Surgical management of holocord intramedullary spinal cord astrocytomas in children

Report of three cases

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This report describes the therapeutic approach to extensive multisegmental spinal cord astrocytomas in three pediatric patients. It is concluded that radical resection (gross total removal) is compatible with neurological recovery.

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

INTRAMEDULLARY spinal cord gliomas account for 4% of central nervous system tumors in the pediatric age group. Of these, 60% are astrocytomas and 20% to 30% ependymomas. These tumors are invariably multisegmental, and may rarely extend from the cervicomedullary junction to the conus (holocord). We have recently encountered three holocord astrocytomas, and have come to the conclusion that a gross total removal is technically feasible and compatible with neurological recovery. The therapeutic approach used is the subject of this report.

Case Reports

Case 1

This 15-year-old boy was admitted to our institution with the chief complaint of progressive weakness in the right upper extremity of 2 months' duration. The weakness was first noted after he had been thrown from a horse, and was interpreted as a brachial plexus injury. Only a few days before admission, weakness of the right lower extremity evolved and intraspinal pathology was suspected.

Neurological examination disclosed a right hemiparesis as well as some impairment of sensation on the left side of the body. Lumbar myelography revealed a widened spinal cord from the cervicomedullary junction to the conus, but there was no complete obstruction to the flow of the contrast material. In the hope of finding a cyst, a cervical laminectomy and posterior myelotomy from C-5 to C-7 was performed. A partially calcified intramedullary mass was noted, and biopsies were obtained. As there was no obvious cyst within the limits of the myelotomy, a diagnosis of a solid glioma was made, and the dura was left open.

Postoperatively, the neurological deficit was unchanged, and the patient was given a course of spinal axis radiotherapy. Despite this, his neurological status continued to deteriorate, and 2 months later he was severely triparetic, with bowel and bladder dysfunction as well as steroid-dependent. A follow-up myelogram disclosed that the tumor had increased in size, and there was now a complete block in the midthoracic region. Thoracic laminectomy with a posterior myelotomy from T-4 to T-7 was performed. Once again, an extensive tumor was encountered and a radical excision was carried out within the margins of the myelotomy. Postoperatively, there was some transient improvement in lower extremity weakness as well as in bowel and bladder function. However, 2 months later (4 months after the initial presentation) he once again became severely triparetic. At this time it was elected to attempt a radical excision of the entire tumor.

At staged surgical procedures, the lower thoracolumbar spinal cord was exposed, and tumor radically excised through a posterior myelotomy. It was noted
that the lower three segments of the spinal cord were filled by a cyst which was adjacent to the tumor. At cervical myelotomy performed 2 weeks later, the tumor was again radically excised, and a cyst was noted to extend from the cervicomedullary junction to C-4. At this last surgical procedure, a Cavitron ultrasonic device* was used to remove the nonsuckable, firm, and partially calcified cervical tumor.

Postoperatively, the patient's bowel and bladder function have returned to normal. His triparetesis has improved slowly, but he remains dependent, and there is a dense hypesthesia over the entire trunk and left lower extremity. Pathological examination disclosed that the tumor was a low-grade astrocytoma.

Case 2

This 5-year-old boy was admitted to our care with the chief complaint of progressive paraparesis and impairment of bowel and bladder function of 2 months' duration. This patient had undergone a T4-6 myelotomy for an intramedullary astrocytoma 5 months earlier. Only a small amount of tissue was removed for biopsy, and a dural decompression was performed, after which local radiotherapy was administered. Although he did well for a period of 6 weeks, the preoperative paraparesis recurred, and he was unable to walk unassisted at the time of admission. In addition, there was severe pain in the neck as well as neck stiffness. Sensory examination was normal. Myelography via lumbar and cervical injection disclosed a holocord tumor extending from the cervicomedullary junction to the conus. There was a complete block at T4-5.

On the basis of our experience with the previous case, we believed that there was no substitute for surgical excision, and no advantage to staged procedures. For this reason, a laminectomy was performed from C-3 to L-2 (inclusive of the previous thoracic laminectomy). When the dura was opened, the entire spinal cord was noted to be discolored and distended with tumor (Fig. 1). A myelotomy was performed from C-4 to just above the conus, and, using the Cavitron ultrasonic device, gross total removal of a very firm, partially calcified tumor was accomplished. There were large intramedullary cysts extending from the cervicomedullary junction to C-4, and from T-9 to the conus.

Because the cord was relaxed, the dura was closed and, postoperatively, there was immediate improvement in the paraparesis. In addition, the patient's bowel and bladder function became normal, and over a period of several weeks he became ambulatory. Sensory examination was also normal. He has continued to improve over the few months since surgery, and only manifests mild spasticity in the lower extremities which does not interfere with normal function. Pathological examination disclosed that the tumor was a low-grade astrocytoma.

Case 3

This 4-year-old boy was admitted to our institution with the chief complaint of recurring weakness in the right upper extremity. This child had been operated on 4 months earlier for an intramedullary cervical astrocytoma. At that time, only a small amount of tissue was obtained for biopsy, and, following a dural decompression, a full course of spinal axis radiotherapy was administered. Although transient improvement was noted, he once again developed severe weakness of the right upper extremity. The neurological examination was otherwise normal, and there was normal bowel and bladder function.

Myelography disclosed a holocord tumor extending from the cervicomedullary junction to the conus. There was a complete block at T2-3. The C1-2 puncture entered a cyst within the spinal cord which extended from the foramen magnum to C-4 (Fig. 2).

Laminectomy was performed from C-3 to L-1, and the spinal cord was noted to be markedly swollen and had a yellowish discoloration. A myelotomy was performed from C-4 to T-4. At the caudalmost portion of

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*Ultrasonic device manufactured by Cavitron Lasersonics, Division of Cavitron Corp., Stamford, Connecticut.
Excision of spinal cord intramedullary tumors

FIG. 2. Case 3. Contrast substance injected directly into a cyst via a C1–2 puncture delineates extension from the foramen magnum to C-4 (cyst inadvertently punctured).

the myelotomy, a cyst was entered which extended to the conus. Gross total removal of solid tumor which extended from T-4 to C-4 was accomplished with a Cavitron ultrasonic device, and a large cyst cavity was entered at the rostralmost portion of the tumor which extended to the foramen magnum (Fig. 3). The dura was closed as the spinal cord was decompressed.

Postoperatively, the right upper extremity recovered normal function over a period of 48 hours. There was no motor or sensory deficit, and bowel and bladder function remained normal. This patient is now asymptomatic; he has no demonstrable neurological deficit and is totally independent. Pathological examination disclosed that the tumor was a low-grade astrocytoma.

Discussion

Although intramedullary astrocytomas are frequently extensive, extending over 19 to 20 segments, “holocord” tumors occupying an average of six spinal segments have been described. Proper surgical management of these extensive intramedullary astrocytomas is contingent on the neurodiagnostic delineation of the multisegmental nature of the lesion as well as the recognition that these tumors are not only operable but perhaps curable.

It is possible that these extensive neoplasms may be more common than previously recognized. Earlier myelographic examinations were often terminated in the presence of a complete block. The present series of patients demonstrates that a C1–2 myelogram is a mandatory adjunct to definitive investigation, as in only one of these cases would the rostral extent of the lesion have been recognized in the absence of this study.

The experience with this small group of patients, in whom deterioration occurred despite dural decompression and radiation therapy, suggests that radical surgical excision may be the only therapy that offers the potential of future neurological recovery in patients with such extensive intramedullary neoplasms.

Elsberg first recognized that intramedullary spinal cord neoplasms were surgically excisable. Since his classic description of a two-stage procedure, his observations have been confirmed by many neurosurgeons. The operating microscope and evolving instrumentation have since obviated the necessity for more than one surgical procedure. Most contemporary neurosurgeons would agree that surgical exploration is mandatory in any child with a progressively symptomatic intramedullary mass. We do not believe that radiation therapy should be offered without a definitive tissue diagnosis. Furthermore, in most situations, surgery should be directed toward radical removal of tumor.

FIG. 3. Case 3. The gross total tumor excision complete. Note region of cyst at caudal and rostral margins of tumor (arrows).
Although these surgical principles have been generally accepted for relatively well localized intramedullary tumors,\textsuperscript{3,4,6-10,14,20} they have not been applied to the multisegmental or holocord astrocytomas.\textsuperscript{5,8} Obviously, the risk of producing a permanent neurological deficit as a result of an extensive dissection within the entirety of the spinal cord has been regarded as unacceptable.

It is widely recognized that ependymomas are relatively easy to remove in toto\textsuperscript{9,9,10,14,20} as a result of their well demarcated cleavage plane; however, astrocytomas present additional technical problems. They are often very firm, and frequently contain calcium deposits. There is no obvious cleavage plane, and removal must commence from within the tumor. Suctioning and blunt dissection are relatively inefficient and cause considerable movement and traction of normal cord elements. Whereas an extensive posterior myelotomy does not cause a neurological deficit, it is likely that excessive manipulation of the entire spinal cord might result in permanent injury. We have found the Cavitron ultrasonic surgical aspirator very helpful for excision of holocord astrocytomas.\textsuperscript{7} This is a vibrating suction device that fragments and aspirates tissue within a 1- to 2-mm radius of the tip. The vibration is imperceptible, although the ultrasonic frequency is 23 kHz. It emulsifies the firmest tissue and permits atraumatic aspiration of all visible tumor. There is no associated movement of adjacent normal spinal cord, and no need for conventional suction or blunt tumor dissection. For these reasons, a gross total tumor removal is more readily accomplished. Despite the extensive tumor resection and massive residual intramedullary tumor cavity, neurological function has not been compromised by this procedure in this small series of patients.

It seems likely that a holocord neoplasm is, in most situations, not entirely solid tumor. In all of our cases, there were large cysts at the most caudal and rostral portions of the neoplasm. The cysts were white and smooth-walled, contained deeply xanthochromic fluid, and were within spinal segments that were subserving normal neurological function. Solid tumor spanned the anatomical segments that related to the neurological deficit. Therefore, it was apparent that there was no direct relationship between the holocord widening and discernible neurological dysfunction.

The dramatic clinical improvement in two of these patients suggests that the tumor is not diffusely infiltrating, but seems to compress local intramedullary structures, and it is on this basis that internal decompression improved neurological function. Obviously, orthopedic follow-up review is essential inasmuch as an evolving spinal deformity is a near certainty in a growing child who has had an extensive laminectomy as well as an intramedullary spinal cord tumor.

Conclusions

This report is intended to document the fact that radical surgical excision of extensive, multisegmental intramedullary astrocytomas may be accomplished with little or no neurological morbidity. It would be premature to make any claims concerning a permanent cure, which can only be based on neurological stability for a period of many years.

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