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Intramedullary spinal cord lesions in children

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Abstract

Paediatric intramedullary spinal cord lesions are uncommon pathologies, prone to result in dismal prognosis if not managed promptly and aggressively. While children usually present in good functional grades compared to adults, early recognition and treatment is important to improve outcomes. In this review, we present tumour demographics, patient factors, and treatment modalities of intramedullary spinal cord lesions in paediatric patients.

Keywords: intramedullary spinal cord tumors, pediatric, ependymoma, astrocytoma

Introduction

Tumours of the Central Nervous System (CNS) are the second most common malignancies in children, after leukaemias.[1] Intramedullary spinal cord tumours (IMSCT), however, are uncommon and account for only 3-10% of all CNS tumours, and 30-40% of all spinal cord tumours in the paediatric population.[1-5] These tumours can be localized or holocord — the latter involving almost the entire spinal cord from the cervico-medullary junction to the conus medullaris, and representing a rarer and more challenging condition to treat.[2,4] Clinically, the tumours present insidiously, are slowly progressive, and may lead to profound neurologic deficits.[2]

Low grade astrocytoma represents most of the childhood IMSCT, closely followed by ependymomas, haemangioblastomas and gangliogliomas — the latter more common below age three.[2,6] Developmental tumours such as teratoma, dermoid and epidermoid cysts are rarer, occurring mostly during first year of life.[2] With advancement in radiology, intraoperative monitoring and surgical technique; safe and effective treatment strategies for these tumours are now available.[3,4] Surgery remains the mainstay of treatment and may or may not be supplemented by adjuvant therapy.[1] Most of the available evidence on the management of IMSCT in children is based on case series, limiting our ability to completely understand tumour demographics and outcomes.[4]

Herein, we have reviewed the literature to assess the incidence, clinical and radiological characteristics, and management strategies in children with IMSCT and other spinal cord lesions.

Review of Evidence

In one of the most cited case series of IMSCT published in 1996, Constantini et al., carried out a retrospective analysis of 27 children under 3 years of age, who presented with IMSCT at their centre. They reported pain to be the commonest presenting symptom, followed by motor weakness, torticollis, and progressive kyphoscoliosis. Gross total resection (GTR) was achieved in 72% of the cases, and no surgical mortality was reported. Twenty-four low-grade (LG) lesions were observed on histopathology, with astrocytoma being the commonest (n= 12). Out of the three reported high-grade (HG) lesions, two were

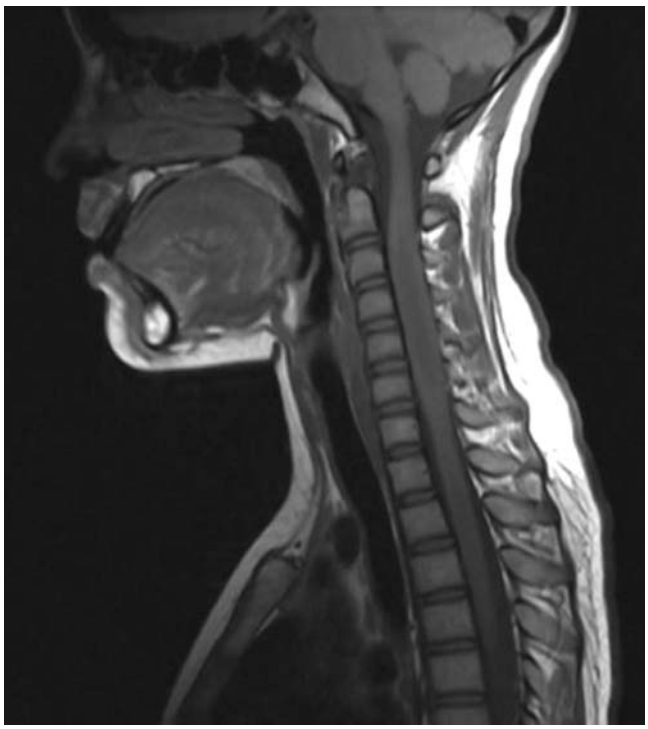

Figure-1: T1 weighted MR image, sagittal section of cervical spine of a 15 years old girl, showing loss of cervical lordosis, and a hypo-intense signal change in the spinal cord, extending from C3 till C6, with swelling of the cord.

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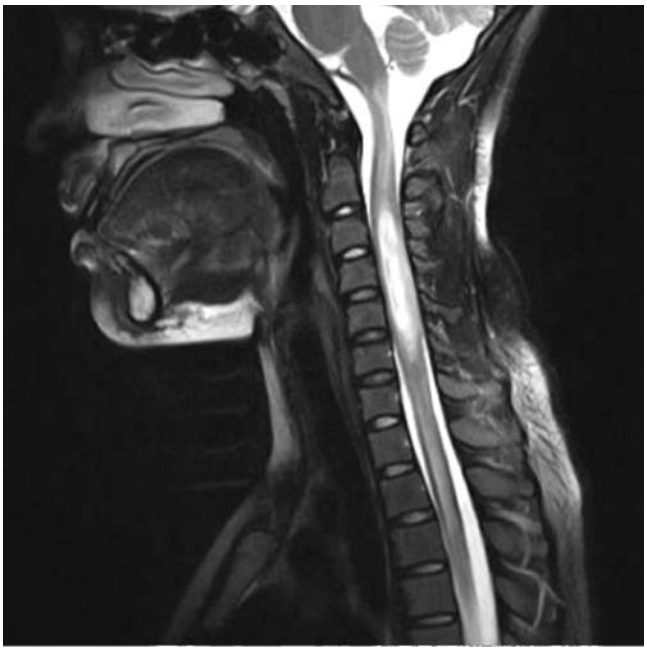

Figure-2: T2 weighted MR image of the same girl, showing a hyperintense, sausage shaped intramedullary lesion, extending from C3 to C6. It is causing expansion of the spinal cord, and there are hyperintense signal changes in the cord, proximal and distal to the lesion.

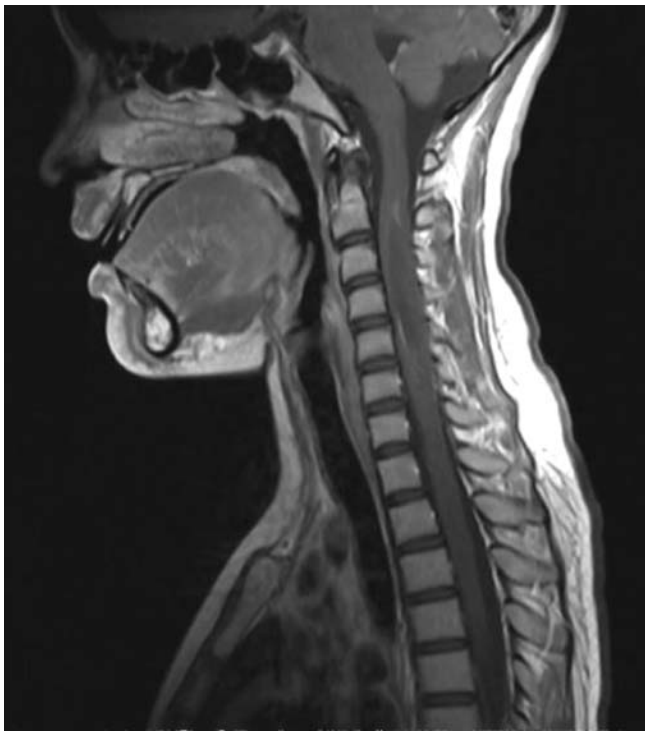

Figure-3: T1 post-contrast image of the same girl, showing subtle peripheral enhancement. This lesion was resected, and histopathology was reported as diffuse astrocytoma grade II.

anaplastic astrocytoma, and one glioblastoma. Encouraging functional outcomes were seen at 76 months of follow-up, concluding that IMSCT can be radically and safely removed in children less than 3 years of age.[2,6]

In 2012, Bansal et al., carried out a retrospective analysis of 82 paediatric patients with IMSCTs, out of which 69.5% were males (n=57). Epidermoid cysts constituted the most common type of tumour, followed by low-grade gliomas and ependymoma. Ependymomas had a higher amenability for GTR as compared to pilocytic astrocytoma (43.7% vs. 22.2% respectively). Near-total resection was achieved for most other pathologies. At the last follow-up, most patients had improved (n=8) or unchanged (n= 46) functional status, while two patients had worsened. The authors concluded that pre-operative neurological status was more predictive of post-operative functional outcomes than histological grading.[7]

Babu et al., published a retrospective analysis in 2014, of 46 patients treated for astrocytoma of the spinal cord at their center. The median age was 15 years, with a male predominance observed. Pilocytic astrocytoma was the commonest pathology (41.6%), followed by grade 2 diffuse astrocytoma (21.7%), anaplastic astrocytoma grade 3 (19.6%) and glioblastoma in the rest. Most patients presented with pain (45.7%), while dysesthesia/paresthesia and scoliosis constituted the rest of the presenting symptoms. While surgery was the mainstay of treatment, GTR could only be achieved in 12.5% of the patients. The study revealed a direct relationship between high tumour grade, radical resection and significant post-operative neurological deficits.[8]

In another retrospective series by Ahmed et al., 55 children with IMSCT were reviewed. Most children presented with sensory deficit, with cervicomedullary junction the commonest site on radiology. Low-grade astrocytoma was the commonest pathology (53%), followed closely by ependymoma. Most children underwent sub-total resection for their tumours (62%), followed by an additional procedure of shunt insertion in 22 patients, for post-operative hydrocephalus. While no surgical mortality was observed, 27% patients reportedly developed new or progressive neurological deficit and 47% had a recurrence of the tumour, at a mean of three months after surgery. Extent of resection and tumour grade were conclusively reported to influence patient outcomes.[9] In 2015, Kutluk et al., evaluated the pathologic factors and treatment modalities in paediatric patients with IMSCTs. In their retrospective review of 36 patients, aged 7.9 years on average, they reported ependymoma (52.8%), and astrocytoma (44.4%) as the commonest tumours. Only three high grade tumours were reported.

The tumour was primarily located in the thoracic (47%) and cervical (28%) segments, and surgery was the mainstay of treatment, with STR and GTR achieved in most patients (45% and 33% respectively). Adjuvant radiation and chemotherapy was used to supplement treatment in 26 and 15 patients, respectively. The 3, 5 and 10 year overall survival rates were 72%, 63% and 56%, and event free survival rates were 43%, 40% and 40%, respectively. The authors reported significantly higher survival rates in patients with low grade tumours, especially those in the ependymoma group.[1]

Luksik et al., carried out a large retrospective assessment, exploring factors that may influence survival in 348 paediatric patients with spinal cord astrocytoma, recorded in the surveillance, epidemiology, and end result (SEER) database. Average age for the group was 9.3 years, with a gender predominance for males. Tumours were majorly WHO grade 2 (20.7%) followed by grades 4, 1, and 3 tumours. Surgery was the primary treatment modality, with partial resection achieved in 51.7% of the patients and gross total resection in 28.7%. Radiotherapy followed as an adjunct in 28.4% patients and the survival rate for patients at 5 years was 74.8%. Surgical intervention, especially with GTR, improved survival, while high grade lesions, application of radiation therapy, and distant and invasive extension of the tumour significantly decreased survival. Younger age was shown to have a protective effect. The authors conclusively highlighted the need for early diagnosis for better prognosis.[10] In another major study in 2019, Bhimani et al., analyzed the Paediatric American College of Surgeons National Surgical Quality Improvement Programme database, retrospectively for acute surgical risk. In the group of 139 patients from the registry, predominantly males (n= 81), and at a mean age of 8.7 years, tumours mostly arose from the thoracic spine, followed by the cervical and lumbar spinal regions. Worse preoperative health status, and a greater proportion of malignant tumours were noticed with cervical and thoracic IMSCT populations compared with the lumbar IMSCT population. Of the 12 readmissions, patients with cervical IMSCTs returned to the operating room at a significantly greater rate than did the thoracic and lumbar IMSCT populations, mainly due to respiratory problems and hydrocephalus management. However, the overall acute surgical risk reportedly remained low.[11]

Conclusion

Astrocytoma and ependymoma, predominantly low-grade, constitute the most common intramedullary IMSCT in children. While modern surgical adjuncts such as microscopes and intra-operative monitoring have resulted in improved outcomes over the years, preoperative neurologic status, remains the most important predictive factor of prognosis in children, even more so than tumour grade.

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