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Pediatric Intramedullary Spinal Cord Tumors

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Pediatric intramedullary spinal cord tumors (IMSCTs) are rare and account for only 4% to 6% of all central nervous system (CNS) tumors [1]. Approximately 100 to 200 cases are diagnosed each year in the United States [2], representing an incidence of less than 1 in 100,000. IMSCTs in children are usually located in the cervical or thoracic spine (46%) and occur infrequently in the lumbar spine [3,4]. There is equal male and female distribution in the reported pediatric series [5–7]. Astrocytomas and ependymomas represent most IMCSTs in children. Other less frequent tumors in this location include gangliogliomas, oligodendrogliomas, subependymomas, neurocytomas, hemangioblastomas and, rarely, metastases.

Presenting symptoms can be subtle in the pediatric population, often leading to delay in recognition. Parents usually report that mild symptoms were present for months to years before diagnosis. Signs and symptoms include pain, weakness, paresthesias, spinal deformity, sphincter disturbance, and cervicomedullary symptoms [4]. Slow deterioration of neurologic function can also occur [8]. Because of their rarity, treatment recommendations are extrapolated from those for similar tumor types in other CNS locations or from small cohort studies. Nevertheless, it seems that the definitive treatment, if possible, for primary IMSCTs is surgical resection. This approach may leave a patient with severe neurologic deficits, however. Effective treatment requires an interdisciplinary approach, with specialists in pediatric neurosurgery, neuro-oncology, child neurology, pediatrics, and nursing.

Ependymoma

Epidemiology and pathogenesis

Ependymomas can arise from the cranial ventricular system or within the spinal cord. Those originating in the spinal cord presumably arise from the remnants of the central canal and can be seen in children and young adults. Ependymomas are commonly found in the cervical region in children and have a lower incidence as compared with astrocytomas [9,10]. In a study by Miller [11], ependymomas were found in 19 (16%) of 117 pediatric patients with IMSCTs. Ependymomas tend to increase in frequency with age. A recent study by Constantini and colleagues [8] showed that there were no ependymomas reported in a series of IMSCTs in patients less than 3 years of age.

Ependymomas (World Health Organization [WHO] grade II) have been found to contain SV40 virus, large, T-antigen–related DNA sequences. This finding has received a great deal of attention, because widespread use of SV40-contaminated polio vaccines may have occurred between 1955 and 1962 [12]. These findings have not been corroborated by other investigators, however, and the role of SV40 in the pathogenesis of ependymomas still remains to be proven [13]. Intramedullary spinal cord ependymomas are associated with neurofibromatosis type 2 (NF2) [14], a genetic syndrome inherited in an autosomal dominant pattern. In fact, a recent study showed that mutations of the NF2 transcript occurred in

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most sporadic intramedullary spinal cord ependymomas [15]. In this study, five of seven intramedullary spinal cord ependymomas had mutations in the region of the transcript homologous to previously identified cytoskeletal proteins and resulted in truncation of the predicted protein product, probably rendering the protein product inactive [15].

Pathologic findings

Grading

Eighty-nine percent of IMSCTs in children are low-grade lesions [8]. Four different entities are delineated according to the most recent WHO classification of tumors [16]. The subependymoma (WHO grade I) is a benign and slowly growing intraventricular tumor that is often detected incidentally and carries a good prognosis. These tumors have histologic features of subependymomas and ependymomas. The myxopapillary type (WHO grade I) arises in the filum terminale or conus medullaris almost exclusively and carries a good long-term prognosis. Classic ependymoma (WHO grade II) is a common intramedullary neoplasm found in the spinal cord in children. Anaplastic ependymomas (WHO grade III) are thought to arise from a malignant transformation of low-grade ependymomas [16].

Histopathology

The myxopapillary ependymoma has a pseudopapillary architecture and abundant mucin production. This variety of ependymoma is characterized by glial fibrillary acidic protein (GFAP)–expressing tumor cells with a cuboidal or elongated morphology arranged radially in a papillary manner around vascularized stromal cores. Mitotic activity is almost absent, and a mucoid matrix material accumulates between tumor cells and blood vessels. Ependymomas are highly cellular tumors regardless of their grade. Their characteristic features include pseudorosettes and perivascular grouping. The high cellularity of ependymomas can cause disagreement among neuropathologists on the grade of a particular specimen [17]. In particular, there is a tendency to attribute a higher grade to low-grade tumors.

Classic ependymomas contain perivascular pseudorosettes which are immunoreactive for GFAP, and, uncommonly, true epithelial-lined ependymal rosettes. The perivascular pseudorosettes originate from tumor cells arranged radially around blood vessels and occur in most of these tumors. These tumors are well delineated, and mitotic figures are rare. An occasional nonpalisading focus of necrosis is sometimes observed and is compatible with lower grade histology. Unusual variants, such as the clear cell ependymoma and the tanycytic ependymoma, can mimic oligodendroglioma or astrocytoma, respectively [4].

The anaplastic ependymoma displays histologic and behavioral characteristics that are distinct from the classic ependymoma. Although the cells of the classic ependymoma are morphologically similar to normal ependymal cells, those of the anaplastic ependymoma have clear histologic features of malignancy. More specifically, the anaplastic ependymoma is characterized by aggressive mitotic activity and increased cellularity, which are often associated with microvascular proliferation and pseudopalisading necrosis.

Molecular biology and cytogenetics

A study by von Haken and coworkers [18] reported a 50% incidence of allelic losses on the short arm of chromosome 17 in 18 pediatric ependymomas. The tumor suppressor gene TP 53 was excluded; therefore, a candidate gene has yet to be identified. Recognizing the increased incidence of ependymomas in patients with NF2, some investigators have reported evidence for mutations of the NF2 gene, which is located on chromosome 22q12. One group analyzed 62 ependymal tumors for loss of heterozygosity (LOH) 22q, LOH 10q, and for mutations of the NF2 tumor suppressor gene. Six of the tumors revealed mutations of NF2, all of which came from patients with IMSCTs [19]. This may suggest that spinal intramedullary ependymomas constitute a distinct phenotypic variant of an altered NF2 gene. Additional cytogenetic changes have been found in a significant number of ependymomas. In a series of 22 childhood ependymomas, loss of chromosome 22 was found in two cases, deletion of chromosome 17 in two cases, and rearrangements or deletions of chromosome 6 in 5 tumors [20]. In anaplastic ependymoma, the genetic alterations remain largely undefined. A recent study of 23 anaplastic ependymomas revealed LOH 10q in four cases, but the significance of this finding is unknown [19].

Clinical features

Spinal cord ependymomas occur commonly in the cervical region. Children most often present with pain, which is reported in 42% of cases.

Motor regression with weakness is present in 36% of cases, gait abnormality or deterioration in 27%, torticollis in 27%, and progressive kyphoscoliosis in 24% [8]. Hydrocephalus occurs with greater frequency than in adult patients and may require treatment with cerebrospinal fluid diversion [21].

Diagnostic imaging

MRI has greatly improved the preoperative evaluation of spinal cord tumors. The exact histologic diagnosis of these tumors, however, cannot be made by MRI features and patterns of enhancement (Table 1). Tumor tissue is still required in virtually all cases to establish a definitive diagnosis. Because ependymomas originate from the ependymal walls, they tend to be more central in location when compared with astrocytomas. On average, ependymomas span three to four vertebral bodies [22]. The pattern of enhancement is homogeneous, usually with well-defined borders (Fig. 1). A "cap sign" is often seen, which corresponds to a low signal intensity area on either side of the tumor mass itself. The cap represents hemosiderin deposits secondary to chronic hemorrhage [22]. These tumors can present with subarachnoid hemorrhage on occasion. Ependymomas are also frequently associated with large intramedullary satellite cysts. These can extend many levels above or below the solid component of the tumor.

Miyazawa and colleagues [23] reported that the typical MRI characteristics of ependymoma include an enhancing border, sharply defined margins, and a central location in the spinal cord. Axial postcontrast sequences demonstrate symmetric expansion of the spinal cord, unlike astrocytoma, in which the pattern of expansion is often asymmetric or nodular. The study also concluded that hemorrhage within the tumor, hemosiderin deposits, or calcifications are more frequent in ependymomas, which may be attributed the presence of a highly vascular connective tissue stroma.

Astrocytoma

Epidemiology and pathogenesis

Astrocytomas are the largest group (approximately 60%) of pediatric IMSCTs [3,24,25]. They are typically large and mostly located in the thoracic region. A cystic component may be present, although such cysts are commonly intratumoral. Satellite cysts and secondary hydromyelia may also be observed [22]. The histology in all age groups is usually low grade, and high-grade tumors occur in only 10% to 15% of cases [1,26]. In children, the most common IMSCT is the pilocytic astrocytoma, which a well-circumscribed and often cystic tumor that has an indolent pattern of growth. Other related tumors that can occur within the spinal cord include ganglioglioma (Fig. 2), mixed glial tumors, and primitive neuroectodermal tumors.

Low-grade astrocytomas most often present in the first 2 decades of life, with no clear gender predilection. In adults, they are mostly encountered in younger patients (mean age of 29 years), with a predominance in male patients (63%) [22]. Previous irradiation is known to play a causative role in the formation of CNS gliomas. Nevertheless, few case reports of radiation-induced intramedullary astrocytomas exist in the literature. Grabb and colleagues [27] reported a case of a radiation-induced spinal cord anaplastic astrocytoma in a 20-year-old female patient who presented with neck pain and new significant neurologic deficits 17 years after resection of a posterior fossa medullomyoblastoma and craniospinal irradiation.

Familial clustering of astrocytomas is frequently described. These tumors are associated

Table 1
MRI of intramedullary spinal cord tumors

	Ependymoma	Astrocytoma
Location	Centrally located in the spinal cord; mostly arise in the cervical spine but can also occur in the conus in children	Eccentrically located and usually widens the spinal cord; 75% of astrocytomas arise in the cervicothoracic region, 20% arise in the distal spinal cord, and 5% arise in the filum terminale
T1	Spinal cord symmetrically expanded; isointense/hypointense to the spinal cord	Spinal cord asymmetric and "lumpy" in appearance; isointense/hypointense to the spinal cord
T1 with contrast	Enhances with contrast but less than astrocytomas	Ill-defined borders; heterogeneous with partial contrast enhancement

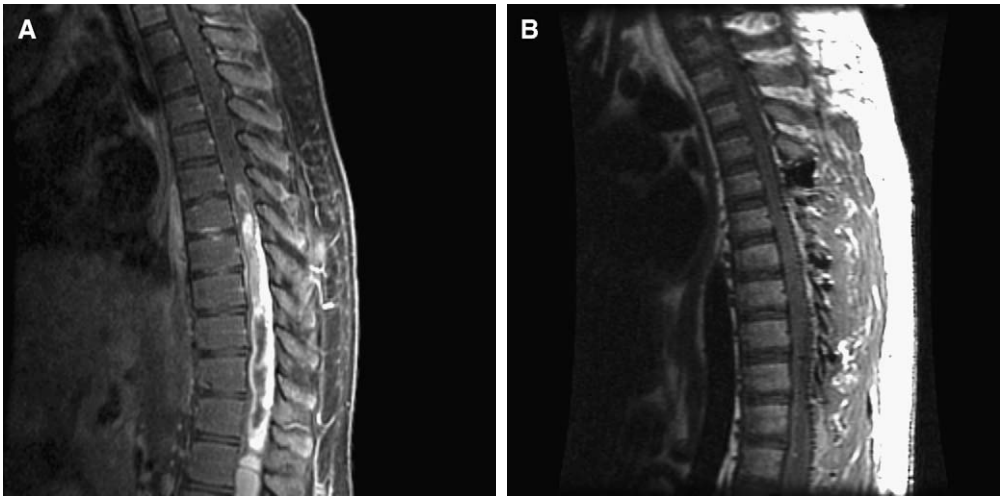

Fig. 1. A 13-year-old male patient presented with back pain and bilateral lower extremity weakness. (*A*) Sagittal T1-weighted MRI scan with gadolinium revealed a partly cystic, contrast-enhancing, intramedullary tumor extending from T7 to L3. (*B*) Postoperative MRI scan confirms that a gross total resection was achieved. The pathologic diagnosis was consistent with an ependymoma.

with inherited syndromes, such as Li-Fraumeni syndrome, Turcot syndrome, tuberous sclerosis, multiple enchondromatosis (Maffucci/Ollier disease), and neurofibromatosis (NF) [28–30]. The typical picture of spinal involvement with NF is the presence of multiple, extramedullary, spinal nerve root neurofibromas. These tumors can cause spinal cord compression but are anatomically outside the spinal cord itself. Lee and coworkers [31] have reported a series of nine patients (age range: 4–31 years) with NF who had developed IMSCTs. Three patients had neurofibromatosis type 1 (NF1), five patients had NF2, and one patient had an undefined variety. The pathologic findings of the tumors were also variable, with five ependymomas, three astrocytomas, and one intramedullary schwannoma reported [31]. Based on these limited data, it is unclear whether IMSCTs are a subgroup of NF-associated neoplasms or simply represent higher tumor predisposition in these patients.

Pathologic findings

Grading

The WHO grading system divides astrocytomas into four groups based upon the area with the highest degree of anaplasia: grade I (pilocytic astrocytoma), grade II (diffuse astrocytoma), grade III (anaplastic astrocytoma), and grade IV (glioblastoma multiforme).

Histopathology

The pilocytic astrocytoma is characterized by elongated "hair" cells with a coarse cytoplasmic process. Intracytoplasmic Rosenthal fibers and eosinophilic granular bodies are characteristic but not specific features. Cellular pleomorphism, vascular proliferation, infiltration of the meninges, and occasional mitoses can also be present but have no prognostic value and do not represent malignant features. The diffuse fibrillary astrocytoma is an infiltrative tumor that produces fusiform enlargement of the cord. The typical features include hypercellularity, nuclear atypia, and an infiltrative growth pattern. Mitotic activity is mostly absent. Three major variants can be distinguished based on the appearance of the astrocytes: fibrillary (fibrillary neoplastic astrocytes), gemistocytic (a conspicuous although variable fraction of gemistocytic astrocytes), and protoplasmic (small cell body with few flaccid processes, a low content of glial filaments, and scant GFAP expression). Higher grade tumors, such as anaplastic astrocytoma and glioblastoma multiforme, have increased cellularity, anaplasia, and marked mitotic activity.

Molecular biology and cytogenetics

Some astrocytomas are known to lose chromosome 17q, which includes the region encoding the NF1 gene [32]. Screening of NF1 coding sequences,

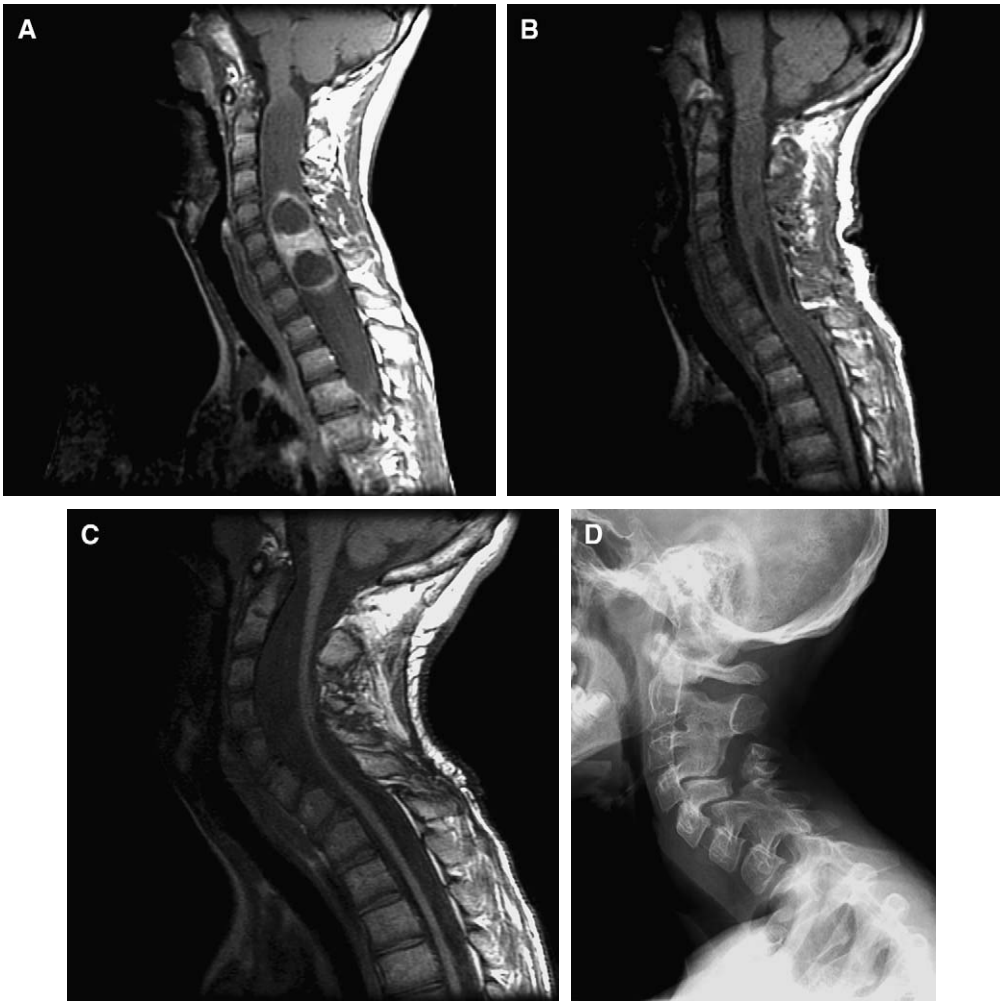

Fig. 2. A 10-year-old male patient presented with decreased dexterity and weakness of the right upper extremity. (*A*) Preoperative T1-weighted MRI scan with contrast revealed a cystic intramedullary cervical cord mass from C5 to C7. A substantial amount of spinal cord edema was present rostral and caudal to the mass. (*B*) Immediate postoperative T1-weighted MRI scan confirmed a gross total resection, although the spinal cord edema persisted. The pathologic diagnosis was consistent with a ganglioglioma. (*C*) Postoperative MRI scan obtained 2 years after surgery shows a diminished volume of spinal cord tissue, although neurologic function remained excellent. (*D*) Despite a laminoplasty being performed at the time of surgery, a gradual lordotic curve in the cervical spine is developing as seen on this plain radiograph of the cervical spine 2 years later.

such as the GRD region (guanine triphosphate activating protein-related domain), the only functional region of the NF1 mRNA transcript identified to date, has failed to detect any mutations. Current evidence has failed to support a role for the tumor suppressor gene NF1 in the oncogenesis of pilocytic astrocytomas [16].

Cytogenetic studies of pilocytic astrocytomas have revealed a variety of aberrations, but no specific pattern suggestive of a specific tumor suppressor gene has been reported. Ransom and colleagues [33] attempted to locate tumor suppressor genes on chromosome 10 by correlating cytogenetic studies and LOH analysis in human astrocytomas and mixed oligoastrocytomas. Of 53 specimens, 45 were diffuse astrocytomas, 1 was an astroblastoma, and 7 were mixed oligoastrocytomas. By cytogenetic analyses the most common numeric chromosome abnormalities were +7, −10, −13, −14, −17, +19, −22, and −Y.

The most common structural abnormalities involved chromosome arms 1p, 1q, 5p, and 9p. By LOH and dosage analysis, the most common molecular genetic abnormalities were of chromosome arms 5p, 6p, 7q, 9p, 10p, 10q, 13q, 14q, 17p, and 19p. When the results of all methods were combined, the most commonly abnormal chromosomes were, in descending frequency, 10, Y, 17, 7, 13, and 9. In 80% of cases, the cytogenetic and molecular genetic studies were concordant. The authors concluded that based on the genetic analyses of these tumors, there may be two regions on chromosome 10 that may contain tumor suppressor genes [34].

Clinical features

The clinical features of intramedullary astrocytomas are similar to those of intramedullary ependymomas. Gait disturbance, pain, and sphincter disturbance are the most common presenting symptoms, whereas reflex changes, paralysis, and sensory impairment are the most frequent physical findings [1]. Spinal deformity can be present in up to 30% of patients [35]. In a series of 152 spinal cord astrocytomas, Epstein [36] reported that scoliosis was the most common early sign in 34 cases of thoracic tumors, whereas torticollis was present in 21 cases of cervical spine astrocytomas. Tumors involving the cervicomedullary junction can present with unusual symptoms. If the medulla is involved, children can develop (1) failure to thrive because of nausea, vomiting, choking, and dysphagia; (2) frequent respiratory tract infections from chronic aspiration; (3) dysphonia and dysarthria; and (4) sleep apnea from respiratory center involvement. If the tumor arises from the cervical cord, children can present with neck pain, torticollis, slowly evolving motor deficits, muscle atrophy, dysesthesias, hyperreflexia and, occasionally, hydrocephalus [37,38].

Diagnostic imaging

As with all spinal cord tumors, MRI is the diagnostic tool of choice. Astrocytomas are commonly located eccentrically within the spinal cord (see Table 1). On imaging, there is heterogeneous, moderate, and partial contrast enhancement after administration of gadolinium, with ill-defined borders [22]. Approximately 75% of astrocytomas occur in the cervicothoracic region, 20% occur in the distal spinal cord, and 5% occur in the filum terminale [39]. Whereas the mean size of an ependymoma corresponds to a mean extent of 3 to 4 vertebral bodies, astrocytomas are, in general, much more extensive, with some series reporting a mean span of 5.6 vertebral bodies (minimum of 2 vertebral bodies, maximum of 19 vertebral bodies) [22].

Management of ependymomas and astrocytomas

The treatment of choice for intramedullary tumors, ependymomas, and astrocytomas is surgical resection. The resection is aided by motor and sensory evoked potential monitoring. Gross total resection can be achieved in most ependymomas and is likely to result in cure [21]. In contrast, astrocytomas are infiltrating neoplasms, and gross total resection is only occasionally possible in the pediatric population. The outcome for low-grade astrocytomas is better in children than in adults, but the outcome is not as favorable as that for ependymomas. The role of radical resection of low-grade fibrillary astrocytomas of the spinal cord in children has not been definitively demonstrated in the literature [21].

Surgery

Surgical approach

The operative exposure is centered on the solid part of the tumor as identified by preoperative MRI. The preferred technique in children is osteoplastic laminotomy, removal of the laminar roof in one piece, and replacement at the end of the case [21]. This preserves the posterior tension band, restores normal anatomy, and may result in bony fusion of the reapproximated lamina [40]. There is evidence that this technique results in a reduced incidence of postoperative spinal deformity, although it does not guarantee the avoidance of this complication (see Fig. 2) [2]. Intraoperative ultrasound is crucial to verify that the rostral-caudal bony exposure is sufficient to expose the entire solid component of the tumor [41,42]. More important, intraoperative ultrasound was shown to reduce the extent of the laminectomy, dural opening, and myelotomy needed for resection [43].

Neuromonitoring-assisted tumor removal

Neuromonitoring assistance is of value during the resection of pediatric spinal cord tumors;

however, in general, the consistency of recordings can vary, especially in extremely young children. Conventional orthodromic somatosensory evoked potentials (SSEPs) are used to measure dorsal column function during surgery [4]. Excessive traction or injury to the spinal cord results in a reduction of these potentials. Antidromic-elicited SSEPs assist in the planning of the initial myelotomy [44]. This type of recording is performed by stimulating along the posterior surface of the spinal cord while recording evoked responses in the extremities. A stimulating electrode is applied to the area of the dorsal columns, beginning laterally and then moving toward midline. Sites where stimulation evokes no SSEPs are then marked "septum," indicating the optimal electrical site of the myelotomy.

A number of other techniques are used to reduce the likelihood of postoperative neurologic deficits. These include transcranial epidural motor evoked potentials (MEPs) and/or transcranial muscle MEPs [45]. Direct stimulation of the spinal cord is also possible with simultaneous recordings of extremity electromyographic (EMG) recordings. This technique helps to differentiate tumor from spinal cord and prevents damage to the corticospinal tracts [44]. Additional details regarding neuromonitoring are available elsewhere in this issue.

Radiation therapy

Radiation therapy has played a limited role in the primary management of IMSCTs in children because of potentially debilitating side effects. CNS irradiation can cause growth retardation, endocrine dysfunction, decreased IQ, radionecrosis, vasculopathy, and alopecia [46]. Some investigators advocate that radiotherapy should be reserved for incompletely resected tumors and for high-grade astrocytomas.

In a recent review of the literature regarding radiation therapy and the management of IMSCTs, Isaacson [47] concluded that low-grade completely resected astrocytomas should be followed by serial imaging studies. Low-grade astrocytomas with incomplete resection seem to benefit from postoperative radiation. For high-grade astrocytomas of the spine, some authors recommend aggressive surgical resection followed by radiotherapy, but these tumors can relapse early. Isaacson [47] recommended that all high-grade astrocytomas be treated with radiation.

Literature supporting the use of radiotherapy for intramedullary spinal cord ependymoma in children is scarce and inconclusive [47]. Nagib and O'Fallon [48] treated three children aged 7, 8, and 13 years for conus medullaris myxopapillary ependymoma over a 2-year period, with a 24-month follow-up. One of these children received radiation therapy after a recurrence. Based on this patient and on a literature review addressing this type of tumor in 11 other children, the authors concluded that the gross feature of myxopapillary ependymoma allowing for complete resectability seems to be the key prognostic factor and that radiotherapy seemed to have no proven value in completely resected conus medullaris myxopapillary ependymoma tumors in children.

In his review of the literature, Isaacson [47] concludes that patients with gross total resection of low-grade ependymomas with no evidence of disease should be observed with serial imaging studies. High-grade and multifocal benign ependymomas should be given adjuvant radiation therapy. Complete imaging of the neuraxis and cerebrospinal fluid analysis should be obtained to assist in the decision-making process. Isaacson's specific radiation therapy guidelines are as follows: (1) low-grade astrocytoma and low-grade ependymoma residual disease after surgery should be prescribed a total dose of 5040 cGy in 180-cGy fractions over 28 treatment days using external beam radiation therapy (EXBRT); (2) for high-grade astrocytomas, despite complete resection, the treatment recommended is the same (5040 cGy with 180-cGy fractions with EXBRT); and (3) malignant ependymomas and benign multifocal ependymomas are treated with a locally delivered dose of 5040 to 5400 cGy in 28 to 30 fractions, with occasional consideration of radiation to the neuraxis [47].

Chemotherapy

Chemotherapy for the treatment of IMSCTs as an adjunct to surgery, radiotherapy, or both is not standardized. Adjuvant chemotherapy may play an important role in children younger than 3 years of age, mainly in an effort to delay radiotherapy [49]. Chemotherapy guidelines for IMSCTs are mainly based on the clinical experience with intracranial low-grade gliomas. As with radiotherapy, no randomized trials have been performed and only anecdotal reports and small series are reported [50–53].

Lowis and coworkers [52] described two children with astrocytomas. The first child, a 19-month-old with an anaplastic astrocytoma of the cervical spinal cord, progressed rapidly after initial partial resection. Chemotherapy was given according to the United Kingdom Children's Cancer Study Group Baby Brain Protocol [54], with marked clinical improvement. This regimen comprised cycles of chemotherapy (carboplatin, vincristine, cyclophosphamide, methotrexate, and cisplatin) given every 2 weeks, regardless of count, for a period of 1 year. No apparent disease remained at the end of treatment [52]. The second child was 4 years old and had a recurrent low-grade astrocytoma. Chemotherapy was given for 3 months according to an International Society of Pediatric Oncology protocol for low-grade gliomas. This regimen consisted of cycles of carboplatin and vincristine administered every 3 weeks and vincristine administered weekly for the first 10 weeks. This was followed by carboplatin and vincristine administered every 4 weeks up to 1 year. Marked tumor regression was observed, accompanied by neurologic recovery. The authors concluded that these patients demonstrate the potential value and low morbidity of chemotherapy in spinal cord astrocytoma [52].

A study by Hassall and colleagues [51] reported responses to carboplatin in three patients with progressive low-grade spinal cord gliomas. The diagnoses of the tumors were juvenile pilocytic astrocytomas (n = 2) and ganglioglioma (n = 1). With a mean follow-up of 27 months, one patient had a complete response, one patient had a partial response, and one patient had stable disease [51]. The potential role of chemotherapy in the management of spinal cord astrocytoma remains to be defined.

Outcome

Different studies have consistently demonstrated that there are only two significant predictors of outcome in patients with IMSCTs: the histologic grade of the tumor and the preoperative neurologic status at the time of surgery [55]. In general, patients with ependymoma have a more favorable outcome than those with low-grade astrocytoma (WHO grade II). In a series of 21 patients, Sandler and coworkers [56] reported a 5-year survival rate of 57% in patients with grade I or II spinal cord astrocytoma. Patients with pilocytic astrocytoma, the most common spinal cord tumor found in children, have an even more favorable prognosis [21]. Patients with malignant astrocytomas do poorly, with no correlation between the extent of resection and survival. In one study of 19 patients with malignant astrocytomas of the spinal cord, median survival was only 6 months. At the time of publication, 15 (79%) of the 19 patients in the series had died. Although all underwent radical excision, none of the patients improved after their operations. Hydrocephalus and dissemination of disease occurred in most patients [57]. Some data, however, suggest that surgery for malignant spinal astrocytomas is not always futile. In one study of 18 children with spinal astrocytomas, gross total resection was achieved in 5, including 3 with anaplastic tumors. All 5 of the patients in whom gross total resection was achieved were alive and disease-free between 12 and 18 years after treatment [58].

The prognosis and outcome of patients with intramedullary ependymomas mainly depend on the extent of the original resection. Ependymomas are slow-growing tumors, and late recurrence has been seen up to 12 years after surgery as reported at our institution [59]. Gross total resection of benign intramedullary ependymomas more commonly results in long-term tumor control or cure than does subtotal resection and radiation therapy [55,60–62]. Some authors have reported up to 100% recurrence-free survival after gross total resection [62], whereas others have reported recurrence rates of 5% to 10% [63,64]. The current consensus is that gross total resection is the most efficacious treatment and that radiation therapy is unnecessary if complete removal has been accomplished [17,62,63,65,66].

Summary

Although spinal cord tumors are rare, early diagnosis plays an important role in the management of these lesions and is an important factor in prognosis and outcome. In the pediatric population, low-grade astrocytomas predominate; ependymomas increase in frequency with ascending age and are the most frequent IMSCT in adults. Unexplained and intractable lumbar pain in childhood should be investigated with a high-quality MRI scan. Postoperative baseline MRI and regular sequential imaging studies are essential for long-term follow-up in patients who have undergone resection of an IMSCT. The mainstay of treatment to date for IMSCTs is surgery.

Electrophysiologic monitoring has proven to be a useful adjunct in aiding the resection of these lesions. In the future, appropriate management of these lesions is likely to involve a more complete understanding of their molecular genetics.

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