Surgical treatment of spinal cord astrocytomas of childhood

A series of 19 patients

FRED EPSTEIN, M.D., AND NANCY EPSTEIN, M.D.

Department of Neurosurgery, New York University Medical Center, New York, New York

This report describes the first author's surgical experience with a series of 19 consecutive cases of spinal cord astrocytoma treated over the past 2 years, with a follow-up period of 6 to 24 months. The clinical presentation, neurodiagnostic investigation, surgical technique, and results are analyzed. The authors conclude that radical resection (gross total removal of the tumor) is the optimal therapeutic option.

KEY WORDS • spinal cord tumor • intramedullary astrocytoma • ultrasonic surgical aspirator

INTRAMEDULLARY spinal cord astrocytoma is a relatively uncommon neoplasm, accounting for only 4% of central nervous system (CNS) tumors of childhood. The authors have recently cared for 19 young patients with very extensive astrocytomas, many of which involved the entire length of the spinal cord. This unusual series has provided a unique opportunity to study the biology of the neoplasm, as well as its response to different methods of treatment. It is hoped that information derived from this report will contribute to the formulation of a neurosurgical consensus regarding the optimal mode of treatment.

Summary of Cases

Location of Tumor

Fourteen of the 19 patients in this series had widening of the spinal cord, which extended down from the lower brain stem or upper cervical cord to the conus. These children are considered a subgroup (with "holocord astrocytoma"), as there are a number of common features which distinguish them from the patients with relatively localized, although multisegmental, neoplasms of similar histology. Of the latter group of tumors, three were thoracic and two were cervical in location.

Clinical Presentation

One patient was 11 months old, eight patients were between 2 1/2 and 5 years of age, eight patients were between 6 and 10 years of age, and the remaining two were aged 13 and 16 years. In four patients (two with holocord astrocytoma and two with focal cervical neoplasms), symptomatic hydrocephalus required shunting at some time during the course of treatment.

Holocord Astrocytoma. Fourteen patients had holocord astrocytoma. The striking feature of this subgroup was the mode of presentation, which was separable into two syndromes (Groups A and B). Group A contained seven patients, who presented with unilateral upper extremity weakness as the chief complaint. On examination, five patients also manifested spasticity and minimal weakness of the lower extremities which had been previously unrecognized. Severe paraspinal cervical pain was present in four patients. All patients were continent of bowel and bladder. In this group of patients, the solid component of the neoplasm was invariably within the cervical cord, the remainder of the tumor being cystic.

Seven patients comprised Group B. These children presented with mild, progressive paraparesis, and five also had severe pain over the thoracic spine. In five patients, examination disclosed spasticity in the lower extremities.
FIG. 1. Radiographs showing diffusely widened spinal cord extending from C-1 to the conus in a child with holocord astrocytoma.

extremities. Two had absent reflexes, impairment of bowel and bladder function, and a lax anal sphincter. In the former five patients, the solid portion of the tumor was within the thoracic cord, while in the latter two there was also solid tumor extending into the conus. The remainder of the cord was distended by associated cysts. One 5-year-old boy, in whom the solid portion of tumor was within the thoracic cord, presented with progressive scoliosis as the only complaint. On examination, there was spasticity but no weakness of the lower extremities.

Focal Astrocytoma. Five patients had the focal form of astrocytoma. Three patients with cervical neoplasms presented with mild weakness of one upper extremity, and two with thoracic tumors had a mild paraparesis. Spinal axis pain was not a common complaint, and spasticity, although often present, was not prominent. No patients had bowel or bladder dysfunction.

Neurodiagnostic Studies

Holocord Astrocytomas. All of the 14 patients with holocord astrocytomas had some degree of scoliosis, varying from mild to moderate. Diffuse widening of the spinal canal from the cervical to lower thoracic regions was invariably present. Plain spine x-ray films disclosed erosion of pedicles in eight patients, and scalloping of vertebral bodies in four patients. This invariably corresponded to the segments of the spinal cord that were spanned by solid tumor as compared to the diffuse widening of the spinal canal which was secondary to associated cysts. Myelography disclosed that the widened spinal cord extended from the lower brain stem to the conus in nine patients (Fig. 1), and from C-2 to the conus in five patients. In nine patients, there was a complete subarachnoid block, and delineation of the full extent of the tumor required an injection of contrast material at Cl-2 in combination with the lumbar myelogram. In one patient, a cervical cyst, which extended into the medulla, was inadvertently punctured during this study.

The spinal computerized tomography (CT) scan has proven to be a useful adjunct to myelography, but is not yet sufficiently useful to replace it. Although we have occasionally observed the delayed appearance of intrathecal contrast material within the cyst cavity, this was not a consistent observation in this series of patients. Occasionally, the spinal CT scan supplemented with intravenous contrast material has demonstrated an enhancing neoplasm.

Focal Astrocytoma. Two of the five patients with focal astrocytoma had mild scoliosis; local erosion of the pedicles was present in only one patient. Widening of the spinal cord varied in length from four to six segments, and was an accurate reflection of the extent of these non-cystic neoplasms.

Treatment

Holocord Astrocytoma. Eight patients with holocord astrocytoma were initially treated with biopsy and radiation therapy (conservative treatment). In each case, there was progressive neurological deterioration over 2 to 6 months, which ultimately mandated radical surgery (gross total removal). At the time of the secondary procedure, one patient had become paraplegic, and another was essentially triplegic, having only trace movement in the involved extremities. Three patients were paraparetic but able to walk with assistance, and two patients were able to contract all muscle groups in the lower extremities against gravity but not against resistance. Four of the patients had impaired bowel and bladder function.

Six patients were treated primarily with radical surgery. All of these patients had relatively minor neurological dysfunction involving one upper extremity or both lower extremities.

Focal Astrocytoma. All five patients with focal astrocytomas underwent radical tumor excision. Two patients were treated primarily with biopsy and radiation therapy. In both cases, there was progressive neurological deterioration culminating, 4 to 6 months following the primary procedure, in paraplegia in one patient and quadriplegia in the other.

F. Epstein and N. Epstein
Spinal cord astrocytomas of childhood

Surgical Technique

In the first patients operated on for what was presumed to be a holocord neoplasm, a total laminectomy was carried out from C2–L1. It was subsequently recognized that since much of the cord widening was secondary to associated rostral and caudal cysts, it was only necessary to perform a limited laminectomy over the solid component of the neoplasm, and this procedure was employed in 10 patients. The surgical technique used for tumor excision was identical in both groups of patients.

After midline myelotomy over the limits of the neoplasm, pial traction sutures were placed to facilitate exposure and thereby obviate the need for spinal cord retraction. A plane of cleavage between the tumor and adjacent normal neural structures was found only rarely, so tumor excision was initiated within the neoplasm. With the aid of the Cavitron ultrasonic surgical aspirator* (CUSA) and high magnification, it was consistently possible to identify a glia-tumor interface and accomplish a gross total surgical excision of the neoplasm (Fig. 2). In 13 patients with holocord widening, the longitudinal extent of the solid tumor varied from four to 16 segments. In one 16-year-old girl, the solid component of the neoplasm was only 1.5 cm in length. In 11 of the 14 patients with holocord astrocytoma, there were rostral and caudal cysts which were easily drained once the solid component of the neoplasm had been excised. In the two patients in whom the solid tumor extended into the conus, there was only a rostral cyst and, in another, there was only edema rostral and caudal to a five-

---

* Cavitron ultrasonic surgical aspirator manufactured by Cavitron Lasersonics, Division of Cavitron Corp., 88 Hamilton Avenue, Stamford, Connecticut.

Results of Treatment

In all patients, there was a direct relationship between the preoperative neurological deficit and the degree of neurological recovery. Patients with mild to moderate dysfunction fared well, while those with advanced disability made incomplete recoveries.

Four patients who had previously received 4500 to 5000 rads of spinal axis radiation therapy had transient cerebrospinal fluid fistulae secondary to poor wound healing. On the basis of this experience, wounds in previously irradiated patients are now closed with muscle transposition flaps to obviate this occurrence.

Holocord Astrocytoma. Eleven of the 14 patients with holocord astrocytoma eventually made an excellent postoperative recovery, regaining full neurological function. Patients with minor preoperative deficits recovered quickly, while those with more pronounced

---
dysfunction required many weeks to accomplish the same result. They have been followed for 6 to 24 months, and have no clinical evidence of recurrence. Three patients with profound preoperative deficits (triplegia, paraplegia) did not improve, although there has been no further deterioration. Although some transient increase in the preoperative neurological dysfunction occurred in six patients, no patient was made permanently worse by the procedure.

**Focal Astrocytoma.** The paraplegic and severely quadriparietic patients with focal astrocytoma did not improve as a result of surgery, while the three patients with evolving neurological dysfunction have enjoyed a complete or nearly complete neurological recovery. They are fully functional, with no evidence of recurrence 12 to 24 months postoperatively.

**Discussion**

It is clear that holocord astrocytoma forms a large subgroup among spinal cord astrocytomas. In fact, it is apparently a more common occurrence than the more limited neoplasm, and accounted for 75% of our entire series of radically treated astrocytomas (14 among 19 patients). Although described in occasional case reports, its prevalence has not previously been recognized. This is a result of the earlier tendency not to carry out a complete neurodiagnostic study when there was a complete block due to an intramedullary mass.5,4

It has been a consistent observation that, despite the widened spinal cord extending from the cervicomedullary junction to the conus, the presenting symptomatico is relatively minor. In this series of 14 patients with holocord tumors, the primary complaint was a weak arm or mildly weak legs, and often associated pain somewhere along the spinal axis. The signs and symptoms were relatively trivial vis-à-vis the apparently diffuse nature of the pathological process. It is because of this apparent paradox that a temporizing and conservative surgical approach is most often employed, and that ultimate total neurological disability is accepted as a strong possibility. There are, however, a number of observations relating to gross and microscopic pathology which suggest that these neoplasms are, in fact, grossly excisable and perhaps curable in many cases.

A nearly uniform occurrence in 14 holocord tumors was a finding of enormous cysts rostral and caudal to the solid component of the neoplasm. These occupied the central part of the cord, contained deeply xanthochromic fluid, and were lined by glistening white matter. The centrally located cysts contributed to pain along the spinal axis, probably as a result of distension of the dural tube, but apparently did not cause primary impairment of neurological function, which consistently had a direct relationship only to the segments of spinal cord which were spanned by solid neoplasm. Evolving weakness in an upper extremity correlated with a cervical neoplasm, while primary paraparesis occurred with thoracic tumors. In the two patients presenting with bowel and bladder dysfunction, solid tumor extended into the conus. In three other patients with only a cyst in the conus, bowel and bladder dysfunction occurred only in association with advanced neurological disability. From a surgical perspective, it was only necessary to expose and remove the solid component of the neoplasm and drain the associated cysts to obtain an excellent clinical result. Therefore, a limited laminectomy over the solid portion of the neoplasm was sufficient. The segments of bone to be removed were defined by the combination of neurological deficit, eroded pedicles, scalloped vertebral bodies, and the area of maximal spinal cord widening.

In three patients the tumor was soft and easily suctioned, and there was a very clear plane of cleavage. In 16 of the 19 patients, the tumor was firm, did not aspirate with suction and bipolar cautery, and did not have a clear plane of cleavage between the neoplasm and adjacent neural tissue. It was in this group of children that the CUSA was an indispensable adjunct to radical tumor excision. With this instrumentation and high magnification, tumor excision was started within the bulk of the neoplasm. In this way, the tumor was gradually removed from the “inside out” until the consistently present glia-tumor interface was identified. Frequently, additional tumor fragments were identified along this interface, and these were vaporized with the bipolar cautery. It would seem likely that use of the laser for vaporizing residual tumor fragments will be a more satisfactory technique. Clearly, the combination of the CUSA and laser instrumentation will facilitate surgical excision of these neoplasms.

The microscopic pathology of this group of neoplasms was remarkably uniform, since they were all low-grade astrocytomas. In three patients there were foci of calcification, and in six there were abundant numbers of Rosenthal fibers. As a group, these tumors are pathologically indistinguishable from pilocytic astrocytomas of the cerebellum. Although gross total removal was accomplished in all patients, there can be little doubt that residual fragments often remain. Hopefully, these may remain dormant or involute, as has been described with the “sister” tumors of the cerebellum.5,7

The presence of the enormous rostral and caudal cysts suggests that these are congenital fluid-producing neoplasms which have their inception during gestation, at which time the fluid dissipates and the spinal cord in the region of least resistance, namely, the central canal. This anatomic relationship may explain the paucity of neurological dysfunction related to the segments of the spinal cord that contain cysts. One might also speculate that, in some cases, the classical symptoms of syringomyelia may in fact be a late manifestation of such a cyst, in which the tumor either has involuted or is not anatomically obvious. Perhaps the centrally located cyst may grad-
Spinal cord astrocytomas of childhood

Usually increase in size over many years and compress the surrounding cord. In this regard, it is of interest that one patient had an exceedingly small neoplasm (1.5 cm) associated with huge cysts.

On the basis of this entire series, in which 10 of 19 patients were treated primarily with only biopsy and radiation therapy, it is clear that progressive neurological dysfunction is a strong possibility in the absence of radical tumor excision. Furthermore, there is no convincing evidence that low-grade astrocytomas are sensitive to radiation therapy, and there is abundant evidence that irradiation has a most deleterious effect on young and developing nervous tissues. Although there may be occasional cases in which an apparent favorable long-term response to radiation therapy is recorded, this may be more related to the indolent biology of the tumor than the response to treatment; in any event, such a response is probably only found in relatively focal tumors without the associated cysts. Therefore, it is the opinion of the authors that these tumors should be viewed as surgically excisable lesions, and supplementary radiation therapy should be withheld unless there is future recurrence.

Hydrocephalus was manifested by headaches and lethargy in four patients. In one case, it antedated the overt symptoms of spinal cord dysfunction, while in the other three, it occurred within 2 to 4 weeks of the primary surgical procedure. It did not occur in any case of focal thoracic tumor, and seemed to be associated with arachnoiditis at the cervicomedullary junction.8

Children who have undergone extensive laminectomy and also have denervation of paraspinal muscles, as a result of both the neoplasm and of muscle retraction, are likely to develop severe spinal deformities as they pass through periods of rapid growth. For this reason, they are treated with body braces which are to be worn during waking hours for the first few years after surgery. With this regime, deformities have not yet occurred in this patient group.

Conclusions

This series of patients suggests the following conclusions:

1. In the pediatric age group, holocord widening is a relatively common occurrence with astrocytoma.

2. Conservative therapy is commonly associated with continued neurological deterioration and eventual total disability.

3. A constellation of findings, which include a relatively minor neurological deficit in association with a diffusely widened spinal canal and spinal cord, is indicative of a very slowly expanding congenital neoplasm. This tumor is quite analogous to cystic astrocytoma of the cerebellum, which may have an excellent long-term prognosis if surgical excision can be accomplished.

4. The magnitude of the preoperative neurological deficit has a direct correlation with the degree of postoperative recovery. Complete or nearly complete dysfunction does not improve, while mild to moderate dysfunction has potential for complete recovery.

References


Manuscript received April 12, 1982.

Address reprint requests to: Fred Epstein, M.D., Department of Neurosurgery, New York University Medical Center, 550 First Avenue, New York, New York 10016.