Magnetic resonance imaging of intramedullary spinal cord lesions

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Learning objectives

Spinal cord disease is often viewed as having a poor outcome. Although in certain conditions this is true, non-traumatic myelopathy encompasses a vast array of diseases some of which are exquisitely responsive to treatment. Accurate diagnosis becomes important as damage is often progressive and long-term disability and morbidity is related to the degree of neurological impairment when the diagnosis is reached.

The radiological evaluation of spinal cord lesions has undergone a dramatic change since the introduction of magnetic resonance (MR) imaging.

In this educational exhibit, we propose to:

1) To describe the normal appearance of the spinal cord and the different spaces within the spinal canal.

2) To define the typical magnetic resonance imaging protocol for evaluation of the spinal cord lesions.

3) To review the benign and malignant intramedullary spinal cord lesions and to present the MR signal characteristics of each entity.

4) To elucidate the pitfall in the differentiation of tumors from nonneoplastic disease of the spinal cord.
Background

The normal appearance of the spinal cord

It is important to be able to recognize the normal appearance of the spinal cord and the different spaces within the spinal canal.

The normal spinal cord should be an oval shape with smooth contour. The cord signal has intermediate signal on both T1- and T2-weighted images. It is surrounded by cerebrospinal fluid (CSF) within the subarachnoid space which is separated from the epidural space by the dura. (Fig 1)

Diagnostic approach and imaging protocols:

Out-of-hours, MR imaging is the current imaging modality of choice in the evaluation of patients presenting with myelopathic symptoms in the search for spinal cord lesions. It allows for the noninvasive investigation of spinal cord and identification of internal structural abnormalities.

The typical MR imaging protocol for evaluation of the spinal cord lesions includes unenhanced sagittal and axial T1-weighted and T2-weighted images, as well as post-gadolinium-enhanced sagittal and axial T1-weighted images.

The combination of sagittal and axial imaging planes is invaluable for the analysis of intramedullary spinal cord lesions. Sagittal images delineate the longitudinal extent of a lesion, and axial images show its cross-sectional distribution and allow its assignment to the gray matter, white matter, or both.

Contrast-enhanced images are helpful in determining the solid portion of an intramedullary neoplasm, tumoral vs nontumoral cysts, other enhancing pathologic entities, or other features that may modify the differential diagnosis.

According to Koeller and al, when using MR imaging in the evaluation of patients suspected of having an intramedullary spinal process, there are 3 principles to appreciate to facilitate differentials.

- The essential imaging criterion for an intramedullary spinal neoplasm is cord expansion.

If the cord expansion is absent, then a nonneoplastic etiology including demyelinating disease, sarcoidosis, dural arteriovenous fistula, cord infarction, or myelomalacia may be suggested.
It is essential to obtain contrast-enhanced images following the administration of intravenous gadolinium-based contrast material in at least 2 different planes, not only for limiting the differential diagnosis but also for planning surgery.

However, the absence of enhancement does not exclude an intramedullary neoplasm in the presence of cord expansion.

- Frequent association of intramedullary spinal tumors with tumoral and nontumoral cysts.

The nontumoral cysts are located at the rostral or caudal pole of the solid portion of the tumor. These nontumoral cysts usually represent simple reactive dilatation of the central canal (syringomyelia) and do not demonstrate enhancement with contrast.

By contrast, tumoral cysts are part of the tumor itself and frequently exhibit peripheral enhancement. Tumoral cysts are commonly seen in astrocytomas, hemangioblastomas, and ependymomas.

**Etiologies**

**I. Benign intramedullary spinal abnormalities:**

Benign intramedullary spinal abnormalities includes syringohydromyelia, contusion, infarction, myelitis, vascular anomalies and spondylitic myelopathy.

*1- Syringohydromyelia (Fig 2)*

Syrinx is a general term used to describe a cystic intramedullary spinal cavity.

The term syringomyelia refers to a cystic cavity that is eccentric to the central spinal canal and lined by gliotic parenchyma.

Hydromyelia refers to ependymal-lined cystic central canal dilatation. Often the 2 entities cannot be distinguished; in which case, the term syringohydromyelia is used.

Primary forms may be related to basilar invagination, or Chiari 1 or Chiari 2 malformation.

Secondary forms can be associated with trauma (usually developing at the specific site of injury and extending rostrally) or may develop in response to obstruction from an intramedullary or extramedullary spinal cord tumor.

Clinical symptoms include pain and temperature sensory loss in a "cape-like" distribution over the upper extremities and back with preservation of position sense, proprioception,
and light touch. Patients may experience mechanical spinal pain, radicular pain, spastic paraparesis, or scoliosis.

There is no association between size of the syrinx and severity of clinical symptoms.

On MR imaging, a cystic cavity is seen within the spinal cord, which follows cerebrospinal fluid signal on all sequences. There may be associated expansion of the spinal cord or cerebrospinal fluid pulsation artifact within the syrinx.

When a syrinx is secondary to an obstructing tumor, it may be challenging to differentiate tumoral from nontumoral cyst formation.

No enhancement on postcontrast images and absence of cerebrospinal fluid pulsation helps to distinguish a benign syrinx from nontumoral and tumoral cysts. The cystic cavity of a tumoral cyst is contained within the tumor itself and frequently shows peripheral enhancement.

2- Spinal Cord Contusion (Fig 3)

Spinal cord contusion usually occurs in conjunction with other findings such as fracture, dislocation, ligamentous injury, or hematoma. Rarely, cord contusion may be present without these findings, as in SCIWORA (Spinal Cord Injury without Radiographic findings), which is seen mainly in the pediatric population.

Clinically, these lesions can cause varying degrees of motor and sensory dysfunction.

MR imaging, plays a decisive role in:

• Assessing the spatial relationship between bony fragments and spinal cord.

• Visualizing concomitant intraductal and paravertebral soft tissue and vascular or nerve injuries.

• Visualizing primary and secondary spinal cord changes in the acute and post-acute phases.

Susceptibility-sensitive, blood-sensitive gradient echo sequences are very useful. They can visualize the ligaments, distinguish between traumatic vertebral prolapse and osteophytes, and can render spinal cord anatomy and the blood vessels.

Findings of spinal cord injury include hemorrhage, edema, and swelling.
In the acute phase of spinal cord hemorrhage, focus of deoxyhemoglobin appears hypointense on T2 and gradient images. Identification of spinal cord hemorrhage of greater than 10 mm in diameter indicates a complete neurologic injury.

Spinal cord edema appears as an area of hyperintense T2 signal reflecting focal accumulation of intracellular and interstitial fluid in response to injury.

Spinal cord swelling invariably accompanies spinal cord edema.

In the absence of other traumatic injury findings, the differential diagnosis of a focal simple cord edema with swelling includes demyelinating disease, acute disseminated encephalomyelitis (Adem), transverse myelitis, sarcoidosis, and low-grade glioma. Ependymoma, hemangioblastoma, and cavernous malformation should be considered if there is evidence of hemorrhage associated with the cord edema.

3- Spinal Cord infarction (Fig 4)

The key to distinguishing spinal cord infarct from other entities that can have a similar appearance on imaging is the clinical history. The loss of neurologic function is rapid, and deficits typically occur in an hour on less; less frequently, the deficit may develop over a period of hours. The cardinal symptoms are sudden onset of flaccid para- or quadriaparesis, sphincter paralysis, and dissociated sensory loss below the level of the lesion.

Other important clinical information includes a history suggesting aortic disease such as dissection, severe atherosclerotic disease. Numerous other causative disorders including syphilis, vasculitis, coagulopathy, hypotension, vascular malformation, embolism and meningitis have been described.

Infarction occurs most commonly in the thoracic and thoracolumbar cord, which has a tenuous blood supply. The lesion usually spans more than 1 vertebral segment. Infarction in the conus can result from occlusion of the artery of Adamkiewicz in the absence of adequate collateral flow.

Spinal cord infarcts most commonly affect the territory of the anterior spinal artery, involving the corticospinal and spinothalamic.

On MR imaging, T2 hyperintensity can be seen in the central gray matter or the entire cord cross-section. The "owl's eye" appearance may be seen on axial T2 images due to involvement of the anterior grey matter.

As in the brain, restricted diffusion in infarct can be detected within hours and persists up to 1 week. Once the blood-cord barrier is disrupted, enhancement of the infarcted region is seen after contrast administration.
Concomitant infarction of the vertebral body may also be seen if the more proximal segmental artery is also involved and may help to localize the level.

Lack of enhancement will distinguish this lesion from spinal tumor. Demyelinating disease tends to be located more peripherally than infarction and involves less than 2 vertebral segments. Transverse myelitis may be indistinguishable from infarction on imaging studies but varies in clinical presentation.

4- Myelitis

a- Multiple sclerosis (MS) (Fig 5)

MS is considered the most common primary demyelinating disease of the central nervous system (CNS), characterized by distinct episodes of neurological deficits separated in time, which are caused by white matter lesions that are separated in space. There is a female predominance, young age of onset (10-50 years), and a higher prevalence in temperate climates. The clinical presentation varies depending on the spinal cord segment affected and the degree of involvement.

According to the modified McDonald criteria for MS, the diagnosis requires objective evidence of lesions disseminated in time and space.

Spinal cord plaques can be found at any level, most commonly the cervical spine. They are primarily eccentric and peripherally located and generally do not respect boundaries between white and gray matter. The lesions typically involve the dorsolateral regions of the cord and most commonly distributed less than half the cross-sectional area of the spinal cord. Most of the lesions are smaller than 2 vertebral body segments. Acute lesions usually demonstrate prolongation of both T2 and T1 times. Most MS plaques appear as well- or ill-defined hyperintensities on T2-weighted images with larger active lesions associated with cord expansion and extensive edema. They usually demonstrate homogeneous, nodular, or ring-enhancement during the acute and subacute phase. Enhancement may last 2-8 weeks, while chronic lesions do not demonstrate contrast enhancement.

The differential diagnosis include spinal cord neoplasm, Adem, transverse myelitis, infarction, sarcoidosis, and arteriovenous malformation, especially dural fistula.

b- Acute disseminated encephalomyelitis (ADEM) (Fig 6)
ADEM is a severe, monophasic illness of children and young adults. It is characterized by a rapid onset of symptoms and signs often preceded by a viral illness. As the name suggests both the brain and spinal cord are affected. The clinical picture is that of severe headache, drowsiness and the rapid development of neurological signs. ADEM is seen in the cord as multiple, flameshaped, high T2 signal, intra-axial lesions. These tend to be more extensive than MS. There is often cord swelling and patchy peripheral enhancement, which can make these lesions difficult to differentiate from cord tumours on imaging grounds alone.

Brain lesions often coexist and are more useful in differentiating this condition from other causes of cord inflammation as they are more distinctive. Unlike MS the lesions often involve the basal ganglia and thalamus.

c- Infection (Fig 7)

On the whole, infectious myelopathy due to viral myelitis is equivalent in imaging terms to idiopathic transverse transverse myelitis. The diagnosis is reached by laboratory testing of the cerebrospinal fluid, serum serology, or by exclusion of other causes. Occasionally, however, the imaging is specific for other infectious causes.

Being aware of and differentiating these is important as early treatment is vital.

These typically include bacterial (Staphylococcus, Streptococcus, and Mycobacterium are the most frequently reported organisms, Lyme disease), viral (enteroviruses, human immunodeficiency virus (HIV), human T-lymphotrophic virus -1 (HTLV-1), Epstein-Barr virus (EBV), cytomegalovirus (CMV), varicella zoster virus (VZV)…), fungal and parasitic.

d- Other myelitis:

- Acute Transverse Myelitis
- Autoimmune myelitis (systemic lupus erythematosus, Bechet's syndrome, Sjogren's syndrome, Wegener's syndrome, sarcoid, Neuromyelitis Optica)

5- Cavernous Malformation (Fig 8)

Cavernous malformation (CM) is also named cavernous angioma, cavernous hemangioma, and cavernoma. Cavernous angioma, which is known as a "blood sponge," is a slow-flow vascular lesion without shunting.
The spinal cord is an uncommon site of occurrence with 3%-5% of all CMs involving the spine, most often distributed in the intramedullary compartment.

The peak presentation is in the fourth decade and there is a female predominance.

Ten percent to 30% of spinal cord CMs are associated with multiple (familial) cavernous malformation and carry a higher risk of hemorrhage.

The clinical presentation is variable. The classic presentation is episodic sensorimotor symptoms with intermittent recovery.

Findings on MR imaging are often characteristic and usually provide a relatively specific diagnosis. CM typically demonstrates speckled "popcorn" heterogeneous signal intensity on both T1- and T2-weighted images due to blood products in various stages of evolution. The lesion may demonstrate a peripheral rim of hemosiderin of low signal intensity on both T1- and T2-weighted images. Gradient echo sequences demonstrate "blooming" susceptibility artifact from the presence of blood products.

With the presence of acute hemorrhage and edema, MR appearance of CM may become less specific and other etiologies such as neoplasm, including ependymoma, astrocytoma, hemangioblastoma and metastasis, MS, and AVMs, should be considered in the differential diagnosis. Enhancement is either absent or minimal. CMs are angiographically occult vascular lesions; thus, conventional angiography is unrevealing.

6- Spondylitic myelopathy  (Fig 9)

Cervical spondylitic myelopathy results from segmental or generalized degenerative disease of the cervical spine. It has been proposed that a congenitally narrowed cervical spinal canal, progressive spondylosis, mechanical spinal cord compression, or alterations in vascularity of the spinal cord contribute to the development of cervical spondylitic myelopathy.

The diagnosis is usually easy at MRI demonstrating an ill-defined hyperintensities on T2-weighted images interesting all or a part of the spinal cord next to a degenerative spinal stenosis causing an anteroposterior spinal flattening.

II. Neoplasm:

Intramedullary neoplasms include dermoid tumor, astrocytoma, ependymoma, hemangioblastoma, and metastases.

1. Dermoid Tumor
Dermoid tumors or dermoid cysts are rare congenital benign, slow-growing tumors composed of more than 1 of 3 primitive germ cell layers that produce skin and its appendages. They represent 1%-2% of intraspinal tumors.

The lumbosacral region is the most common site to be affected (60%), followed by upper thoracic (10%) and cervical (5%) spine.

**Extramedullary location (60%) is more common than intramedullary site (40%). (Fig 10)**

They can be associated with dermal sinus (20%), vertebral abnormalities, and closed dysraphism. Dermoid cysts usually cause symptoms before 20 years of age and there is no sex predilection.

On MR imaging, dermoid cysts can appear hypointense or hyperintense on T1-weighted images. They contain 2 different components, a lipid and a more solid or more fluid. The lipid component appears hyperintense focus on T1-weighted images and exhibits low signal intensity on postgadolinium fat-suppression T1 weighted images. On T2-weighted images, the lesions usually have hyperintense signal, but may be very hypointense depending on the amount of water and fat contents. The soft-tissue component usually enhances after administration of intravenous contrast.

Differential diagnosis of intramedullary dermoid cysts includes syringomyelia with hemorrhage and neoplasm with hemorrhage. The fat-suppression technique of MR imaging would make the diagnosis fairly easy.

2. **Astrocytoma (Fig 11)**

Approximately 36% to 54% of gliomas are astrocytomas. The peak incidence of spinal astrocytomas is in the third and fourth decade with the average age of 31 years with a slight male predominance. Seventy-five percent to 92% of cord astrocytomas are benign in adults.

Astrocytomas are most often located in the thoracic cord and much less frequent in the lower thoracic and lumbar regions. They are frequently located eccentrically in the posterior columns and over multiple segments.

The presenting symptoms are frequently nonspecific.

On T1-weighted images, astrocytomas demonstrate low signal intensity. Cord enlargement is virtually always present.
On T2-weighted images, the tumors and associated edema are hyperintense. The lesions almost always enhance immediately with contrast; however, enhancement can be inhomogeneous.

The margins of the lesions are often poorly defined and irregular. Intratumoral or peritumoral cysts are often seen.

3. Ependymoma (**Fig 12**)

Ependymomas tend to be intracranial rather than intraspinal with approximately 31%-36% of reported ependymomas being intraspinal. The lesions usually present during the fourth and fifth decade of life.

Ependymomas are the most common primary cord tumor of the lower spinal cord, conus medullaris, and filum terminale.

Myxopapillary ependymoma, a subtype of ependymoma, is particularly common in the conus and filum and constitutes about 30% of all ependymomas.

Cysts are seen in 50% of the cases. Calcification is extremely uncommon in spinal ependymomas.

Ependymomas cause expansion of the cord typically over 3-4 segments. The lesions are hypointense or isointense relative to normal cord on T1-weighted images and are typically heterogeneous on T2-weighted images.

Ependymomas tend to enhance intensely, but heterogeneously and often have well-defined margins. Intratumoral and peritumoral cysts are often identified.

It is often difficult to differentiate ependymomas from astrocytomas by imaging criteria but there are a few suggestive features:

- Ependymomas occur more often in the lower cord and conus than astrocytomas.
- Ependymomas tend to be central, whereas astrocytomas arise eccentrically within the posterior cord.
- Ependymomas are more frequently hemorrhagic
- Ependymomas have a thin pseudocapsule separating the tumor from the normal cord.

4. Hemangioblastoma (**Fig 13**)


Hemangioblastomas are low-grade capillary-rich neoplasms of the cerebellum and spinal cord that occur either sporadically or in association with von Hippel-Lindau (VHL) syndrome. These benign neoplasms account for 1%-7% of all spinal cord neoplasms and are most commonly intramedullary (75% of cases) in location.

The peak of age is 30-40 years.

The thoracic cord is most commonly involved (50% of cases) followed by the cervical cord (40% of cases).

At MR imaging, hemangioblastomas demonstrate diffuse cord expansion and variable signal intensity on T1-weighted images, most commonly isointense. T2-weighted images characteristically exhibit a hyperintense focus with intermixed flow void. Associated cyst formation or syringohydromyelia is very common, as well as considerable edema. Contrast-enhanced MR typically shows an intense, homogeneously enhancing subpial tumor nodule, allowing differentiation of the small nidus form the adjacent edematous cord and peritumoral cyst, as well as from arteriovenous fistulae, cavernomas, and other hypervascular intramedullary spinal cord neoplasms.

5. Other primitive spinal cord tumor

Ganglioglioma; ganglioneuroblastoma; lymphoma; lipoma.

6. Intramedullary Spinal Cord Metastasis (Fig 14)

Metastatic intramedullary tumors are exceedingly rare compared to extradural metastases, accounting for only 0.9%-2.1% of autopsied cancer patients. They are most commonly seen in the cervical and thoracic cord with lung carcinoma being the most common primary site, followed by breast, melanoma, renal, colorectal, and lymphoma. Most patients with a spinal cord metastasis have symptoms for less than 1 month before diagnosis.

MR imaging usually shows expansion of the spinal cord on T1-weighted images and focal areas of contrast enhancement that may be homogeneous or ring enhancing.

On T2-weighted images there is usually high signal and cord expansion although there also may be smaller areas of lower or intermediate signal. The degree of cord expansion and high signal on the T2-weighted images is often much more extensive than the smaller focal enhancing area, suggesting that the enhancing area represents tumor and the more extensive high signal represents edema.

These imaging characteristics are no specific to clearly distinguish from other intramedullary lesions.
Differential considerations include hemangioblastoma, demyelinating disease (MS and Adem), arteriovenous malformation, sarcoidosis, and transverse myelitis.
Fig. 1: Normal cervical spine MRI. Sagittal T1 (A) and sagittal T2 (B) images demonstrate bright CSF surrounding the cord within the subarachnoid space.

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**Fig. 2:** Chiari I malformation with hydromyelia. Sagittal T1 (B) and T2 (A) and axial Gradient Echo images (C) demonstrate a large cystic central canal dilatation of the cervical and thoracic spinal cord, consistent with a hydromyelia. CSF flow-related artifact is noted within the syrinx. There is protrusion of the cerebellar tonsils below the foramen magnum (arrow), consistent with Chiari I malformation.

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Fig. 3: Sagittal STIR and Gradient Echo spine MRI: A severe cervical sprain with traumatic disc herniation. The spinal cord has increased T2 signal (arrow).

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**Fig. 4:** Spinal cord infarction. A 25-year-old woman presented with acute onset lowerlimb weakness. She was found to have a flaccid paraparesis with up going plantar responses. Sagittal T2 (A), axial, GRE (E) and postcontrast T1 (F) images demonstrate diffuse central T2 hyperintensity with mild cord expansion, involving the cervical cord. The area of infarction has mild enhancement on postcontrast images. Diffusion images (B, C, D) demonstrate diffusion restriction consistent with cytotoxic oedema.

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Fig. 5: MS. A 56-year-old woman with known history of MS presents with exacerbation of sensory and motor symptoms of her upper extremities. Sagittal T2 (A, B) and postcontrast T1 (C) images of the cervical spine demonstrate a short, well-defined, eccentric lesion within the cervical cord with ring-enhancement. Imaging of the brain (D, E) demonstrated ovoid high T2 lesions in a juxtaventricular and subcortical location.

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Fig. 6: ADEM. A 6-year-old boy presented with confusion after a viral illness. T2-weighted sagittal image (A) of the cord demonstrates an increase in signal, with fusiform swelling of the cervical cord. On contrast-enhanced images (B) there was a mild enhancement. The axial T2 images of the brain (C,D) demonstrate multifocal areas of increased T2 signal in pons, left middle cerebellar peduncle and medulla oblongata.

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**Fig. 7:** Myelitis. A 62-year-old patient presents with acute bilateral lower extremity weakness and numbness. Sagittal T2 (A) and postcontrast T1 (B,C,D) images demonstrate an expanded and edematous cord with diffusely increased T2 signal and enhancement extending over 4 vertebral segments.

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**Fig. 8:** Cavernous malformation. Cervical spine MRI showing a diffuse cervical heamatomyelia. The second MRI practiced 2 months later (second T2 sagital slice and axial Gradient Echo axial slice) shows a significative regression of heamatomyelia and presence of spinal cavernous angioma.

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**Fig. 9:** Cervical spondylitic myelopathy. Sagittal T1 (A) and T2 (B) images demonstrate intramedullary high signal intensity involving both white and gray matter next to a degenerative spinal stenosis. Flattened spinal cord

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**Fig. 10:** Intradural-extramedullary dermoid Tumor. Sagittal T1 (A), T2 (B) and postcontrast T1 (C) images demonstrate a mixed hypointense and hyperintense T1 signals due to presence of both lipid and water content. There is also mild enhancement on postcontrast sagittal T1 image (C).

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**Fig. 11:** Astrocytoma. A 18-year-old man presents with back pain and urinary leakage. Sagittal T1 (A), T2 (B), and postcontrast T1 (C,D) images show a centromedullary lesion within the thoracic cord. The lesion demonstrates hypointense T1 and high T2 signals with inhomogeneous enhancement.

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Fig. 12: Cervical spine MRI in sagital and axial T2 and axial T2 fat sat GADO: Mixopapillary ependymoma appearing as a cervical cord expansion with multiple cystlike lesions.

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**Fig. 13:** Hemangiblastoma. A 54-year-old man presents with heaviness of the lower limbs. Sagittal T1 (A,B), and postcontrast T1 (C) images show an intensely enhanced nodule (arrow head) of isointense T1 signal at the posterior aspect of the cord. A large area of hypointense T1 and hyperintense T2 signal (black and white arrow), extending from the medulla to upper thoracic cord and with expansion of the cord, is consistent with edema and an associated syringomyelia.

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**Fig. 14**: Intramedullary metastasis (primary lung). A 22-year-old man presents with bilateral lower extremity paresthesias and urinary retention. Postcontrast images show multiple nodular lesions (black arrow) with homogeneous enhancement the lower thoracic cord.

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Findings and procedure details

The MR examinations were performed on a 1.5-T superconducting magnet (General Electric, Milwaukee, WI).
Conclusion

MR imaging is the modality of choice for evaluation of intramedullary lesions of the spinal cord. Knowledge of the structural organization of the spinal cord and its blood vessels, as well as the pathophysiology and temporal evolution of many diseases affecting the cord, frequently allows the radiologist to make definitive diagnoses using MR imaging. An important benefit of this knowledge is that the biopsy of nonsurgical lesions (e.g., infarction of the conus medullaris, transverse myelitis) can be avoided.

An early diagnosis and therapy is essential with intramedullary spinal cord diseases to hold residual neurological deficits as low as possible.
References