Spinal Cord tumors: Radiologic Findings and Differential Diagnosis

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Learning objectives

To describe the radiological findings of the different types of intramedullary spinal cord tumors.

Background

Spinal cord tumors are classified into three groups based on their anatomical location: epidural, intradural extramedullary and intradural intramedullary types. In this paper we will review the intradural intramedullary spinal cord tumors.

Intramedullary spinal cord tumors are uncommon, representing only 4-10% of all central nervous system tumors.

The most common intramedullary spinal tumors are ependymoma (which represents 60% of all spinal cord tumors), astrocytoma (30%) and hemangioblastoma (3%).

Other much less frequent intramedullary spinal neoplasms are ganglioglioma, paraganglioma, metastasis, lymphoma, primitive neuroectodermal tumor (PNET), etc.

Magnetic resonance (MR) imaging is the technique of choice for the diagnosis and evaluation of spinal cord tumors. The most useful sequences are T1 and T2-weighted sagittal images and gadolinium-enhanced axial and sagittal T1-weighted images.

Imaging findings OR Procedure details

There are three important principles in the use of MR imaging to evaluate patients suspected of having an intramedullary spinal cord tumor:

1. **Spinal cord expansion** is the essential imaging criterion for an intramedullary spinal neoplasm. Its absence should suggest a nonneoplastic etiology of the lesion such as demyelinating disease, sarcoidosis, medullary infarction, dural arteriovenous fistula, cystic myelomalacia, etc.
2. The majority of spinal cord tumors show some degree of enhancement following the intravenous administration of gadolinium-based contrast material; in contrast to intracranial tumors, the majority of spinal cord tumors, even low-grade neoplasms, enhance after the administration of contrast material. Nevertheless, the absence of enhancement does not exclude an intramedullary spinal neoplasm in the presence of medullary expansion.

3. Cysts are a common associated finding in the setting of intramedullary spinal tumors. There are two basic types of cysts: tumoral and nontumoral.

Nontumoural cysts tend to be located at the poles of the solid portion of the tumor. They are not part of the tumour itself and should not enhance after the administration of contrast material. Approximately 60% of all intramedullary spinal tumors demonstrate these type of cysts, which are also known as polar, satellite, or rostral or caudal cysts. Only the solid component of the tumour should be resected; the polar cysts with decompress upon removal of the solid portion or they can be aspirated by the surgeon at resection.

Tumoral cysts are contained within the tumor itself and frequently show peripheral enhancement. Identification of the location of the solid enhancing portion of the tumor (including tumoral cysts) is essential in order for the neurosurgeon to be able to perform a laminotomy or laminectomy limited to this area.

CLASSIFICATION

As we already mentioned above, the most common intramedullary spinal cord tumors are the following:

- Ependymoma (60%)
- Astrocitoma (30%)
- Hemangioblastoma (3%)

Other much less common intramedullary spinal cord tumors are ganglioglioma, paraganglioma, metastasis, lymphoma, PNET, etc.

EPENDYMOMA
They are the most common intramedullary spinal neoplasms in adults.

They are believed to arise from ependymal cells that line the central spinal cord canal.

The most frequent location is the cervical spinal cord (44%), followed by the cervicothoracic (23%) and thoracic cord (26%).

They tend to manifest in young adults, with an average age at presentation of 38.8 years, and are more frequent in male patients. They occur with greater frequency in patients with type 2 neurofibromatosis (NF).

They typically present with dorsal or cervical pain (67%), sensory deficits (52%), motor weakness (46%) or intestinal or bladder dysfunction (15%).

**Imaging findings** (Fig 1-3)

Most spinal cord ependymomas are seen as centrally located, well-defined, enhancing lesions. The vast majority of spinal cord ependymomas (84% of cases) enhance to at least some degree and enhancement is typically focal, intense and homogeneous (in some series, however, the majority of tumors showed heterogeneous enhancement).

The presence of a hypointense rim at the poles of the tumor on T2-weighted images is a frequent finding (20-33% of cases). It represents residual hemosiderin due to hemorrhage and it is common in ependymomas and other highly vascular tumors such as hemangioblastomas and paragangliomas.

Polar cysts are a common feature (62% of cases). Tumoral cysts are seen less frequently (22% of cases).

Spinal cord edema is also a frequent finding (60%).

Most spinal cord ependymomas are isointense or hypointense relative to the spinal cord on T1-weighted MR images; rarely they may be hyperintense due to hemorrhagic component of the mass. They are typically hyperintense on T2-weighted MR images, although in the largest review of spinal ependymomas, isointense tumors were as common as hyperintense tumors.

The mean number of vertebral segments involved with abnormal signal intensity is 3.6.
MYXOPAPILLARY EPENDYMOMA

Myxopapillary ependymoma is a subtype of ependymoma that constitutes about 13% of all spinal ependymomas and shows characteristic findings.

It has a marked predilection for the conus medullaris or filum terminale, representing up to 90% of the neoplasms in this region. They are thought to arise from the ependymal glia of the filum terminale. In rare instances, they may arise in the subcutaneous tissue of the sacrococcygeal region, normally without connection to the spinal cord. Primary subcutaneous sacrococcygeal myxopapillary ependymomas are believed to arise from heterotopic ependymal cell rests or vestigial remnants of the distal neural tube.

These tumors tend to have an earlier clinical presentation (mean age, 35 years) and are more common in male patients. They usually manifest with low back pain, sciatica, sacral pain and weakness, and sphincter dysfunction.

**Imaging findings** (Fig 4-6)

They usually present as a lobulated mass in the filum terminale with possible extension to the conus medullaris.

The smaller tumors tend to displace the nerve roots of the cauda equina while the larger tumors frequently compress them.

They are typically isointense relative to the spinal cord on T1-weighted MR images (occasionally they may be hyperintense, a finding that reflects mucin content or hemorrhage) and hyperintense on T2-weighted MR images. Myxopapillary ependymomas are said to have the greatest tendency to bleed of all subtypes of ependymoma; in fact, they may be a source of subarachnoid hemorrhage that occasionally leads to superficial siderosis of the subarachnoid space surrounding the surface of the brainstem, cisterns and cortical fissures. Superficial siderosis is not specific for myxopapillary ependymomas, as it has been noted in association with other highly vascular tumors.

Similar to other ependymoma subtypes, myxopapillary ependymomas show intense enhancement.

ASTROCYTOMA
They are the most common intramedullary spinal tumors in children and the second in prevalence in adults.

The most frequent site of involvement is the thoracic cord (67%), followed by the cervical cord (49%). Unlike ependymomas, they are rarely seen in the filum terminale. Involvement of the entire spinal cord (holocord presentation) is common in children (up to 60% in some series) but rare in adults.

The mean age at presentation is 29 years, a decade younger than in ependymomas, with a slight male predominance. They occur with greater frequency in patients with type 1 NF.

Similar to ependymomas, they most commonly manifest with pain and sensory deficits (53.6% of cases) and motor dysfunction (41.4%). Bowel and bladder deficits are uncommon.

Most astrocytomas (75- 96%) are low-grade tumors (grade I or II) and are considered benign.

**Imaging findings** (Fig 7-10)

Astrocytomas generally show an eccentric location within the spinal cord (given that they arise from the cord parenchyma and not from the central canal), poorly-defined or infiltrating margins and virtually all of them show at least some enhancement, that is most commonly patchy and irregular.

Hemorrhage is uncommon.

Both polar and tumoral cysts are a common feature.

They are typically isointense or hypointense on T1-weighted MR images and hyperintense on T2-weighted MR images.

The average length of involvement is 7 vertebral segments.

DIFFERENTIAL DIAGNOSIS BETWEEN EPENDYMOMA and ASTROCYTOMA
**TABLE 1**

<table>
<thead>
<tr>
<th>EPENDYMOMA</th>
<th>ASTROCYTOMA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adults</td>
<td>Children</td>
</tr>
<tr>
<td>Central location</td>
<td>Eccentric location</td>
</tr>
<tr>
<td>Well-defined margins</td>
<td>Poorly-defined margins</td>
</tr>
<tr>
<td>Hemorrhage common</td>
<td>Hemorrhage uncommon</td>
</tr>
<tr>
<td>Focal, intense, homogenous enhancement</td>
<td>Patchy, irregular enhancement</td>
</tr>
<tr>
<td>Predilection for the conus medullaris or filum terminale</td>
<td>No predilection for the conus/filum</td>
</tr>
</tbody>
</table>

Even with these features it may not be possible to differentiate between these two types of tumors based on imaging findings alone. Differentiating between astrocytoma and ependymoma before surgery is important for the surgeon since ependymomas are usually well-circumscribed lesions and are candidates for total resection while the more infiltrating astrocytomas are difficult to resect completely.

**HEMANGIOBLASTOMA**

Hemangioblastomas are the third intramedullary spinal cord neoplasms in prevalence. They can occur either sporadically (70-80% of cases) or as part of Von Hippel-Lindau (VHL) disease (16-25% of cases). In patients with VHL disease, hemangioblastomas are frequently multiple.

Sporadic cases occur typically in patients between 30-40 years of age. Hemangioblastomas associated with VHL disease typically occur in younger patients (20-30 years of age).

They most commonly involve the thoracic cord (50%), followed closely by the cervical cord (40%). Although the majority of hemangioblastomas (75%) are intramedullary, they may also involve the intradural extramedullary or extradural spaces; 10-15% of cases have an intra and extramedullary component.

They usually present with sensory deficits (39%), motor dysfunction (31%) and pain (31%).

*Imaging findings* (Fig 11-15)
The most common finding consists of a superficially located, solid, well-demarcated nodule, most frequently at the posterior aspect of the spinal cord (reflecting the subpial location) that shows intense and homogeneous enhancement. Cysts and syringomyelia are also common findings (64% of cases). Although syrinx is not specific to hemangioblastomas, as it may be associated with other spinal tumors such as ependymoma and astrocytoma, a small, superficially located tumor with a large syrinx is considered characteristic of hemangioblastoma.

Surrounding edema is a common but variable finding; extensive edema despite the small size of the tumor may be seen.

Occasionally, a hypointense rim representing hemosiderin may be seen at the poles of the tumor on T2-weighted MR images.

Vascular flow voids in or around the tumor that reflect distended feeding arteries or draining veins are characteristic. MR angiography is useful for detecting these anomalous vascular structures. Enlarged feeding arteries, a densely staining tumor nodule and prominent draining veins are observed at angiography.

Hemangioblastomas show variable signal intensity. Small tumors (10 mm or less) are most commonly isointense on T1-weighted MR images and hyperintense on T2-weighted MR images and show homogeneous enhancement, whereas larger tumors tend to be hypointense or mixed hypo- and isointense on T1-weighted images and of heterogeneous intensity on T2-weighted images, with heterogeneous enhancement.

OTHER INTRAMEDULLARY SPINAL CORD TUMOURS

Other much less common intramedullary spinal tumors are ganglioglioma, paraganglioma, metastasis, lymphoma and PNET.

**Gangliogliomas** are more common in children and young adults. Characteristically, they involve more than eight vertebral segments. Most of them show mixed signal intensity on T1-weighted images (a unique feature for spinal tumors) and hyperintensity on T2-weighted images. They usually have an eccentric location within the spinal cord and contain tumoral cysts in 46% of the cases. Surrounding edema is less common than in ependymomas and astrocytomas. The majority of gangliogliomas enhance to at least some degree after the administration of contrast material; patchy enhancement is the most common pattern (65%) although enhancement of the pial surface is also common (58%).
**Paragangliomas** are neoplasms of neuroendocrine origin that show a definite affinity for the filum terminale and cauda equina. MR imaging studies typically reveal a well-circumscribed mass that is isointense relative to the spinal cord on T1-weighted MR images and iso- to hyperintense on T2-weighted images. Like hemangioblastomas, these hypervascular lesions may show prominent serpentine flow voids along the surface and within the tumor nodule. Hemorrhage is common and a hypointense rim on T2-weighted images representing residual hemosiderin may be seen. Some lesions may demonstrate the characteristic salt-and-pepper appearance, which is so common in neck and skull base paragangliomas. They almost always show intense enhancement after the administration of contrast material. Associated syringohydromyelia has been reported in some cases.

Most intramedullary spinal metastases are solitary. Lung carcinoma is the most common primary site, followed by breast carcinoma, melanoma, renal cell carcinoma, colorectal carcinoma and lymphoma. Lesions typically produce mild spinal cord expansion, generally over 2-3 vertebral segments. They usually show low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted images. They demonstrate intense and homogeneous enhancement with prominent surrounding edema, often disproportionately increased for the size of the lesion. In contrast to primary intramedullary neoplasms, cysts are uncommon. (Fig 16-18)

There are only a few reports of the radiological findings of intramedullary lymphomas. Most of them are solitary lesions and all of them show high signal intensity on T2-weighted MR images.

There are few cases of primary spinal PNETs reported in the literature. They have an affinity for the filum terminale and cauda equina. MR imaging findings are non-specific.

**DIFFERENTIAL DIAGNOSIS**

Apart from intramedullary spinal neoplasms, there are numerous nonneoplastic causes of intramedullary lesions such as demyelinating, infectious, inflammatory or vascular diseases. Medullary infarctions, multiple sclerosis, venous hypertension secondary to vascular malformations, hemorrhage and transverse myelitis can cause spinal cord expansion, and some of these lesions may also enhance, representing a potential diagnostic dilemma, especially in the acute stage of multiple sclerosis. It is important to take into account the clinical history of the patient in order to make a proper differential diagnosis.
Fig. 1: Ependymoma in a 40 year-old-male. Sagittal T2-weighted MR image shows expansion of the entire cervical spinal cord, cystic lesions and increased intramedullary signal intensity consistent with edema proximal and distal to the lesion.
**Fig. 2:** T1-weighted sagittal MR image of the same patient shows expansion of the spinal cord with hypointense lesions that represent polar cysts and a isointense intramedullary lesion at the C5-C6 level.
Fig. 3: T1-weighted sagittal MR image after the administration of gadolinium-based contrast agent shows intense and homogeneous enhancement of the solid component of the tumor. Surgery was performed with anatomopathological diagnosis of stage II ependymoma.
**Fig. 4:** Myxopapillary ependymoma in a 86-year-old male. T2-weighted sagittal MR image shows an expansive hyperintense lesion in the conus medullaris.
Fig. 5: T1-weighted sagittal MR image of the same patient shows that the lesion is isointense relative to the spinal cord.
Fig. 6: T1-weighted sagittal MR image after the administration of gadolinium-based contrast agent shows intense enhancement of the lesion.

Fig. 7: Astrocytoma in a 74-year-old female. T2-weighted sagittal MR image shows a mildly hyperintense lesion that expands the spinal cord from the C5-C6 to D1 level, with cystic areas and medullary edema proximal and distal to the lesion.
Fig. 8: T1-weighted sagittal MR image of the same patient shows that the lesion is hypointense relative to the spinal cord.
**Fig. 9:** T1-weighted sagittal MR image after the administration of gadolinium-based contrast agent shows heterogeneous enhancement of the lesion.
**Fig. 10:** T1-weighted gadolinium-enhanced axial MR image shows the eccentrically located intramedullary lesion with poorly-defined margins. The pathological diagnosis revealed grade II astrocytoma.
Fig. 11: Hemangioblastoma in a 70-year-old woman. T1-weighted sagittal MR image shows an intradural nodular lesion that shows intermediate signal intensity at the L1 level.
Fig. 12: T2-weighted sagittal MR image of the same patient shows serpentine flow voids adjacent to the lesion.

Fig. 13: T1-weighted gadolinium-enhanced axial MR image shows intense and homogenous enhancement of the lesion.
Fig. 14: Angiography of the same patient shows a tortuous anterior spinal artery that arises from the left T10 intercostal artery and descends to the L1 vertebral level where an intensely enhancing nodular image is seen.
Fig. 15: Another angiographic image of the same patient shows a prominent draining vein.

Fig. 16: Intramedullary spinal cord metastasis in a 71-year-old male with lung carcinoma. T2-weighted sagittal MR image shows a hyperintense intramedullary lesion at the C5 level associated with extensive spinal cord edema.
**Fig. 17:** T1-weighted sagittal MR image of the same patient shows that the lesion is isointense relative to the spinal cord.

**Fig. 18:** T1-weighted gadolinium-enhanced axial MR image shows intense enhancement of the lesion.
Conclusion

Intramedullary spinal cord neoplasms are rare.

The most common intramedullary spinal tumors are ependymoma (the most common in adults), astrocytoma (the most common in children) and hemangioblastoma. The majority of tumors of the filum terminale and conus medullaris are myxopapillary ependymomas.

MR imaging is the technique of choice for the diagnosis and evaluation of spinal cord tumors. Spinal cord expansion, tumoral enhancement and associated cysts are the three main MR findings that allow differentiating spinal cord tumors from other intramedullary nonneoplastic lesions. Different types of tumors have characteristic imaging findings; awareness of these findings allows the radiologist to make a correct diagnosis.

Ependymomas usually manifest as centrally located tumors with well-defined margins and focal, intense and homogeneous enhancement; hemorrhage is a common finding.

Astrocytomas usually present as eccentrically located tumors with poorly defined margins and patchy and irregular enhancement; hemorrhage is not a common finding.

Hemangioblastomas typically manifest as solid, well-defined nodules at the posterior surface of the spinal cord that show intense and homogeneous enhancement and an associated large syrinx. They show flow voids in or around the tumor.

There are other less frequent types of intramedullary spinal cord tumors and nonneoplastic spinal cord lesions such as demyelinating, inflammatory, infectious and vascular diseases that occasionally can pose diagnostic problems and must be taken into account.

Personal Information

References


