Spinal Cord Tumors

New Views and Future Directions

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KEYWORDS

• Intramedullary spinal tumors • Ependymoma • Hemangioblastoma • Astrocytoma • Ganglioglioma • Spine cysts • MRI • DTI

KEY POINTS

- About 90% of spinal intramedullary tumors are ependymomas or astrocytomas.
- Ependymomas commonly cause central cord syndrome because of their location.
- Myxopapillary ependymomas compose 90% of filum terminale tumors.
- Astrocytomas compose 90% of all primary spinal cord tumors in those less than 10 years of age and up to 60% of primary spinal cord tumors in adolescents.
- Astrocytomas typically present in an asymmetric eccentric location within the cord.
- Contrary to intracranial astrocytomas, low-grade spinal astrocytomas enhance commonly.
- Spinal cord hemangioblastomas are associated with Von Hippel-Lindau disease in 25% and typically show flow voids in the periphery of the tumor.

INTRODUCTION

Spinal cord tumors are uncommon neoplasms that, without treatment, can cause significant neurologic morbidity and mortality. The historic classification of spine tumors is based on the use of myelography with 3 main groups as schematically depicted in **Fig. 1**: (1) extramedullary extradural, (2) intradural extramedullary, and (3) intradural intramedullary. Using this scheme, spinal tumors are characterized as either arising inside the dura mater (intradural) or outside (extradural). This article focuses on *intramedullary spinal cord tumors*, with an emphasis on new diagnostic modalities[1] and treatment options. Because of the rarity of these lesions, the

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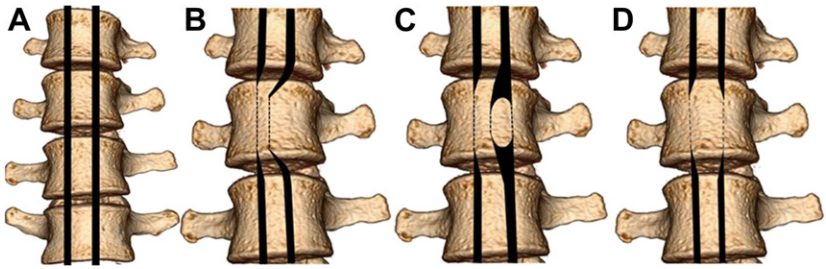

Fig. 1. Historic classification of spine tumors based on myelography. Myelographic spine tumor classification. (*A*) Normal, (*B*) extradural extramedullary, (*C*) intradural extramedullary, and (*D*) intradural intramedullary.

heterogeneous clinical presentation, and varied treatment strategies, it is not feasible to perform a prospective randomized study; a multicenter prospective case series or cohort study would require the standardization of treatment and outcomes.[2] Data from national registries and improved imaging capabilities have allowed spine tumor specialists the opportunity to study and treat these unusual and rare tumors with more confidence and better results.

EPIDEMIOLOGY OF INTRAMEDULLARY SPINE TUMORS

Intramedullary spinal tumors account for 5% to 10% of all spinal tumors in adults and approximately 35% in children. About 90% of the tumors of the spinal cord are glial tumors, of which most of these neoplasms are ependymomas and astrocytomas. Ependymomas represent about 60% and astrocytomas 30%.[3] Of the remaining 10%, hemangioblastomas account for 2% to 8%[4] and 2% are intramedullary metastases.[5] Intramedullary tumors are more common in children, with extramedullary tumors being more common in adults.

The rarity of these tumors has significant ramifications on potential outcomes because it impacts research allocation and treatment decisions. Most descriptive epidemiologic studies in primary spinal cord tumors include intradural extramedullary tumors, such as meningiomas, and nerve sheath tumors. Some recent studies have focused on the actual frequency of intramedullary tumors. Data from the central registries in the National Program of Cancer Registration and Surveillance, Epidemiology, and End results programs for 2004 to 2007 and 1999 to 2007 were analyzed.[1] This study provided the first comprehensive population-based incidence of primary spinal cord tumors, covering approximately 99.2% of the US population from 2004 to 2007.

Men had higher rates than women for ependymomas, lymphomas, and nerve sheath tumors but lower rates than women for meningiomas. The most common intradural spinal tumors histologically are meningiomas (33%), nerve sheath tumors (27%), and ependymomas (21%).[1] The overall incidence of spinal tumors, malignant and nonmalignant combined, was 0.97 per 100 000.[3] Seventy-eight percent of the primary spinal tumors were nonmalignant, accounting for most incident cases diagnosed between 2004 and 2007. As a result of mandating the collection of nonmalignant primary spinal tumors in conjunction with malignant primary spinal tumors in a 2004 population-based surveillance of cancers, the authors were able to capture the burden of the disease more completely in the US population.[1,2]

Tumors of the spinal cord are much less frequent than intracranial tumors. Overall prevalence is about 4 intracranial lesions for every 1 spinal tumor, which varies based