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Intramedullary ependymoma of the spinal cord

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✓ A consecutive series of 23 patients underwent operative removal of an intramedullary spinal cord ependymoma between January, 1976, and September, 1988. Thirteen women and 10 men between the age of 19 and 70 years experienced symptoms for a mean of 34 months preceding initial diagnosis. Eight patients had undergone treatment prior to tumor recurrence and referral. Mild neurological deficits were present in 22 patients on initial examination. The location of the tumors was predominantly cervical or cervicothoracic. Radiological evaluation revealed a wide spinal cord in all cases. Magnetic resonance (MR) imaging was the single most important radiological procedure. At operation, a complete removal was achieved in all patients. No patient received postoperative radiation therapy. Histological examination revealed a benign ependymoma in all cases.

The follow-up period ranged from 6 to 159 months (mean 62 months) with seven patients followed for a minimum of 10 years after surgery. Fourteen patients underwent postoperative MR imaging at intervals ranging from 8 months to 10 years postoperatively. No patient has been lost to follow-up review and there were no deaths. No patient showed definite clinical or radiological evidence of tumor recurrence during the follow-up period. Recent neurological evaluation revealed functional improvement from initial preoperative clinical status in eight patients, no significant change in 12 patients, and deterioration in three patients. The data support the belief that long-term disease-free control of intramedullary spinal ependymomas with acceptable morbidity may be achieved utilizing microsurgical removal alone.

KEY WORDS • **spinal neoplasm** • **ependymoma** • **radiation therapy** • **outcome** • **intramedullary tumor**

EPENDYMOMAS account for only 4% to 6% of primary central nervous system tumors, but about one-third arise within the spinal canal where they represent the most common intramedullary neoplasm in the adult population.[4,35] Although ependymomas are unencapsulated glial neoplasms, the vast majority of spinal ependymomas exist in a histologically benign form with little infiltrative potential, and exhibit slow biological growth evidenced by a prolonged average symptom duration preceding diagnosis, preservation of neurological function in patients with extreme spinal cord compression, and characteristically long survival times following various forms of treatment.[24,25,36] These features of spinal ependymomas, coupled with their relatively low incidence, render the evaluation of treatment difficult to assess.

It has been the authors' opinion that long-term disease-free control of intramedullary ependymomas with optimum preservation of neurological function is best achieved with gross total removal at initial operation.[15,28,38] This retrospective study was undertaken to critically assess this bias through the careful evaluation and follow-up monitoring of a consecutive series of patients with spinal ependymoma treated with microsurgical removal alone.

Clinical Material and Methods

Clinical Methods

A consecutive series of 23 patients underwent operative resection of an intramedullary spinal ependymoma between January, 1976, and September, 1988. Patients with filum terminale ependymomas were not included in this study. The location of the tumors was: cervical or cervicothoracic (17 cases), thoracic (two cases), or conus medullaris (four cases). Two conus tumors (Cases 6 and 10) had both intra- and extramedullary components. At operation, a grossly complete tumor removal was believed to be accomplished in each patient. No patient received radiation therapy following

TABLE 1

Clinical/functional classification scheme

Grade	Definition
I	neurologically normal; mild focal deficit not significantly affecting function of involved limb; mild spasticity or reflex abnormality; normal gait
II	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
III	more severe neurological deficit; requires cane/brace for ambulation or significant bilateral upper extremity impairment; may or may not function independently
IV	severe deficit; requires wheelchair or cane/brace with bilateral upper extremity impairment; usually not independent

definitive surgery but six patients had undergone surgery and radiation therapy prior to clinical tumor recurrence and referral to this institution.

Preoperative clinical evaluation was carefully performed and each patient was assigned a clinical grade according to the clinical/functional classification scheme listed in Table 1. Neurological function was assessed during the immediate postoperative period and at approximately 3 months and 1 year following surgery. Long-term follow-up monitoring, consisting of office visits and completion of a detailed questionnaire, has recently been performed. Fourteen patients have had postoperative magnetic resonance (MR) imaging at intervals ranging from 8 months to 10 years following operation. There have been no deaths and no patient has been lost to follow-up review. The follow-up period ranged from 6 to 159 months (mean 62 months), with seven patients followed for a minimum of 10 years after surgery.

Clinical Summary

The patient population consisted of 13 women and 10 men ranging in age from 19 to 70 years (mean 43 years) at the time of initial diagnosis (Table 2). Pain was the initial complaint in 15 patients (65%) and was present an average of 16 months before the onset of objective neurological symptoms. The pain was localized in the back or neck in 12 patients and usually corresponded to the level of the tumor. Interscapular or shoulder pain was also common in patients with cervical or cervicothoracic tumors. Two patients had associated pain radiating to the arm or leg, and one patient presented with isolated leg pain. Six patients described a sensory disturbance, consisting of numbness, paresthesiae, or dysesthesiae, as the initial symptom. In four patients the sensory change began distally

TABLE 2

Clinical summary of 23 patients with intramedullary ependymoma

Case No.	Age (yrs),* Sex	Initial Symptom	Duration of Symptoms†	Cord Enlargement	Tumor Level	Clinical/Functional Grade					Net Grade Change
						Preop‡	Immediate Postop	3 Mos Postop	1 Yr Postop	Recent Follow-Up§	
1	50, M	paresthesiae, hand	23 mos	C1–T1	C2–7	I (III)	IV	III	III	III (159 mos)	0
2	19, M	pain, back	16 mos	C2–T1	C3–T1	I (III)	III	III	III	IV (159 mos)	−1
3	35, F	pain, neck	86 mos	C2–7	C3–7	I (III)	III	II	II	II (156 mos)	+1
4	46, M	pain, neck & arm	38 mos	C2–7	C4–7	I	II	I	I	I (146 mos)	0
5	48, F	pain, leg	6 mos	T12–L1	T12–L1	I	I	I	I	I (135 mos)	0
6	24, M	pain, back & leg	14 mos	T8–L2	T10–L2	II (IV)	IV	IV	IV	IV (125 mos)	0
7	41, F	pain, neck	13 mos	C2–T2	C3–T1	I	II	II	II	I (121 mos)	0
8	36, M	pain, neck	96 mos	C1–T3	C3–7	I (IV)	IV	IV	IV	IV (89 mos)	0
9	43, F	pain, neck	24 mos	C2–T3	C5–7	III	III	II	I	I (66 mos)	+2
10	33, F	pain, back	92 mos	T11–L2	T11–L2	I (II)	III	III	II	II (63 mos)	0
11	32, F	pain, back	35 mos	C4–T1	C7	II	II	I	I	I (59 mos)	+1
12	37, M	pain, back	72 mos	C4–L2	T8–L2	III	IV	III	III	III (54 mos)	0
13	32, M	gait difficulty	7 mos	C4–T9	C5–T2	III	III	III	II	II (43 mos)	+1
14	49, F	pain, neck & back	17 mos	C3–T1	C5–7	II	II	II	I	I (42 mos)	+1
15	64, F	numbness, hands	30 mos	C2–5	C4–5	I	II	II	II	II (39 mos)	−1
16	70, F	numbness, hands	37 mos	C3–7	C5–7	II	III	II	II	II (36 mos)	0
17	52, F	pain, neck	46 mos	C4–T1	C4–7	I	II	I	I	I (33 mos)	0
18	42, M	paresthesiae, arms	30 mos	C3–T1	C5–6	I (II)	II	II	II	I (23 mos)	+1
19	48, M	weakness, arm	14 mos	C3–7	C4–7	I (III)	IV	II	II	II (21 mos)	+2
20	47, M	paresthesiae, legs	17 mos	C5–T8	T3–4	I	II	I	I	I (20 mos)	0
21	61, F	pain, arm	26 mos	C4–T1	C5–7	III	III	III	II	II (14 mos)	+1
22	54, F	dysesthesiae, feet	26 mos	C7–T3	T1–3	I	II	II	—	II (8 mos)	−1
23	35, F	pain, back	10 mos	C3–T1	C4–7	I	II	I	—	I (6 mos)	0

* Age at time of initial diagnosis.

† Duration of symptoms prior to initial diagnosis.

‡ Cases 1, 2, 3, 6, 8, 10, 18, and 19 had previous operation with or without radiation therapy. Grade in parentheses indicates clinical status prior to most recent treatment.

§ Length of follow-up since completion of most recent treatment is given in parentheses.

and later progressed to involve the proximal limb. Weakness was the initial complaint in two patients. The duration of symptoms preceding initial diagnosis ranged from 6 to 96 months (mean 34 months, median 26 months).

Objective neurological deficit was present in 22 patients at the time of initial diagnosis. The deficit was usually mild with 21 patients (91%) demonstrating independent ambulation and 19 patients (83%) classified as having clinical Grade I or II. Eight patients had received treatment prior to tumor recurrence and referral. The specific therapy, initial preoperative grade, time to clinical recurrence, time from recurrence to recent treatment, and preoperative clinical grade prior to most recent treatment are listed in Table 3.

The pattern and progression of neurological deficits were variable and related to tumor location. The cervical or cervicothoracic tumors (17 cases) tended initially to produce isolated deficit of the upper extremities of a predominant sensory (eight cases) or motor (four cases) nature. Two patients complained of pain and noted no neurological deficit on admission interview. Five patients, with a mean symptom duration of 22 months, had neurological deficits confined to one or both arms. Five patients, with a mean symptom duration of 39 months, denied lower-extremity symptoms but had mild leg deficit on admission examination. Seven patients complained of lower-extremity weakness and/or numbness as the first noticeable deficit (one patient), concurrently with the onset of upper-extremity deficit (one patient), or occurring an average of 23 months after the onset of upper-extremity deficits (five patients).

Atrophy of one or both hands was particularly common with tumors of the cervical spinal cord and was noted in eight patients. Proximal muscle atrophy was present in two patients. Only one patient noted bowel or bladder difficulty. Six patients had predominantly unilateral neurological deficits, with two patients demonstrating an incomplete Brown-Séquard syndrome.

Both patients with thoracic ependymomas (Cases 20 and 22) presented with bilateral leg numbness. Minimal leg weakness or stiffness occurred 14 months and 2 years after the onset of numbness, respectively. One patient noted bowel and bladder difficulty. One patient with a T3–4 ependymoma (Case 20) had dissociated sensory loss in one arm secondary to a rostral cyst extending to the C-6 level.

All four patients with conus ependymomas presented with pain as the initial complaint. Neurological examination in one patient was normal. In two patients progressive weakness of one or both legs was the predominant neurological complaint. Unilateral leg numbness followed by leg weakness was noted by the remaining patient. One patient also complained of bilateral hand numbness and had dissociated sensory loss in the arms. A rostral cyst extending to the C-4 level was present. Three patients noted bowel or bladder difficulty.

The clinical course was slowly progressive in all patients. Several patients, however, noticed an accelerated progression in the 6 months to 1 year preceding diagnosis. Two patients had an acute worsening of neurological deficit due to an intratumoral hemorrhage confirmed at surgery.

Radiological Investigation

Preoperative radiological evaluation consisted of plain films in 12 patients, myelogram in seven patients, myelogram followed by computerized tomography (CT) in eight patients, and MR imaging in 13 patients. Metrizamide or iohexal myelography followed by delayed CT was performed in five patients. Plain films revealed mild scoliosis of the upper or mid thoracic spine in three patients. An enlarged spinal canal with vertebral body scalloping, medial pedicle erosion, and thinning of the laminae was noted in two patients.

Spinal cord enlargement was identified in all patients, ranging in length from two to 18 spinal segments (mean seven). Magnetic resonance imaging was particularly

TABLE 3

Previous treatment in eight patients in this series

Case No.	1st Operation & Diagnosis*	Radiation Therapy	Time to Recurrence†	Time to 2nd Op‡	Clinical/Functional Grade at:		
					1st Op	2nd Op	Recent Follow-Up
1	laminectomy (tumor)	5000 cGy	13 mos	11 mos	I	III	III
2	laminectomy, cyst aspiration (tumor)	—	6 mos	61 mos	I	III	IV
3	laminectomy (tumor)	5000 cGy	37 mos	13 mos	I	III	II
6	laminectomy, biopsy (astrocytoma)	5000 cGy	13 mos	296 mos	II	IV	IV
8	laminectomy, cyst aspiration (tumor)	4500 cGy	15 mos	27 mos	I	IV	IV
10	laminectomy, subtotal removal (ependymoma)	3000 cGy	26 mos	16 mos	I	II	II
18	laminectomy (spondylosis)	—	3 mos	10 mos	I	III	I
19	laminectomy, biopsy (astrocytoma)	2500 cGy	30 mos	14 mos	II	III	II

* The diagnosis is given in parentheses. No tissue was obtained in Cases 1, 2, 3, 8, and 18. Diagnosis of tumor was made based on observations of the operating surgeons. In Cases 6, 10, and 19 the diagnosis was made from tissue obtained at initial operation.

† Elapsed time from completion of radiation therapy or following surgery (Cases 2 and 8) to clinical tumor recurrence.

‡ Elapsed time from clinical recurrence to reoperation.

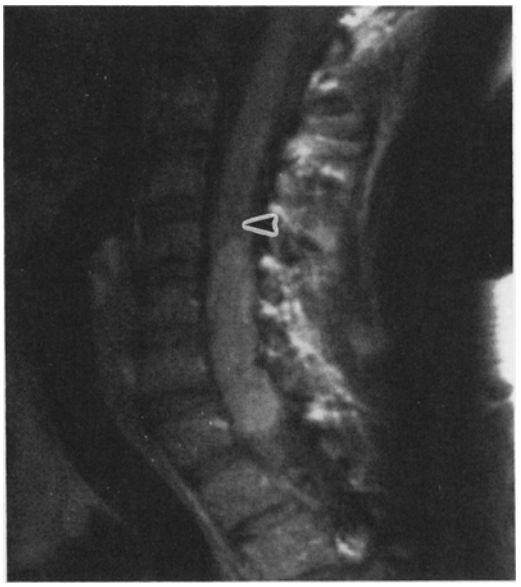

FIG. 1. T_1-weighted gadolinium-enhanced sagittal magnetic resonance image demonstrating a large intramedullary ependymoma of the cervical cord with a small cyst *(arrow)* capping the rostral pole of the tumor.

useful in defining the level of the tumor and in identifying associated spinal cord edema or cysts (Fig. 1). Delayed CT scanning following water-soluble myelography did not reveal contrast uptake into the spinal cord in any patient.

Surgical Technique

Following routine induction of anesthesia and oropharyngeal intubation, the patient is placed prone and positioned to maintain a perpendicular orientation between the entire extent of the tumor and the surgeon. The prone position is preferred because it comfortably allows assistance of a second surgeon, which facilitates tumor removal. With the patient in the prone position, movement of the spinal cord secondary to respiratory excursions does not present a significant technical problem since the enlarged spinal cord is minimally affected by cerebrospinal fluid (CSF) pulsations, and the use of pial sutures further restricts spinal cord movement. Perioperative steroids and anti-staphylococcal antibiotics are administered. Somatosensory evoked potential monitoring during tumor removal has been of limited practical value and is employed primarily for investigative purposes.

A standard midline incision and subperiosteal reflection of the paraspinal muscles is performed. The laminectomy should include the medial facet joints only in the mid and lower thoracic spine to minimize the risk of delayed cervical instability, particularly of the lower cervical spine. In recent cases, laminoplasties have allowed anatomical restoration of the bone canal, which may further reduce the incidence of delayed instability.

A midline dural incision is made leaving the arachnoid intact to prevent inadvertent injury to the under-

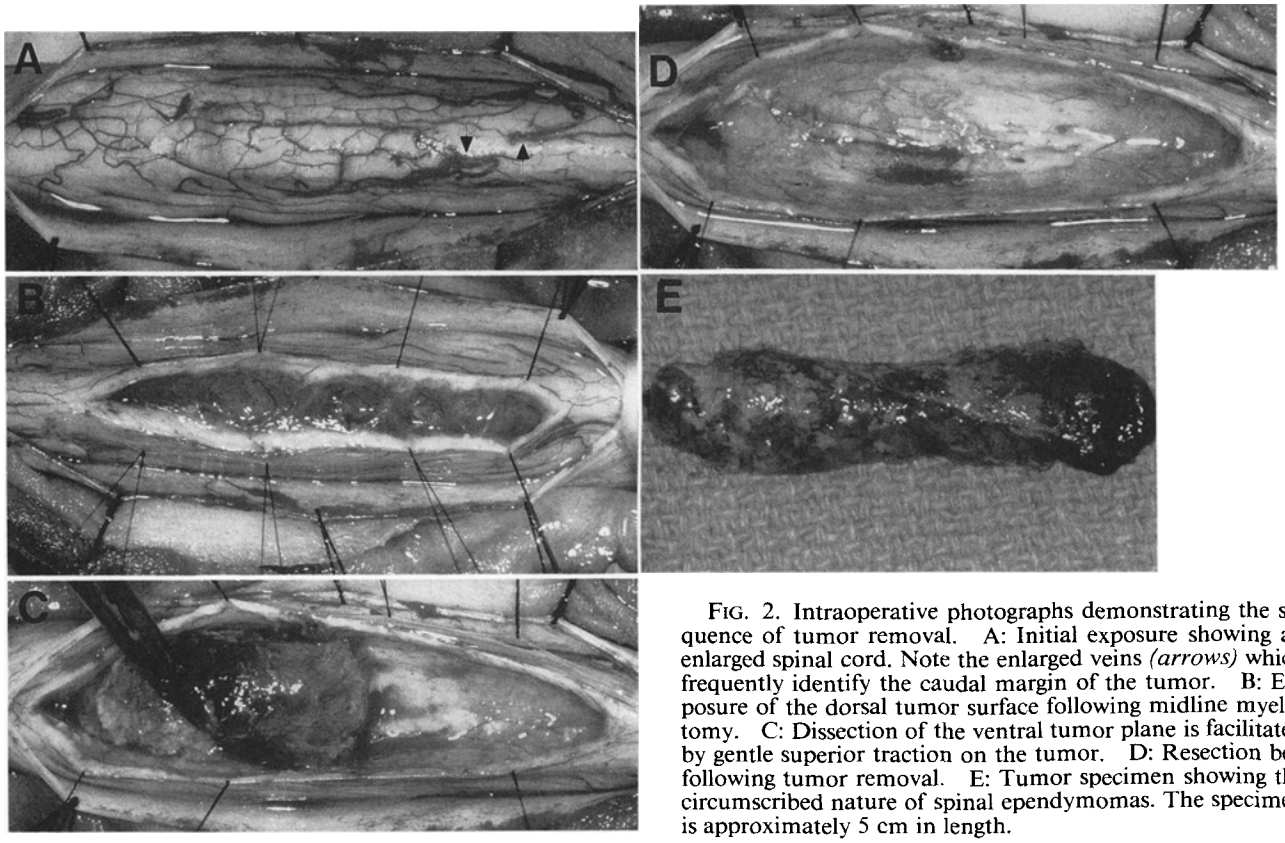

FIG. 2. Intraoperative photographs demonstrating the sequence of tumor removal. A: Initial exposure showing an enlarged spinal cord. Note the enlarged veins *(arrows)* which frequently identify the caudal margin of the tumor. B: Exposure of the dorsal tumor surface following midline myelotomy. C: Dissection of the ventral tumor plane is facilitated by gentle superior traction on the tumor. D: Resection bed following tumor removal. E: Tumor specimen showing the circumscribed nature of spinal ependymomas. The specimen is approximately 5 cm in length.

lying vessels. The spinal cord is inspected and the dorsal midline is grossly estimated by noting the dorsal root entry zones bilaterally. The arachnoid is then opened sharply with the aid of the microscope. A relatively constant fold, the septum posterius, which attaches to the pia in the posterior median septum, further assists delineation of the dorsal cord midline. The midline pia is cauterized and cut beginning in the region of maximum cord swelling and carried rostrally and caudally as far as necessary to expose the polar regions of the tumor (Fig. 2). Small crossing pial vessels are cauterized and cut, but larger longitudinally oriented vessels are dissected if possible, and swept laterally.

The myelotomy is deepened by gentle spreading of the posterior columns using microforceps or dissectors. Identification of a longitudinal array of penetrating pial vessels on the medial surface of each posterior column assures that the midline orientation has been maintained. The tumor will usually be encountered at a depth of about 2 mm. Gentle but constant superior and lateral traction on each dorsal hemicord is achieved with fine pial sutures.

It is at this point that determination of the gross and histological characteristics of the tumor is made. If no clear demarcation exists between tumor and spinal cord and if frozen section identifies the tumor as an astrocytoma, it is the authors' opinion that aggressive removal is not warranted. If, however, there appears to be a plane between the tumor and spinal cord, regardless of tumor histology, operative removal should continue. The myelotomy is lengthened and deepened to fully expose the entire rostrocaudal extent of the tumor and should actually continue a few millimeters above and below the tumor margins, allowing greater lateral retraction and visibility while minimizing tension on the spinal cord.

Ependymomas appear as a soft red or grayish-purple mass with a variable number of vessels crossing the tumor surface. Although these tumors are somewhat friable, ependymomas are sharply circumscribed and gentle blunt manipulation will not violate the tumor surface. The rostral tumor is frequently rounded and often projects into a cyst which aids dissection. The caudal pole is usually more tapered since inferior cysts are less common, but there is often a tough fibrous connection between the caudal tumor pole and the central canal (Fig. 3A).

The dorsal and lateral tumor margins are established by gentle traction on the tumor against the countertraction provided by the pial sutures. Spreading with microforceps parallel to the long axis of the tumor easily develops the dissection plane owing to the differences in texture and consistency between the tumor surface and the surrounding gliotic margin of the spinal cord. Feeding vessels and more fibrous attachments are cauterized and divided close to the tumor. The decision to

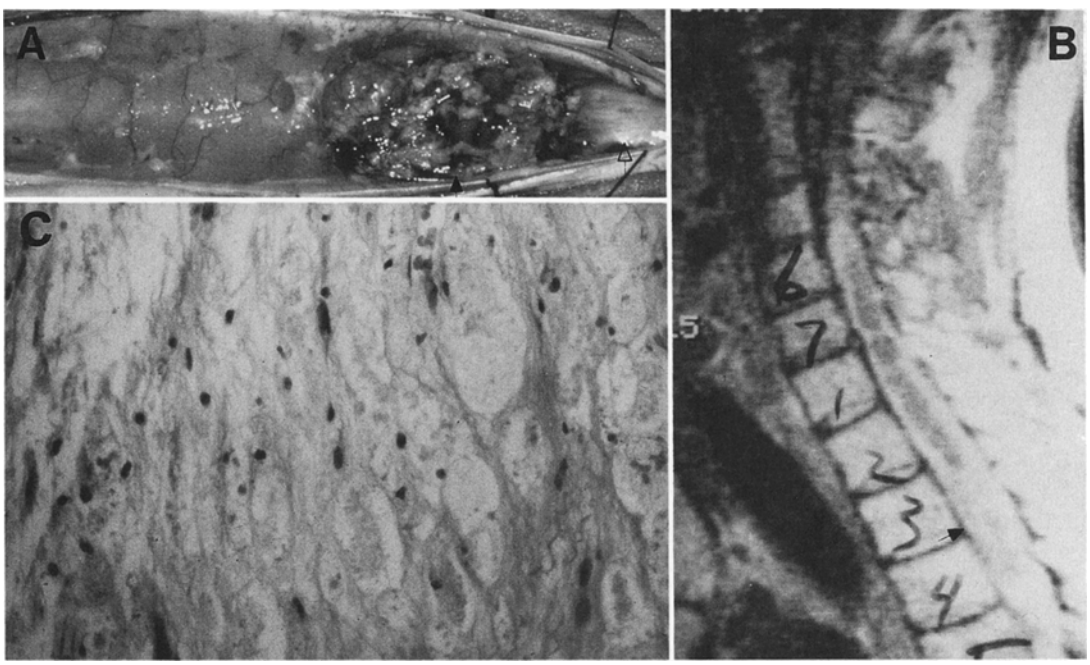

FIG. 3. A: Operative photograph following myelotomy demonstrating a well-circumscribed tumor *(closed arrow)*. Note the fibrous attachment at the inferior tumor pole *(open arrow)*. A swollen edematous-appearing segment of spinal cord rostral to the tumor can be appreciated. B: Preoperative T_2-weighted sagittal magnetic resonance image showing focal cord enlargement at the T3–4 level *(arrow)* with what appears to be a rostral cyst extending to the C-6 level. C: Photomicrograph from biopsy material of the swollen cord tissue demonstrating mild gliosis and swollen degenerated axons. H & E, × 70.

internally debulk the tumor is usually made once the dorsal half of the tumor is exposed. It is generally preferable to maintain the tumor intact during the entire dissection, but in some cases the bulk of the tumor hinders exact visualization of the dissection plane, requiring a prohibitive amount of spinal cord retraction. In these cases the dorsal tumor surface is incised and internal decompression of the tumor is performed with the ultrasonic aspirator. Too much internal tumor removal, however, may cause fragmentation of the tumor surface and obscuration of the correct dissection plane, resulting in an undesirable piecemeal removal.

Dissection of the ventral plane is the most difficult aspect of tumor removal for several reasons. First, less advantage can be taken of the difference in texture between the tumor and spinal cord because the pial sutures do not transmit effective countertraction to the ventral cord/tumor interface. Thus, the tumor margin appears less distinct and requires sharp dissection techniques. Second, the anterior median fissure extends almost to the central canal and the tumor is frequently in close approximation to the anterior spinal artery and branches. Finally, the major vascular supply to the tumor is derived from penetrating branches of the anterior spinal artery. Nevertheless, with superior traction on the tumor directed perpendicular to the long axis of the spinal cord, the tumor can be separated from the anterior spinal vessels which are easily identified, systematically cauterized, and divided.

Following tumor removal, the resection bed is inspected. Any bleeding is controlled with warm saline irrigation or application of oxidized cotton. The pial traction sutures are removed and the cord assumes its normal position. No attempt is made to reapproximate the dorsal hemicords with pial sutures. The dura is closed primarily, if possible, since dural substitute patch grafts increase the risk of postoperative CSF leak. The remainder of the wound is closed in a standard fashion.

Tumor Characteristics

Intramedullary ependymomas are circumscribed lesions of modest vascularity. The tumors are usually soft and somewhat friable but a firmer, more nodular appearance is occasionally encountered, particularly in patients previously treated by operation followed by radiation therapy. Evidence of prior intratumoral hemorrhage was noted in four patients. An unusual edematous-appearing portion of spinal cord above the tumor was encountered in three patients (Fig. 3A). This tissue, while having a cystic appearance on MR imaging (Fig. 3B), was solid and gelatinous with histological characteristics of swollen degenerated axons, edematous white matter, and gliosis (Fig. 3C).

Cysts were identified in 19 patients and were usually related to the rostral pole of the tumor (13 patients). Five patients had both rostral and caudal cysts. One patient had a cyst capping the caudal tumor pole only. Xanthochromic fluid filled these cysts and cytological examination of this fluid did not reveal tumor cells in any patient. Histological examination from biopsy of the cyst wall routinely revealed gliosis and Rosenthal fibers. In no case has tumor been identified in the lining of the cyst, and biopsy of the cyst wall is no longer performed.

Histological examination of the tumor revealed a benign ependymoma in all cases. Tumor margins appeared smooth and regular, with a variable amount of surrounding compressed gliotic white matter included in the surgical specimen (Fig. 4). Gross or microscopic evidence of previous hemorrhage was noted in seven specimens. Necrosis was identified in nine tumors.

The tumors were classified into the following histological types: cellular (14 cases), tanacytic (three cases), epithelial (two cases), myxopapillary (one case), and mixed (three cases: two cellular and subependymoma and one cellular and epithelial).

Complications

Four patients had complications related to the surgical wound. Three of these patients developed a CSF leak requiring wound revision. A ventriculoperitoneal shunt was required in one patient with a persistent CSF leak. One patient developed a wound abscess and was treated with drainage, secondary closure, and antibiotics. Three of these patients had undergone previous operation and radiation therapy. One patient developed meningitis which was successfully treated with a 14-day course of antibiotics.

There were no operative deaths. Neurological morbidity is discussed under outcome.

Outcome

Neurological evaluation in the immediate postoperative period revealed either worsening of an existing deficit or onset of a new deficit (usually related to the posterior column) in 20 patients. This resulted in a deterioration of the clinical grade in 12 patients. In

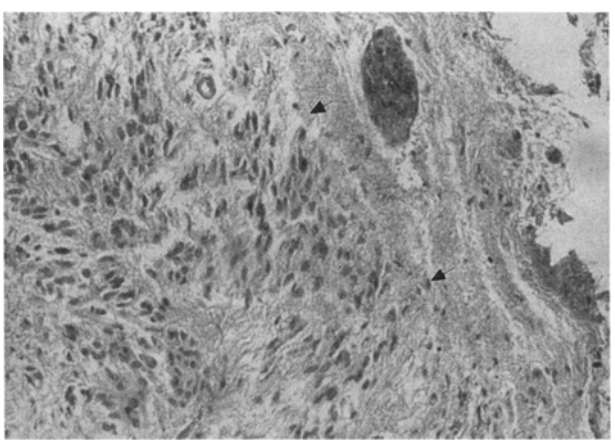

FIG. 4. Photomicrograph of tumor specimen showing clear demarcation of the tumor margin *(arrows)* from a small amount of surrounding neural tissue. H & E, × 50.

general, however, significant recovery from the operative deficit was achieved within 3 months postoperatively. Only two patients (Cases 15 and 22) experienced a persistent net worsening in clinical grade caused by surgery. Both of these patients were in Grade I preoperatively. One patient, neurologically unchanged following surgery, developed a severe dysesthetic pain syndrome. The other patient, currently in Grade II at 8 months postoperatively, continues to show slow improvement.

Recovery from a preoperative deficit resulting in an ultimate improvement in clinical grade was seen in eight patients. The timing of recovery was variable, with relief of spasticity occurring early (< 6 months) postoperatively while recovery of lower motor-neuron and sensory deficits tended to appear later (> 6 months) with continued improvement noted up to 2 years postoperatively in two patients. Bowel and bladder function improved in all patients with cervical, cervicothoracic, or thoracic tumors but in only one patient with a conus tumor. Two patients with conus tumors had permanent worsening of bowel and bladder function following surgery.

One patient (Case 2), with severe neurological deficit prior to definitive tumor removal, experienced delayed neurological deterioration 10 years postoperatively. Magnetic resonance imaging demonstrated a huge cervical intramedullary cyst just above the level of previous resection. Operative exploration revealed a dense adhesive arachnoiditis in addition to the cyst, but no gross tumor recurrence was observed. No improvement in neurological function occurred following cyst drainage.

A frequent phenomenon following surgery was the appearance of a dysesthetic syndrome. While subjective complaints of numbness, out of proportion to objective sensory deficit, were also common and usually of little functional consequence, dysesthetic complaints were perceived to be particularly annoying and often debilitating to the patient. The severity of the dysesthesiae and the patient's emotional response were quite variable, ranging from intermittent and clinically insignificant "pins and needles" sensations to persistent distressing, often causalgia-like, complaints of "itching," "crawling," or "burning" dysesthesiae. This syndrome generally appeared early in the postoperative period and frequently seemed to follow a posterior column or radicular distribution. Patients with preoperative dysesthesiae seemed especially prone to develop this syndrome postoperatively, but no other predictive factors could be identified. The dysesthesiae were refractory to various forms of medical therapy but were usually self-limited, with resolution or amelioration of complaints several months following surgery. Only two patients had significant dysesthetic complaints at 1 year postoperatively. The physiological basis of this syndrome is unclear but the anatomical distribution of the dysesthesiae suggests injury to the posterior columns or dorsal root entry zone.

With the exception of one patient (Case 2), no patient has shown any clinical evidence of tumor recurrence during the follow-up period ranging from 6 to 159 months (mean 62 months). A comparison of the most recent preoperative clinical grade and the current follow-up evaluation revealed an improvement in clinical grade in eight patients, no significant change in 12 patients, and deterioration in three patients.

Follow-up MR imaging has now been performed on 14 patients. Spinal cord atrophy extending varying degrees above and below the level of tumor resection, dorsal tethering of the spinal cord to the posterior dura with a large ventral subarachnoid space, and diminution but persistence of cysts were the most common findings (Fig. 5A and B). No patient showed definitive evidence of tumor recurrence, although one patient had faint gadolinium uptake in the walls of the resection bed on MR images obtained 6 months postoperatively (Fig. 5C). Repeated MR imaging over 3 years has shown no change. Although residual tumor cannot be ruled out, it seems more likely that this appearance represents a persistent postoperative change.

Discussion

In 1911, Elsberg and Beer[7] reported the first successful removal of an intramedullary spinal cord tumor. Frazier[11] also commented on the potential for removal of encapsulated intramedullary neoplasms; however, early attempts at removal of intrinsic spinal cord tumors were associated with prohibitive operative morbidity and mortality which led even Elsberg[6] to adopt a more pessimistic attitude towards these tumors. For the next several decades, radiation therapy evolved as the major treatment modality for intramedullary tumors.[27,42] Surgery was employed primarily for diagnosis (frequently by visual inspection alone), cyst aspiration, and external decompression.[36,41,42]

Shortly after Bailey and Cushing[2,3] formally classified ependymomas as a distinct neoplastic entity, Kernohan, *et al.*,[20] described three histological types and later proposed further classification of ependymomas into four grades.[21] This grading system, similar to the grading system for astrocytomas, stressed a correlation between histological criteria for progressive anaplasia and clinical prognosis. This classification effectively inferred a close association between astrocytomas and ependymomas, serving to further limit the role of surgery in the treatment of ependymomas. Recent studies have suggested, however, a rather poor correlation between this grading system and prognosis.[19,24,29,30]

In 1954, Greenwood,[17] utilizing bipolar cautery and loupe magnification, reported six cases of complete removal of intramedullary ependymomas and, by 1963, he had accumulated nine cases treated by surgical removal alone.[16] The mortality rate was 22% but there was no tumor recurrence in the surviving patients during follow-up periods averaging 9 years. With further refinement in microsurgical techniques, it has become clear that the majority of intramedullary spinal epen-

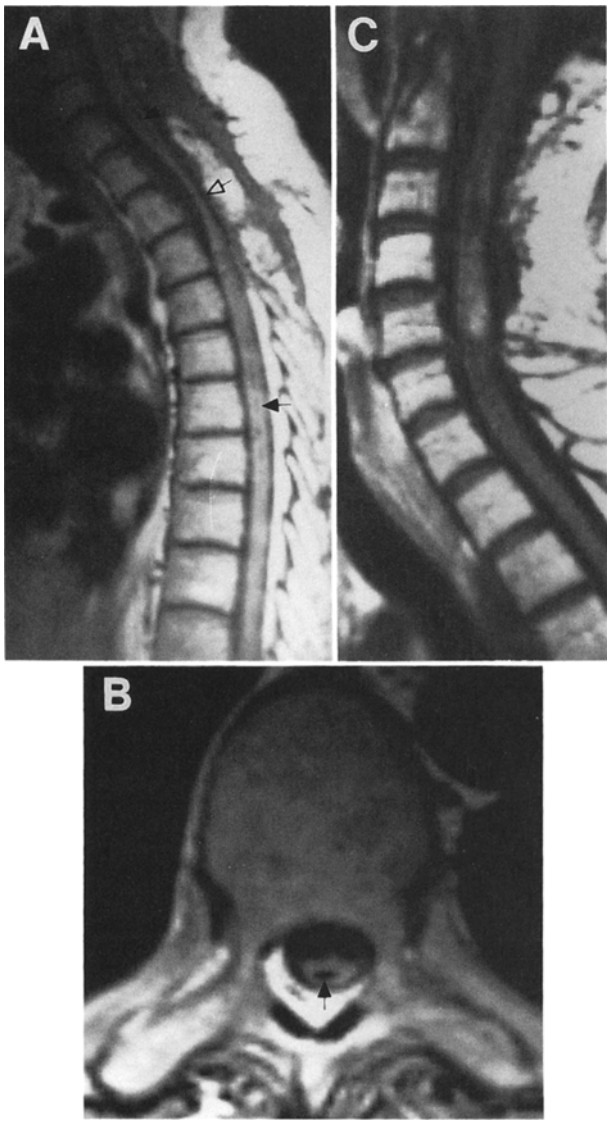

FIG. 5. A: Postoperative T_1-weighted sagittal magnetic resonance (MR) image showing spinal cord atrophy, most marked at the level of tumor resection *(open arrow)*. There is dorsal tethering of the spinal cord and a large ventral subarachnoid space at this level. Persistence of both rostral and caudal cysts can be seen *(closed arrows)*. B: T_1-weighted axial MR image performed three spinal segments below the level of tumor resection in the same patient better demonstrates the persistent intramedullary cyst *(arrow)*. C: Postoperative T_1-weighted gadolinium-enhanced MR image in another patient demonstrating an area of increased signal intensity at the level of previous tumor resection. This area has not changed on serial MR images performed over a 3-year period.

dymomas are amenable to complete surgical excision with acceptable morbidity and mortality and a low incidence of recurrence.[4,9,15,18,22,28,29,38]

Despite these results, recent reports demonstrate that radiation therapy in the management of spinal ependymomas remains prevalent.[4,13,14,19,23,31–33,35,37,40] Although radiation therapy may have a beneficial effect for incompletely removed cauda equina ependymomas,[5,34,36] studies advocating irradiation following subtotal or total removal of intramedullary ependymomas are difficult to evaluate due to small patient populations, limited follow-up periods, inadequate or no matched cohorts treated without radiation therapy, the use of survival as the only evaluation, inclusion of intracranial ependymomas in the study, and the limited or absent discussion of surgery and clinical follow-up data. Despite these limitations, the accumulated data in these series seem to suggest that radiation therapy may be beneficial in patients after subtotal removal of an intramedullary ependymoma. Because there will be eventual clinical tumor recurrence in a significant number of these patients up to 10 years following treatment, results of studies comparing gross total removal and subtotal removal followed by radiation therapy clearly indicate a more favorable outcome in patients treated with complete removal alone.[1,4,8,10,18,24,29,36,37] No benefit of radiation therapy following gross total removal of spinal ependymomas has ever been demonstrated and, considering the risks of radiation therapy,[23,26,39] its use to theoretically sterilize or control residual microscopic tumor following complete removal cannot be supported.

A number of aspects concerning the surgical management of intramedullary spinal cord tumors must be stressed. First, the surgeon should assume that the majority of all intramedullary tumors are benign and potentially resectable. Second, intraoperative judgment to the contrary should be made according to the gross rather than the histological tumor characteristics since many low-grade astrocytomas are also well circumscribed and amenable to resection. An inadequate myelotomy may fail to reveal a clear resection plane, and histological interpretation of small tumor specimens obtained through a limited myelotomy frequently results in an erroneous tissue diagnosis, as was seen in two patients in this series. This is particularly true for the tanacytic variant of ependymoma.[12] Conversely, if histological examination clearly demonstrates a benign ependymoma, then removal must continue. This is particularly important for large tumors which initially may seem infiltrative or severely compress the surrounding spinal cord into an almost unrecognizable configuration. It seems remarkable but this thin ribbon-like spinal cord may not only function reasonably well but have the capacity for a certain amount of recovery following tumor removal. Thus, even extremely large or extensive ependymomas should not deter the surgeon from complete removal. Finally, reoperation for recurrent tumor is extremely difficult, particularly if the patient has received previous radiation therapy. Dissection planes are obscured by gliosis, spinal cord plasticity is more tenuous, and complications of wound-healing are frequent. Considering as well that patients with recurrent tumor usually present at a lower clinical grade (Table 3) and that recovery is poorer in patients with

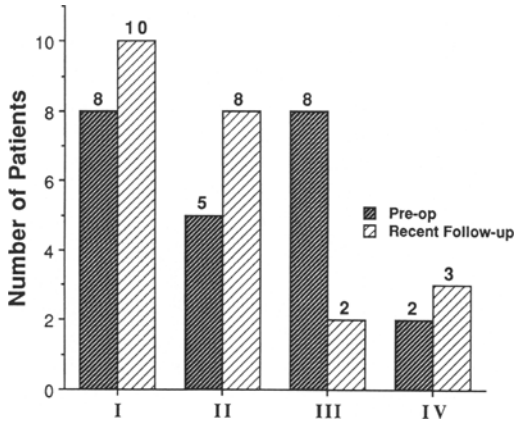

FIG. 6. Functional grade changes following the most recent surgery in this series.

more severe preoperative deficit (Fig. 6), it seems logical that complete removal be performed at the initial operation.

Intramedullary ependymomas occur predominantly in young and middle-aged adults with a small but significant incidence in the pediatric population.[5,40] Although they are benign noninfiltrative tumors their unencapsulated friable nature poses a continued risk of recurrence, regardless of therapy. The overall treatment goal in these patients should be long-term management aimed at optimal preservation of neurological function. The results of the present study indicate that this is best achieved by surgical removal. Currently, our patients are followed with serial MR imaging and periodic clinical evaluation. Should clinically symptomatic tumor recur, it is the authors' opinion that reoperation should be considered.

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