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Radiographic Features of Intramedullary Spinal Cord Tumors

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The advent of MRI revolutionized the characterization of intramedullary spinal cord lesions. By providing detailed resolution of the spinal cord in multiple planes, MRI has become a key tool for the differentiation of operative tumors from nonoperative lesions, such as multiple sclerosis (MS) plaques, transverse myelitis, or cord infarction. Within the tumor subpopulation, MRI has allowed for detailed preoperative planning through understanding the precise limits of the tumor and surrounding edema, the presence of cysts, or evidence of preexisting hemorrhage.

Retrospective series demonstrate that approximately 90% of intramedullary spinal cord tumors are of glial origin [1]. Together, ependymomas and astrocytomas account for 95% of these lesions [1]. The balance between astrocytomas and ependymomas is age dependent. In the adult population, ependymomas account for 60% of all glial tumors, whereas astrocytomas account for 30% [2]. In the pediatric population, the situation is reversed, with astrocytomas accounting for 90% of intramedullary tumors in children less than 10 years of age [3]. The remaining glial lesions are composed of oligodendrogliomas and the more malignant anaplastic astrocytomas as well as glioblastoma multiforme. Nonglial tumors found within the spinal cord include hemangioblastomas, gangliogliomas, metastases, subependymomas, and an assortment of rare tumor types.

MRI reveals crucial information about intramedullary lesions. Evidence of cord expansion, contrast enhancement, or syrinx favors the diagnosis of tumor. Despite the improved resolution offered by MRI, current technology is still not sufficient to allow preoperative diagnosis and, in particular, the identification of astrocytoma versus ependymoma.

Imaging of the spinal cord

With the exception of MRI, most imaging modalities play a limited role in imaging the spinal cord. Radiographs fail to provide visualization of the cord, although scalloping of the posterior aspect of the vertebral body may occur in some cases of long-standing intramedullary lesions with substantive cord expansion. CT and/or CT myelography allows for visualization of the cord and identification of areas of gross enlargement but does not provide resolution of the internal structure. MRI allows for a detailed evaluation of the spinal cord, providing information on the substance of the cord, including enlargement, atrophy, syringomyelia, or edema as well as the nature of the tumor, including the presence of cysts and hemorrhage.

Baseline MRI of the spinal cord should include T1, T1 with gadolinium, and fast spin echo T2 sequences obtained in the axial and sagittal planes. Obtaining sequences in more than one plane is important in understanding the full extent of the lesion and differentiating subtle findings from artifact. T1-weighted sequences are ideal for identifying regions of cord enlargement or other anomalies in the contour of the cord and provide a baseline for comparison with the postgadolinium images. Most intramedullary spinal cord tumors are isointense or hypointense on T1

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images. T1-weighted sequences taken after administration of gadolinium demonstrate tumor enhancement in most cases. The postgadolinium images allow for the differentiation of tumor from cyst and, in many cases, demonstrate a clear tumor boundary. Fast spin echo T2-weighted sequences demonstrate tumor and surrounding edema as regions of hyperintensity. Cysts are also frequently hyperintense and may be difficult to differentiate from tumor. Hemosiderin appears hypointense on T2-weighted sequences and is particularly pronounced on gradient echo sequences.

Certain characteristics are common to spinal cord tumors; most enhance with contrast despite being low-grade lesions [4], and the higher it is located along the spinal axis, the more frequently is a tumor associated with a syrinx [5]. Cysts associated with tumors may vary in T2 intensity based on the level of protein in the cyst content. High-protein cysts may be indistinguishable from tumor based on T2 sequences alone. Cysts may be reactive in nature or directly associated with tumor. Reactive cysts are found at the poles and do not enhance with contrast. Tumor-associated cysts are located in the body of the tumor and may demonstrate rim enhancement.

A key advantage of MRI is the ability to gain substantial information as to whether a lesion represents a tumor or another type of spinal cord lesion. MS, transverse myelitis, and cord infarction represent the primary nonoperative lesions that must be differentiated from tumor. All three lesions are focal, without significant mass effect in most cases [6]. In rare instances, acute lesions may be associated with cord enlargement and surrounding edema. When doubt remains after initial imaging, the brain may be imaged to look for further evidence of active MS or follow-up imaging may be obtained in 4 to 6 weeks. Interval imaging should reveal decreased signal and size in the case of transverse myelitis and regression of surrounding edema in the instance of spinal cord infarction.

Ependymoma

Together, ependymomas and astrocytomas compose 90% of intramedullary spinal cord tumors [1]. Ependymomas are predominantly a tumor of adulthood, becoming the most frequent tumor type after the age of 30 years, a predominance that persists in the elderly [7,8]. Clinically, ependymomas are discrete lesions that have well-defined borders with the surrounding spinal tissue. This clearly delineated margin frequently allows for gross total resection. In some instances, plain films may reveal scoliosis and bony changes associated with a chronically widened canal. Myelography may reveal a complete or partial block at the affected level [2].

MRI typically reveals enlargement of the cord centered around the lesion on T1-weighted images. Approximately 65% of lesions are associated with syringomyelia, because most arise in the cervicothoracic region [5]. Rostrally and caudally located cysts are present with great frequency, although they are not specific. Most lesions span multiple vertebral segments [9]. Intramedullary ependymomas are almost always centrally located [10]. This central location results from the believed origin of ependymomas from the ependymal cells that line the central canal [11]. Ependymomas are frequently associated with hemorrhage, revealed as hemosiderin on T2 and gradient-recalled echo (GRE) sequences (Fig. 1) [9]. In a series of 28 cases of intramedullary ependymomas scanned with 1.5-T MRI, 26 lesions were noted to be cervical and 2 were thoracic. All were T2 bright, 17 presented with rostral and/or caudal cysts, 6 presented with intratumoral cysts, and 27 of 28 were central in location. All demonstrated some enhancement, with 13 in a heterogeneous pattern, 10 in a homogeneous pattern, 3 with a heterogeneous pattern and cyst wall enhancement, and an enhancing nodule in a cyst wall in 2 [12]. A separate look at the enhancing characteristics in another study revealed enhancement in 23 of 23 lesions, of which 15 were heterogeneous and 8 homogeneous [10]. Twenty of 23 lesions revealed sharply defined borders [10]. This pattern of T2 brightness, central location, various patterns of contrast enhancement, and cyst association is supported by other series [10].

Astrocytoma

Intramedullary astrocytomas are the dominant tumor type in children and make up one third of intramedullary glial tumors in adults [13,14]. In patients younger than 10 years of age, they represent 90% of tumors, and they represent 60% of tumors in adolescents [7]. Astrocytomas lose their predominance in adulthood, with ependymomas becoming more frequent after the age of 30 years [7]. Intramedullary astrocytomas have

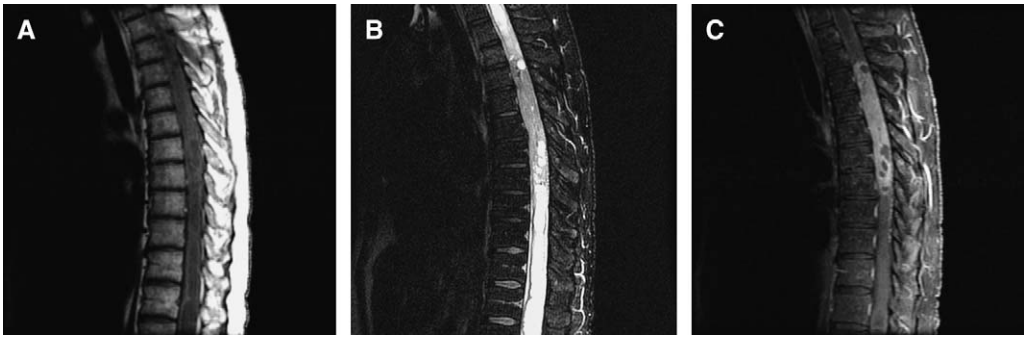

Fig. 1. Ependymoma. Precontrast sagittal T1-weighted (*A*), sagittal T2-weighted (*B*), and post contrast sagittal T1-weighted (*C*) images of the thoracic spine demonstrate a large, expansile, heterogeneous intramedullary mass involving multiple vertebral body levels. (*C*) The mass shows avid enhancement after the intravenous administration of gadolinium contrast agent, with a few areas of cystic degeneration.

a documented association with neurofibromatosis type I, and several series have demonstrated a male predilection [15–17].

Several imaging characteristics may suggest the presence of an astrocytoma over an ependymoma. In most cases, astrocytomas are eccentric in location and, unlike ependymomas, are frequently infiltrative, presenting with ill-defined borders. Most lesions present with a cervical location; a series of 17 astrocytomas revealed 11 cervical, 2 cervicothoracic, and 4 thoracic lesions [18]. Rostral and/or caudal cysts are less frequently associated with astrocytomas than with ependymomas, although this is not significant enough to assist with diagnosis [15]. Consistent with most intramedullary spinal cord tumors, astrocytomas are hypointense to isointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 2). A series of 11 astrocytomas demonstrated cord enlargement in all 11, all were bright on T2-weighted images, 36% presented with peritumoral cysts, 1 demonstrated hemorrhage, and 8 (73%) of 11 enhanced [19]. Astrocytoma borders are less distinct than ependymomas, reflecting their invasive nature. Correspondingly, T1 postgadolinium sequences reveal less well-defined patchy borders [13].

Most intramedullary astrocytomas are low-grade lesions, although World Health Organization (WHO) grade III and IV lesions occur in approximately 10% to 15% of cases [8,13]. Given the enhancement of most intramedullary astrocytomas, unlike the case with intracranial lesions, tumor grade is not evident on imaging. Less than 2% are pathologically diagnosed as oligodendroglioma [1].

Hemangioblastoma

The next most common intramedullary tumor after ependymoma and astrocytoma is hemangioblastoma. Hemangioblastomas compose 1.6% to 6.8% of all intramedullary tumors [20–23]. Approximately 60% to 75% of spinal hemangioblastomas are purely intramedullary, with another 10% to 15% having intramedullary and extramedullary components [24]. Twenty percent to 30% of intramedullary hemangioblastomas are associated with von Hippel-Lindau disease (VHL), an autosomal dominant genetic syndrome featuring central nervous system (CNS) hemangioblastomas, retinal angiomas, renal and pancreatic cysts, pheochromocytomas, and renal cell cancer [20,25]. The age of onset of symptoms is most commonly the fourth decade of life for sporadic lesions and a decade earlier for lesions associated with VHL. A male predominance is typical, accounting for 78% of tumors in a recent series [21,22,25]. Most hemangioblastomas are found in the cervical region (56%), followed by 40% in the thoracic cord and 4% in the conus [22].

As with all intramedullary tumors, MRI remains the imaging modality of choice. The tumor is isointense on T1-weighted images. T1-weighted images with gadolinium reveal a homogeneously enhancing tumor nodule. Rostral and caudal cysts are present in a large number of cases, and high protein content may make these indistinguishable from tumor or surrounding edema. T2-weighted images reveal extensive surrounding edema and associated syrinx in 83% of patients [22]. Flow voids are frequently noted and are distinctive, although they may be present in other

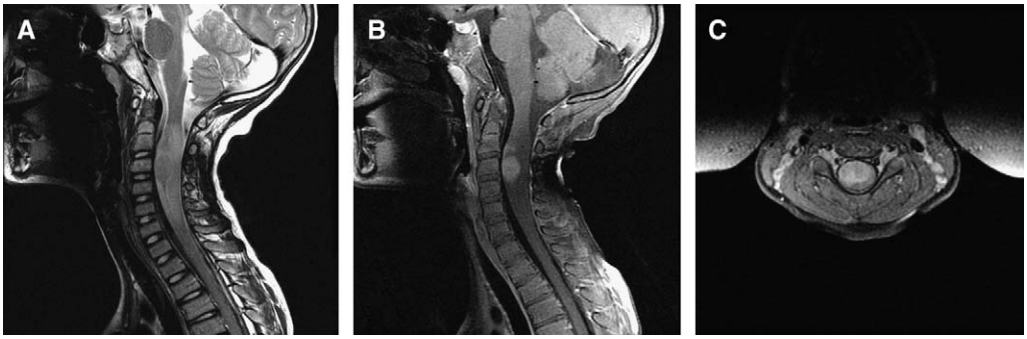

Fig. 2. Astrocytoma. Sagittal T2-weighted (*A*), post contrast sagittal T2-weighted (*B*), and post contrast saggital T1-weighted images (*C*) images of the cervical spine demonstrate a ventral intramedullary mass centered at the C3 vertebral body level. There is a moderate amount of tumor-related edema, which extends superiorly to the level of the medulla and inferiorly to the C6 level as seen on sagittal T2-weighted images. The mass demonstrates homogeneous enhancement.

intramedullary tumors [22,26,27]. Because of their comparative rarity, hemangioblastomas may be difficult to differentiate from ependymomas in some instances, given that both may exhibit a pattern of nodular enhancement. The lack of hemosiderin and the presence of flow voids in hemangioblastomas may provide clues to the diagnosis. Angiography reveals a distinctive vascular pattern with intense staining and dilated feeding arteries and draining vessels [28].

Preoperative angiography may facilitate the resection of intramedullary hemangioblastomas. The posterior and lateral spinal arteries are the most common feeding vessels, with anterior spinal artery involvement only when the tumor is in an eccentric ventral position in the cord. The usefulness of preoperative embolization is a matter of debate. Case series of successful superselective embolization have been reported throughout the spinal axis; however, experienced surgical centers report good outcomes without the addition risk entailed by embolization [28,29].

Ganglioglioma

Intramedullary gangliogliomas are extremely rare entities, with a reported occurrence of approximately 1% of all spinal cord tumors (Fig. 3) [30–33]. They occur predominantly in children and young adults, with a mean age of 12 years [34]. In a review of 27 reported cases by Patel and colleagues [34], gangliogliomas were found to extend over more vertebral segments than astrocytomas or ependymomas (8 to 4 segments). Forty-eight percent of lesions were localized to the cervical cord, 22% to the thoracic cord, 15% to the conus and/or filum, and holocord involvement was found in 15%. Tumors were

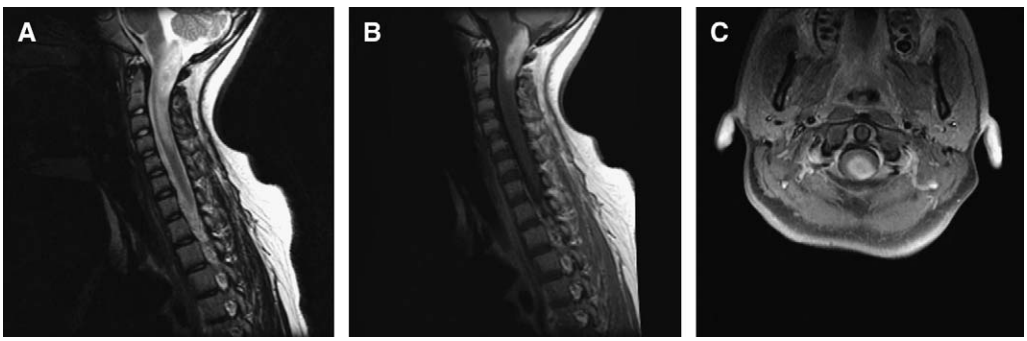

Fig. 3. Ganglioglioma. Sagittal T2-weighted (*A*), post contrast sagittal T2-weighted (*B*), and post contrast saggital T1-weighted images (*C*) images of the cervical spine demonstrate a dorsal intramedullary mass centered at the level of the cervicomedullary junction. The mass enhances avidly but heterogeneously. There is tumor-related edema that extends inferiorly to the C4 level as seen on T2-weighted imaging.

eccentrically located in 100% of cases. Tumoral and polar reactive cysts were common. Forty-six percent of intramedullary gangliogliomas demonstrated tumoral cysts, a result the authors found to be statistically significant when compared with ependymomas and astrocytomas [34]. T1-weighted images of gangliogliomas revealed a mixed pattern of hyperintensity and hypointensity in 82% of cases. This pattern of mixed signal intensity is distinct from the pattern noted for astrocytomas or ependymomas. T2-weighted images revealed hyperintense signal in all cases; however, gangliogliomas were significantly distinct in the absence of peritumoral edema. Contrast-enhanced T1-weighted images revealed patchy enhancement in 65% of cases and none in 15%.

Subependymoma

Intramedullary subependymomas are extremely rare entities, with approximately 40 cases reported in the literature [35]. A review of reported cases revealed a mean age of diagnosis in the fifth decade, a 2:1 male/female preference, and fairly equal distribution along the spinal axis. MRI findings revealed hypointensity on T1-weighted images, hyperintensity on T2-weighted images, and contrast enhancement in a homogeneous or nodular pattern in 55% of cases [35,36]. Because intramedullary subependymomas lack imaging features distinctive from the more common glial neoplasms, diagnosis is based on pathologic findings.

Metastasis

Intramedullary spinal cord metastases are relatively rare, with autopsy series in patients with cancer demonstrating a frequency of intramedullary metastasis of 0.9% to 2.1%, representing 8.5% of all CNS metastases [37,38]. In a review of 138 patients with intramedullary spinal cord metastases, lung cancer accounted for 54%, with breast cancer (9%) and melanoma (8%) a distant second and third, respectively [38]. The advent of MRI has been a key tool in the timely diagnosis of intramedullary metastases, allowing intervention early in the progression of neurologic deficit. Before MRI, myelography was the only imaging modality available. When positive, myelography revealed a fusiform enlargement of the cord. Reported series demonstrate that approximately 40% of myelograms may be falsely negative, however [37–40].

MRI reveals a cervical predominance for intramedullary metastases, with 42% originating in the cervical spine, 32% in the lumbar spine, and 26% in the thoracic spine [38]. Cord enlargement is found in roughly half of intramedullary metastases. MRI of 18 patients with a total of 26 intramedullary metastases demonstrated no evidence of spinal cord deformation in 54% [41].

In the series by Crasto and coworkers [41], 24 of 26 lesions were isointense on T1-weighted images, with the caveat that this may have been secondary to volume averaging. Ninety-four percent enhanced with the administration of gadolinium. Two patterns of contrast enhancement were noted: a homogeneous enhancement of the entire nodule in 21 (81%) of 26 cases and a ring-like enhancing pattern in 5 (19%) of 26 cases, likely corresponding to a region of central necrosis. On T2-weighted images, all lesions were hyperintense. The regions of hyperintensity did not correspond to the regions of enhancement, indicating the presence of surrounding edema. Of note, 33% had multiple lesions, highlighting the need to image the entire cord.

Uncommon intramedullary tumors

Numerous other reports of an intramedullary presentation of a variety of tumors exist. Approximately 40 cases of intramedullary schwannoma have been reported, with most being localized to the cervical spine [1]. Rare cases of primary intramedullary germinoma, lymphoma, primitive neuroectodermal tumor, meningioma, and melanoma have also been reported [1,14]. In addition, reported nonneoplastic lesions include epidermoid and/or dermoid cysts, lipomas, cavernous malformations, abscesses, and arteriovenous malformations [14,42].

Summary

MRI allows the detailed resolution of intramedullary spinal lesions, serving as a valuable tool for neurosurgeons contemplating surgical intervention. MRI provides significant guidance as to whether a lesion is surgical or nonsurgical in nature and, for surgical lesions, crucial information for preoperative planning. Based on multiplanar imaging, information regarding the nature and presence of cysts and a central or eccentric location of the lesion allows optimal localization of the myelotomy. Despite its utility in preoperative planning, MRI does not allow the

neuroradiologist to distinguish astrocytoma from ependymoma or other rare tumor types. Pending advances in technology, final diagnosis is still dependent on an open biopsy.

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