SPINAL TUMORS

CLASSIFICATION OF THE SPINAL TUMORS: /by World Health Organization/

SPINAL TUMORS

I EXTRADURAL TUMORS

A. Extravertebral tumors /outside from the bony elements of the vertebral column/

B. Intravertebral tumors /between the thecal sac and the vertebral column/

II INTRADURAL TUMORS /SPINAL LESION/

A. Extramedullar (2/3) variants

Nerve sheath tumor
Meningioma
Filum ependymoma
Miscellaneous / includes paraganglioma, metastases and granuloma/

B. Intramedullary (1/3) variants

Ependimoma - 45%
Astrocytoma - 40% / includes oligodendroglioma, ganglioglioma, neurocytoma, and subependymoma/
Hemangioblastoma - 5%
Miscellaneous - 10% /includes metastatic tumor, inclusion tumor (e.g., lipoma), inflammatory lesions (e.g., abscess, tuberculosis, sarcoid), and vascular lesions (e.g., cavernous malformation, aneurysm)/.

INCIDENCE AND PATHOLOGY

Tumors of the spine are anatomically classified by their relationship to the dura and spinal cord parenchyma. **Extradural tumors** arise outside of the thecal sac or from the bony elements of the vertebral column and are most often metastatic. **Intradural tumors** e intra- or extramedullar and account for roughly three-fourths of all spinal tumors. radural neoplasms account for approximately 10% of primary central nervous system NS) tumors in adults.

Extramedullary tumors consist about two-thirds of all spinal column neoplasms, histologically benign and well circumscribed. Meningioma, schwannoma and filum terminale ependymoma are the most common histopathological lesions in this location.

Intramedullary Tumors. Intramedullary spinal cord tumors account for approximately from 2% to 4% of all CNS neoplasms and about 20% to 25 % of spinal tumors. If primary and metastatic vertebral column and epidural tumors are excluded, intramedullary spinal half comprise about one-third of all intradural neoplasms in adults and about onehalf in the pediatric population. Over 90% of intramedullary spinal cord neoplasms have a glial origin.

GLIAL TUMORS
ASTROCYTOMA

About 3% of CNS astrocytomas arise within the spinal cord. These tumors can occur at any age but are most prevalent in the first three decades of life. They are also the mo common type of pediatric intramedullary spinal cord tumor, comprising about 90% of cases in patients under 10 years of age and about 60% in adolescents. Ependymoma become slightly more common than astrocytomas when the population is about 30 years old and increasingly predominate in the middle decades of life (McCormick P.C., Stein B.M., 1990). After the sixth decade of life, astrocytomas and ependymomas are encountered with about equal frequency. Nearly 60% of spinal astrocytomas occur in the cervical and cervicothoracic segments **(Fig.** 1).

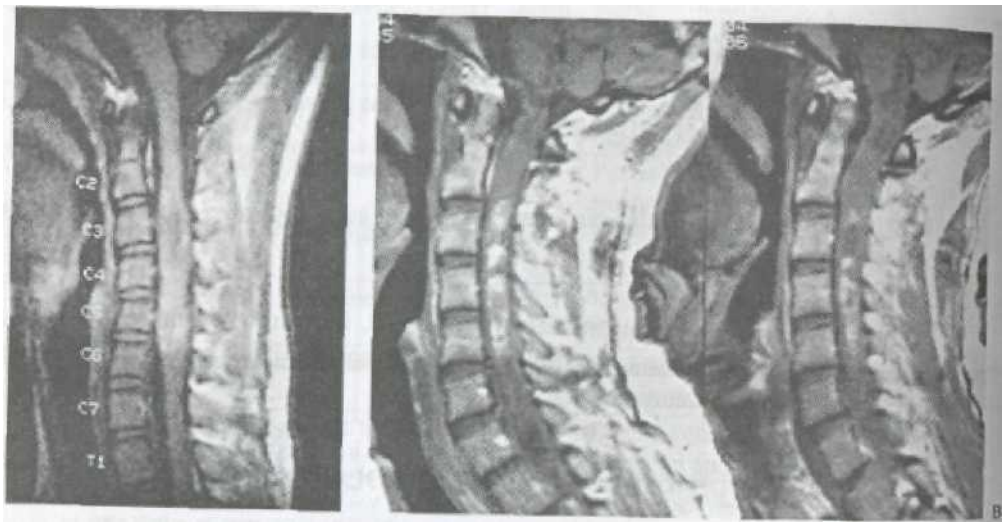

Fig. 1. Astrocytoma. (A) Tl-weighted sagittal magnetic resonance image (MRI) without intravenous contrast reveals fusiform widening of the spinal cord- (B) Sagittal and (C) axial MRI after intravenous contrast (from a different patients). The tumor displays patchy enhancement and an irregular interface with the surrounding spinal cord.

Spinal cord astrocytomas represent a heterogenous group with respect to histotogy,' gross characteristics, and natural history. These tumors include the low-grade fhrillary and pilocytic astrocytoma, ganglioglioma, malignant astrocytoma, and glioblastoma. Oligodendrogliomas also occur in the spinal cord but are very rare. About Q0% of pediatric astrocytic tumors are benign. Most of these are fibrillary astrocytomas. However, up to one-third represent juvenile pilocytic astrocytomas or gangliogliomas, hoth of which are associated with a particularly indolent natural history. Malignant astrocytomas and glioblastomas account for about 10% of intramedullary astrocytomas. These lesions are characterized by rapidly progressing clinical course, high incidence of cerebrospinal fluid (CSF) tumor dissemination, and poor survival. Fibrillary astrocytomas prevail in the adult. Juvenile pilocytic astrocytomas and gangliogliomas are rare and usually limited to early adulthood. The designation of a pilocytic astrocytoma in the adult

usually reflects an abundance of pilocytic features that occur as secondary structures in an otherwise typical fibrillary astrocytoma (Russel D.S., Rubinstein L.J., 1989). It is unclear whether these pilocytic features have an age-independent prognostic significance. About *25%* of adult astrocytomas are malignant (McCormick PC, Stein B.M., 1990).

EPENDYMOMA

Ependymomas are the most common intramedullary tumor in adults. They occur throughout life but are most frequent in the middle adult years. Men and women are equally affected. Although the spinal cord and filum terminale account for only 3% of the CNS by weight, nearly half of all CNS ependymomas originate within the spinal canal. The cervical region is the most common level of true intramedullary occurrence; however, 40% of intradural ependymomas arise from the filum **(Fig.** 2). For anatomical and surgical reasons, these lesions will be considered with extramedullary tumors.

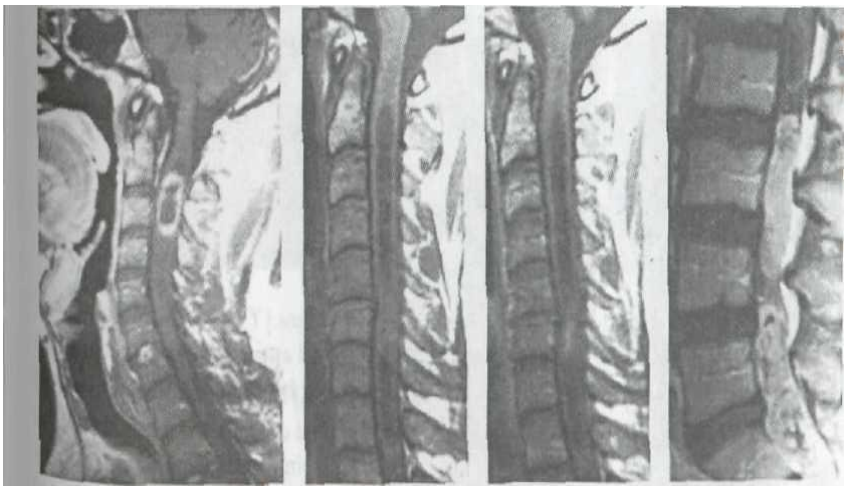

Fig. 2. Ependymoma. (A) Tl sagittal magnetic resonance image (MRI) after administration of intravenous contrast shows a rim-enhancing intramedullary lesion in the cervical spinal cord. The border of the lesion is distinct. An apical polar cyst is present.
Sagittal pre- (B) and postcontrast (C) MRI of another patient show a minimally enhancing sion at C4-5 with extensive syringomyelia. (D) Postcontrast sagittal MRI of the lumbar spine showing extensive involvement of the cauda equina by tumor. Histopathological diagnosis was myxopapillary ependymoma.

Histological differentiation from astrocytoma may be difficult, but the presence of perivascular pseudorosettes or true rosettes establish the diagnosis. Most spinal ependymomas are histologically benign, though necrosis and intratumoral hemorrhage are frequent. Although unencapsulated, these glial tumors ar usually well circumscribed and do not infiltrate adjacent spinal cord tissue.

A number of previous reports have included the classification of spongioblastoma unde the heading of glial tumor. Although this term is no longer employed in the typing of astrocytomas these tumors are interesting because of their frequently well-circumscribed nature, potential for complete resection, and long postoperative survival. Although the histological criteria used for typing of spongioblastoma have not been specified, these neoplasms probably represent either the tanycytic variant of ependymoma, subependymoma, or a type of juvenile pilocytic astrocytoma, which may be quite indolent or even regressive in behavior.

HEMANGIOBLASTOMA

Hemangioblastomas are benign tumors of vascular origin that are sharply circumscribed but not encapsulated. Almost all have a pial attachment and are dorsally or dorsolaterally located (Fig. 3). They are distributed evenly throughout the spinal cord but show a cervical predominance when they occur in association with von Flippel-Lindau (VHL) syndrome. This syndrome is autosomal-dominant with variable and incomplete penetrance. Affected individuals also have a predisposition to developing cerebellar lesions and cysts of other organs. Spinal hemangioblastomas account for 3% to 8% of intramedullary tumors and arise in any age group but are rare in early childhood. Most patients present before the age of 40 years. Lesions are generally sporadic, but up to 25% of patients will have evidence of VHL syndrome. Patients with VHL syndrome tend to become symptomatic at an earlier age and occasionally have multiple tumors (Neumann H.P., Eggert H.R,WeigelK., 1989).

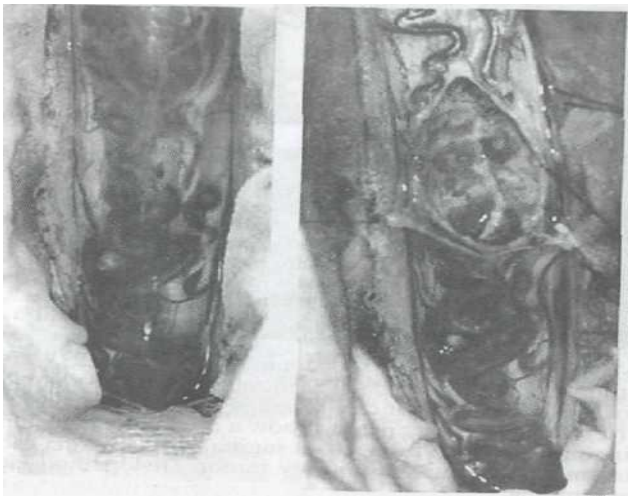

Fig. 3. Hemangioblastoma. (A) Intraoperative photograph of the dorsal surface of the cervical spinal cord. An intramedullary hemangioblas-toma is evident at the superior aspect of the exposure. Large veins drain interiorly. (B) Excision of the pial attachment from the lesion facilitates removal.

MISCELLANEOUS PATHOLOGY

Metastases account for about 2% of intramedullary tumors. This low prevalence is probably due to the small size of the spinal cord and its poor vascular accessibility to hematogenous tumor emboli. **Lung and breast are the most common primary lesions.**

Melanocytoma, melanoma, fibrosarcoma and myxoma have also been reported as intramedullary metastases (Pasaoglu A., Patiroglute, Orhon C, 1988).

Other nonneoplastic entities can present as intramedullary spinal lesions. Vascular malformations, particularly cavernous angiomas, may occur in the spinal cord. Inclusion tumors and cysts are rarely intramedullary. Lipomas are the most common dysembryogenic lesion and account for about 1 % of intramedullary masses (Fig. 4). These are not true neoplasms but probably arise from inclusion of mesenchymal tissue within the spinal cord itself. They typically enlarge and produce symptoms in early and middle adult years through increased fat disposition in metabolically normal fat cells. Lipomas are often considered juxtamedullary because they occupy a subpial location.

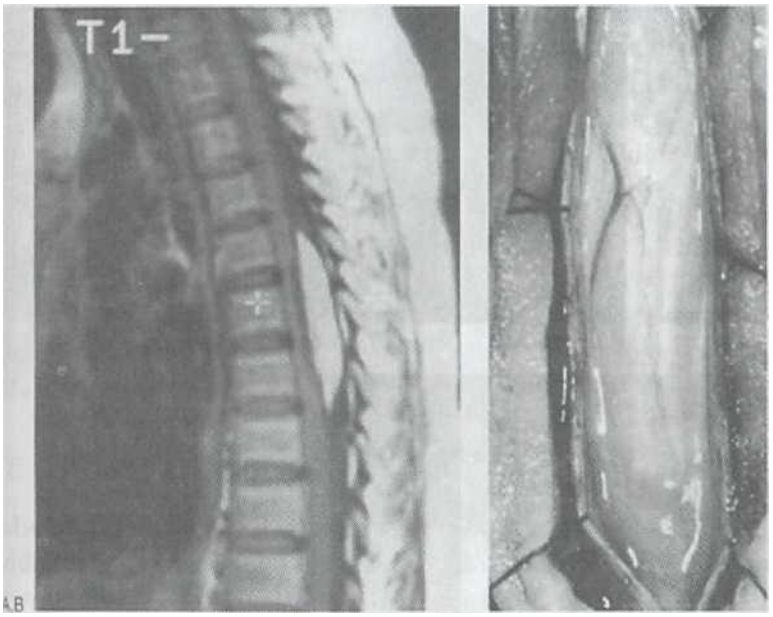

Fig. 4. Lipoma. (A) Tl sagittal magnetic resonance image of the thoracic spine without intravenous contrast shows a hyperintense intradural lesion posterior to the spinal cord at T5-7. The cord is compressed. (B) Intraoperative photograph confirms the subpial location of the lipoma.

EXTRAMEDULLARY TUMORS

Meningiomas and nerve sheath neoplasms account for 80% of extramedullar spinal cord tumors **(Table 1).** Filum terminale ependymomas accounts for 15% of these lesions. The remaining 5% includes paragangliomas, drop metastases and granulomas, all rare entities.

MENINGIOMA

Meningiomas arise from arachnoid cap cells imbedded in the dura near the nerve root sleeve, reflecting their predominant lateral location and meningeal attachment. Other hypothetical cells of origin include fibro-blasts associated with the dura or pia, which may account for occasional ventral or dorsal locations of these tumors. Meningiomas occur in all age groups, but the majority arise in individuals between the fifth and seventh decades of life. From 75% to 85% occur in women and about 80% are thoracic. The upper cervical spine and foramen magnum are also common sites **(Fig.** 5).

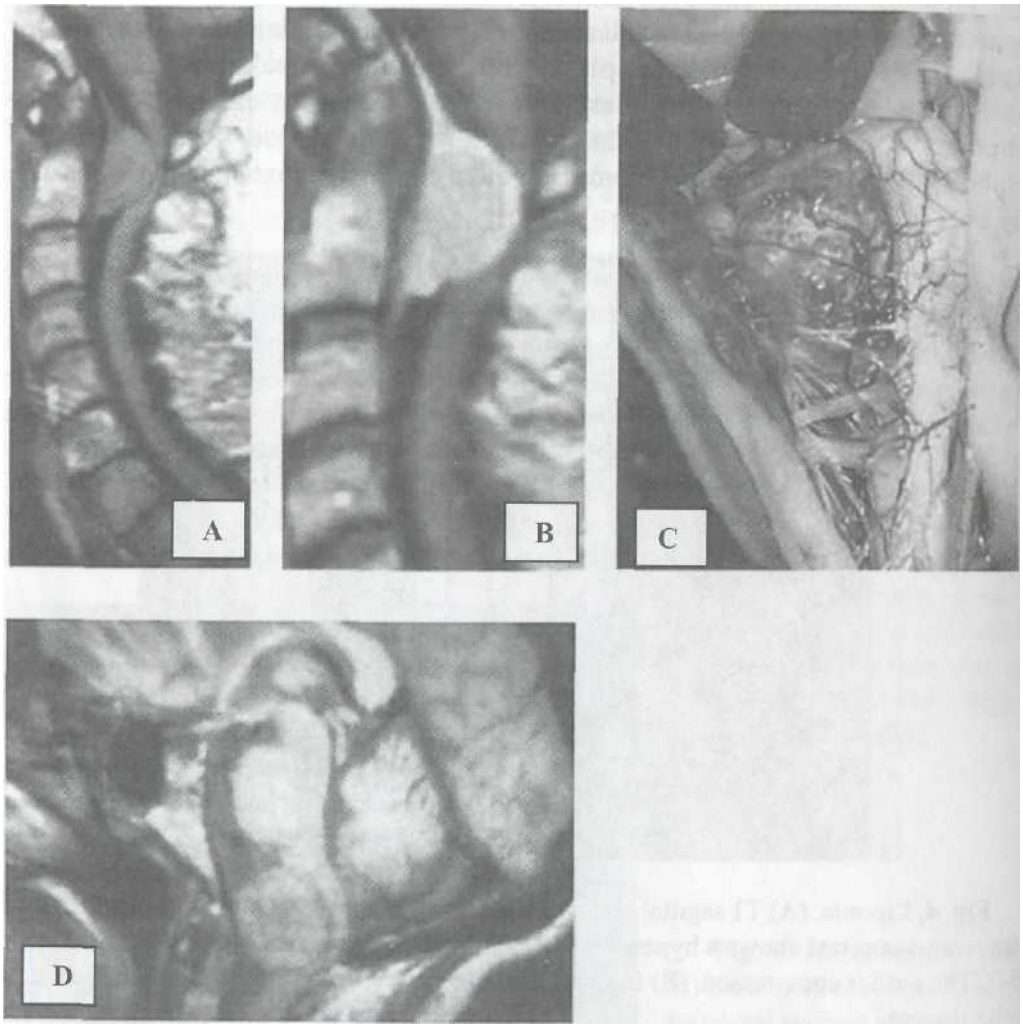

Fig. 5. Meningioma. Pre- (A) and post-contrast (B) sagittal magnetic resonance images (MRI) of the cervical spine demonstrate a homogeneously enhancing, ventral and extraaxial tumor at Cl-2. A clear dural tail is evident at the superior aspect of the lesion. Precontrast TI sagittal MRI (C) and intraoperative photograph after posterolateral exposure (D) show a foramen magnum meningioma. The surface of the tumor is irregularly lobulated and displays moderate neovascularity (D).

Here, meningiomas often occupy a ventral or ventro-lateral position and may adhere to the vertebral artery near its intradural entry and initial intracranial course. Low cervical and lumbar meningiomas are infrequent. The majority of spinal meningiomas are entirely intradural; however, about 10% may be both intradural and extradural or entirely extradural. Meningiomas are generally solitary, but multiplicity can be observed in patients with neurofibromatosis **(Fig.** 6). The overall incidence of multiplicity in the spine is 1% to 2%. The gross characteristics range from smooth and fibrous to the more frequent variegated, fleshy, and friable appearance. Microscopic calcification may occur. The dural attachment is often broader than expected, but en plaque examples are unusual. Bony involvement does not occur in the spine because of the well-defined epidural space. Psammomatous meningioma is the most frequent histological subtype (McCormick P.C., Post K.D., Stein B.M., 1990).

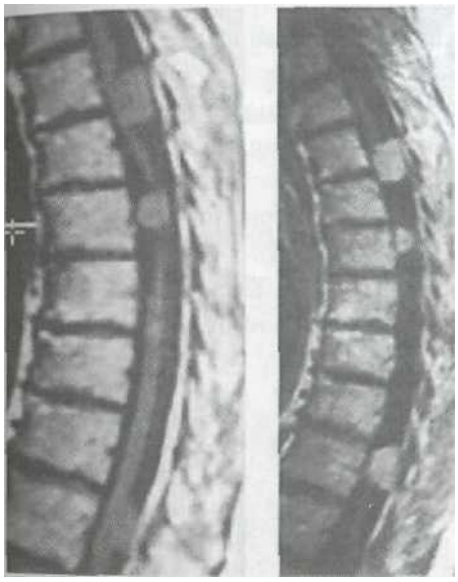

Fig. 6. Multiple meningiomas. Pre- (A) and postcontrast (B) sagittal magnetic resonance images of the thoracic spine of a patient with von **Recklinghausen's disease** reveals multiple intradural extramedullary enhancing tumors.

NERVE SHEATH TUMORS: SCHWANNOMA AND NEUROFIBROMA

Nerve sheath tumors are categorized as either schwannomas or neurofibromas. Although evidence from tissue culture, electron microscopy, and immuno-histochemistry supports a common Schwann cell origin of the neurofibroma and schwannoma, the morphological heterogeneity of neurofibromas suggests participation of additional cell types, such as the perineural cell and fibroblast. Neurofibromas and schwannomas merit separate consideration because of distinct demographic, histological, and biological characteristics. The histological appearance of neurofibromas consists of an abundance of fibrous tissue and the conspicuous presence of nerve fibers within the tumor stroma (Russel D. S., Rubinstein L.J., 1989). Grossly, the tumor produces fusiform enlargement of the Evolved nerve, which makes it impossible to distinguish between tumor and nerve. Multiple neurofibromas establish the diagnosis of neurofibromatosis, but this syndrome should be considered even in patients with solitary involvement. Schwannomas appear grossly as a smooth globoid mass that does not produce enlargement of the nerve but is suspended eccentrically from it by a discrete attachment **(Fig.** 7). The histological appearance consists of elongated bipolar cells with fusiform, darkly staining nuclei arranged in compact, interlacing fascicles that tend to palisade (Antoni A pattern). A loosely arranged pattern of stellate-shaped cells (Antoni B pattern) is less common.

Nerve sheath tumors account for about 25% of intradural spinal cord tumors in adults (Levy W.J., Latchaw J., Hahn J.F., 1986). Most are solitary schwannomas that occur throughout the spinal canal. The fourth through sixth decades of life represent the peak incidence of occurrence. Men and women are equally affected. The majority of nerve sheath tumors arise from a dorsal nerve root. Ventral root tumors are more commonly neurofibromas. Most nerve sheath tumors are entirely intradural (Fig. 7), but 10% to 15% extend through the dural root sleeve as a dumbbell tumor with both intradural and extradural components (Fig. 8). About 10% of nerve sheath tumors are epidural or paraspinal in location. One percent of nerve sheath tumors are intramedullary and are believed to arise from the perivascular nerve sheaths that accompany penetrating spinal cord vessels. Centripetal growth of a nerve sheath tumor may also result in subpial extension. This occurs most often with plexiform neurofibromas. In these cases both intra- and extramedullary tumor components will be apparent. Brachial or lumbar plexus neurofibromas may extend centrally into the intradural space along multiple nerve roots. In contrast, the retrograde intraspinal extension of a paraspinal schwannoma usually remains epidural. About 2.5% of intradural spinal nerve sheath tumors are malignant. At least one-half of these occur in patients with neurofibromatosis. Malignant nerve sheath tumors carry a poor prognosis; survival is generally less than 1 year. These tumors must be distinguished from the

rare cellular schwannoma, which has aggressive histological features but is associated with a favorable prognosis.

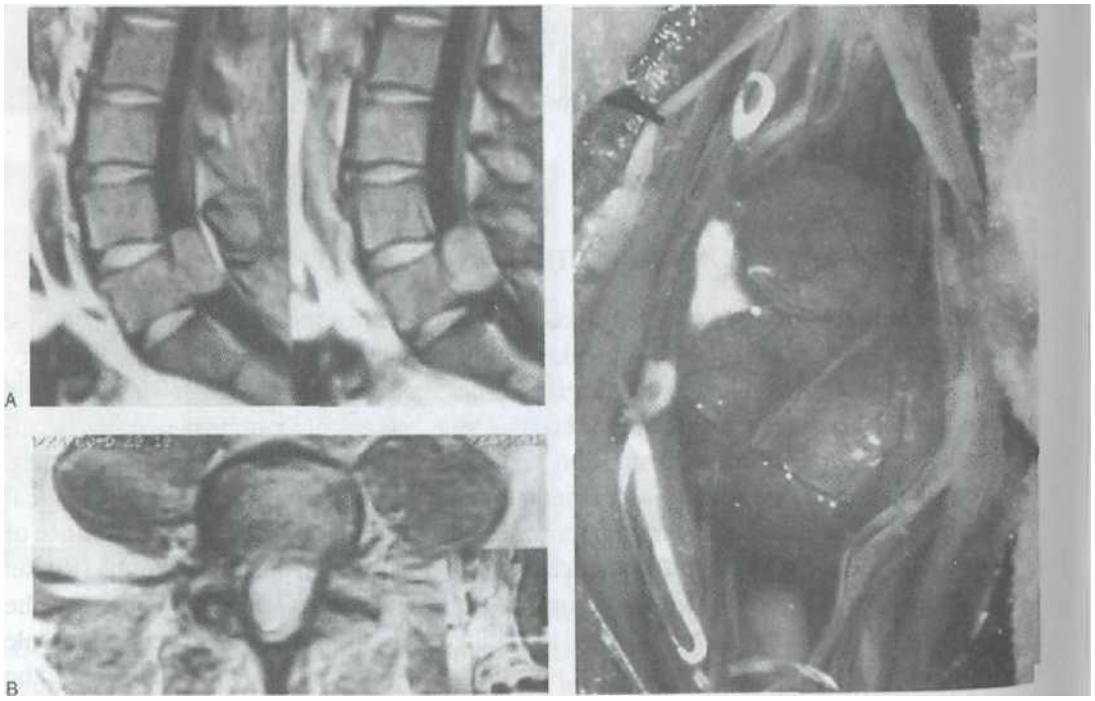

Fig. 7. Schwannoma. Sagittal pre- and postcontract lumbar magnetic resonance images (MRI) (A) and postcontrast (B) axial MRI shows a globoid and homogeneously enhancing intradural lesion at L-5. (C) Intraoperative photograph showing a well-circumscribed tumor with a smooth capsule that is adherent to the cauda equina

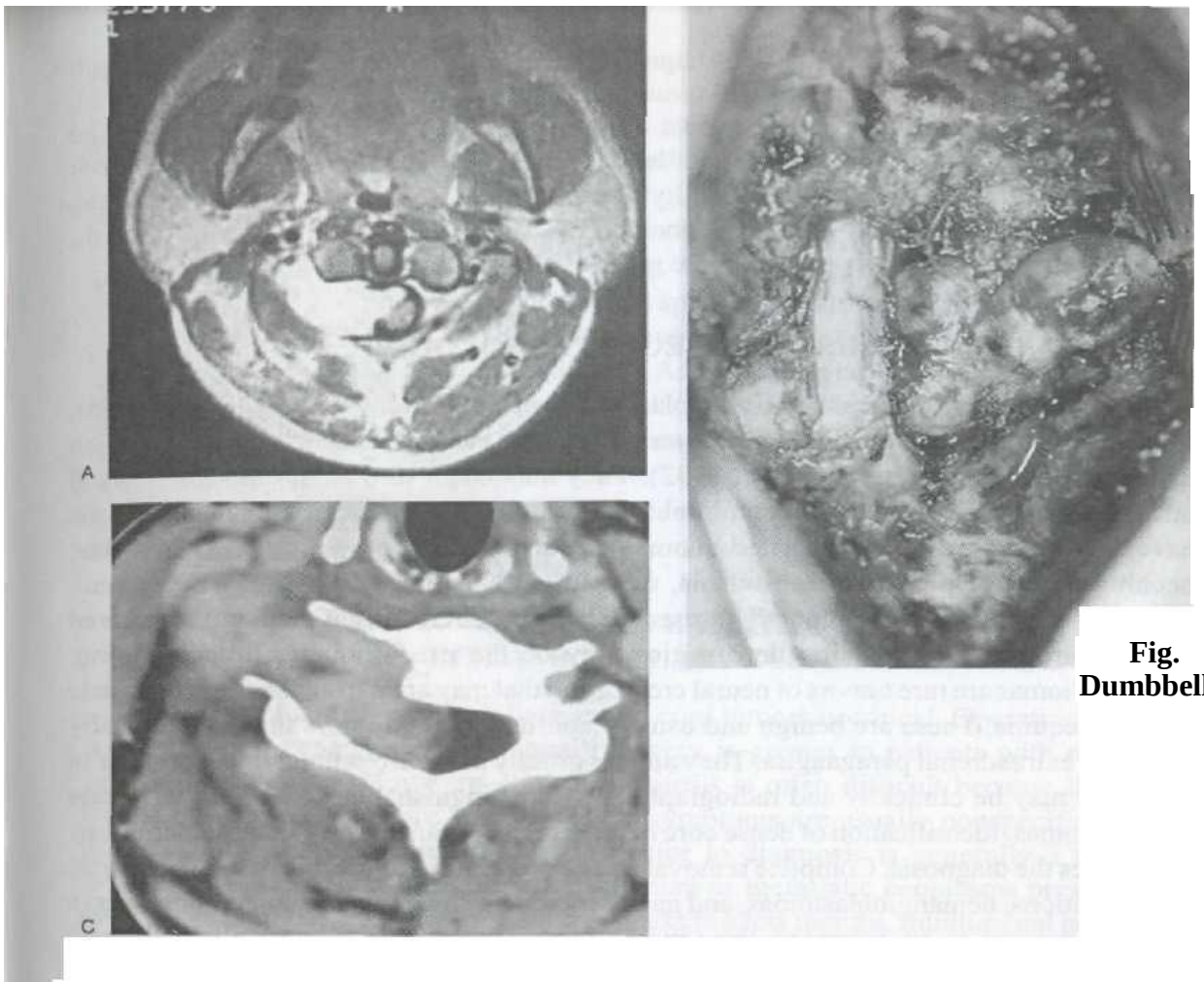

Fig. 8. Dumbbell

schwannoma. (A) Axial magnetic resonance image through the CI-*2* level after intravenous contrast. A homogeneously enhancing intradural lesion compresses the spinal cord toward the patient's left and extends through the right neural foramen. (B) Intraoperative photograph after removal of the posterior arches of C-l and C-2 shows extensive involvement of the C-2 nerve root. (C) Bone-windowed axial computed tomographic image through C3-4 from a different patient reveals bony erosion of the neural foramen by a dumbbell tumor.

FILUM TERMINATE EPENDYMOMA

Although filum ependymomas have been classified as intramedullary lesions by virtue of the neuroectodermal derivation of the filum terminate, it is appropriate to consider them with extramedullary tumors from an anatomical and surgical perspective (McCormick PC., Post K.D., Stein B.M., 1990). About 40% of spinal canal ependymomas arise within the filum terminale. Most occur in its proximal intradural portion. Extensive involvement of the cauda equina is sometimes present **(Fig.** 2D). Astrocytoma, oligodendroglioma, and paraganglioma may also originate in the filum but are rare. Filum terminale ependymomas occur throughout life but are most common in the third to fifth decades.

Lesions are typically red, sausage-shaped growths with moderate vascularity. Although unencapsulated, they are usually well circumscribed and may be covered by arachnoid. Myxopapillary ependymoma is the most common histological type encountered. The microscopic appearance consists of a papillary arrangement of cuboidal or columnar tumor cells surrounding a vascularized core of hyalinized and poorly cellular connective tissue. Nearly all are histologically benign (Sonneland P.R.W., Scheithauer B.W., Onofrio B.M., 1985). These tumors, however, tend to be more aggressive in younger age groups.

MISCELLANEOUS PATHOLOGY

Extramedullar masses may be neoplastic or nonneoplastic. Dermoids, epidermoids, lipomas (Fig. 4), teratomas, and neurenteric cysts are inclusion lesions that result from disordered embryogenesis (Pang D., 1992). They may occur throughout the spinal canal but are more common in the thoracolumbar and lumbar spine. Intramedullary locations have also been reported. Associated anomalies, such as cutaneous lesions, sinus tracts, occult anterior or posterior rachischisis, or split cord malformations, may be present. Inclusion tumors and cysts generally present as masses, but recurrent meningitis, tethered cord syndrome, or congenital deformities may be the predominant clinical finding. Paragangliomas are rare tumors of neural crest origin that may arise from the filum terminale or cauda equina. These are benign and usually nonfunctioning tumors that histologically resemble extraadrenal paraganglia. They appear grossly as well-circumscribed vascular tu mors and may be clinically and radiographically indistinguishable from filum terminale ependymomas. Identification of dense core neurosecretory granules on electron microscopy establishes the diagnosis. Complete removal can be accomplished in most cases. Cavernous malformations, hemangioblastomas, and ganglioneuromas may involve an intradural nerve root and present as an extrameduUary mass. These lesions may present clinically as a nerve sheath tumor with early radicular symptoms. Ganglioneuromas may present as dumbbell tumors in pediatric patients. Nonneoplastic lesions may also present as extrameduUary masses. Arachnoid cysts are a well-known example. These are most common in the thoracic spine and are usually dorsal to the spinal cord (Nabors M.W., Pait T.G., Byrd E.B., 1988). Intraspinal aneurysms are extremely rare. Herniated intervertebral discs have occasionally been reported to rupture through the dura and present as an intradural extramedullary mass.

Inflammatory pathologies, such as sarcoidosis, tuberculoma, subdural empyema, or transverse myelitis, rarely present as intradural mass lesions **(Fig.** 9). While spinal carcinomatous meningitis frequently complicates systemic cancer, secondary metastatic mass lesions of the intradural extramedullary compartment are rare. Malignant intracranial neoplasms that appose the subarachnoid space or ventricles are the most likely intracranial tumors to

demonstrate CSF drop metastasis into the spinal subarachnoid space. Systemic cancer accesses the subarachnoid space either through direct dural root sleeve penetration or, more commonly, hematogenously via the choroid plexus.

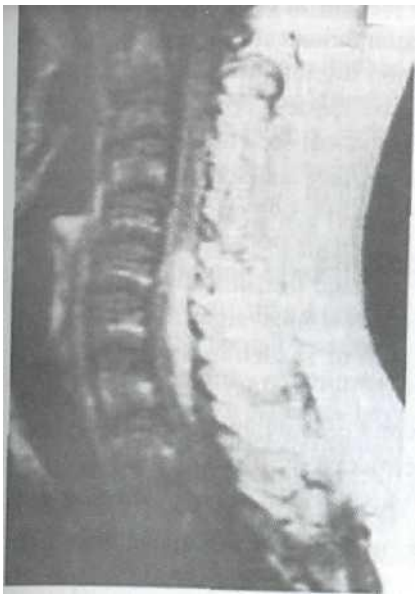

Fig. 9. Transverse myelitis. Postcontrast cervical spine sagittal magnetic resonance image in a patient with a subacute myelopathy (after a viral illness). An intramedullary enhancing mass is evident at C6-T1. Spinal cord biopsy revealed inflammation but no neoplastic cells.

CLINICAL FEATURES INTRAMEDULLARY TUMOR

Early diagnosis of intramedullary spinal cord tumors is critical. Operative morbidity is reduced and capacity for functional recovery is greater in patients with only minor neurological deficits. However, clinical diagnosis is often difficult because the clinical features of these lesions are variable. Early symptoms are usually nonspecific and may progress only subtly. Symptom duration prior to diagnosis is generally 2 to 3 years (McCormick P.C.. Stein B.M., 1990). Malignant or metastatic neoplasms present with a much shorter course: typically from several weeks to a few months. Intratumoral hemorrhage is more often associated with ependymomas.

Descending character of the neurological disorders is typical to the intramedullary spinal cord tumors (according to the law of the eccentric neural pathways disposition in the spinal cord).

Pain is the most common presenting symptom of intramedullary spinal cord tumors **in adults,** while **motor and gait disturbances** predominate **in children.** The pain typically occurs at the level of the tumor and is rarely radicular. Sensory or motor complaints are the initial symptom in about one-third of patients. The distribution and progression of symptoms are related to tumor location.

Upper extremity complaints predominate with cervical neoplasms. Unilateral or asymmetric involvement is typical. Dysesthesias are more common than numbness. A central cord syndrome may be present on examination.

Thoracic cord tumors produce spasticity and sensory disturbances. Peculiarity, what numbness is a more common complaint and typically begins in the legs with subsequent descending progression. Symptoms are related to both spasticity and sessory dysfunction.

Tumors of the lumbar enlargement and conus medullaris often present with back and leg pain. The leg pain is often multiradicular. Urogenital and anorectal dysfunction tend to occur early in the clinical course.

Mild deficit with significant radiographic spinal cord enlargement is characteristic of the most benign intramedullary tumors. In contrast, malignant tumors are associated with a much more rapid clinical course and there may only be modest cord enlargement at diagnosis. Increasingly, the diagnosis of an intramedullary spinal cord tumor is made prior to the occurrence of neurological deficits due to the availability and sensitivity of **MRI.**

EXTRAMEDULLARY TUMORS

Extramedullar)-' spinal cord tumors cause a variety of clinical signs and symptoms, the more common three groups by stages of grow: **(1) pain or radicular symptom,** early symptom followed by progressive neurological deficit is the clinical course most often encountered; the type of pain and specific deficits encountered depend mainly on tumor location and rate of growth; **(2) Brown-Sequard hemicord syndrome; (3) complete transverse cord syndrome** of the spinal cord conductibility disorder. It's typical **ascending character of the neurological disorders** (according to the law of the eccentric neural pathways disposition in the spinal cord).

Despite the classic syndrome historically ascribed to **intradural extramedullary tumors** consists of progression through segmental, hemicord, and transverse cord dysfunction, this presentation is rarely seen in current clinical practice and is not specific to extramedullary lesions. Generally, the clinical features of most extramedullary tumors reflect a slowly growing intraspinal mass. Specific presentations are variable and mainly determined by tumor location. Upper cervical and foramen magnum tumors are often ventral and frequently present with suboccipital pain, distal arm weakness, and hand intrinsic muscle weakness and atrophy, causing clumsiness. The etiology of this well-known syndrome is uncertain but most likely results from venous insufficiency. Increased intracranial pressure and hydrocephalus may rarely occur with an extramedullary tumor at any level, but they are more common with upper cervical lesions. The mechanism is probably related to elevation of the CSF protein and the resultant impaired CSF flow and absorption. Segmental motor weakness and long tract signs are the hallmarks of low and mid-cervical tumors. Early signs and symptoms are typically asymmetric, which reflects the predominantly lateral location of most intradural tumors.

A **Brown-Sequard hemicord syndrome,** characterized by dysfunction of the ipsilateral corticospinal spinal tract and posterior column, and dysfunction of the contralateral spinothalamic tract are common. Thoracic tumors frequently present with long tract signs. The corticospinal tracts are particularly vulnerable. Initial signs of stiffness and early muscle fatigue eventually give way to spasticity. Weakness usually begins distally, particularly with dorsiflexion of the ankle and large toe. A gait ataxia of sensory origin may result from bilateral posterior column compression by dorsal midline tumors. Bowel and bladder functions are not significantly impaired until late in the clinical course. Filum ependymomas present most frequently with back pain and subsequent asymmetric radiation to both legs. Increased pain on recumbency, an important clinical feature of extramedullary tumors, is most often associated with large cauda equina lesions. Subarachnoid hemorrhage has also been reported as the presenting feature of an extramedullary tumor.

RADIOGRAPHIC EVALUATION

The mainstay of radiographic diagnosis for all spinal cord tumors is MRI. MRI provides spatial and contrast resolution of neural structures, which is unattainable by any other imaging modality. Plain x-rays have little role in the modern diagnosis of spinal cord tumors as they do not image soft tissue adequately. However, the effects of intraspinal tumors on the vertebral elements are some times evident. Nerve sheath tumors can cause enlargement of the intervertebral foramina. Chronic intramed-ullary lesions can produce erosion or scalloping of the posterior vertebral bodies and widening of the interpe-dicular distance. At present, myelography has a limited role in the diagnosis of spinal cord tumors. It is seldom performed without subsequent computed tomography (CT). Intradural extramedullary tumors typically produce rounded filling defects of the dye column on a plain myelogram. Intramedullary

lesions characteristically cause focal widening of the spinal cord shadow. For extramedullary tumors, CT myelography provides excellent visualization of rumors arising in the region of the neural foramen. Accompanying bony changes are well demonstrated. Intramedullary tumors are more difficult to demonstrate because they are generally confined to the cord tissue.

INTRAMEDULLARY TUMORS

Most intramedullary tumors are isointense or slightly hypointense to the surrounding spinal cord on Tl-weighted images (Amour TE, Hodges S.C., Laaktnan R. W., Tamas D.E., 1994). Often, only subtle spinal cord enlargement is evident. T2-weighted images are more sensitive because most tumors are hyperintense to the spinal cord on these pulse sequences. T-2 studies, however, are not particularly specific and may not distinguish the solid tumor from polar cysts. Nearly all intramedullary neoplasms will enhance on T2-weighted contrast examinations.

Ependymoma. Ependymomas usually demonstrate uniform contrast enhancement and are symmetrically located within the spinal cord. Polar cysts are identified in the majority of cases, particularly with cervical and cervicothoracic tumors. Heterogeneous enhancement from intratumoral cysts or necrosis can also be seen. In some cases, contrast enhancement of a cystic ependymoma may be minimal. In these cases, it is difficult to distinguish with certainty these tumors from intramedullary astrocytomas.

Astrocytoma. The MRI appearance of astrocytomas is variable. These tumors tend to be less well defined than ependymomas on contrast-enhanced MRI because of their irregular tumor margins. Contrast uptake may be minimal, uniform, or patchy. Heterogeneous uptake is more commonly seen with astrocytomas because of intratumoral cysts or necrosis. Patchy, irregular rumor margins that extend over several spinal segments are also common.

Hemangioblastoma. Hemangioblastomas typically show marked and uniform contrast enhancement on MRI. Vascular flow-voids may be seen on noncontrast images. Associated polar cysts are also common. Lesions rarely span more than one spinal segment. Despite characteristic MRI patterns, there is enough variability and overlap in appearance of intramedullary tumors to preclude competent histological diagnosis based on MRI characteristics alone.

EXTRAMEDULLARY TUMORS

Signal abnormalities of the tumor, CSF capping, and spinal cord or cauda equina displacement will identify most extramedullary masses on a technically adequate MRI study. The diagnosis of lipoma, neurenteric cysts, dermoid or epidermoid, arachnoid cysts, or vascular pathology may be established on the basis of imaging characteristics alone. Gadolinium-enhanced images markedly increase the sensitivity of MRI, particularly for small tumors. Most extramedullary tumors are isointense or slightly hypointense with respect to the spinal cord on Tl-weighted images. Nerve sheath tumors are more likely to be hyperintense to the spinal cord than meningiomas on T2-weighted images. Cauda equina tumors usually demonstrate increased signal intensity with respect to CSF on both Tl and T2 pulse sequences. Small cauda equina tumors are easily overlooked, however, on noncontrast scans. Virtually all extramedullary spinal tumors demonstrate some degree of contrast enhancement. Meningiomas typically exhibit intense uniform enhancement, although nonenhancing calcifications or intratumoral cysts may be seen. Enhancement of the adjacent dura, a «dural tail,» strongly supports the diagnosis of meningioma. Although most nerve sheath tumors and filum ependymomas also demonstrate uniform contrast uptake, heterogenous enhancement from intra-tumoral cysts, hemorrhage, or necrosis is frequent.

Myelography and postmyelographic CT (myelo-CT) Mt not often utilized for the evaluation of intradural pathology. Nevertheless, the spatial resolution of a myelo-CT remains superior to MRI. For tumors that are closely applied to the surface of the spinal cord in which the MRI is equivocal with respect to an intra- or extramedullary location, myelo-CT can provide better

resolution. The intra- or extradural distribution of a paraspinal or dumbbell tumor is also better visualized with myelo-CT than MRI.

MANAGEMENT

After clinical and radiographic evaluations reveal a lesion that is thought to be a spinal cord tumor, tissue diagnosis is necessary. The surgical objective for most spinal cord tumors is gross total removal. Surgical planning must proceed accordingly. Immediately preoperatively, patients are given high-dose glucocorticoids and intravenous antibiotics. Most tumors are accessible with the patient in the prone position. For cervical lesions, stabilization of the head and neck in pins is necessary. Adequate exposure is crucial and is dictated by the location and extent of the lesion. Intraoperative monitoring with somatosehsory evoked potentials (SSEP) and motor evoked potentials (MEP) should be routine. Intraoperative ultrasound is often useful to localize and delineate the extent of intramedullary pathology. The extent of resection is guided by the anatomy of the lesion, intraoperative monitoring, the surgeon's experience, and the preliminary histological diagnosis on frozen sections of the lesion. A watertight dural closure is essential. Steroids are tapered postoperatively and early mobilization and rehabilitation are encouraged.

INTRAMEDULLARY TUMORS

Surgical Considerations. Surgery represents the most effective treatment for most intramedullary spinal cord tumors. The goal of surgery is determined primarily by the nature of the tumor-spinal cord interface. Gross total removal of a benign tumor should be accomplished when, clear demarcation exists between the tumor and the surrounding spinal cord. This can be achieved routinely with minimum morbidity, utilizing contemporary microsurgical techniques. Micro-surgical removal alone provides long-term tumor control or cure for hemangioblastomas, nearly all ependymomas, and some well-circumscribed astrocytomas. Determination of the resectability of an intramedullary tumor is best achieved by direct intraoperative inspection of the tumor-spinal cord interface. This requires an adequate myelotomy that extends over the entire rostrocaudal extent of the tumor. Reliance on intraoperative biopsy obtained through a tiny myelotomy should be avoided because the results are frequently inconclusive or inaccurate. The role of surgery for malignant intramedullary tumors is limited. Although significant palliation may be achieved with removal of an intramedullary metastasis, surgery provides no therapeutic benefit for primary malignant spinal cord tumors. Aggressive surgical removal of a malignant glial tumor is associated with significant morbidity. In these situations, an open biopsy with an unequivocal intraoperative histological verification of a malignancy signals an end to the operation. For many inclusion tumors and cysts, a clear plane of separation from the normal spinal cord is impossible to develop. In these cases, total removal should not be attempted. Small residuals present after removal of dermoids and epidermoids pose only a minor risk of recurrence. Subtotal internal decompression is usually definitive treatment for an intramedullary lipoma.

Radiation Therapy. The role of radiation therapy in the management of benign intramedullary spinal cord tumors is uncertain. In many instances, the benefits ascribed to radiation therapy may simply be due to the indolent natural history of most benign intramedullary tumors. Nevertheless, radiation therapy may provide a degree of tumor control in some patients with low-grade ependymomas or astrocytomas (Whitaker S.J., Bessel E.M., Ashley S.E., 1991). This response is neither uniform nor predictable. It is now well established, however, the gross total resection of intramedullary ependymomas more consistently provides long-term tumor control or cure than subtotal or near total resection followed by radiation therapy (Stein B.M., McCormick P.C., 1992). Further, there is no evidence of any therapeutic value of radiation therapy following gross total removal. Therefore, the role of radiation therapy in the management of benign intramedullary ependymomas is limited. Radiation therapy for ependymomas should be viewed as salvage therapy and employed in cases of an aggressive

benign tumor that cannot be totally removed, the rare malignant ependymoma, or CSF dissemination (Davis C, Barnard R.O., 1985). For hemangioblastomas, where total removal is achieved in almost all cases, postoperative radiation therapy has no role. Inclusion tumors and cysts are benign lesions; postoperative radiation therapy is not necessary for these entities. The role of postoperative radiation therapy for intramedullary astrocytomas is equivocal. For low-grade tumors that have been substantially resected, it is probably not necessary and may complicate the prospects for any future surgery. Radiation therapy is the primary treatment for malignant astrocytomas.

Operative Technique. Intramedullary tumors are approached via standard laminectomy with the patient in the prone position. SSEP and direct MEP are routinely employed. Although the number of times during an operation that evoked potentials influence surgical decisions is small, reduction in amplitude or increase in latency of potentials during dissection and retraction should be used as a guide for temporarily suspending those activities and waiting until the potentials improve before proceeding. Only rarely do they influence surgical decisions or technique. Laminoplasty is performed in pediatric patients but does not guarantee long-term stability. The dura is opened in the midline and tented laterally to the muscle with suture. The operating microscope is brought into the field for the remainder of the operation. The arachnoid is sharply opened and the spinal cord inspected. The strategies for intramedullary tumor removal depend on the relationship of the tumor to the spinal cord. Most tumors are totally intramedullary and are not apparent upon surface inspection. Intraoperative ultrasound may be used to localize and determine ros-trocaudal tumor extent. Tumors that do not present to the surface of the cord are best exposed through dorsal midline myelotomy. The myelotomy may be placed directly over an eccentrically located intramedullary tumor that extends to the pial surface. Hemangioblastomas originate from the pia. Resection of the pial attachment as part of the lesion will facilitate removal.

A midline myelotomy is performed through the posterior median septum and should extend over the entire rostrocaudal extent of the tumor. The tumor is first encountered in the area of maximum cord enlargement. The dissection continues on the surface of the tumor until the entire rostrocaudal extent has been identified. Polar cysts are entered and drained when present. The technique of tumor removal is determined by the surgical objective, tumor size, and gross and histological characteristics of the lesion. If no plane is apparent between tumor and surrounding spinal cord, it is likely that an infiltrative neoplasm is present. Biopsy is obtained to establish a histological diagnosis. If an infiltrating or malignant astrocytoma is identified and is consistent with the intraoperative findings, further tumor removal is not warranted. In most cases, however, a reasonably well-defined benign glial tumor will be present.

Ependymoma. Ependymomas have a smooth, reddish gray, glistening tumor surface that is sharply demarcated from the surrounding spinal cord. A variable vascular network crosses the tumor surface, distinguishing these tumors from astrocytomas, which rarely display these surface characteristics. The plane between an ependymoma and the surrounding spinal cord is usually well defined and easily developed. Large tumors may require internal decompression with an ultrasonic aspirator or laser. The tumor margins are then developed and feeding arteries from the anterior spinal artery are easily identified, cauterized, and divided.

Astrocytoma. Benign astrocytomas present with varying degrees of demarcation from the surrounding spinal cord. About one-third of adult patients have benign, infiltrative tumors without an identifiable tumor margin. Decompression is achieved with an ultrasonic aspirator or laser and proceeds systematically from the center of the tumor radially to the surface. Although a clean plane does not exist for the majority of astrocytomas, there is frequently a difference in the color of the tumor and the spinal cord. Changes in MEP or SSEP or uncertainty of the location of the tumor-spinal cord interface should signal an end to tumor resection.

Hemangioblastoma. Removal of hemangioblastomas is facilitated by excision of the pial attachment as part of the tumor mass. Cauterization of the surface tumor vessels is followed by a circumscribing incision around the pial base. The buried portion of the tumor within the spinal

cord is easily dissected and delivered, with traction on the pial base. A small polar myelotomy may provide better visualization for larger tumors. Internal decompression cannot be performed with these neoplasms, but cautery on the tumor surface usually shrinks it to a manageable size.

Following the removal of an intramedullary tumor, the resection bed is inspected and any bleeding points are controlled with warm saline or oxidized cotton. Closure of the myelotomy is not performed. The dura is usually closed primarily, although a dural patch graft may prevent dorsal tethering of the spinal cord at the operative site, a potential cause of morbidity in the postoperative period. An autologous fascia lata or thoracodorsal fascial patch graft can be utilized. The remainder of the wound is closed in standard fashion. Meticulous closure techniques are especially important in reoperations orpreviously radiated cases, which present a high risk of postoperative CSF fistula. Early mobilization in the postoperative period is encouraged.

EXTRAMEDULLARY TUMORS

Surgical Considerations. The optimal treatment of intradural extramedullary tumors is surgical excision. For nerve sheath lesions this can be accomplished in nearly all cases through a standard laminectomy. Recurrences are rare when gross total removal has been achieved. Most nerve sheath tumors are dorsal or dorsolateral to the spinal cord and are easily seen after opening the dura. Ventral tumors may require dentate ligament section to achieve adequate visualization. Lumbar tumors may be covered by the cauda equina or conus medullaris. The nerve roots must be separated to provide adequate visualization.

Laminectomy provides adequate exposure for spinal meningiomas in most cases. Unilateral laminectomy and facetectomy can be utilized for eccentrically located or ventral tumors. Large ventral tumors may also be approached satisfactorily through standard posterior exposures because the tumors have already provided the necessary spinal cord retraction. Suture retraction on a divided dentate ligament or noncritical dorsal root provides additional ventral exposure. Depression of the paraspinal muscle mass with table-mounted retractors further facilitates ventral access. Alternatively, a costotransversectomy or lateral extracavitary approach may be utilized for ventral thoracic tumors. The extreme lateral approach is used when there is a significant ventral tumor component above the foramen magnum.

Gross total en bloc resection should be attempted whenever possible. This can usually be accomplished for small and moderately sized tumors that remain well circumscribed within the fibrous coverings of the filum terminale and are easily separable from the cauda equina nerve roots. A portion of uninvolved filum terminale generally is present between the tumor and spinal cord. Amputation of the afferent and efferent filum segments is required for tumor removal. Internal decompression is not utilized for small and moderately sized tumors because this may increase the risk of dissemination of tumor into the CSF. Recurrences following successful en bloc resection are rare.

Radiation Therapy. The effects of postoperative radiation therapy on spinal meningiomas and nerve sheath tumors have not been extensively studied. Radiation treatment may be given for recurrent tumors that are histologically and clinically aggressive. It is probably best to reoperate and then to give radiotherapy. However, biologically aggressive filum ependymomas, which are more common in the younger population, may recur quickly; therefore, if resection is incomplete or if CSF dissemination is present, radiotherapy can be given following the initial surgery. While the response of spinal cord ependymomas to radiation therapy is unpredictable, there is some evidence that long-term control can be achieved with radiation therapy in some patients. This response cannot be predicted individually.

Operative Technique. Nerve Sheath Tumors. Surgical exposure of nerve sheath tumors depends on the specific anatomy of the lesion. Tumors that are small and strictly intradural may be approached via laminectomy. Ventrally located rumors may require facetectomy or a transthoracic or far lateral approachs. Once exposure is achieved, a plane of dissection directly

on the tumor surface must be identified. There is usually an arachnoid membrane tightly applied to the tumor surface. This is the fenestrated arachnoid layer, which separately ensheathes each dorsal and ventral nerve root within the subarachnoid space. This layer is sharply incised and reflected off the tumor surface. The tumor capsule is cauterized to diminish vascularity and shrink tumor volume. Tumor removal requires identification and division of the proximal and distal nerve root tumor attachments. These may not be immediately apparent with large tumors. Internal decompression with a laser or ultrasonic aspirator is utilized in these cases. Sacrifice of the nerve root of origin is usually required for tumor removal. Occasionally, some fascicles of the nerve root may be preserved, especially with smaller tumors. It is usually possible, however, to preserve the corresponding intradural nerve root because the fenestrated arachnoid sheaths allow anatomical separation of the dorsal and ventral nerve roots to a point just distal to the dorsal root ganglion. In a typical case of a dorsal root tumor origin, for example, it is possible to preserve the ventral root, which is tightly applied to the ventral tumor surface. Dumbbell extension through the root sleeve, however, usually necessitates resection of the entire spinal nerve (McCormick P.C., 1994). This rarely causes significant deficit, even at the cervical and lumbar enlargements. The function of the involved root has probably already been compensated for by adjacent roots. A very proximal tumor origin may be partially embedded in the epipial tissue or may elevate the pia to occupy a subpial location. The tumor-cord interface may be difficult to develop in these cases and may require resection of a segment of pia to effect complete removal.

Meningioma. Dorsal and dorsolateral lesions are delivered away from the spinal cord with traction on the open dural margins. A circumscribing excision of the dural origin completes the removal. For lateral and ventral tumors, the arachnoid over the exposed portion of the tumor is incised and reflected so that the dissection may proceed directly on the tumor surface. The rostral and caudal tumor poles should be identified. Small cottonoid pledgets may be placed in the lateral canal gutters on either side of the tumor to minimize blood spillage into the subarachnoid space. The exposed tumor surface is then cauterized to diminish tumor vascularity and shrink the tumor mass. Large tumors are bisected and debulked through a central trough. The tumor segment apposing the spinal cord is then delivered into the resection cavity with gentle traction and surface dissection. The remaining dura-based tumor is amputated from the dural attachment. The attachment is then extensively coagulated. Alternatively, the dural base may be excised and replaced with a thoracodorsal fascial patch graft. All blood and debris are irrigated from the subarachnoid space with warm saline. Arachnoid adhesions, which hold the cord in a deformed position, are divided. These maneuvers may diminish the risk of postoperative complications, such as spinal cord tethering, arachnoiditis, delayed syrinx formation, and hydrocephalus, that occasionally complicate extramedullary tumor removal. Rarely, a spinal meningioma may extend through a dural nerve root sleeve to present as a dumbbell tumor. The techniques for removal are similar to those already described for nerve sheath tumors.

Filum Ependymoma. Exposure for resection of a filum ependymoma is a laminectomy over the involved levels. Removal consists of developing a clean arachnoid plane around the lesion and separating it from the involved nerve roots. Resection of large filum terminale ependymomas, however, can present significant problems. Large tumors have probably been present for many years, and their resection carries a significant risk of dissemination into the CSF. Unencapsulated soft neoplasms may insinuate among the roots and within the arachnoid sheaths of the cauda equina, compartmentalized by innumerable arachnoid septae. Filum ependymomas may also spread as contiguous tumor sheaths along the arachnoid septae, which act as a scaffolding for surface growth. CSF dissemination may occur because of the subarachnoid location. Tumor removal in these cases is necessarily piecemeal and will almost always be subtotal. Dense tumor attachments to the roots of the cauda equina present significant risks of postoperative deficits because of the manipulation required for removal.

OUTCOME

INTRAMEDULLARY TUMORS

The results of surgery are strongly related to the preoperative neurological status of the patient and tumor location. In general, most patients note sensory loss in the early postoperative period, which, in many cases, is a result of the midline myelotomy. Subjective complaints are often out of proportion to objective findings. The sensory dysfunction tends to improve with time. Patients with significant or long-standing deficit rarely demonstrate any significant recovery and are more likely to worsen following surgery than patients with milder symptoms. Preservation, rather than restoration, of neurological function is the most reasonable expectation for intramedullary tumor surgery. The greatest benefit and the least risk of surgery for intramedullary tumors is experienced by patients who are minimally symptomatic. This fact underscores the importance of early diagnosis and treatment.

Ependymoma. Total microsurgical removal alone is optimal treatment for intramedullary spinal cord ependymomas. In one series of 23 patients treated in this manner, no clinical or radiographic evidence of tumor recurrence existed with a mean follow-up time of 5 years. Seven of these patients were followed for more than 10 years (McCormick P.C., Torres R., Post K.D., 1990). The use of radiation therapy following subtotal removal is prevalent. Its effects are difficult to interpret because many studies have few patients, lack of adequate tissue diagnoses, and limited follow-up. Favorable results following radiotherapy may reflect the natural history of the disease. As a general guideline, postoperative radiotherapy should be reserved for patients with histologically malignant ependymomas or benign lesions where the surgical anatomy precludes total resection. No benefit from radiotherapy following gross complete removal of a spinal ependymoma has ever been demonstrated.

Astrocytoma. The treatment of intramedullary spinal cord astrocytomas has been difficult to evaluate because of their rarity and biological variability. Age appears to be the most significant prognostic factor. Pediatric astrocytomas are associated with a particularly indolent behavior, which is partly explained by their predominantly benign histology. Approximately 60% of patients below 21 years of age at diagnosis are alive without recurrence 10 years after treatment, while only 40% of patients over the age of 21 at diagnosis are alive without recurrence 5 years following treatment (Sandier H.M., Papadopoulous S.M., Thuntan A.F., 1992). This age-related behavior is reflected in other studies (Rossitch E., Zeidman S., Burger P.C., 1990). Astrocytomas in the adult are typically more aggressive. There is a higher incidence of malignant tumors in the adult, and low-grade tumors have also been associated with more frequent recurrence and diminished survival. Despite numerous factors that may correlate with overall clinical course, such as age, gross tumor characteristics, and various histological parameters, it is not possible to predict individual tumor behavior. Results for specific treatment regimens are variable. In one study, long-term tumor control was achieved in two patients treated with biopsy and radiation therapy. However, two other patients treated in this manner demonstrated tumor recurrence 2 years postoperatively. Long-term tumor control was also seen in two patients who underwent biopsy alone. Similar variability of outcome with different treatment strategies has been repeated elsewhere. Studies suggest that outcome may be improved if gross total removal has been performed. No tumor recurrence following gross total resection in three patients at 39, 57, and 134 months postoperatively has been reported. Radical removal has resulted in an average survival of up to 173 months, almost three times that of patients treated with biopsy or subtotal removal. In another series, only one of four patients treated with gross total resection had tumor recurrence that did not become clinically apparent until 38 years postoperatively. Two of these four patients received radiation therapy. It is possible that the indolent behavior and benign natural history of the tumor are reflected in better demarcation from the spinal cord, which allows gross total resection. This may also explain why a significant percentage of patients have **long-term** tumor control regardless of the degree of resection or treatment with radiation therapy. Parameters that predict which patients will benefit from aggressive removal have iot been identified.

Although gross total removal is theoretically optimal, it is gradually becoming clear that it alone is not always definitive treatment for intramedullary astrocytomas. 20% 5-year recurrence rate in 100 pediatric patients treated with radical gross total removal has been reported. The event-free 5-year survival of 47 patients undergoing gross total removal of low-grade gangliogliomas is only 36%. Therefore, unlike spinal cord ependymoma, radical gross total removal does not represent the primary goal of surgery. The treatment priority should be long-term tumor control with preservation of neurological function.

Hemangioblastoma. Since hemangioblastomas are so well circumscribed, subtotal removal is uncommon (Neumann H.P., Eggert H.R., Weigel K., 1989). The risk of recurrence after gross total resection is extremely small. If there is concern that a total resection was not obtained, the patient should be followed frequently, both clinically and by MRI. Symptom recurrence with radiographic evidence of recurrent tumor is an indication for reoperation.

EXTRAMEDULLARY TUMORS

The results of surgery for intradural extramedullary spinal cord tumors are usually excellent. Neurological morbidity is typically less than 15%. Mortality is extremely uncommon (Epstein F.J., Farmer J.P., 1990; McCormick P.C., Post K.D., Stein B.M., 1990). Complications are generally related to wound healing and CSF leakage. Management with lumbar drainage is sufficient to stop CSF leak in most of these cases. Neurological complications, such as new deficits or exacerbation of existing ones, are uncommon; they are most often associated with manipulation of the cauda equina. Motor and sensory deficits typically improve postoperatively. Return of bladder function is variable. Improvement of preoperative deficits is typical and may be dramatic early in the postoperative period. Recovery is related to the duration and severity of the existing deficit and the age of the patient.

Nerve Sheath Tumors. Total removal of neurofibromas and schwannomas that are not associated with neurofibromatosis is generally curative. However, tumors with extensive paraspinal involvement that are subtotally resected have a definite propensity to recur. Deficits resulting from sacrifice of the involved nerve roots are usually minor and well tolerated. Patients with multiple lesions from neurofibromatosis should usually be observed. Resection is reserved for progressive and symptomatic focal lesions.

Meningioma. Recurrence after total resection of spinal meningiomas is about 1% at 5 years and 6% at 14 years. Subtotally resected lesions have average recurrence rates of approximately 15%. Meningiomas with extradural spread or en plaque lesions are more difficult to remove and tend to recur more frequently. These lesions also are associated with greater degrees of postoperative morbidity. These factors must be balanced when planning the extent of resection.

Filum Ependymoma. Neurological deterioration following removal of filum ependymomas is more frequent than that associated with nerve sheath tumors and meningiomas. Lesions that involve the conus medullaris or that are intimately adherent to many roots of the cauda equina carry the highest risk of postoperative morbidity. Recurrence after gross total resection is rare. Subtotally removed lesions recur in approximately 20% of cases. Survival after total removal is almost 100% (McCormick P.C., Post K.D., Stein B.M., 1990; Sonneland P.R.W, Scheithauer B.W., Onofrio B.M., 1985). Incompletely resected lesions treated with postoperative radiation therapy are associated with 5- and 10-year survival rates of 69% and 62%, respectively. Subtotally removed lesions should be frequently followed by MRI. In summary, treatment of intradural spinal cord tumors remains a gratifying area of neurosurgery. Advances in imaging sensitivity and refinement of microsurgical techniques have allowed removal alone to become definitive management for most spinal neoplasms. Early diagnosis and surgical excision usually comprise optimal treatment for most intradural spinal tumors.