Minimally Invasive Approaches for the Treatment of Intramedullary Spinal Tumors

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KEYWORDS

• Minimally invasive spine surgery • Intramedullary tumors • Ependymoma • Astrocytoma
• Juvenile pilocytic astrocytoma • Hemangioblastoma

KEY POINTS

- Identify and differentiate between the various types of intramedullary tumors including astrocytomas, ependymomas, juvenile pilocytic astrocytomas (JPAs), and hemangioblastomas.
- Determine surgical treatment options for the different types of intramedullary tumors including ependymomas, astrocytomas, JPAs, and hemangioblastomas.
- Describe the surgical procedure for minimally invasive resection of an intramedullary spinal cord tumor as well as the potential advantages.

INTRODUCTION

Primary tumors of the spinal cord are 10 to 15 times less common than primary intracranial tumors, and overall represent 2% to 4% of all primary tumors of the central nervous system (CNS). There are an estimated 850 to 1700 new adult cases of primary spinal cord tumors diagnosed each year in the United States.[1] The histology of spinal cord tumors is similar to that of their intracranial counterparts; however, unlike primary intracranial tumors, spinal cord tumors show no association between increasing grade of malignancy and age at diagnosis. Most primary spinal cord tumors are classified as low grade (grades 1 and 2) according to the World Health Organization (WHO) pathologic classification.

Primary spinal cord tumors are divided into 3 categories based on anatomic location: intramedullary, intradural extramedullary, and extradural.[2] Intramedullary spinal cord tumors (IMSCT) constitute 8% to 10% of all primary spinal cord tumors, with the majority comprising gliomas (80%–90%), of which 60% to 70% are ependymomas and 30% to 40% are astrocytomas.[3] The third most common IMSCT is hemangioblastoma, representing approximately 3% to 8% of all IMSCTs, of which 15% to 25% are associated with von Hippel–Lindau (VHL) syndrome.[4–6]

The clinical presentation of primary spinal cord tumors is determined in part by the location of the tumor, and in nearly all clinical instances pain is the predominant presenting symptom. In a recent series of IMSCT, pain was the most common presenting symptom (72%), manifesting as back pain (27%), radicular pain (25%), or central pain (20%). Motor disturbance was the next most common presenting symptom (55%), followed by sensory loss (39%).[7] Diagnosis of a primary spinal cord tumor requires a high index of suspicion based on clinical signs and symptoms, in addition to spine-directed magnetic resonance imaging (MRI).

INTRAMEDULLARY SPINAL CORD TUMORS

Astrocytomas and ependymomas represent the most common intramedullary neoplasms. It is estimated that the intracranial to spinal ratio for astrocytomas and ependymomas are 10:1 and 3:1 to

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20:1, respectively (depending on the histologic variant).[8] The clinical presentation of an intramedullary tumor is variable, but pain and a mixed sensorimotor tract disturbance (segmental sensory level, upper motor neuron signs) are usually present.

MRI of the spine is the diagnostic modality of choice; however, patients unable to undergo MRI (ie, patients with a cardiac pacemaker) may require computed tomography (CT) myelography. An intramedullary tumor is radiographically recognized by focal, and sometimes holocord, spinal cord expansion with associated T2-weighted (T2W) and fluid-attenuated inversion recovery (FLAIR) image hyperintensity, T1-weighted (T1W) hypointensity or isointensity, variable contrast enhancement, and occasional tumor-associated syrinx.[9]

Ependymoma

Ependymomas are the most frequently encountered intramedullary spinal cord tumor in adults.[1,10] Histologically there are 2 distinct pathologic types: cellular (WHO grades 2 and 3) and myxopapillary (WHO grade 1). Cellular (classic) ependymoma arises from the intraspinal canal of the cervical and thoracic cord. Myxopapillary ependymomas arise from the filum terminale and occur almost exclusively at the conus medullaris. The treatment and prognosis for spinal cord ependymomas is often excellent, as these tumors may be resected completely and in such instances manifest a low risk of recurrence.[7,11,12]

On MRI ependymomas appear as a focal enlargement of the cord and hyperintense on T2W and FLAIR images, and hypointense or isointense to normal spinal cord on T1W images with heterogeneous contrast enhancement.[9] These tumors may also be associated with cystic changes, hemosiderin suggestive of previous hemorrhage, and syrinx (**Fig. 1**).

Ependymomas most often are of low grade with a benign indolent course, although malignant histologic subtypes (anaplastic ependymoma; WHO grade 3) rarely occur. Surgery is the most effective treatment, with complete surgical resection yielding reported local control rates of 90% to 100%, although gross total resection is not achieved in most patients.[12–14] Intraoperative monitoring of motor and somatosensory evoked potentials are often used to assist in achieving a more safe and complete resection.[15–17] Involved-field external beam radiotherapy at a dose of 45 to 54 Gy is indicated for partially resected or biopsied WHO grade 2 ependymomas or malignant WHO grade 3 tumors.[14,18,19] Overall, spinal cord ependymomas are associated with prolonged progression-free and overall survival, with a median of 82 months and 180 months, respectively.[20]

Astrocytoma

Approximately 40% of IMSCTs are astrocytomas.[10,21] The majority (75%) are low-grade (WHO grade 2) fibrillary astrocytomas with 5-year survivorship exceeding 70%.[7,10,22] Histology is the most important prognostic variable.[23–25] Juvenile pilocytic astrocytoma (JPA) is a low-grade (WHO grade 1) variant that more commonly presents in younger patients. High-grade spinal cord gliomas (WHO grades 3 and 4, 25%) are less common and associated with a poor survival. Regardless of WHO grade, spinal cord astrocytomas are

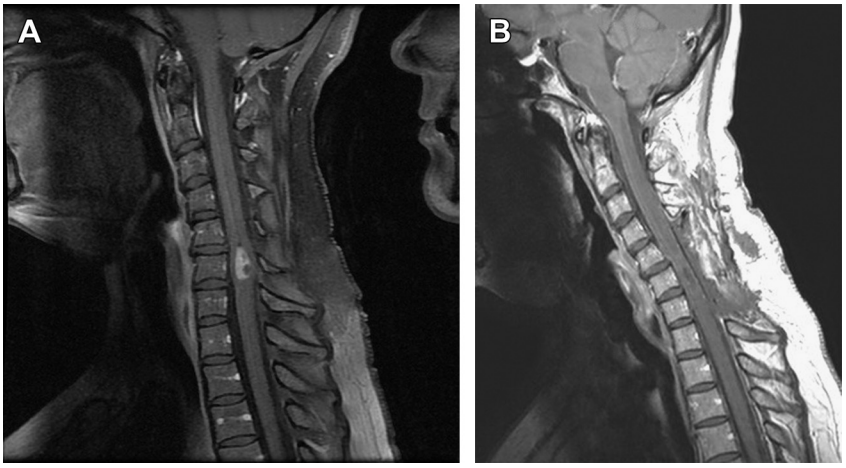

Fig. 1. (*A*) Preoperative sagittal T1-weighted magnetic resonance (MR) image with gadolinium, showing characteristics of an intramedullary ependymoma. (*B*) Postoperative sagittal T1-weighted MR image with gadolinium, showing complete resection of ependymoma with associated postoperative changes.