



Neoplastic and Paraneoplastic Involvement of the Spinal Cord

John Michael Hazenfield, MD, and Mary F. Gaskill-Shipley, MD

Neoplasia of the spinal cord, including both primary and metastatic tumors, is relatively rare, representing 4%-10% of all central nervous system tumors, and can present a diagnostic challenge to the radiologist. More than 90% of primary spinal cord neoplasms are derived from the glial cell lineage, including the 2 most common tumors ependymoma and astrocytoma. However, less common spinal cord tumors, including metastatic disease, as well as nonneoplastic and paraneoplastic processes should be considered in the diagnosis of intramedullary spinal cord lesions.

Semin Ultrasound CT MRI 37:482-497 © 2016 Elsevier Inc. All rights reserved.

Introduction

The imaging approach to spinal neoplasms, like other central nervous system (CNS) lesions, can be initially organized using a location-based system to derive a reasonable and refined differential diagnosis. Classifying lesions as extradural, intradural extramedullary, and intramedullary in location is the first step in forming a diagnosis.¹ One caveat to this classification is the conus medullaris region and its fibrous continuation, the filum terminale. Distinction between the 2 intradural categories in this region is often not possible; therefore, it may be the best to consider this area and the tumors that arise here as a separate category.

Intramedullary spinal cord tumors are rare and represent 4%-10% of all CNS tumors.²⁻⁴ The vast majority of primary intramedullary neoplasms arise from glial cell lines, the 2 most common of which are astrocytoma and ependymoma. Some imaging features favor one tumor type over the other, but much overlap exists and distinction between these tumors may not always be possible by imaging alone.⁵ Mixed glial and nonglial neoplasms including gangliogliomas, hemangioblastomas, and paragangliomas are much less common, but characteristic imaging features have been described for these entities. Metastatic disease and lymphoma can present with either an intraparenchymal or pia-arachnoid pattern of involvement. There are a few tumors, which are unique to the conus

medullaris or filum terminale region, such as myxopapillary ependymomas and paragangliomas. Not specific to this region, but nevertheless considered in the differential diagnosis, are nerve sheath tumors and pia-arachnoid metastatic disease.

Neoplasia can also indirectly affect the spinal cord.⁶ Spinal cord abnormalities not caused by extrinsic compression, metabolic or nutritional deficits, coagulopathy, or side effects of cancer treatment may be related to paraneoplastic involvement, a rare process with recently characterized imaging features.

The current review focuses on the common intramedullary neoplastic tumors, tumors of the conus medullaris or filum terminale region and a few tumor mimics. We also review the recently described imaging findings of the relatively rare paraneoplastic spinal cord syndrome.

Common Intramedullary Spinal Cord Tumors

Gliomas

In adults, gliomas account for 90% or more of all primary spinal cord tumors.^{7,8} However, in terms of all CNS glial tumors, spinal cord neoplasms are very rare and make up only 2%-4% of tumors.² Ependymoma is the most common primary intramedullary spinal cord tumor in adults and is 2 times more prevalent than the second most common tumor, astrocytoma.^{9,10} In children the incidence is reversed, with astrocytomas representing the most common pediatric spinal cord neoplasm.^{3,9,11,12}

Ependymomas are tumors arising from the ependymal cell lining of the central canal and can pathologically be subdivided

Division of Neuroradiology, University of Cincinnati Medical Center, Cincinnati, OH.

Address reprint requests to J. Michael Hazenfield, MD, Division of Neuroradiology, University of Cincinnati Medical Center, 234 Goodman St, Cincinnati, OH 45267. E-mail: Hazenfjm@ucmail.uc.edu

into 4 types, with the cellular subtype being the most common.^{5,11} The myxopapillary subtype uniquely and almost exclusively arises in the conus medullaris or filum terminale region.⁵ Most ependymomas are sporadically occurring World Health Organization (WHO) grade 2 tumors, with a typical age range of 35-45 years at presentation. Ependymomas may also occur in Neurofibromatosis (NF) type 2 patients. NF2 is characterized by Multiple Inherited Schwannomas, Meningiomas, and Ependymomas (MISME).¹³ Anaplastic ependymomas are uncommon WHO grade 3 tumors that may not be differentiated by imaging.

Astrocytomas are tumors arising from astrocytes, which make up the support structure of the CNS. A total of 80%-90% of all spinal cord astrocytomas are low grade and include both fibrillary astrocytomas (WHO grade 2) and pilocytic astrocytomas (WHO grade 1).¹¹ Most of higher grade tumors consist of anaplastic astrocytomas (WHO grade III), whereas glioblastomas (WHO grade IV) of the cord are uncommon. Unlike in the brain, most of the low grade lesions enhance.⁸ Most of spinal cord astrocytomas arise sporadically, although there is also an association with NF 1, particularly WHO grade 1 pilocytic astrocytomas.

As spinal cord gliomas make up the most of the spinal cord neoplasms, the radiologist would often be challenged to differentiate ependymoma from astrocytoma. Although both ependymomas and astrocytomas have radiographic features that are more common to each, there is considerable overlap in imaging findings and tumor type cannot always be readily differentiated by imaging alone.¹⁴

As ependymomas arise from the central lining of the canal, the mass tends to be located in the center of the cord and result in symmetric expansion (Fig. 1). This is in contrast to astrocytomas which tend to be more peripherally or eccentrically located. Astrocytomas also can have an exophytic growth

portion (Fig. 2), which is a helpful delineating characteristic.² Both ependymomas and astrocytomas are most common in the cervical cord followed by the thoracic region; however, astrocytomas are unlikely to involve the caudal aspects of the cord.^{2,15} Both tumors typically span a cord length of approximately 4 vertebral bodies.⁵ However, pilocytic astrocytoma may have more extensive holocord involvement.¹⁶ Both tumors are T2 signal hyperintense masses, but hemorrhage and necrosis are more common in ependymomas than astrocytomas (Fig. 3). A "cap sign" of hemosiderin staining has been reported in 50% of ependymomas.¹⁷

Cysts associated with primary glial neoplasms can be seen in 60% of all tumors.² These are divided into polar and tumoral cysts. Polar cysts are located at the rostral and caudal ends of the neoplasm and, while the exact mechanism is not known, they are postulated to represent focal dilatations of the central canal because of altered CSF flow or from mechanisms of fluid production and egress from the tumor into the subarachnoid space (Fig. 1). These are not considered neoplastic and should not have any associated enhancement.² Tumoral cysts on the contrary, usually are located within or near the enhancing portion of the lesion (Fig. 4). These are lined by neoplastic cells and should have peripheral enhancement. Most of ependymomas have polar cysts which may extend for greater lengths cranially and caudally through the spinal cord. Rostral cysts associated with cervical ependymomas can extend above the pyramidal tract decussation, elevating the floor of the fourth ventricle, thought to be a pathognomic sign.⁵

Differential enhancement patterns of ependymomas vs astrocytomas can be helpful. Tumor enhancement in ependymomas tends to be intense, uniform, and circumscribed (Fig. 1), whereas in astrocytomas, enhancement is more typically ill-defined, patchy, or heterogeneous (Fig. 2).¹⁸ These enhancement patterns reflect the histologic tendency



Figure 1 Cervical ependymoma, WHO grade II. (A-C) Sagittal T2 and pregadolinium and postgadolinium T1-weighted images demonstrate a homogeneously enhancing intramedullary mass within the upper cervical spinal cord. On the T2-weighted image the mass appears to surround the central canal (arrow). Syringobulbia is noted above the enhancing lesion. (D) Axial postgadolinium image confirms the central location of the mass.

Download English Version:

<https://daneshyari.com/en/article/2739080>

Download Persian Version:

<https://daneshyari.com/article/2739080>

[Daneshyari.com](https://daneshyari.com)