Learning Objectives

The purpose of this pictorial review is:

1. To illustrate the MRI characteristics of a broad spectrum of intramedullary spinal cord lesions.
2. To discuss the clinical presentation and differential diagnoses of these intramedullary spinal cord lesions.
3. To discuss the utility of MRI and to provide the radiologist with an approach to accurately differentiating between these lesions.

Background

Magnetic resonance imaging (MRI) is the modality of choice for the evaluation of intramedullary spinal cord lesions. A detailed understanding of the anatomy of the spinal cord and the predilection of various conditions for different patterns of cord involvement is essential for the reporting radiologist. MRI is able to provide a diagnosis or limited differential diagnosis in the majority of cases, providing superior contrast differentiation between cord, epidural space and CSF compared to CT and thus usually allows discrimination between intra and extra medullary lesions.

This article presents a range of both benign and malignant non traumatic, intramedullary conditions of the spinal cord that are likely to be encountered by radiologists, and are broadly categorised as congenital, demyelination, vascular and neoplastic. The pertinent clinical features of each condition and their MRI appearance are discussed with an emphasis on differentiating features.

Imaging Findings OR Procedure Details

CONGENITAL ABNORMALITIES:

SYRINX (not necessarily congenital):

The term syrinx is used to collectively describe the presence of a cystic intramedullary cavity. The term syringomyelia refers to a cystic cavity that is around the central canal of the spinal cord and is distinct from hydromyelia in which there is simply dilatation of
the central canal. When these two conditions cannot be distinguished the collective term syringohydromyelia is used.

The predominant symptoms are dissociated pain and thermal sensory impairment [1] in a cape like distribution over the back and upper limbs. Causes can be divided into primary, or congenital, such as the Chiari 1 and 2 malformations, and secondary causes as a result of trauma or tumours. [2].

On MRI, a syrinx appears as a cystic cavity within the spinal cord that typically follows CSF signal on all sequences. There may be associated CSF pulsation artefact that is best visualised on the T2 weighted sequence and most common in the largest syrinx cavities [2]. Syrinxes are most commonly found at the cervicothoracic junction and are variable in length. They do not enhance on post contrast studies.

See Figures 1 - 2.

**VENTRICULUS TERMINALIS:**

The ventriculus terminalis, or fifth ventricle, is a simple CSF filled cavity of the conus medullaris that forms in early embryogenesis and usually regresses in the first week after birth. It may, however persist as an open cavity and is usually an incidental finding. Rarely, it can become large enough to cause bilateral sciatica or cauda equina syndrome. It can be observed at any age and is typically smallest in middle age and largest in early childhood and advanced age [3]. The ventriculus terminalis follows CSF signal on all sequences and does not enhance. It can measure up to 20 mm in length and 2-4 mm in diameter with rare cases of cystic dilatation reported. Differentials include syringohydromyelia, cystic myelomalacia or cystic neoplasm [4].

See Figures 3 - 4.

**DEMYELINATING CONDITIONS:**

**IDIOPATHIC ACUTE TRANSVERSE MYELITIS:**

Acute transverse myelopathy is a term used to describe any disorder that acutely affects function on both sides of the spinal cord regardless of longitudinal extent. The
term encompasses a broad range of conditions including multiple sclerosis, neoplasm, infarction and trauma. Idiopathic acute transverse myelitis (IATM) should be distinguished from the broader category of acute transverse myelopathy. The clinical criteria for the diagnosis of IATM include a rapid onset of bilateral motor, sensory and autonomic dysfunction in the absence of other known neurological disease and is, therefore, a diagnosis of exclusion [5]. Its aetiology is uncertain but is most often suggested to be an autoimmune process. On MRI, IATM demonstrates low T1 and high T2 signal that most commonly extends over multiple vertebral body segments, at least 2, and typically involves the majority of the cord in cross section. Variable enhancement is reported, 38% in one study and 47% in another [6]. The cord may be of normal or slightly increased calibre. There is often a dramatic response to steroid treatment [4].

Clinically, the condition can be distinguished from acute spinal cord infarction based on the absence of a sudden onset of symptoms. Typically spinal cord infarction also spares the posterior columns of the cord.

See Figure 5.

**SUBACUTE COMBINED DEGENERATION:**

Vitamin B12 deficiency is an uncommon systemic disease that can affect the nervous system, of which cord involvement is a manifestation. The most common cause is malabsorptive states, of which pernicious anaemia predominates. The spinal condition is clinically characterised by systemic dysaesthesia, disturbance of position and vibration sense, and spastic paraparesis or tetraparesis that most commonly affects the lower limbs [7]. The term subacute denotes the course of the clinical features in which symptoms evolve over weeks to months and can cause severe disability [8]. It most commonly presents in the fifth to eight decades.

The disease most commonly affects the posterior columns of the cord, manifest as increased T2 signal intensity without contrast enhancement within the dorsal columns. The lateral columns may also be affected. The lesion is of variable length and typically begins in the thoracic region.

The final diagnosis can be made with clinical and biochemical correlation.

See Figure 6.
MULTIPLE SCLEROSIS:

Multiple sclerosis (MS) is a primary demyelinating condition of the central nervous system characterised clinically by distinct neurological deficits that are separated in both time and space. It has a female predominance of 2:1 with a peak age of onset of 20 - 40 years. It is thought to be autoimmune in aetiology.

The natural history of MS is variable, and the most common of three patterns of disease, relapsing-remitting MS, is typified by acute episodes with partial or complete recovery. The symptoms depend on the location of the affected cord segment and may include spastic paraparesis or paraplegia, bladder or bowel dysfunction or sensory disturbance.

On MRI of the spinal cord, T2 hyperintense lesions have a cigar shape on the sagittal plane and on the axial plane are most frequently eccentric with a propensity for the posterior columns [9]. The lesions rarely occupy more than half of the cord or exceed two vertebral body segments in length. Lesions may be distributed throughout the neuraxis however predominate within the cervical cord [10], most commonly the mid cervical region.

Enhancement may be seen in acute lesions and typically lasts between 2 - 6 weeks. The pattern of enhancement is variable but is often patchy, peripheral and incomplete [11]. Acute lesions are often associated with mild cord swelling.

The presence of associated intracranial lesions, visualised in 90% [4], in a typical distribution for MS significantly assists diagnosis. The Modified McDonald Criteria 2010 provides both a high degree of sensitivity and specificity for early detection of the condition.

See Figures 7 - 8.

NEUROMYELITIS OPTICA:

Neuromyelitis Optica (NMO), previously called Devic's disease, is considered to be a separate neurological syndrome to MS [12] and is most likely immune mediated. The syndrome is characterised by a severe myelopathy associated with acute unilateral or bilateral optic neuropathy. There is typically no clinical involvement beyond the optic nerves or spinal cord and the condition may be either monophasic or multiphasic in its course [13]. If there are lesions within the brain they are atypical for MS [14]. Some patients with NMO develop lesions within the hypothalamus that have been described
in one article as being characteristic, if not specific to NMO or its spectrum of disorders [15]. The lesions are hyperintense on T2, predominantly involve the hypothalamus and occasionally extend into parenchyma surrounding the third and fourth ventricles[15].

In contrast to MS, the T2 hyperintense spinal cord lesions in NMO tend to extend over multiple vertebral body segments (more than 3) and are frequently associated with cord swelling [12]. NMO primarily involves the central portion of the spinal cord on axial sections [14] and a large proportion of the lesions are hypointense on T1 weighted scans as opposed to MS where lesions are typically isointense. Enhancement is reported from none to intense depending on the phase of the condition.

See Figures 9 - 10.

**VASCULAR:**

**SPINAL CORD INFARCTION:**

Spinal cord infarction is a rare condition. It most frequently occurs in the anterior spinal artery (ASA) territory or its terminating branches. The ASA supplies the anterior two thirds of the spinal cord, and is the main supply to the grey matter.

Patients with ASA territory infarction present with an acute, rapidly progressive sensorimotor deficit. There is often severe pain irrespective of the extent of the infarction. In the uncommon case of PSA infarction, proprioception, light touch and vibration sense with be affected.

Aetiologies include aortic or vertebral artery dissection, atherosclerosis, hypotension, embolism, vasculitis, and spinal or vascular surgery [16]. Some cases are idiopathic. It most commonly occurs in the thoracic and thoracolumbar spine [4, 17]. On MRI, infarction appears a 'pencil-like' hyperintensity on sagittal T2 weighted sequences [16] that may be accompanied by cord swelling and typically spans over several segments. On axial T2 weighted sequences, an 'owl's eye' appearance may be seen due to involvement of the grey matter, however both grey and white matter may be involved. Restricted diffusion may be seen early but, unlike the brain, may show a more rapid signal normalisation [17]. There may be patchy ill-defined enhancement in the subacute phase.
Extra spinal findings, such as the presence of an abdominal aortic aneurysm, vertebral dissection or abnormal signal in the vertebral body supplied by the same segmental artery as the affected region of the cord [4] may aid diagnosis.

See Figures 11 - 12.

**ARTERIOVENOUS MALFORMATION:**

Arteriovenous malformations (AVM) of the spine are rare lesions that encompass any arteriovenous shunt along the axis of the spinal cord. Depending on the type of malformation, symptoms may vary between acute intramedullary or subarachnoid haemorrhage and subacute venous congestion leading to progressive myelopathy. There are various classification schemes described, with the conventional scheme dividing AVMs into 4 types [18].

The first type is the most commonly encountered (up to 80%), and is a spinal dual arteriovenous fistula (AVF) which typically consists of a single radiculomedullary artery that drains into one or more low-pressure pial veins. Sometimes, there may be multiple feeding arteries. They most commonly occur in the lower thoracic cord or conus medullaris and typically present in male patients (5:1) in the fifth or sixth decade. They typically present with an insidious onset of lower extremity weakness and sensory deficit with non radicular pain and very rarely associated with acute haemorrhage.

Type II (Glomus) and III (Juvenile) AVMs are true arteriovenous malformations that are intramedullary in location and are fed by branches of the anterior and posterior spinal arteries into a nidus and then into spinal veins. Type IV AVMs are intradural, extramedullary arteriovenous fistulas and comprise of a direct fistula between anterior and posterior spinal arteries and a draining vein with no intervening nidus.

On T2 images, peri-nidus increased signal may be due to oedema (with cord expansion) or gliosis (with volume loss). Serpentine, low signal intensity flow voids indicating the tortuous feeding vessels and draining veins are best depicted on T2 weighted sagittal sequences [19] however may be obscured by CSF pulsation artefact. Variable T1 hyperintensity may be visualised due to the presence of blood products. Variable enhancement of the nidus and enhancement of the vessels may be identified post contrast administration.

Although MRI is the best non invasive modality for the evaluation of this condition, the definitive diagnostic modality remains catheter angiography.
See Figures 13 - 14.

NEOPLASTIC:

EPENDYMOMA:

Ependymomas are the most common spinal cord tumour in adults, accounting for 60% of cases. They typically present in the fourth and fifth decade of life with a slight male predominance. Cord ependymomas most commonly occur in the cervical region and may extend to involve the upper thoracic cord [20]. The myxopapillary ependymoma subtype (13% of cases of spinal ependymomas) has a distinct predilection for the conus medullaris or filum terminale and is the most common histological cause of neoplasm in that region (83%) [20].

Patients most frequently present with an insidious onset of back or neck pain, radicular pain, unsteady gait, paraesthesia and progressive paraplegia due to the slow growth of the tumour [20], with a slight predominance of sensory symptoms.

Centrally located with symmetrical cord expansion, ependymomas are typically isointense on T1 although may have areas of hyperintensity due to haemorrhage, a complication that is uncommon with astrocytomas. On T2 weighted sequences, they are typically hyperintense, although may be isointense relative to the cord. 20 - 30% of cases demonstrate the 'cap sign', a rim of haemosiderin manifest as very low T2 signal at the poles of the tumour [20]. The average length of the tumours is 3 - 4 vertebral segments [4]. On post contrast T1 weighted images, ependymomas typically enhance homogeneously (75%) and have a clearly defined margin [21], a useful differentiating feature from astrocytomas which are typically poorly defined.

Tumour associated cysts are common [20], the most common being non-tumoural cysts located at the rostral and caudal poles of the tumour that do not enhance and are not part of the tumour itself. Syrinxes, due to disturbance of CSF circulation, may also be seen [20]. Extramedullary findings include scalloping of the adjacent vertebral body and canal widening due to the slow growing nature of the neoplasm.

See Figures 15 - 16.
**ASTROCYTOMAS:**

Astrocytomas are the second most common intramedullary neoplasm in adults, typically presenting in the third and fourth decade [20]. Like ependymomas, there is a slight male predominance and the presenting symptoms are similar.

Approximately 85% are low grade, slow growing with symptoms progressing for many months to years before diagnosis [22]. High-grade astrocytomas have a more rapid symptom progression.

On MRI, astrocytomas are T1 hypointense and T2 hyperintense masses with cord expansion and patchy enhancement less intense than ependymomas. They are more likely to be eccentrically located within the cord compared to ependymomas [22] and typically have less well defined margins. The average length is seven vertebral segments [20], however they may rarely be multisegmental or involve the entire cord (holocord). Associated haemorrhage is uncommon. Non-tumoural cysts (caudal and rostral cysts) are commonly identified. Tumoural cysts may also be present, are contained within the tumour, demonstrate peripheral enhancement and are more frequently than in ependymomas. Vertebral body erosion and widening of the interpedicular distance is less commonly visualised in astrocytomas compared to ependymomas.

See Figures 17 - 18.

**HAEMANGIOBLASTOMA:**

Haemangioblastomas are benign, WHO grade 1, neoplasms that constitute approximately 1.6 - 6.4% of spinal cord tumours [23]. The mean age of presentation is approximately 30 years with no gender predilection.

Approximately 75% of cases of haemangioblastoma are intramedullary, however they may also be extramedullary, intradural or extradural. Most are solitary (80%). Approximately 25% of cases occur in patients with Von Hippel Lindau (VHL) disease and the presence of multiple lesions is almost exclusively found in this condition [23]. The remainder are sporadic.

Typically a long clinical course to diagnosis is observed although they may rarely present with subarachnoid haemorrhage [20] or polycythaemia secondary to upregulation of erythropoietin.
Hamangioblastomas vary in size from a few millimetres to several centimetres in length. They are isointense to hypointense to normal spinal cord on T1 weighted images and isointense to hyperintense on T2 weighted sequences [24]. Prominent flow voids representing dilated tortuous feeding arteries and draining veins may be visualised [21] as well as peritumoural oedema. They are commonly associated with peritumoural cysts and syrinxes (55% in one study) [20, 24] however unlike ependymomas, the rostral or dorsal cysts are often identified at the pial surface of the cord rather than the centre [21]. Haemangioblastomas typically demonstrate intense, homogeneous enhancement. Some cases may have the classic 'cyst with an enhancing nodule' appearance characteristic of cerebellar haemangioblastomas [20].

The presence of a well-defined enhancing mass assists in differentiating haemangioblastomas from AVMs [24].

See Figures 19 - 20.

**SPINAL CORD METASTASES:**

Intramedullary spinal cord metastases are rare and typically produce rapid neurological dysfunction. The most common cause is lung carcinoma followed by breast carcinoma.

They are typically small lesions however associated oedema and cord expansion may be disproportionately large. They are usually T1 hypointense and T2 hyperintense and may demonstrate evidence of intrinsic haemorrhage. In contrast to the primary intramedullary neoplasms, metastases are rarely associated with peritumoural cysts [4]. They tend to enhance intensely.

See Figure 21.

**OTHER RARE NEOPLASTIC CONDITIONS:**

Ependymomas and astrocytomas account for approximately 90% of intramedullary tumours. A wide variety of neoplastic conditions account for the remaining 10% and include lymphoma, oligodendrogial tumours, neuronal and mixed neuronal-glial tumours (such and gangliogliomas) and primitive neuroectodermal tumours.

**OTHER:**
HIRAYAMA'S DISEASE:

Hirayama's disease is a benign, juvenile spinal muscular atrophy of the upper extremities characterised by unilateral or asymmetric wasting of the C7, C8 and T1 innervated muscles [25]. It is rare, typically presents in the second and third decade and affects males more than females. Most cases are sporadic with rare familial cases reported.

On T1 weighted MRI, there is focal cord atrophy in the C4 - C7 distribution that may be accompanied by increased T2 signal in the anterior horns of the grey matter. With flexion MRI, anterior displacement and indentation of the cord occurs with widening of the posterior epidural space. The neuroradiological findings are not specific with differentials including trauma or amyotrophic lateral sclerosis [25] and hence the clinical presentation is useful in the differentiation.

See Figures 22 - 23.

Images for this section:
Fig. 1: Syrinx in a 31 year old woman with a history of posterior fossa decompression for Chiari 1 malformation. Sagittal T2-weighted MR image demonstrates cystic dilatation of the central canal from C2 to T8.
Fig. 2: Sagittal T2-weighted MR image from C1 to T5 in a 50 year old woman demonstrates a Chiari 1 malformation with tonsillar descent and an extensive syrinx extending from the medulla to the mid thoracic cord. In addition, there is an 18 mm mass within the inferior aspect of the fourth ventricle that is hyperintense to brainstem, histologically proven to be an ependymoma.
Fig. 3: Ventriculus terminalis in a 44 year old man with back pain. T1-weighted MR image demonstrates an intramedullary cystic lesion at T11/T12 measuring 40 mm in craniocaudal dimension, that follows CSF signal.
Fig. 4: Sagittal T2-weighted MR image demonstrates the cystic lesion to follow CSF signal. There is mild increased T2 signal within the cord immediately superior to the cyst.
Fig. 5: Idiopathic transverse myelitis in a 34 year old woman with bilateral lower limb weakness. Sagittal T2-weighted MR image from C1 to T5 demonstrates a long segment of abnormal increased signal within the cord from C3 to T5 involving the majority of the cord volume.
Fig. 6: Subacute combined degeneration of the cord in a 52 year old lady with upper limb sensory disturbance. Figure 5a - Sagittal T2-weighted MR image demonstrates abnormal increased T2 signal within the posterior column from the C2/3 junction to C6 without cord expansion. A disc protrusion at C5/6, indenting the anterior aspect of the cord, is not associated with focal signal alternation or high grade spinal canal narrowing.
**Fig. 7:** Multiple sclerosis in a 47 year old lady. Sagittal T2-weighted MR image demonstrates two T2 hyperintense foci within the cord at C2 and C3, with the largest lesion at C3 associated with mild cord expansion. Neither lesion measures more than one vertebral body in craniocaudal length.

**Fig. 8:** Axial T2-weighted MR image at C3 of the same patient shows the T2 hyperintense lesion to be eccentrically position within the cord on the right and associated with mild cord expansion.
**Fig. 9:** Neuromyelitis optica in a 45 year old woman with a 1 month history of progressive lower limb weakness and left optic neuritis. Sagittal T2-weighted MR image demonstrates abnormal increased T2 signal within the central portion of the cord from the cervicothoracic junction to at least T9 associated with mild cord expansion.
**Fig. 10:** Contrast enhanced sagittal T1-weighted MR image demonstrates faint, patchy amorphous enhancement within the lower thoracic cord that corresponds in position to altered signal on the T2 weighted sequence.
**Fig. 11:** Spinal cord infarction in an 80 year old man with sudden onset paraparesis. Sagittal T2-weighted MR image demonstrates abnormal increased signal within the cord from T8 to the conus associated with lower cord expansion. Superiorly, the altered signal is anterior in location, sparing the posterior columns, whilst inferiorly it involves the majority of the cord in cross section.

![Fig. 11](image1.png)

**Fig. 12:** Axial T2-weighted MR image at T9 in the same patient demonstrates increased T2 signal within the anterior grey matter giving an 'owl's eye' appearance.

![Fig. 12](image2.png)
**Fig. 13:** Spinal dural arteriovenous fistula in a 68 year old woman with progressive lower limb weakness and increased tone. Sagittal T2-weighted MR image demonstrates extensive abnormal cord signal from T8 to the conus associated with cord expansion. Abnormal leptomeningeal flow voids are identified on the surface of the cord extending superiorly to the cervicothoracic junction. The diagnosis of a spinal dural arteriovenous fistula was made.

**Fig. 14:** Axial T2-weighted MR image at T10 more clearly demonstrates the abnormal leptomeningeal flow voids in keeping with a spinal dural arteriovenous malformation.
**Fig. 15:** Ependymoma in a 34 year old woman with a diagnosis of Neurofibromatosis Type 1 and progressive upper and lower limb weakness. Sagittal T2-weighted MR image demonstrates the presence of a hyperintense intramedullary mass at C2 with cord expansion. There is an associated syrinx visualized around the mass extending from upper medulla to the lower aspect of C3. There is no low T2 signal within the mass to suggest the presence of haemosiderin.
**Fig. 16:** Contrast enhanced sagittal T1-weighted MR demonstrates homogeneous enhancement of the mass with a well defined peripheral margin. Note the presence of a small enhancing mass within the falx superior to the torcula in keeping with a meningioma.
**Fig. 17:** Astrocytoma in a 67 year old man with a history of progressive upper and lower limb weakness. Sagittal T2-weighted MR image demonstrates an 18mm mass within the central aspect of the cord at T1 with central necrotic or cystic components and peripheral low signal intensity. There is extensive cystic change within the cord extending from the medulla to T4 that demonstrates increased CSF signal intensity likely reflecting the presence of proteinaceous material. There is also increased T2 signal within the thoracic cord below the cystic change from T4 inferiorly.
Fig. 18: Contrast enhanced sagittal T1-weighted MR in the same patient demonstrates heterogeneous enhancement of the mass with ill-defined margins superiorly. There is also enhancement of the periphery and within several septations of the associated cyst in keeping with the diagnosis of a tumoural cyst.
**Fig. 19:** Haemangioblastoma in a 25 year old man with progressive lower limb weakness and sensory disturbance. Sagittal T2-weighted MR image demonstrates an intramedullary mass at T2 that is hyperintense to cord with areas of low signal intensity in keeping with flow voids. Small cysts are identified above and below the mass and there is abnormal increased T2 signal within the cord extending from the cervicomedullary junction to T6 with cord expansion.
**Fig. 20:** Contrast enhanced sagittal T1-weighted MR image in the same patient demonstrates intense, homogeneous enhancement of the mass with central hypointense foci in keeping with flow voids. Further small enhancing lesions are identified at C3 and at C7/T1 that are likely pial based.
**Fig. 21:** Cord metastasis in a 48 year old woman with a history of breast carcinoma and rapidly progressive lower limb weakness. Contrast enhanced sagittal T1 weighted MR image demonstrates the presence of a small, ovoid mass at T11/12 measuring 14mm in craniocaudal length. The mass demonstrates intense enhancement with central hypointensity in keeping with necrosis or cystic change. The margins are well defined. There is abnormal enhancement within the T11 vertebral body in keeping with a metastasis.
**Fig. 22:** Hirayama disease in a 27 year old woman with a progressive history of upper limb weakness and muscle wasting. Sagittal T1-weighted MR image demonstrates loss of normal cervical lordosis in the neutral position with fusiform narrowing of the cord at C6/7.

**Fig. 23:** Sagittal T2-weighted MR image in flexion demonstrates anterior movement of the cord which is flattened anteriorly, the greatest at C6/7, with associated widening of the posterior epidural space. The degree of cord narrowing at C6/7 is more pronounced in flexion.
Conclusion

There are a wide variety of conditions that result in similar imaging findings on MRI, often distinguished by subtle variations that, coupled with an accurate clinical history, examination and/or biochemical studies, can be narrowed to an accurate diagnosis or limited differential. It is essential that the radiologist have a detailed understanding of spinal cord anatomy and the particular imaging features of specific intramedullary lesions to aid clinicians to make a prompt diagnosis.

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References


