Adult intramedullary spinal cord ependymomas: the result of surgery in 38 patients

FRED J. EISEN, M.D., JEAN-PIERRE FARMER, M.D., AND DIANA FREED

Division of Pediatric Neurosurgery, New York University Medical Center, New York, New York, and The Montreal Children's Hospital, Montreal, Quebec, Canada

Thirty-eight patients underwent surgery for an intramedullary spinal cord ependymoma. In 37 patients, postoperative magnetic resonance imaging confirmed that the tumor was totally removed. The morbidity of surgery was directly related to the preoperative neurological condition. Patients who were normal or nearly normal preoperatively were rarely worse after surgery, and those who had significant disability preoperatively were at greatest risk of being more impaired after surgery. There has been no tumor recurrence in any patient after a mean follow-up period of 24 months, and radiation therapy has not been employed as a surgical adjunct.

Key Words • ependymoma • spinal cord neoplasm • intramedullary tumor

Intramedullary spinal cord ependymomas have long been recognized as neoplasms that may be totally removed, and long-term remission or cure may be anticipated in many patients. However, most of the relevant literature antedates the magnetic resonance (MR) imaging era and there are few large contemporary series that provide an opportunity to analyze the clinical presentation and neurodiagnostic studies as well as the surgical results. Over the past 6 years, the senior author (F.J.E.) has operated on 38 patients with spinal cord ependymomas, which has afforded an opportunity to further understand the biology of this often indolent neoplasm.

Clinical Material and Methods

Patient Population

Between 1985 and 1991, 38 patients were surgically treated for spinal ependymoma at New York University Medical Center. There were 26 men and 12 women. The age at diagnosis ranged from 19 to 66 years (mean 37 years). The initial symptoms were almost uniformly sensory. In 33 patients, the predominant sensory presentation was a dysesthetic syndrome which varied in location depending on the tumor site; cervical tumors were associated with dysesthesias involving the arm or hand, high thoracic lesions with chest wall dysesthesias, and low thoracic lesions with dysesthesias involving the lower extremities. Only five patients presented primarily with motor dysfunction. Of the 38 tumors, 12 were exclusively cervical, 12 bridged cervical and thoracic segments, 10 were exclusively thoracic, and four involved the conus.

Preoperative Course

The disease onset was usually insidious and the duration of symptoms prior to diagnosis was variable but usually lasted several months. In 32 patients for whom the information was available, the average duration of symptoms prior to diagnosis was 14 months. Thirty of these patients had only subjective sensory symptoms (dysesthesias), which was the reason for the delay in diagnosis.

Magnetic Resonance Imaging

All patients were evaluated preoperatively with an MR image, which is virtually diagnostic of an intraspinal ependymoma. Thirty-five of the 38 lesions enhanced homogeneously and “brightly” with gadolinium and had sharply defined rostral and caudal margins. In cross-sectional images, because ependymomas originate from the region of the central canal, they expanded the cord symmetrically and, in most cases, the surrounding cord was reduced to a few millimeters thick (Figs. 1, 2, and 3).

Clinical Grade

Case records were reviewed retrospectively and patients were assigned a preoperative clinical grade based on the grading system of McCormick, et al. (Table 1). Eighteen patients were classified as Grade 1, 11 as
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Fig. 1. Preoperative magnetic resonance images, sagittal view, in an ependymoma patient. *Left*: Unenhanced image showing diffuse widening of the cervical cord from C-4 to C-7 (arrows). *Right*: Gadolinium-enhanced image demonstrating that the tumor is sharply demarcated (arrows) and less extensive than might have been suspected from the unenhanced image (left). Note that the homogeneous enhancement is typical of an ependymoma.

Table 1

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<th>Grade</th>
<th>Definition</th>
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<tr>
<td>I</td>
<td>Neurologically normal; mild focal deficit not significantly affecting function of involved limb; mild spasticity or reflex abnormality; normal gait</td>
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<tr>
<td>II</td>
<td>Presence of sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty; severe pain or dysesthetic syndrome impairing patient's quality of life; still functions and ambulates independently</td>
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<tr>
<td>III</td>
<td>More severe neurological deficit; requires cane/brace for ambulation or significant bilateral upper-extremity impairment; may or may not function independently</td>
</tr>
<tr>
<td>IV</td>
<td>Severe deficit; requires wheelchair or cane/brace with bilateral upper-extremity impairment; usually not independent</td>
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Grade II, eight as Grade III, and one as Grade IV. Immediate and late postoperative evaluations consisting of a telephone questionnaire were used to assign the same grading system to the patients following definitive treatment.

Previous Surgery

Fourteen patients had undergone surgery elsewhere prior to definitive treatment at New York University Medical Center. In nine cases "subtotal" resection was described, while in five patients only a biopsy was attempted. In these latter patients, a microscopic diagnosis of astrocytoma was made as a result of obtaining tissue only from the periphery of the tumor. Nine patients underwent local radiation therapy to the tumor. All of these patients had progressive neurological deterioration over the 6 to 18 months prior to the definitive surgical procedure. Fascial transfer closures described elsewhere* were used for previously irradiated cases.

Surgical Techniques

Following induction of anesthesia and removal of posterior bone elements by osteoplastic laminotomy or laminectomy, all patients underwent intraoperative ultrasonography prior to dural opening in order to ascertain that the rostral-caudal dimension of the laminectomy was of sufficient length to expose the entire tumor. Ependymomas were invariably echogenic neoplasms and the rostral and caudal poles of the tumor were well
defined by the ultrasound study (Fig. 4 left). On cross-sectional films, the tumors were almost identical in appearance to the same view of the MR images, expanding the cord symmetrically and filling at least two-thirds of the cross-sectional diameter (Fig. 4 right).

Following dural opening, a midline myelotomy was made over the length of the tumor with a CO2 laser or fine blade (Fig. 5A). Pia traction sutures then opened the myelotomy and exposed the tumor along its entirety (Fig. 5B and C). In the absence of a rostral or caudal cyst, the tumor resection was initiated in the middle of the neoplasm, which was the most voluminous area and therefore the region where there was the minimum threat to surrounding normal neural structures. The most important technical point is that there was no effort to carry out an en bloc resection. The "centrum" of the tumor was removed with the Cavitron ultrasonic aspirator and the lateral margins were gradually "folded in," establishing the interface between tumor and cord (Fig. 5D, E, and F). Ependymomas were invariably adherent to the cord in the region of the anterior median raphe where small branches of the anterior spinal artery enter the neoplasm. These were coagulated and divided, and the residual fragments of tumor were removed "piecemeal" in most cases. In some, adhesions and small feeding vessels caused the tumor to appear to be infiltrating. However, once these vessels were meticulously lysed, a very well-developed interface between the tumor and the neural tissue invariably appeared.

The resection of ependymomas associated with rostral or caudal cysts was similar but was initiated at the cyst-tumor junction where the interface between tumor and normal neural tissue was more easily identified. In all cases, intraoperative ultrasonography was utilized at the end of the tumor removal to confirm completeness of resection (Fig. 6).

Follow-Up Study

All 38 patients underwent immediate postoperative and follow-up MR imaging evaluations. The latest recorded MR image was used as an assessment of the surgical result vis-à-vis tumor removal. In 35 patients, the follow-up period varied from 6 to 60 months (mean 24 months) and three patients were lost to follow-up review. No patient died during the follow-up period.

Results

Postoperative Neurological Status

Immediate postoperative examination disclosed that 14 of 18 Grade I patients remained at the Grade I level; three deteriorated to the Grade II level and one to the Grade III level. Six months after surgery, three of these four patients returned to a Grade I functional level, and one remained at the Grade II functional level. In summary, 17 of the 18 Grade I patients continued to be at the Grade I level at follow-up evaluation.

Six of the 11 patients in the Grade II category preoperatively had preserved Grade II status immediately postoperatively, while four deteriorated to Grade III status and one to Grade IV status. At longer follow-up examination, one patient who was at the Grade III level immediately postoperatively returned to Grade II status and the Grade IV patient improved to Grade III status. Therefore, at long-term follow-up review, four of 11 patients preoperatively in Grade II had lost one grade of function.

There were eight preoperative Grade III patients in this series. At the immediate postoperative evaluation, five patients had deteriorated to a Grade IV status, two patients remained at Grade III, and only one patient improved to a Grade II status. At last follow-up examination, two patients had improved from a preoperative Grade III to a Grade II status (one from an immediate postoperative Grade IV), four patients remained paraplegic (Grade IV), and one patient remained at the Grade III functional level; the remaining Grade III patient, who was unchanged immediately postoperatively, was lost to follow-up monitoring. Therefore, of eight Grade III patients, two improved, four deteriorated, and two remained unchanged.

The only preoperative paraplegic patient (Grade IV) has remained so postoperatively.

Postoperative Magnetic Resonance Imaging

All patients underwent immediate and late (range 5 months to 36 months, mean 14 months) MR imaging. One patient with an extensive cervicomедullary tumor had a residual tumor fragment, while no evidence of residual or recurrent tumor was disclosed on the other 37 imaging studies.

Postoperative Dysesthesias

Postoperative dysesthesias occurred in all patients. Invariably the intensity of the pain was directly related to how severe it had been preoperatively. Patients in whom dysesthesia was the most prominent complaint prior to surgery were at greater risk for it to be even more severe after surgery (usually in the identical anatomical location). In addition, all patients described diffuse dysesthesias caudal to the spinal segment from which the tumor was removed. In most patients, the
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Fig. 5. Diagrammatic illustrations of the surgical technique for the removal of an ependymoma. A: Myelotomy is carried out with a fine blade or laser, staying within the median raphe to avoid damage to the dorsal columns. B: After completion of the myelotomy, the middle of the posterior surface of the tumor is exposed. However, there is not adequate lateral visualization to safely remove the tumor without excessive retraction of normal neural tissues. C: Pia traction sutures increase the lateral exposure of the tumor as the posterior columns are separated from it. D: The Cavitron ultrasonic aspirator is utilized to "debulk" the central tumor. E: The residual tumor extending circumferentially around the cavity is gently separated from the lateral neural tissues and "folded" into the cavity. F: The remaining tumor is separated from the anterior surface where perforating vessels (vascular supply from the anterior spinal artery) are divided. The residual tumor fragments are then removed "piecemeal."

latter symptoms resolved within a few days to a few months. Patients with intense dysesthesias in the same area as preoperatively were at greatest risk for persistent and severe subjective sensory dysfunction.

Discussion

Ependymoma vs. Astrocytoma

This series of patients strongly suggests that intramedullary ependymomas are a group of spinal cord tumors with a well-defined clinical presentation and MR imaging appearance, and favorable outcome after total removal. Over the past 10 years, the senior author (F.J.E.) has operated on 300 spinal cord astrocytomas and this experience has made it clear that these neoplasms are quite different from ependymomas in many aspects. These differences will be addressed in the context of the overall discussion.

Clinical Presentation

It was striking that patients with ependymoma not only had a relatively long antecedent history, but that the sensory phenomena manifested by dysesthesias were present as the first symptom in the great majority. We have speculated that this is in some way related to the origin of the tumor being around the central canal.

Fig. 6. Ultrasound film disclosing the residual cavity after tumor removal.
which seems to expand symmetrically and circumferentially and, in all likelihood, compresses or interrupts crossing spinothalamic tracts. Interestingly, in most cases the dysesthias were present without objective evidence of sensory dysfunction. This was quite different from patients with spinal cord astrocytomas in whom the presenting problem has usually been spinal axis pain followed by progressive motor dysfunction; if sensory phenomena were part of the original symptomatic presentation, they were usually manifest as loss of sensation (numbness) but only rarely as dysesthias.

We have also noted that in ependymoma patients in whom treatment was withheld until there was significant motor dysfunction, the tumor had invariably thinned the surrounding spinal cord to a few millimeters in width. In other words, when there was significant motor dysfunction, the tumor was of such enormous proportions and the spinal cord so attenuated that surgical dissection was extremely hazardous as a result of the vulnerability of the thinned out neural elements to manipulation. This is the reason why Group II and Group III patients were at greatest risk to be injured by the surgical procedure. It also re-emphasizes our most important point: surgery is least hazardous when the neurological deficit is minimal or nonexistent.

**Magnetic Resonance Imaging**

The great majority of ependymomas have a "typical" appearance: homogeneous enhancement of the tumor with sharply defined rostral and caudal poles. About 30% of the tumors were associated with a rostral or caudal cyst similar to that noted for spinal cord astrocytomas. However, we have consistently observed that, unlike astrocytomas, the gadolinium enhancement in an ependymoma clearly demarcates what is in reality the rostral and caudal poles of the tumor. In the area where occasional astrocytomas may enhance quite homogeneously, the expansion of the cord rostral or caudal to that area is usually related directly to the tumor in that region. With an ependymoma, however, contrast enhancement clearly demarcates the upper and lower poles, and the small amount of expansion that is invariably present rostral or caudal to this area is secondary to edema, not to the tumor. In addition, when viewed on cross-sectional images, the ependymoma is also "classical" in appearance. The gadolinium enhancement is obvious but, most important, the cord is expanded symmetrically unlike the astrocytoma in which it is usually asymmetric ("lumpy"). This is because the ependymoma grows from the region of the central canal and is not an infiltrating neoplasm. The astrocytoma may originate anywhere in the cord; for this reason, the growth pattern is very different from that of an ependymoma.

**Surgical Treatment**

It has been a consistent observation that intraoperative ultrasonography is almost diagnostic of an ependymoma. This is because the tumors have been, without exception, echogenic and, as on MR imaging, very sharply defined both rostrally and caudally. In addition, what is demonstrated on the intraoperative cross-sect}

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Junction is carried out. We have utilized different techniques, such as fine blade or laser, in an effort to carry out anatraumatic myelotomy but have not been able to correlate a more favorable outcome with the surgical technique. Therefore, patients are informed of this in some detail preoperatively to prevent the sense of alarm in patients with loss of proprioception postoperatively. When proprioception in the lower extremities is interrupted, the patient will often believe that the involved extremity is paralyzed insomuch as he/she does not perceive movement. In any event, proprioception often improves over a period of a few weeks or months and physical therapy is very effective in overcoming any handicap resulting from it. Four patients in our series have permanent impairment of proprioception, all four extremities in one, both upper extremities in one, and the lower extremities only in two.

Radiation Therapy

No patient operated on primarily has received adjuvant therapy. In addition, no patient in whom the postoperative MR image disclosed that the tumor had been totally removed has suffered a recurrence of the tumor (mean follow-up period 24 months). It is the opinion of the authors that ependymoma is a tumor that may be permanently cured with surgery, and adjuvant therapy is not appropriate unless there is future recurrence or some specific circumstance that may make it mandatory.

Conclusions

In 37 of 38 patients in this series, a gross total removal of an intramedullary spinal cord ependymoma was accomplished and confirmed by postoperative MR imaging. There has been no clinical or radiographic evidence of recurrence after a mean follow-up period of 24 months in any patient in whom total removal was accomplished. The morbidity of the surgical procedure is directly related to the preoperative neurological status. Patients with little or no disability preoperatively are at little risk to sustain an injury, while patients with more advanced neurological dysfunction have a greater risk of being impaired by surgery.

On the basis of this overall experience, we have reached the following conclusions: 1) gross total removal of a spinal cord ependymoma may be accomplished in the overwhelming majority of patients; 2) ependymomas do not infiltrate adjacent neural tissues and a surgical interface is invariably present; 3) it is important that surgery be carried out immediately after the diagnosis and when the patient is in good neurological condition, as the hazards increase in relation to neurological dysfunction; and 4) radiation therapy is not a necessary adjunct following gross total removal of the tumor.

References


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Address for Dr. Farmer: The Montreal Children's Hospital, Montreal, Quebec, Canada.
Address reprint requests to: Fred J. Epstein, M.D., Division of Pediatric Neurosurgery, New York University Medical Center, 550 First Avenue, New York, New York 10016.