Introduction

Spine neoplasms are classified based on their location as either extradural, intradural extramedullary (IDEM), or intramedullary. This review will focus on intradural neoplasms including both extra- and intramedullary. While neoplasms are relatively uncommon compared with other types of spine pathology, recognition of their imaging appearance and understanding the principles of spine tumor imaging is necessary as MRI is the primary tool for establishing a differential diagnosis, guiding the diagnostic workup, and formulating a treatment plan.

Spine Tumor Imaging

MRI is the imaging modality of choice for assessing the spine because of its high soft tissue contrast; therefore, its use will be emphasized in this review. The first step in forming a pertinent differential diagnosis is assessing the location of the abnormality. Intramedullary neoplasms are characterized by abnormal cord signal and expansion. IDEM tumors are contained within the thecal sac but displace rather than expand the spinal cord. A CSF cleft may be seen separating the tumor from the cord. Use of gadolinium based contrast agents should be standard in suspected spinal tumors. Enhancement is the rule, and unlike most intracranial neoplasms, even low-grade spinal tumors characteristically enhance. Syringohydromyelia and cyst formation is common. Two types of cysts exist: tumoral cysts and rostral/caudal cysts. The former are lined with neoplastic cells and typically demonstrate rim-enhancement; the latter are reactive and do not contain neoplastic cells or enhance. Distinction is important as the former need to be surgically resected while the latter can simply be aspirated.

Intramedullary neoplasms share many common imaging features. Abnormal cord signal intensity and expansion, whether from tumor itself or from cord edema, commonly spans several spinal levels, and may involve the entire spinal cord. Contrast enhancement is the rule, and unlike most intracranial neoplasms, even low-grade spinal tumors characteristically enhance. Syringohydromyelia and cyst formation is common. Two types of cysts exist: tumoral cysts and rostral/caudal cysts. The former are lined with neoplastic cells and typically demonstrate rim-enhancement; the latter are reactive and do not contain neoplastic cells or enhance. Distinction is important as the former need to be surgically resected while the latter can simply be aspirated.

The majority of intramedullary neoplasms are low grade and slow-growing. Therefore, patients typically have a prolonged clinical course extending over many years prior to diagnosis. Common symptoms include back pain and sensory disturbances. In general, treatment consists of surgical resection, with or without adjuvant radiation and chemotherapy.
Ependymoma.

Ependymomas are the most common intramedullary neoplasm in adults and the second most common in children. Mean age of presentation is in the 4th to 5th decades of life, and there is no gender predilection. They arise from ependymal cells lining the central canal, and are therefore typically centrally located. The cervical cord is the most common location. While they share many of the histologic characteristics of intracranial ependymomas, they are considered genetically different and carry a better prognosis. Gross total resection results in extremely low recurrence rates.

Five histologic subtypes of ependymoma exist, including cellular, clear cell, papillary, tanycytic, and myxopapillary (discussed in the IDEM section). The cellular subtype is most common and is typically WHO grade II. Sensory symptoms predominate in the clinical presentation, possibly from involvement of the central crossing spinothalamic tracts. Ependymomas can be seen as part of neurofibromatosis (NF) type 2 (“MISME” syndrome — multiple inherited schwannomas, meningiomas, and ependymomas), though the majority are sporadic.

Ependymomas are characteristically well-circumscribed masses in the central portion of the cord. They generally demonstrate T1 iso- to hypointensity and T2 hyperintensity relative to the spinal cord. Cyst formation is common. Due to their highly vascular nature, profound low T2 signal at the cranial or caudal margins, known as the “hemosiderin cap”, is a distinguishing feature (Fig. 1). While the classic enhancement pattern is homogeneous and intense, this was seen in only 38% of cases according to one series, with heterogeneous, rim, and minimal enhancement patterns collectively making up the remaining majority.

Astrocytoma.

Intramedullary astrocytomas arise from astrocytic cells within the cord and represent the most common cord neoplasm in children and the second most common in adults. There is a slight male predominance. The majority are low-grade WHO I and II, pilocytic and fibrillary variants, respectively. WHO grade III and IV tumors, anaplastic and glioblastoma multiforme (GBM) subtypes, together

Figure 1. Ependymoma. 34 year-old patient with a centrally located mass enlarging the cervical cord. Sagittal T1W (A) and T2W (C) images demonstrate a heterogeneous, solid intramedullary mass with rostral and caudal cyst formation and cord edema. Low T2 signal along the caudal margin is the “hemosiderin cap” (arrow). Diffuse heterogeneous enhancement is seen on post-gadolinium (B) sagittal T1-weighted sequences.

Figure 2. Astrocytoma. Cervical spinal cord intramedullary mass in this child causes fusiform enlargement of the cord spanning several vertebral levels. Pre- (A) and post-gadolinium (B) sagittal and axial (C and D) T1W images demonstrate heterogeneous, irregular ring-enhancement with eccentric involvement of the cord and poorly defined rostral margins. Pathology revealed a WHO grade III anaplastic astrocytoma.
form only 10-15%.\textsuperscript{7} GBM’s are rare, representing a minority of the higher grade cord astrocytomas. Expectedly, high-grade tumors have a higher rate of recurrence and metastasis and carry a worse prognosis with average survival on the order of months.\textsuperscript{8} All subtypes demonstrate infiltrative growth along non-neoplastic scaffolding of the cord. Cyst formation is frequently seen. Cervical and thoracic cord involvement predominates.\textsuperscript{1}

Many imaging features suggest a diagnosis of astrocytoma over ependymoma. As opposed to ependymomas, astrocytomas are more characteristically eccentrically located and are less well-defined secondary to their infiltrative growth pattern (Fig. 2). MRI signal characteristics are similar, with T1 iso- to hypointensity and T2 hyperintensity compared to cord, though hemorrhage is less common. Partial mild-moderate intensity heterogeneous enhancement is characteristic, though no enhancement was seen in 20-30% in one series.\textsuperscript{9} Despite the lack of enhancement, neoplasm should still be suggested based on characteristic cord expansion and signal abnormality.

Hemangioblastoma.

Hemangioblastomas are far less common than ependymomas and astrocytomas, accounting for 3.3% of spinal cord tumors.\textsuperscript{10} They represent a low-grade (WHO grade I) vascular neoplasm, though their cell of origin is yet to be delineated. Association with von Hippel-Lindau (VHL) syndrome ranges from 17-38% in the literature,\textsuperscript{11,12} in conjunction with cerebellar hemangioblastomas, retinal angiomas, and endolymphatic sac tumors in the CNS and head and neck. There is a slight male predominance, and the mean age of diagnosis is in the 4\textsuperscript{th} decade of life, though they tend to present about a decade earlier when associated with VHL.\textsuperscript{13}

MR imaging characteristics include a well-delineated, uniformly T2 hyperintense mass in smaller lesions. Location is subpial, typically along the posterior aspect of the cord. Cyst and syrinx formation is characteristic, and the cyst-mural nodule appearance classically seen in cerebellar hemangioblastomas may be seen with larger lesions (Fig. 3). Due to the highly vascular nature of this entity, enlarged feeding vessels and flow voids in larger masses may be seen. Uniform and intense enhancement is characteristic of smaller masses.

Metastases.

Spinal cord metastases are uncommon, seen in only 0.9-2.1% of autopsied cancer patients in one series.\textsuperscript{14} They represent approximately 3% of metastases to the spine.\textsuperscript{15} The most common primary tumors are lung\textsuperscript{15} and breast and arise either through hematogenous dissemination or CSF spread through the central canal. The cervical cord is the most common site of involvement.\textsuperscript{16} Intratumoral hemorrhage may be seen with certain types of primary neoplasm. While they may be solitary, multiple lesions or multifocal involvement, including the osseous spine, may be key
to distinguishing the diagnosis. Unlike primary spinal cord neoplasms, metastases tend to have a more rapid onset and disease progression, on the order of weeks to months rather than years. Prognosis is uniformly poor with life expectancy on the order of months, as this represents an advanced stage of disease.

Imaging features vary based upon the primary tumor. Common features include a relatively small mass with extensive edema that is out of proportion for that expected based upon the size of the mass. Cord enlargement, if present, is not pronounced, and cysts are uncommon.\(^\text{16}\) Enhancement is common (Fig. 4). T1 hyperintensity can be seen with melanoma metastases and with hemorrhage.

Lymphoma.

Lymphoma has a variety of disease patterns involving the spine, including osseous, epidural, leptomeningeal, and spinal cord involvement. Intramedullary spinal involvement is the least common, accounting for only 3.3% of CNS lymphoma cases.\(^\text{17}\) As in other areas of the CNS, non-Hodgkin lymphoma predominates over Hodgkin disease, though some of the reported cases of spinal lymphoma are of T-cell lineage,\(^\text{18}\) an atypical feature of CNS lymphoma. Cord involvement favors the cervical spine.

The MRI appearance is nonspecific with respect to other intramedullary neoplasms, typically demonstrating T1 isointensity and T2 hyperintensity to the spinal cord. The T2 hyperintensity reported in cases within the literature\(^\text{18}\) is in distinction to the typical T2 hypointensity seen in primary lymphoma of the brain because of the high cellular content.\(^\text{19}\) Contrast enhancement is heterogeneous.

Ganglioglioma.

Ganglioglioma is uncommon, accounting for only 1.1% of spinal neoplasms.\(^\text{20}\) It is a low grade slow growing tumor composed of a mixture of neoplastic neuronal and glial elements. Children are more commonly affected with a classic clinical presentation of painful scoliosis. Cervicothoracic cord involvement predominates but holocord involvement is not uncommon.

Several MRI findings are suggestive of gangliolioma.\(^\text{20}\) Length of involvement tends to be larger with an average of 8 vertebral segments, and they are eccentrically located within the cord. Enhancement is patchy with more than half demonstrating extension to the pial surface. Tumoral cysts are common. Signal intensity on T1-weighted images is heterogeneous, and the amount of cord edema is relatively unimpressive based upon the size of the tumor.

Intradural Extramedullary Neoplasms

IDEM tumors are more common in adults. Schwannomas and meningiomas are the most common, together accounting for 45% of all primary spinal cord neoplasms.\(^\text{21}\) The differential diagnosis also includes neurofibroma, malignant nerve sheath tumor, lymphoma, myxopapillary ependymoma, paraganglioma, and metastatic disease.

Schwannoma.

Schwannomas arise from support cells of the nerve sheath and are the most common IDEM spinal neoplasm. Nerve sheath tumors account for 25% of all intradural spinal tumors in adults, with the majority being schwannomas.\(^\text{22}\) While they may occur in any spinal compartment, they are most commonly
Intradural Neoplasms, Carra et al.

Spinal schwannomas can be difficult to distinguish from meningiomas on imaging. They are typically well-circumscribed T2 hyperintense masses with signal intensity approaching that of fluid. Cyst formation is more common than with meningiomas. Their enhancement pattern is typically uniform (Fig. 6), though heterogeneous and ring-enhancement patterns do occur. Imaging features useful in distinguishing schwannoma from meningioma include foraminal enlargement and osseous remodeling, involvement of the cauda equina, fluid intensity T2 signal, and ring-enhancement.23

Meningioma.

Meningiomas are the second most common IDEM spinal neoplasm. They arise from arachnoid cap cells within the dura and are most commonly seen in females in the 5th-7th decades of life.22 The majority are solitary and sporadic; however, there are multiple syndromic associations, the most common of which is NF2 (see “MISME” in ependymoma section above). Thoracic cord involvement is typical and unlike schwannomas, cystic degeneration is uncommon. WHO grade I tumors are by far the most common, and total resection is generally curative.

Meningiomas are typically seen as well-circumscribed masses along the ventral or ventrolateral aspect of the cord (Fig. 7). MR signal intradural and extramedullary in location within the thoracic or lumbar spine.23 The classic “dumbbell” lesion extending through the neural foramen is considered both intra- and extradural, occurring in 10-15% of cases (Fig. 5).24 Most are solitary and sporadic. Syndromic associations include NF2, Carney complex, and schwannomatosis. Schwannomas are typically low grade (WHO grade I) tumors. Total resection of an intradural schwannoma is considered curative.

Figure 5. Dumbbell (intradural extramedullary and extradural) Schwannoma. Axial T2W image (A) shows a large dumbbell schwannoma exiting and widening the right neural foramen consistent with both an intradural and extradural component. CSF cleft (arrow) along the superior margin of the mass on the sagittal T2W image (B) confirms the extramedullary location.

Figure 6. Multiple Schwannomas in NF2. Multiple uniformly enhancing (A thru C) small extramedullary intradural masses are demonstrated along the pial surface of the distal spinal cord and the cauda equina. Multiplicity of the masses suggests the diagnosis of NF2.

Figure 7. Meningioma. Sagittal (A) and axial (B and C) post-gadolinium T1WI sequences reveal an IDEM mass abutting the ventrolateral aspect of the cervical cord which displaces the exiting right nerve root (B and C). There is a broad dural base and the presence of a dural tail (arrows) which suggests the diagnosis of meningioma.
characteristics include T1 iso- and T2 iso-to hyperintensity relative to the spinal cord. A broad dural attachment, or “dural tail”, was seen in 58% of meningiomas in one series, best demonstrated on post-contrast imaging.\textsuperscript{23} In this same series, calcification, which is easily identified on CT, was also identified in 58%. Post-contrast imaging typically reveals diffuse and prominent enhancement. A solitary IDEM thoracic spine mass in a middle-age female should suggest the diagnosis.

Neurofibroma.

Neurofibromas are much less common than schwannomas within the spine. They are most commonly seen in the 3\textsuperscript{rd}-4\textsuperscript{th} decades of life and have no gender predilection.\textsuperscript{1} The classic association of NF1 with plexiform neurofibroma is virtually pathognomonic. However, the majority of lesions are sporadic. They can be intradural and/or extradural and can be seen at any spinal level. Neurofibromas are WHO grade I neoplasms. While total resection is considered curative, tumors with extensive paraspinal involvement and subtotal resection have a propensity to recur and require follow up imaging.

It is often difficult if not impossible to distinguish solitary spinal neurofibromas from schwannomas based on imaging alone. Peripheral T2 hyperintensity, the “target sign,” is more suggestive of neurofibroma but not pathognomonic (\textbf{Fig. 8}). Cystic degeneration and hemorrhage are also less common than schwannoma. Involvement of a ventral nerve root is more suggestive of neurofibroma.\textsuperscript{1} Extradural extension can be prominent as a plexiform or infiltrative soft tissue mass.

\textbf{Malignant Peripheral Nerve Sheath Tumor.}

Malignant peripheral nerve sheath tumor (MPNST) represents a soft tissue sarcoma with histologic features of peripheral nerves either occurring de novo or arising from malignant degeneration of a neurofibroma. They are WHO grade II-IV tumors. Roughly half are associated with NF1 while the remainder are sporadic.\textsuperscript{1} They most commonly involve the paravertebral soft tissues with rare intraspinal involvement. Masses are typically markedly heterogeneous and infiltrating (\textbf{Fig. 9}). Prognosis is poor.

\textbf{Myxopapillary Ependymoma.}

The myxopapillary variant represents roughly 30% of all intraspinal ependymomas.\textsuperscript{25} It can be distinguished based on its characteristic location within the conus medullaris, filum terminale, or cauda equina where it is the most common tumor. Peak incidence is in the 3\textsuperscript{rd}-5\textsuperscript{th} decades of life, and there is a slight male predominance.\textsuperscript{1} Despite its WHO I grading, it may cause subarachnoid seeding. Hemorrhage, calcification, and cyst formation are common. The tumor can become intimately involved with the nerve roots of the cauda equina making total resection difficult. When accomplished, recurrence rates are low.
Figure 11. **CSF Dissemination of Mantle Cell Lymphoma.** 64 year-old woman with known lymphoma presented with cauda equina syndrome. Imaging shows abnormal low T2 (A) and intermediate T1 (B) signal that fills the lumbar spinal canal and encases the conus medullaris and cauda equina. Post-gadolinium T1W images (C and D) best depict the mass which is avidly enhancing.

Figure 12. **Paraganglioma.** Markedly T2 hyperintense (A), circumscribed mass along the dorsal aspect of the distal spinal cord shows avid enhancement on post-gadolinium T1W images (B and C).

Myxopapillary ependymomas are sausage-shaped masses that can occupy the entire lumbosacral canal (Fig. 10). Unlike other histologic variants of spinal ependymoma, the myxopapillary type may have intrinsic T1 hyperintensity secondary to its high mucin content, seen in 67% of cases in one series. Other imaging features, such as the hemosiderin cap and intralesional flow voids, are similar to other ependymoma variants.

**Lymphoma.**

As discussed above, spine lymphoma has a variety of different disease patterns, which include leptomeningeal involvement. The majority occur as spread from intracranial lymphoma. Patient demographics are similar to that of other types of lymphoma, most commonly occurring in middle age adults. Imaging hallmarks include thickened nerve roots and smooth or nodular leptomeningeal enhancement; IDEM mass formation can occur in extreme cases (Fig. 11).

**Paraganglioma.**

Paragangliomas are neural crest tumors of which the intra-adrenal form, pheochromocytoma, is the most common. Extra-adrenal paragangliomas most commonly occur in the head and neck near the carotid bulb, jugular foramen, and within the temporal bone. When they occur in the spine, they are...
characteristically located within the lumbar spine, involving the cauda equina or filum terminale where they are almost always non-secreting.

Paragangliomas typically appear as a well-circumscribed mass within the cauda equina. They are T2 hyperintense and highly vascular (Fig. 12). Hemosiderin cap and flow voids may be seen in larger lesions. MR imaging features are nonspecific and may mimic myxopapillary ependymoma, schwannoma, and meningioma, which are much more common. The diagnosis can be suggested with avid uptake on nuclear medicine metaiodobenzylguanidine (MIBG) scan.

Metastasis.

CSF seeding with malignant cells may occur through a hematogenous route or may represent spread from a CNS primary. The most common primary malignancies outside of the CNS include lung, breast, and melanoma.26 CNS primary tumors with a tendency for CSF dissemination include high-grade astrocytomas, ependymomas, germ cell tumors, medulloblastomas, and choroid plexus neoplasms.

Imaging findings mirror patterns of disease involvement, which include solitary masses (Fig. 13), diffuse smooth or nodular coating of the cord and nerve roots (Fig. 14), and thickening of the cauda equina. Use of intravenous contrast increases the sensitivity for detection of metastatic lesions. MRI is an important adjunct to lumbar puncture with CSF analysis in diagnosing CSF metastasis; cytology alone has a significant false-negative rate and frequently must be repeated. It is important to remember, however, that chemical meningitis can cause enhancement and thickening of nerve roots, mimicking metastatic disease. Imaging should therefore precede lumbar puncture and craniotomy.

Summary

While neoplasms are a relatively uncommon form of spine pathology, recognition of their imaging features plays an essential role in providing a meaningful differential diagnosis and guiding further evaluation and treatment. While many of the imaging characteristics are nonspecific with significant overlap, when tumors are suspected, certain diagnoses can be suggested based upon location, clinical and patient demographics, and certain specific imaging features.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.
References


