Consultation with the Specialist

Extrinsic Spinal Cord Mass Lesions

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Extrinsic spinal cord lesions have been classified via various methodologies. Most recently, spinal cord tumors have been divided anatomically into those that are intramedullary and those that are extramedullary. By definition, intramedullary lesions encompass all tumors that are situated within the parenchyma of the spinal cord. By far the most common of the intramedullary tumors are various types of gliomas. This article focuses on extramedullary lesions, which includes all tumors that lie outside the spinal cord proper. Unlike intramedullary tumors, extramedullary tumors are an extremely heterogenous group and can be divided into several subcategories for both pathologic and prognostic reasons.

As can be seen in the Table, intradural tumors range from histologically benign tumors, including congenital tumors, meningiomas, nerve sheath tumors, and vascular tumors, to malignant lesions. In this case, benign does not mean that the tumor cannot have significant detrimental effects, including death, but rather that it is localized to the one area, as noted by the imaging study, and does not spread diffusely to other portions of the central nervous system (CNS). In contrast, metastatic tumors can be either of neural origin or of extraneural origin and spread diffusely through the cerebrospinal fluid (CSF) pathways, frequently involving the entire CNS. Extradural tumors include derivatives from tissues that normally are closely located in an extradural position. These tumors arose from either precursor cells or cells that have shown malignant transformation, such as neural crest precursors, soft-tissue tumors such as sarcomas, and various types of bone tumors.

Tumors of the CNS are common and constitute approximately 20% of all pediatric malignancies. Ones involving the spinal canal are far less common than ones located intracranially; the ratios of intracranial to intraspinal tumors range from 20:1 to 5:1. Spinal canal tumors appear to account for about 12% to 15% of all nervous system tumors. Consistent with the diversity of tumors is the predominance of extramedullary lesions relative to intramedullary lesions; approximately twice as many tumors are extramedullary as are intramedullary.

Findings

The presentation of extramedullary tumors can be diverse. Symptoms and signs appear to be consistent throughout the different age groups, but children at different ages present differently according to their neurologic function at specific ages. For example, spinal canal tumors frequently are identified very late in their course in very young children because infants cannot present their "complaints" in the usual manner, and toddlers, although they can speak, often do not know how to express the problems that they are experiencing. The degree of erroneous diagnosis is quite high (>50% in most instances) and is affected greatly by patient age.

The most common presentation for extramedullary spinal cord lesions is pain and weakness, with pain being the most common of all initial presenting symptoms. In contrast, radicular pain is quite uncommon in intramedullary spinal cord disease, which may help to differentiate the two types of lesions. The pain of extramedullary spinal cord lesions usually presents initially as either paraspinal muscle spasm, increased pain with motion in the spine, or both. Toddlers frequently present with irritability as a manifestation of the pain. When the lesion is attached to the cauda equina, sciatic type pain may be present.

Motor weakness usually is appreciated only upon gait analysis. A young child may show regression from previously obtained gait capabilities. If a lower motor neuron lesion predominates (ie, a peripheral nerve lesion), reflexes will be decreased. With clear spinal cord

TABLE.
Extramedullary Tumors

Intradural
- Congenital tumors
  - Epidermoids
  - Dermoids
  - Teratomas
- Meningiomas
- Peripheral nerve sheath (PNST) tumors
- Vascular tumors
- Metastatic tumors
  - Extra-central nervous system metastases (leukemia, lymphoma)
  - Intra-central nervous system metastases

Extradural
- Neural crest tumors
- Soft-tissue tumors
- Bone tumors
  - Primary
  - Secondary

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compression, however, reflexes frequently are increased because of the upper motor neuron finding.

Bladder and bowel problems are next most common symptoms. Of course, this is extremely difficult to diagnose in infants and toddlers and may be ascertained by evaluating for periods of dry diapers. Older children usually regress from previously attained toileting milestones. Unfortunately, these presenting symptoms and signs often are ignored or misunderstood and thought to be a behavior problem.

Sensory changes usually are radicular in initial presentation, but as further cord compression occurs, the ipsilateral spinothalamic and corticospinal tracts may be compromised. When compression is unilateral, one sees Brown-Séquard syndrome (ipsilateral weakness, spasticity, ataxia, and contralateral loss of pain and temperature sensation). Eventually this progresses to a more complete lesion. Presentation of Brown-Séquard syndrome is much more common in an extrinsic cord lesion than in intrinsic or intramedullary spinal cord lesions because of the eccentricity of the lesion in extramedullary disease.

Other findings also may suggest a spinal cord tumor. Spinal deformities such as kyphosis or scoliosis, when associated with pain, often can be warning signs of a spinal cord tumor. Congenital lesions frequently are signalled by midlying cutaneous markers such as hemangiomas or a dural sinus tract. Meningiomas, schwannomas, and neurofibromas can be suggested by other neurocutaneous findings.

Radiologic Studies

PLAIN FILMS

In approximately 50% of cases of extramedullary tumors, plain films reveal abnormalities. Congenital tumors often are associated with characteristic vertebral anomalies or deformities. Tumors such as neurofibromas and schwannomas frequently are characterized by enlargement of the neural foramina. Other common findings for an intradural/extramedullary tumor are: scalloping of the posterior vertebral body, increased sagittal diameter of the spinal canal, widening of the interpeduncular distance, and pedicle erosion. In metastatic disease, there may be frank disruption of the vertebral body, occasionally associated with a pathologic fracture. Pedicle involvement is also very common. Primary bone tumors are visualized easily by plain film.

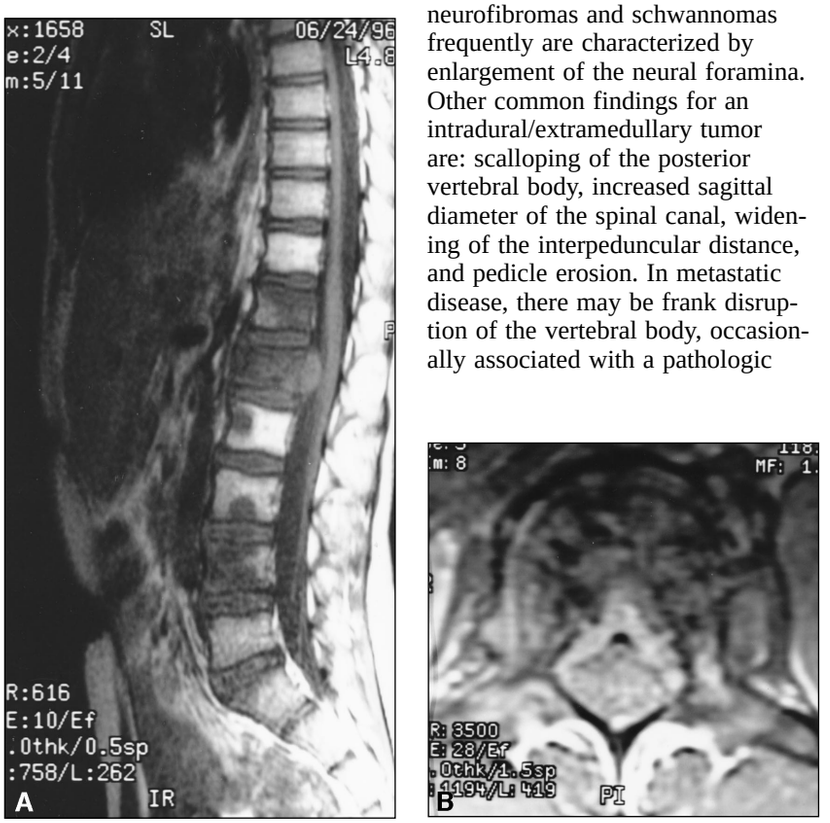

FIGURE. Extradural extramedullary tumor, as visualized in two views on MRI. The patient had a known diagnosis of metastatic neuroblastoma. Note the degree of anterior spinal cord compression (A) and the signal change in the involved vertebral body (B).

MAGNETIC RESONANCE IMAGING (MRI)

MRI is the most useful radiologic study for evaluating the spinal canal and its contents (Figure). It is quite excellent in defining the extent of disease, identifying the possible pathology involved, and evaluating the spinal cord and its surrounding structures in multiple projections. The only facet of spinal cord disease that MRI does not image well is bone anatomy. MRI is absolutely essential whenever a spinal cord tumor is suspected.

COMPUTED TOMOGRAPHY (CT)

The CT scan is particularly useful for defining bony abnormalities, although in isolation it tends to offer very little insight into intramedullary disease. Its delineation of soft-tissue changes is vastly inferior to that seen with MRI. Occasionally CT is combined with myelography, but this usually does not provide as clear an evaluation as that given by MRI scan. CT scanning should be employed primarily only to evaluate the bony anatomy following initial screening and definitive study by MRI scan.

Intradural/Extramedullary Tumors

CONGENITAL TUMORS

Epidermoid and Dermoid Tumors

In a sense, epidermoid and dermoid tumors are not actually true neoplasms; rather, they are malformations and more specifically dysembryonic malformations. These lesions appear to arise from displacement of dermal or epidermal elements into the neural canal during development. Occasionally, they can be created iatrogenically from a lumbar puncture.

Both lesions are lined by stratified squamous epithelium that contains desquamated epithelial surface cells and keratin. Dermoids also contain dermal elements such as hair follicles and sebaceous glands,

which distinguishes them from epidermoids. Epidermoids are distributed throughout the age groups; dermoids are much more common in children. The most common location is in the lower back in the lumbosacral spine, but occasionally they are found in the thoracic and cervical spine. These lesions frequently are associated with other cutaneous abnormalities, including dermal sinuses, hairy patches, port wine nevi, or small hemangiomas. Associated dermal sinus tracts can predispose to infection.

For patients who have these tumors, pain almost always precedes motor and sensory or sphincter changes. If the lesion is in the cervical or thoracic spine, however, upper motor neuron findings may occur before any pain is appreciated.

Plain films show the particular findings already mentioned. Epidermoids usually have CSF intensity, resulting in low signal on T1-weighted and increased signal on T2-weighted MRI images. On CT scan, epidermoids are low in density. Dermoids have a completely distinct pattern, with recorded signal showing the intensity of fat rather than water. They show low attenuation on CT, and on MRI they have high signal on T1-weighted images and moderate-to-high signal on T2-weighted images. Occasionally there may be lower signal on T1-weighted images, but this is secondary to watery secretions from sweat glands that exist in the tumor.

Treatment for these lesions is surgical removal. Those that are not removed completely will recur. The surgeon must be careful to avoid spilling the contents, which can cause a chemical meningitis.

Teratomas

Teratomas contain the elements of all three germ layers. Unlike the dysembryonic malformations mentioned previously, these are true neoplasms that may undergo malignant transformation. Teratomas almost always appear in the sacrococcygeal area, although occasionally they can be located elsewhere within the spinal canal. The sacrococcygeal lesions are not actually within the spinal canal and do not cause neurologic problems. Spinal teratomas account for 3% to 9% of intraspinal tumors in children.

The etiology appears to be migrating germ cells that have left the yolk sac and are supposed to migrate to the fetal gonads. Because of this etiology, these lesions appear predominantly in childhood. These tumors are found most commonly in the lumbar or thoracolumbar region.

Clinical findings are usually nonspecific, and radiologic findings may show the lesion in an intradural/extramedullary location or in the parenchyma of the spinal cord. Plain film findings are as mentioned previously. MRI scanning shows a pattern of multiple tissue signals that can vary substantially, depending on the mix of cystic and solid portions and the diverse tissues that make up the specific lesion. Treatment is complete surgical removal, which can be very difficult because of adherence of the tumor to CNS structures.

MENINGIOMAS

Meningiomas are quite rare in children, comprising only 2% of all tumors found in childhood and approximately 5% of pediatric spinal canal tumors. These generally are found in the thoracic region, but the pattern is not quite as clear as it is in adults. Almost always, these tumors are intradural/extramedullary, but occasionally they are extradural.

These follow the typical pattern of pain as an initial presentation, with later motor and sensory changes. Twenty percent of cases are associated with von Recklinghausen disease. Meningiomas usually are considered benign tumors, but malignant transformation is more common in children than in adults.

Plain films are not of help in defining the type of mass, but MRI is quite useful. These lesions have a signal pattern similar to that of the cord parenchyma and enhance quite well with contrast. Treatment is total removal.

PERIPHERAL NERVE SHEATH TUMORS

The two primary categories within this group are schwannomas and neurofibromas. Both appear to be derived from schwann cells, but neurofibromas also contain fibroblasts and an abundance of collagen fibers. These tumors comprise about 10% of all spinal canal tumors in childhood. They generally occur later than the dysembryonic malformations, usually during the teenage years. They are distributed throughout the spine, with no particular predilection for location.

Pain almost always is the presenting complaint because the tumors usually arise on a sensory nerve root. Only occasionally is motor weakness present. Symptoms appear 6 to 12 months prior to evaluation, and 25% of cases are associated with von Recklinghausen disease, similarly to meningiomas. These tumors are usually benign, but about 10% become sarcomatous. Usually they are intradural, but occasionally they may be extradural, and some may exist as dumbbell tumors (approximately 20%).

Plain films are nonspecific, showing the characteristic findings. MRI scan shows tumor hyperintensity both on T1 and on T2 images relative to skeletal muscle.

Total removal is required to prevent recurrence. Neurofibromas are significantly more difficult to remove than schwannomas. Schwannomas usually arise eccentrically from the nerve and extend outward, which allows the nerve to be dissected free of the mass. On the other hand, neurofibromas have nerve fibers passing through the substance of the tumor, which frequently makes total resection impossible unless the nerve is sectioned.

VASCULAR TUMORS

Hemangioblastomas are quite common among children; 25% are identified in individuals younger than 21 years of age. Typically they present in the cerebellum, but they also can be seen in the medulla and the spinal cord. About 20% occur as part of the von Hippel-Lindau complex. More specifically, 50% of spinal hemangioblastomas are found as part of the von Hippel-Lindau complex. Usually these lesions are isolated, but they may be multiple. Even when intramedullary, these tumors usually are attached only to the pia and are not in the core of the spinal cord.

Presentation is nonspecific. MRI scan tends to show a tumor nodule that is isointense and not well-defined but that becomes markedly hyperintense with gadolinium contrast. Frequently these are associated with syringes, which can be seen clearly on MRI. Serpentine flow voids on MRI scans represent the feeding and exiting vessels from the tumor.

Complete surgical removal is required, but when this is not possible, radiation therapy, particularly focused beam radiation therapy, is the best alternative.

METASTATIC DISEASE

Metastatic disease originates either neurally or extraneurally and spreads via the CSF pathways. The most common extraneural sources are systemic leukemia and lymphoma. A detailed discussion of these metastatic tumors is beyond the scope of this article. Briefly, leukemia is one of the most common systemic malignancies to involve the CNS; 30% of patients who have acute myelogenous leukemia (AML) present with CNS involvement. The frequency is significantly less in acute lymphocytic leukemia (ALL), but it can be greater than 50% if the CNS does not undergo prophylactic treatment.

CNS leukemia can present anywhere throughout the nervous system and appears to gain access to the parenchyma and the meninges via blood vessels. Sometimes these lesions hemorrhage or they grow large enough to produce mass lesions, although it is much more common to have a diffuse infiltration with meningeal irritation or parenchymal involvement. When there is only meningeal disease, meningeal or nerve root irritation is common, whereas intraspinal lesions associated with the parenchyma usually produce mass effect. The presence of the tumor in the CSF frequently induces hydrocephalus.

Diagnosis is made by lumbar puncture. MRI is helpful if a mass is present, but CSF sampling is the most useful method of diagnosis. Treatment involves radiation therapy and chemotherapy. Surgery, unlike with the previously discussed tumors, rarely is indicated unless there is acute mass effect with cord compression.

LYMPHOMA

CNS involvement with a lymphoma almost always is from non-Hodgkin lymphoma when it occurs as a primary lesion. As a secondary lesion, either Hodgkin or non-Hodgkin lymphoma can be the origin. Lymphoma generally is extradural, secondarily invading the epidural space from the paravertebral lymph nodes or from direct bone involvement into the extradural space. The most common location is the thoracic spine, but diffuse spinal involvement also is quite frequent.

Plain films are not specific, and CT and MRI rarely are helpful unless there is clear bony involvement, soft-tissue disease, or a large mass. As with leukemia, the treatment involves radiation therapy and chemotherapy. Surgery is indicated only when a large mass causes acute cord compression.

METASTASIS WITHIN THE CNS

Although not entirely synonymous, by far the most common cause of metastatic intraspinal disease is primitive neuroectodermal tumors. Medulloblastoma metastasizes most frequently, but anaplastic astrocytomas and ependymomas also may involve the CNS. Many other tumors have been noted to show metastases within the CNS. One of the most notorious examples is myxopapillary ependymoma, which occasionally has diffuse spinal subarachnoid spread.

In primitive neuroectodermal tumors, particularly medulloblastoma, metastatic disease is noted when the primary lesion is diagnosed in nearly 50% of cases. The most common means of tumor spread is via CSF pathways. Because posterior fossa tumors are more proximal to subarachnoid or intraventricular CSF collections, these metastasize more frequently than tumors in the cerebral cortex. Drop metastases in the spine are common and multiple. Because of the drop effect, they generally appear in the lumbosacral region.

These patients present with pain similar to that experienced with many of the other tumors and occasionally show meningeal signs. Motor, sensory, and bowel/bladder symptoms appear much later. MRI is useful in evaluation, particularly for a large mass. Spinal MRI is replacing myelography as the study of choice for noninvasive evaluation of metastatic disease. However, lumbar puncture remains the overall diagnostic study of choice. Because of the metastatic nature of the disease, radiation therapy and chemotherapy are absolute treatment choices. A large mass with mass effect can be removed surgically.

Extradural/Extramedullary Tumors

NEURAL CREST DERIVATIVES

Tumors that originate from neural crest tissue account for about 20% of all spinal tumors. The primary constituents of this group are the ganglioneuromas and the neuroblastomas. Ganglioneuromas are the most benign and neuroblastomas the most malignant on this tumor spectrum. Almost always these tumors originate from the adrenal medulla or the paravertebral sympathetic chain, involving the spinal canal and becoming extradural tumors by direct extension. Neuroblastomas occasionally are associated with diffuse metastatic disease to the rest of the body.

Ganglioneuromas comprise about 3% of childhood spinal lesions. They are more common in the older age group, probably because of their more benign nature and slower rate of growth. These tumors are composed of schwann cells interposed between mature ganglion cells. The most common location is the posterior mediastinum, although they can be found in the abdomen. Their slow growth may result in late presentation at which point they usually are quite large. When the spinal canal is involved, it is generally by direct extension. They are treated primarily by surgery.

Neuroblastomas are more common in young children, typically presenting before the age of 10. These tumors consist of sheaths of neuroblasts with very little interspersed stroma. They usually are circumscribed masses with areas of

invasion of the surrounding tissue. Almost 50% of these tumors arise from the adrenal medulla; the rest arise from the sympathetic chain. These very malignant tumors are associated with a better prognosis in children younger than 2 years of age. They metastasize readily, with tumor spread often found at diagnosis. The most common metastatic sites are the lymph nodes, liver, and bone. These tumors usually involve the spine only by direct extension, but occasionally they spread by metastasis.

Findings on plain films are usually nonspecific, indicating only evidence of a mass. CT and MRI are the studies of choice that clearly define extraspinal and intraspinal components of the mass and document bony metastases. Treatment for these tumors involves radiation therapy and chemotherapy, with surgery if there is acute cord compression.

Intermediate between ganglioneuromas and neuroblastomas is the ganglioneuroblastomas, which occur in children at an intermediate age between these other two tumors. Ganglioneuroblastomas are comprised of mature ganglion cells and neuroblasts. They occasionally show invasion, are usually circumscribed, and are found in the same locations as the other two tumors. Their behavior is hard to define because they can act like a ganglioneuroma and be benign or they can progress like a neuroblastoma and become highly malignant. The spinal canal becomes involved only by direct extension or metastasis. For this reason, it is hard to define whether surgery or chemotherapy and radiation therapy are required, but because of the frequent metastatic component, all three modalities often are necessary.

SOFT-TISSUE TUMORS

The most common soft-tissue tumors in the pediatric age group are sarcomas, with rhabdomyosarcoma occurring most frequently. These tumors tend to be parameningeal, involving the base of the skull or the cervical spine. Occasionally they appear in the retroperitoneal space or the urogenital tract. They invade from the paraspinal position, occasionally gaining access into the subarachnoid space. Occasionally neuroblastoma and Hodgkin and non-Hodgkin lymphomas are included in this soft-tissue group. Other, less common tumors include malignant hemangiopericytomas, malignant fibrous histiocytomas, and embryonal mesenchymal tumors.

BONY TUMORS

Bony tumors can be divided into two groups: primary (ie, arising within the bone) or secondary (ie, metastatic to bone). Primary bone tumors can be benign or malignant. The benign tumors are predominantly aneurysmal bone cysts and osteoblastomas, and the malignant forms include Ewing sarcoma, chordomas, chondrosarcomas, and mesenchymal chondrosarcomas.

Aneurysmal bone cysts are not actual neoplasms. Most are recognized before adulthood and are not uncommon. They may occur at any level of the spine and involve any portion of the vertebra. Pain is the most common complaint. Occasionally cord compression occurs and usually is associated with a collapse of the vertebra or hemorrhage into the aneurysmal bone cyst. Radiographs show an expansive destructive process with a soft-tissue mass. The characteristic CT finding is a lytic expansive lesion with egg shell-like peripheral calcification. MRI supports these findings and shows variable signal intensity. Frequently a fluid level is noted, as is high or medium-to-high intensity on T1- and T2-weighted images. Treatment is total resection. If the tumor cannot be removed en bloc, curettage is performed. Recurrence is frequent and more common in children than in adults. Because of the nonneoplastic character of the aneurysmal bone cyst, radiation is indicated only when removal is not total.

Osteoblastoma is the only benign tumor that favors the spine in childhood, with approximately a 40% to 50% occurrence in this area compared with other sites. There is a marked predilection for this tumor among children compared with adults, and males are affected twice as often as females. Usually the lumbar or sacral area is involved, although it can occur at any level. The most common presentation is pain and scoliosis.

Osteoblastomas appear on plain film as radiolucent areas surrounded by a region of new bone formation at various stages. There is often a soft-tissue component. Bone scanning may be of some use, but CT and MRI are preferred because they visualize both the soft-tissue component and bony involvement of the spinal canal. Both studies appear to be indicated.

Treatment involves surgical curettage and supplemental bone grafting. Total removal is required to prevent recurrence. When recurrence does occur, radiation therapy is indicated. Occasionally, osteoblastomas become malignant.

Primary malignant tumors of the spine are quite rare. Even though Ewing sarcoma is a common childhood tumor of bone, it is not common in the spine. Chondrosarcomas and mesenchymal chondrosarcomas are even rarer. Chordomas are very rare in childhood, with only 4% occurring before the age of 20. These tumors arise from a remnant of the notochord and usually occur at either end of the notochordal extent (ie, the skull base or the sacrococcygeal region). Although these tumors grow very slowly, they invariably compress spinal nerves and the cord and may metastasize to other portions of the body.

Plain radiographs document extensive bone destruction and a soft-tissue mass. CT demonstrates the bony destruction extremely well and frequently shows the characteristic randomly scattered amorphous calcification and septated areas of low attenuation. MRI provides a better contrast between the tumor mass and surrounding tissue, particularly on T2-weighted images.

Effective treatment requires total excision, but frequently this is impossible. Radiation therapy has been recommended, but it does not appear to be curative. Rather, it slows tumor growth. Chemotherapy has no role, and proton beam therapy has been attempted recently for these tumors. Gamma knife radiosurgery has shown excellent promise in management of intracranial tumors.

TUMORS THAT ARE METASTATIC TO THE BONE

Twenty-five percent of spinal tumors in children are metastatic. Metastatic bone involvement usually is diagnosed well after the primary tumor has been diagnosed. It is interesting that more than 80% of patients who have bony metastases have some involvement of the spine, with the thoracic spine being the most common site. Symptoms and signs are nonspecific, as are plain films, which only reveal bony destruction. MRI is the ideal study because it can scan the entire spine, recognize bony involvement before symptoms, and document the full extent of intraspinal disease. Treatment depends upon the histopathology of the primary source of metastases. In cases of cord compression, surgery is indicated, but metastatic tumors can be treated nonsurgically with radiation therapy or chemotherapy.

Summary

Tumors that arise outside the substance of the spinal cord can be divided into intradural but extramedullary lesions and extradural and extramedullary lesions. Different groups of tumors appear at different sites and behave quite differently. The presentation, however, can be quite similar and is distinct from that of an intraparenchymal spinal cord mass. The prognosis depends on the histopathology.

In cases of cord compression, surgery is indicated, but for many of the other tumors, radiation therapy and chemotherapy are appropriate, particularly if the lesion is a metastatic process. Both treatment approaches have obvious inherent problems in children. Extensive laminectomies can induce secondary spinal deformity in children, particularly kyphosis, and adverse effects of both chemotherapy and radiation therapy can cause significant stunting or asymmetry of growth or induce endocrine or other problems. It is critical for the pediatrician to recognize the patient who may have an intraspinal mass, to obtain an MRI scan quickly in those suspected of having an intraspinal mass, and to entertain the diagnosis of a psychiatric illness or social problem only after performing a full diagnostic evaluation. With this approach, many children who previously presented very late with diffusely spread disease may have their condition better controlled and possibly cured.