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Intramedullary Spinal Cord Tumors in Pediatric Patients

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Section snippets

Key points

- Pediatric intramedullary spinal cord tumors are rare but clinically important lesions, with astrocytomas and ependymomas predominating. …
- Children often present with months of progressive, nonspecific symptoms—pain, motor deficits, gait disturbance, or scoliosis—leading to diagnostic delays. …
- MRI is essential for diagnosis and surgical planning, highlighting differences between infiltrative astrocytomas and more well-defined ependymomas. …
- Maximal safe resection remains the cornerstone of treatment; …

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Clinical presentation

The clinical presentation of pediatric intramedullary spinal cord tumors (IMSCTs) is highly variable and often insidious. Age influences both the pattern and the recognition of symptoms, contributing to diagnostic challenges and frequent delays, particularly in very young children.

A thorough history typically reveals a prolonged progression of symptoms that may precede diagnosis by months.[3,5] Reported intervals from symptom onset to surgery range from weeks to several years, with mean durations …

Epidemiology

Astrocytomas represent the most common histopathologic subtype of IMSCTs in children, comprising approximately 30% to 60% of pediatric cases.[13,15, 16, 17, 18] The majority are low-grade (WHO grade I–II) lesions that typically present during the first decade of life, with pilocytic astrocytoma being the predominant subtype in children under 5 years of age.[15] High-grade astrocytomas are considerably less frequent, accounting for 10% to 15% of pediatric spinal cord astrocytomas.[13,16]

A slight male …

Epidemiology and Pathogenesis

Ependymomas account for 20% to 30% of pediatric intramedullary tumors, are slightly more common in males and adolescents, and can appear in any part of the spinal cord depending on subtype.40, 41, 42, 43 Historically, ependymomas were the second most common lesion behind astrocytomas; however, the incidence may be changing in pediatric patients.[1,40] …

Molecular Diagnostics and Biology

Ependymomas are from neuroectodermal origin which usually form midline lesions from the central canal's ependymal lining.[44] Recent genetic analysis …

Gangliogliomas, hemangioblastomas, and other tumors

Gangliogliomas are among the more common non-astrocytic intramedullary tumors in children, though they remain less frequent than astrocytomas or ependymomas, accounting for approximately 15% of pediatric intramedullary tumors.[15,38] These mixed neuronal–glial tumors are typically benign, slow-growing, and most commonly located in the cervical or thoracic spinal cord. Because of their indolent course, they may demonstrate holocord involvement at presentation. A distinguishing imaging feature is …

Chemotherapy and radiation

Evidence supporting the efficacy of chemotherapy for pediatric intramedullary spinal cord gliomas remains limited and is largely derived from case reports, small series, and extrapolation from treatment protocols used in intracranial tumors.[66] In low-grade tumors, chemotherapy generally plays a minimal role. It may be considered in cases of tumor progression or recurrence when the goal is to delay or avoid radiotherapy (RT), which carries substantial long-term risks to the developing spinal …

Outcomes/recovery

Pediatric patients with intramedullary spinal cord tumors generally experience favorable long-term survival. In one large series, overall survival (OS) rates were 88% at 5 years, 74% at 10 years, and 64% at 20 years, while PFS rates were 61% at 5 years, 54% at 10 years, and 44% at 20 years.[7] …

Clinics care points

- A high index of suspicion should be maintained in children with subtle, delayed presentations, as intramedullary spinal cord tumors often present insidiously over months with nonspecific symptoms such as pain, gait disturbance, clumsiness, scoliosis, or regression of motor milestones. …
- MRI is critical to delineate the anatomy of the disease and define the safest operative approach and strategy. …
- Maximal safe resection should be prioritized, and neuromonitoring is an invaluable resource during these …

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Disclosures

The authors have nothing to disclose. …

First page preview


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KEYWORDS
• Pediatric neurosurgery • Intramedullary • Spinal cord tumor • Astrocytoma • Ependymoma

KEY POINTS
• Pediatric intramedullary spinal cord tumors are rare but clinically important lesions, with astrocytomas and ependymomas predominating.
• Children often present with months of progressive, nonspecific symptoms—pain, motor deficits, gait disturbance, or scoliosis—leading to diagnostic delays.
• MRI is essential for diagnosis and surgical planning, highlighting differences between infiltrative astrocytomas and more well-defined ependymomas.
• Maximal safe resection remains the cornerstone of treatment; gross-total resection strongly predicts improved outcomes, while adjuvant radiotherapy and chemotherapy are selectively applied.
• Long-term survival is generally favorable, and many children regain meaningful function; molecular markers and extent of resection increasingly shape prognosis and guide management.

INTRODUCTION

Pediatric intramedullary spinal cord tumors are rare central nervous system (CNS) neoplasms that pose significant surgical and medical challenges. These lesions occur in approximately 0.3 per 100,000 children per year and account for less than 10% of all pediatric CNS tumors.[1,2] Despite their rarity, they represent an important cause of morbidity in children with tumors of the CNS and demand a nuanced, interdisciplinary approach.

Adults and children differ in the distribution of intramedullary spinal cord tumors. In the pediatric population, low-grade astrocytomas represent the most common subtype, followed by ependymomas and hemangioblastomas, although recent studies suggest this distribution may be changing.[1,3] Moreover, the 2021 update to the World Health Organization (WHO) classification of CNS tumors introduced a molecularly driven framework that emphasizes genetic and epigenetic markers over purely histopathologic criteria, further refining diagnostic accuracy and prognostic stratification.[4]

In this review, we summarize the contemporary understanding and management of pediatric intramedullary spinal cord tumors, including their clinical presentation, diagnosis, histologic and molecular characterization, surgical and adjuvant treatment strategies, and long-term outcomes. We also highlight emerging trends and future directions that may shape the care of these rare and challenging lesions.

CLINICAL PRESENTATION

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