silent but seen on imaging as herniation of the cerebellar tonsils into the foramen magnum. About a 65% to 80% incidence of syringomyelia is seen with Chiari type I malformation (FIGURE 7-10).[34] More uncommonly, patients with syringomyelia may have Chiari type II malformations, in which both the cerebellum and brainstem herniate into the foramen magnum, often accompanied by myelomeningocele in the lumbar spine. Acquired spinal

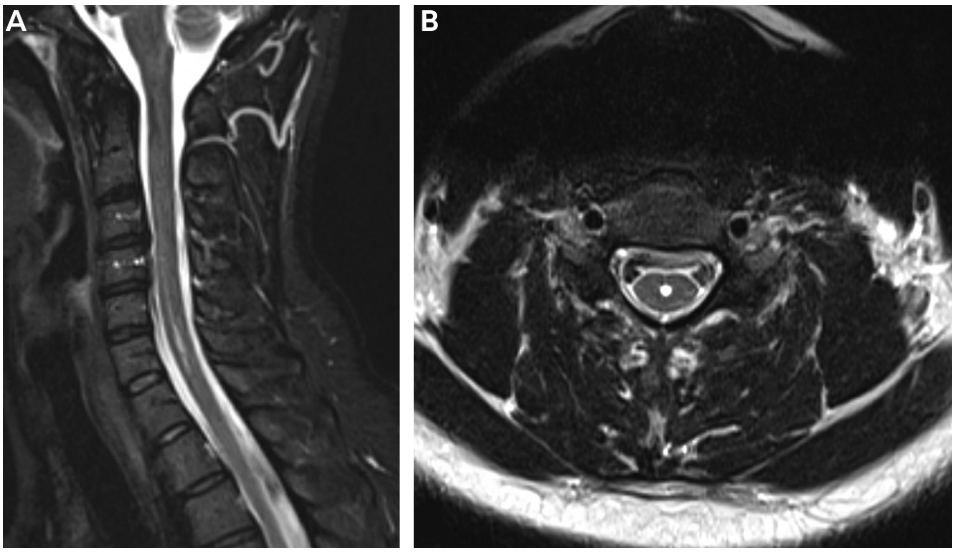

FIGURE 7-9

Enlarged central canal. *A*, Sagittal fat-suppressed short tau inversion recovery (STIR) MRI shows a dilated cystic region at the C6 level. *B*, Axial T2-weighted image shows a rounded central structure without myelomalacia that is most consistent with an enlarged central canal.

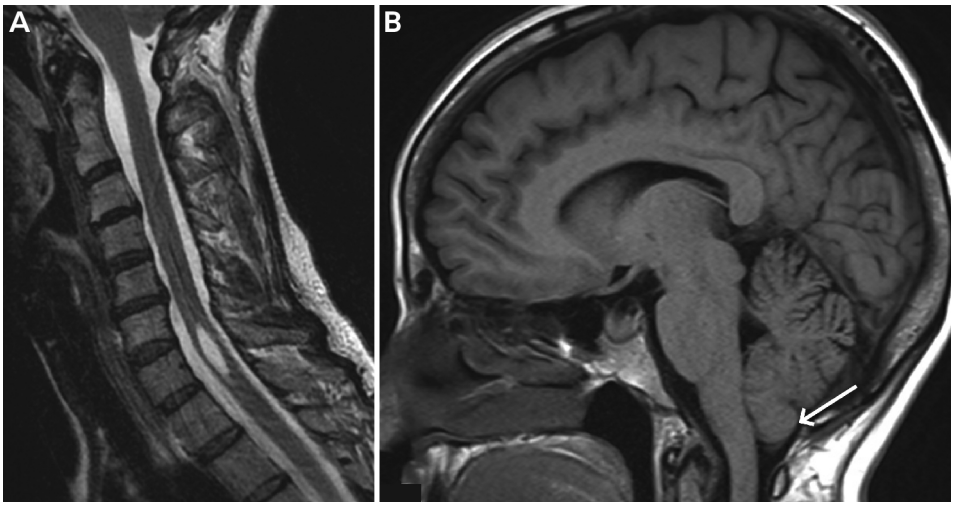

FIGURE 7-10
Syringomyelia with Chiari type I malformation. *A*, Sagittal T2-weighted MRI shows syringomyelia at the C6-C7 level. *B*, Concurrent T1-weighted sagittal brain MRI shows a mild Chiari type I malformation with herniation of the cerebellar tonsils (*arrow*) into the foramen magnum. These findings were incidental for the patient.

KEY POINTS

- 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.

cord injury can also lead to syrinx formation. Traumatic spinal cord injury can predispose to late development of a syrinx. In one study, the mean interval between spinal cord injury and syrinx development was 8.6 years.[35]

Other causes of spinal cord injury, such as spinal cord infarction or myelitis from multiple sclerosis or neuromyelitis optica (NMO), may also infrequently lead to syringomyelia in a delayed fashion. Neoplasms are an important secondary cause of acquired syringomyelia. Midline neoplasms in the spinal cord, such as ependymoma or hemangioblastoma, are primarily associated with syringomyelia. Interestingly, the syrinx is most frequently located above the tumor, followed by both above and below the level of the tumor. It is infrequent to have a syrinx located solely below the level of the tumor. The chances of developing a syrinx are generally higher with a more rostral tumor in the spinal cord.[36] Familial syringomyelia has also been described in small clusters, pointing to as-yet undiscovered genetic causes.

Pathogenesis

Many different theories exist to explain the development and progression of syringomyelia. An early theory hypothesized that syringomyelia represents the cavitation of a glial tumor that became necrotic from insufficient blood supply.[37] However, histopathology did not show tumor cells in most syringes, and tumor-directed treatment with radiation therapy did not resolve the syringes. Others observed that in the nervous system, vascular injury evolved to cavitation. They hypothesized that either ischemic or hemorrhagic injury to the spinal cord evolved with time to have the final appearance of a syrinx. However, these theories do not explain the progressive nature of a syrinx, which often expands with time and causes increasing spinal cord dysfunction. Many patients did not have signs or symptoms of vascular injury to the spinal cord before the development of myelopathy from the syrinx.

Over time, hydrodynamic theories have gained favor as the cause of syringomyelia. Simple occlusion of the central canal and impairment of bulk flow of

CSF are not fully explanatory since most healthy adults have an occluded central canal and do not develop a syrinx. The English neurosurgeon Bernard Williams developed one of the first widely accepted hydrodynamic theories seeking to explain why syringomyelia is more common in Chiari malformations. He hypothesized that activities such as coughing or sneezing transiently displace CSF from the head into the spinal canal. In patients with Chiari malformation and a crowded foramen magnum with impaired circulation of CSF, a longer time passes before pressure in the head and spinal canal equilibrate. During this vulnerable time, CSF is postulated to be sucked into the spinal cord either through the central canal or potentially through the spinal cord itself, causing a cystic cavitation.[37] Although the theory has empirical support in cases of Chiari malformation, the theory does not explain other causes of syringomyelia in which no crowding of the foramen magnum is present. An alternative theory postulates that with interference of CSF circulation in the spinal subarachnoid space, the velocity of CSF in the subarachnoid space increases. Following the Bernoulli law, the increased velocity of the fluid results in lower net subarachnoid pressure, which causes a distending force on the spinal cord. Over time, this gradually results in a cavitary expansion of the spinal cord, causing a syrinx.[38] Many other theories have been postulated to explain the buildup of fluid in the spinal cord. Although we do not yet have a definitive answer, a unifying theme appears to be impairment of free CSF flow in the spinal subarachnoid space. Understanding the mechanism will be important in the future because treatment directed at improving the flow of CSF, such as by lysis of adhesions in arachnoiditis or decompression of the foramen magnum, may be sufficient to prevent the progression of syringomyelia.

Clinical Features

Classically, slow expansion of a syrinx in the central cord causes interruption of the crossing spinothalamic tracts via the anterior commissure. Hence, early on, patients develop a band of numbness limited to pain and temperature sensation in the affected dermatomes with preserved vibration and joint position sense (dissociated sensory loss). This pattern of sensory loss has been called a capelike distribution of sensory loss as the cervical and upper thoracic dermatomes are often affected by syringomyelia. Over time, as the syrinx expands and causes further spinal cord destruction, descending tracts (such as the corticospinal tract) are affected, causing weakness and spasticity in the legs. In clinical practice, many types of syrinx are not located exactly in the midline but are eccentric to one side. Hence, many patients do not have the classic progression from the capelike bilateral sensory loss pattern to weakness. Wide variability can exist in the clinical syndrome, such as starting with a slowly progressive hemicord syndrome.

Because of the interruption of the spinothalamic tracts, neuropathic pain that is often poorly localized is an important early feature. For example, in a case series of midline cord ependymomas, neuropathic pain with sensory loss was the earliest feature, present for a median of 13 months before diagnosis of the tumor.[39] Other important clinical clues are lower motor neuron features that gradually become apparent as the anterior horn cells become injured. This can cause muscle atrophy in the arms with a cervical spinal cord syrinx. When the paraspinal muscles become weak, progressive scoliosis is common as well. Since autonomic fibers descend in the midline, slow expansion of midline lesions as in syringomyelia can cause early autonomic dysfunction. This can take the form of bowel and urinary

sphincter dysfunction, sexual dysfunction, and sympathetic dysfunction such as Horner syndrome with high cervical cord lesions.

Treatment

How to best manage syringomyelia is uncertain, and recommendations derive from small case series rather than randomized trials. The first question is whether anything needs to be done. The natural history of the disease may depend on the age of the patient and the underlying predisposing condition. In a group of 17 pediatric patients with mild neurologic symptoms followed for an average of 3 years, 15 of the 17 patients did not need an intervention.[40] The two children who needed surgery had underlying Chiari malformations. In another natural history study of 19 patients followed for a median of 9 years, eight had no progression of symptoms, another eight had a slowly progressive course, and three had intermittent periods of progression.[41] Finally, in a larger series, 38 patients who did not undergo surgery were followed for an average of about 5 years; about half eventually had progression of symptoms, whereas the other half remained unchanged or even improved.[42] In this series, patients with segmental sensory signs had a more benign prognosis, whereas those with paraparesis from myelopathy or bowel/bladder dysfunction were more likely to progress.

Hence, many patients with mild symptoms will not benefit from interventions, and, despite prominent imaging findings, a conservative approach with symptomatic management and sequential neurologic examinations may be the best course. In patients with progressive myelopathy, surgical intervention is usually the next step. Because of the uncertainty in the pathogenesis, development, and progression of syringomyelia, how best to treat remains unclear as well. One approach is to improve CSF circulation by decreasing resistance in the spinal subarachnoid space. For many patients with Chiari malformation, this takes the form of suboccipital craniectomy and decompression of the posterior fossa. In a surgical series of 75 patients, most of whom had Chiari malformations, craniocervical decompression alone appeared to relieve or stabilize symptoms in about 75%.[43] Additional intervention to improve the syrinx did not confer more benefit. In those with syringomyelia from other causes, spinal subarachnoid circulation can be improved by laminectomy at the affected levels along with lysis of adhesions for those with arachnoiditis.

Despite these steps, many patients continue to progress with enlargement of their syrinx. In these patients, additional interventions should be performed. Most commonly, a syringosubarachnoid shunt is placed, connecting the syrinx with the subarachnoid space (**FIGURE 7-11**). This is a high-risk procedure in which a small incision is made in the spinal cord itself to reach the shunt. In a small series, the shunting procedure appeared to successfully decompress the syrinx and improve symptoms.[44] This procedure may be especially useful in patients with a large syrinx. A caveat with shunting is that although the syrinx may improve rapidly immediately, a high rate of syrinx reaccumulation exists, either from shunt failure or the development of a new syrinx in the cord that does not communicate with the syrinx that was drained.

KEY POINTS

- **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.**

OTHER STRUCTURAL MYELOPATHIES

Other, less common, structural abnormalities of the spine may present with progressive myelopathy. The following sections give brief overviews of a selection of these abnormalities.

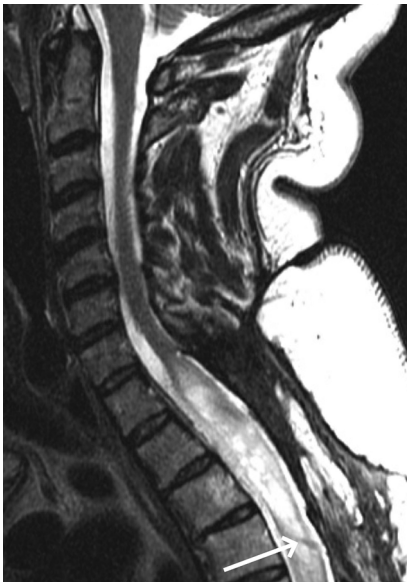

FIGURE 7-11
Sagittal T2-weighted MRI shows a large multiloculated syrinx in the upper thoracic spinal cord. A syringosubarachnoid shunt (*arrow*) did not stop the progression of the syrinx.

Idiopathic Spinal Cord Herniation

Spinal cord herniation is characterized by a defect through which the spinal cord is displaced, typically in the ventral dura. This syndrome primarily affects the thoracic spinal cord. Clinically, patients develop thoracic myelopathy that slowly progresses over the course of months to years. In large case series and meta-analyses, a mild female predominance was seen, with the average age of diagnosis about 51 years.[45] Progressive Brown-Séquard syndrome is the most common clinical syndrome. Diagnosis is typically made by MRI of the spine. The usual finding is adhesion of the spinal cord to the anterior portion of the spinal canal, with deformity in the contour of the spinal cord in sagittal sections (seen as kinking of the cord). Cord T2 signal changes may or may not be seen. With CT myelography, intrathecal contrast is seen in the dorsal aspect of the spinal cord but not in the ventral region. This finding can distinguish spinal cord herniation from a dorsal arachnoid cyst, in which impairment of CSF flow is seen on myelography.

In patients with symptomatic and progressive disease, treatment is with surgical correction. Generally, two different surgical approaches are used to treat spinal cord herniation.[46] The first is a dural patch of the defect with release of the spinal cord to a more natural physiologic state. The second approach is paradoxical widening of the dural defect with the goal of creating enough space to prevent spinal cord strangulation. At present, no clear consensus exists on what the optimal technique is. Most patients improve after surgical correction, and those with Brown-Séquard syndrome have a better prognosis compared to those with a more significant myelopathy.

Arachnoid Cyst

Spinal arachnoid cysts are intradural-extramedullary cystic structures in the subarachnoid space. In many cases, arachnoid cysts are found incidentally on routine imaging of the spine and are clinically asymptomatic. The etiology of the cysts is unclear; many are idiopathic, whereas others are associated with congenital spinal deformities in children or with spinal trauma in adults, such as after epidural hematoma or lumbar myelography. The latter etiologies suggest impairment of subarachnoid fluid flow or arachnoiditis as possible contributors to cyst formation. Overall, spinal arachnoid cysts are located primarily in the thoracic spine dorsal to the spinal cord, although anterior thoracic arachnoid cysts and cervical arachnoid cysts have also been rarely described. MRI is the preferred imaging modality to diagnose arachnoid cysts. Arachnoid cysts have the same signal intensity as CSF, with generally visible boundaries and displacement of the spinal cord (FIGURE 7-12). The boundaries of the cysts do not

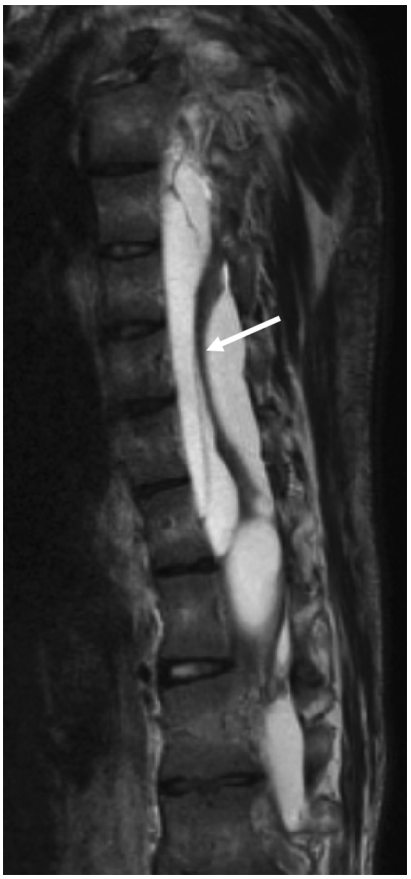

FIGURE 7-12
Sagittal T2-weighted MRI shows multiloculated arachnoid cysts with cord atrophy (*arrow*) in a patient with congenital spinal deformities.

enhance after IV gadolinium injection, and restricted diffusion is not seen within the cyst (more typical in epidermal cysts). In CT myelography, a variable degree of contrast opacification of the cyst is seen.

Clinically asymptomatic cases are followed by serial clinical examinations and imaging. When clinically symptomatic, patients often report symptoms of myelopathy, such as leg weakness, gait instability, or urinary dysfunction, in combination with radicular neuropathic pain.[47] Treatment of symptomatic arachnoid cysts is generally surgical, and multiple approaches have been described in the literature. The simplest procedures are cyst fenestrations. For more refractory cases, the cysts can be excised in combination with laminectomy to increase subarachnoid space.

Arachnoid Web

Like spinal cord herniation and arachnoid cyst formation, arachnoid web primarily affects the thoracic spine, causing progressive myelopathy. Spinal arachnoid webs are intradural bands of arachnoid tissue that usually attach to the dorsal surface of the spinal cord. The cause of web formation is unclear, but based on similar location, some authors speculate that an arachnoid web represents an incomplete or collapsed arachnoid cyst. Presenting symptoms are typically back or neck pain in combination with progressive myelopathy, such as difficulty walking or numbness in the legs. On MRI, the arachnoid web is often not seen directly. Instead, the effects are seen indirectly on the spinal cord, in which a dorsal indentation is seen. T2 cord hyperintensity and, frequently, syringomyelia may be seen. The abrupt change in caliber of the spinal cord with a dorsal cord indentation where the arachnoid web attaches has been called the scalpel sign.[48] Treatment is usually by surgical sectioning of the arachnoid web combined with laminectomy. In a small case series, surgical correction helped improve or stop further progression of symptoms.[49]

Epidural Lipomatosis

Spinal epidural lipomatosis refers to the accumulation of fat in the epidural space. When enough fat is present to encroach on the spinal canal or foramina, patients can have symptoms of myelopathy, neurogenic claudication, or radiculopathy. Spinal epidural lipomatosis can be idiopathic or secondary to other conditions, such as exogenous steroids, hypercortisolism, or obesity. Epidural corticosteroid injections are strongly associated with epidural fat

KEY POINTS

- 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.

proliferation. In a retrospective series of 28,902 consecutive patients undergoing spinal MRI, the prevalence of spinal epidural lipomatosis was estimated at 2.5%.[50] Of these patients, only 5% had symptoms that were attributable to the epidural fat. For the rest, the finding was either incidental or did not explain the clinical syndrome. Hence, for most patients with this finding, intervention for the epidural lipomatosis will not be needed. On imaging, epidural lipomas are hyperintense on both T1 and T2 sequences but suppressed on short tau inversion recovery (STIR) or other fat-suppressive sequences. Epidural fat can span multiple vertebral segments, and whereas the lumbar spine is the most commonly affected single region, the thoracic spine is next most common.

The first step of treatment is to address secondary factors contributing to the accumulation of epidural fat. For example, epidural corticosteroid injections should generally be stopped. Investigations for undiagnosed causes of hypercortisolism should be conducted as well. For symptomatic cases that do not respond to conservative therapy, surgical therapy is generally laminectomy and resection of epidural fat.[51]

Hirayama Disease

Initially described in Japan and other Asian countries, the characteristic feature of this eponymous disease is 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. The weakness is typically asymmetric or unilateral. After progression for several years, the disease stops with static weakness. When initially described, the proposed mechanism was thought to be motor neuron disease of a restricted spinal segment, which the authors called monomelic amyotrophy.[52] Subsequently, Hirayama and colleagues[53] established that in patients who are symptomatic and in whom the disease is progressive, movement of the dural sac occurs in flexion and extension postures. 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. Typically, no movement is seen in the bony elements. The hypothesis is that with repeated neck flexion and extension movements, the anterior horn of the lower cervical spinal cord is injured, accounting for the forearm and hand weakness. Recognition of the clinical syndrome is important because the dynamic movement of the dural sac can be missed entirely on static MRI with the neck extended. How best to manage the disease is unclear. In most patients, conservative therapy with avoidance of neck flexion can stabilize the disease. The natural history of the disease is generally of progression initially with long periods of stability. If patients have progressive disability, cervical decompression and fusion can be performed.

Chronic Adhesive Arachnoiditis

Adhesive arachnoiditis is progressive fibrosis of the arachnoid membrane, usually triggered by physical or chemical injury to the subarachnoid space. Cases have been known to occur after spinal surgery, subarachnoid hemorrhage, meningitis, trauma, or myelography using older oil-based contrast agents. Symptoms are caused by a combination of mechanisms. The arachnoiditis in the subarachnoid space causes radicular injury to traversing nerve roots. This is seen on imaging as clumping and tethering of the nerve roots. Arachnoiditis can

also tether the spinal cord and cause consequent myelopathy. The impairment in free flow of CSF in the subarachnoid space also predisposes to syrinx formation, which can then cause progressive myelopathy. Clinically, arachnoiditis presents after a time delay from the inciting event that can range from weeks to years.[54] In patients with arachnoiditis of the lumbar spine, the presentation is primarily of progressive lumbosacral radiculopathy. In others who have more widespread arachnoid injury, symptoms of slowly progressive myelopathy with radicular findings predominate. Diagnosis is usually by MRI, which shows the clumping of the nerve roots, often with enhancement.[55] Thin septations are often visible, tethering or displacing the spinal cord (FIGURE 7-13). Cord T2 hyperintensity or a cystic cavity may be present. In severe cases, the arachnoid can have calcium deposition (a process called ossification). No clear treatment has been identified for this debilitating and progressive disease. Further intervention to the spine should generally be minimized to decrease further arachnoid fibrosis. Patients are usually treated with a combination of physical therapy and pain management. Anti-inflammatory therapies, including corticosteroids and IV immunoglobulin (IVIg) have been tried without controlled trials to support use. In infrequent cases in which a focal area of adhesion causing progressive symptoms is present, surgery to lyse cord adhesion can be performed. The results, however, are mixed, with benefits offset by further arachnoiditis.

CONCLUSION

In addition to cervical spondylotic myelopathy and syringomyelia, many other structural abnormalities in the spine (which are individually infrequent but as a group common) can compromise the spinal canal and cause myelopathy. These include thoracic spine degenerative disease, arachnoid cysts, spinal arachnoid webs, spinal bony defects causing cord herniation, and Hirayama disease. In patients with acute, subacute, or progressive myelopathy, structural causes should remain high on the list of differential diagnoses and be investigated for urgently. Neurologists have an important role in determining whether the imaged abnormality is relevant to the clinical syndrome and being aware of the appropriate management options and the prognosis with conservative management.

KEY POINTS

- 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.

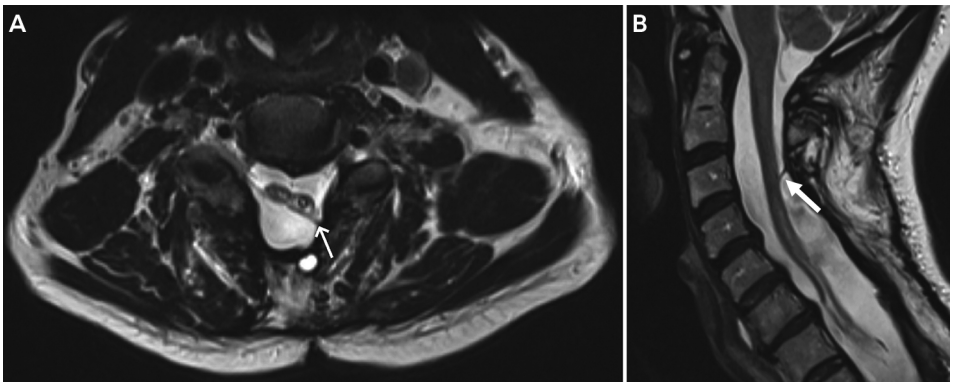

FIGURE 7-13
Arachnoiditis with spinal cord tethering. ***A***, **Axial T2-weighted MRI of the cervical spine shows tethering of the cervical spinal cord (*arrow*) from arachnoiditis.** ***B***, **Sagittal T2-weighted MRI also shows tethering of the cord (*arrow*) through the arachnoid space.**

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