

Owoicho Adogwa and Richard G. Fessler

Although intradural tumors are uncommon, they should be considered in the differential diagnosis of patients presenting with back pain, radicular pain, sensorimotor deficits, or sphincter dysfunction. These tumors can be subclassified into extradural, intramedullary, and extramedullary spinal cord tumors on the basis of their anatomical relation to the spinal parenchyma. The heterogeneous cell composition of the intradural compartment allows the formation of neoplasms, arising from glial cells, neurons, and cells of spinal vasculature. In this chapter, we discuss the epidemiology, radiographic and histological characteristics, as well as the management of intradural extramedullary tumors.

17.1 Introduction

Primary tumors of the spinal cord are ten times less common than their intracranial counterparts and represent 2–4% of all primary tumors of the central nervous system (CNS) [1, 2]. Notwithstanding the lower incidence of spinal cord tumors, the histopathology is similar to primary intracranial neoplasms [3, 4]. The majority of primary spinal cord tumors are classified as low grade according to the World Health Organization (WHO) pathology classification [5–7] (Figs. 17.1 and 17.2).

Spinal cord tumors are subdivided into three categories on the basis of their relationship to the thecal sac that surrounds the spinal spinal cord and cauda equina: extradural, intradural extramedullary, and intramedullary. Extradural tumors are the most common and primarily consist of systemic cancer metastases,

O. Adogwa
Department of Neurosurgery, Rush University Medical Center,
1725 W Harrison St., Suite 955, Chicago, IL, USA

R.G. Fessler (✉)
University of Chicago, Chicago, IL, USA
e-mail: rfessler@rush.edu

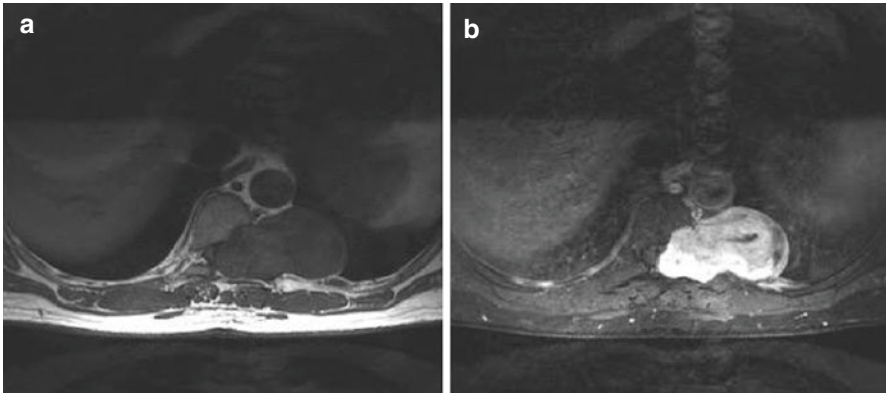


Fig. 17.1 Axial non-contrast T1-weighted (a) and post-contrast (b) images of a thoracic dumb-bell schwannoma

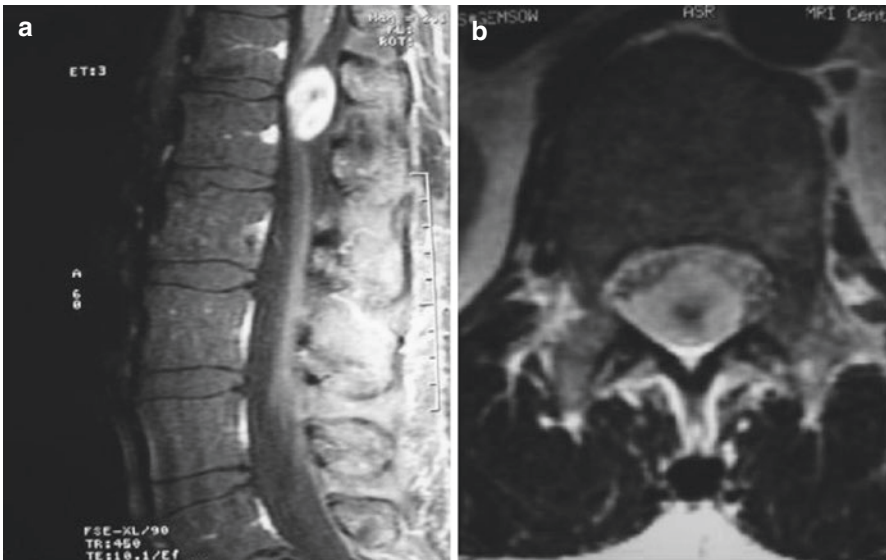


Fig. 17.2 Sagittal post-contrast T1-weighted (a) and axial non-contrast T2-weighted (b) images of a lumbar intradural extramedullary nerve sheath tumor

resulting in epidural spinal cord compression, and are not discussed further in this chapter on primary intradural extramedullary spinal cord tumors [8]. Intradural extramedullary tumors are more common than intramedullary tumors, representing 80% of all intradural tumors in adults and 65% of all intradural tumors in children, with the most common tumors being schwannomas, meningiomas, and neurofibromas [4, 8–10]. Other tumor types, such as hemangiopericytoma, lipoma,

paraganglioma, epidermoid cysts, and dermoid cysts, are less common. Intradural intramedullary spinal cord tumors (IMSCTs) constitute 20–30% of all primary spinal cord tumors. About 90% of IMSCTs are glial tumors, majority of which are ependymomas or astrocytomas [11–13]. Ependymomas represent about 60% and astrocytomas 30% [13, 14]. Of the intramedullary tumors with metastatic origins, 40–60% and 14% arise from primary neoplasms of the lung and breast, respectively [15–17] (Table 17.1).

Differentiation and diagnosis of spinal cord tumors are widely achievable through clinical examination and radiographic techniques. The clinical presentation is determined in part by the location of the tumor. Pain is the most common presenting symptom (72%) and may manifest as back pain (27%), radicular pain (25%), or central pain (20%) [18]. Motor disturbance is the next most common presenting symptom (55%) followed by sensory loss (39%; dermatomal, saddle, or segmental level) [18]. Sphincter disturbance is the least common presenting symptom seen in only 15% of all patients [18].

MRI is the preferred method of radiographic assessment of intradural spinal tumors and can suggest histological subtype. Other radiographic modalities, such as CT and CT myelogram, are useful if contraindications to MRI exist. When tumors are suspected to have a vascular component, magnetic resonance angiogram (MRA) or spinal arteriogram can be beneficial. Histological examination of tumor after biopsy or surgical resection is useful to establish the histogenesis of intradural tumors in almost all cases.

17.2 Evaluation of the Patient with a Spinal Tumor

The evaluation of the patient with a known or suspected spinal tumor begins with a thorough history and physical examination. Patients with vertebral column tumors most frequently present with back pain but may also manifest spinal deformity, neurologic symptoms, and systemic symptoms related to malignancy. Elderly patients with intradural tumors infrequently have radicular or back pain but may present with neurologic deficit from spinal cord or root compromise.

The radiologic evaluation of the patient with a spinal tumor includes MRI for nearly all lesions. CT is particularly helpful for spinal column tumors in assessing the degree of vertebral bone destruction and osteopenia as well as for surgical planning. CT myelography may be useful in patients who are unable to undergo MRI. Plain and dynamic radiographs should also be obtained to assess deformity and instability. Radioisotope bone scanning is highly sensitive for spinal column tumors that demonstrate osteolytic or osteoblastic activity and is most frequently used when searching for small lesions, such as osteoid osteoma, or for metastases in patients with known malignancy. Finally, angiography delineates the vascular supply of a tumor and may also be used to perform embolization to reduce intraoperative blood loss from hypervascular lesions, such as an aneurysmal bone cyst, hemangioma, renal cell carcinoma, melanoma, or chordoma.

Table 17.1 Intradural extramedullary tumors

Tumor	Prevalence (%)	Location	Radiology MR	Treatment	Prognosis
Meningioma	50	Cervical/ thoracic	T1, iso; T2, hyper Contrast enhancing	Resection	Good
Schwannoma	25	Thoracic > cervical	T1, iso/hypo; T2, hyper Contrast enhancing	Resection	Good
Neurofibroma	25	Thoracic, lumbar	Solid or plexiform T1, hypo; T2, hyper Contrast enhancing	Resection	Good
MPNST	Rare	No predilection	Rapid growth on serial MR, contrast enhancing	Resection/ RT/CT	Poor
Leptomeningeal metastasis	Common	Lumbar	T1, iso Contrast enhancing	Intrathecal CT	Poor
Paranglioma	Rare	No predilection	T1, iso; T2, hyper Contrast enhancing	Resection	Fair
Myxopapillary	Most common	Cervical, thoracic	T1, iso/hypo	Surgery; RT/CT	Good
Ependymoma			T2, hyper Contrast enhancing	If unresectable	
Mesenchymal lipomas	Rare	Lumbar	T1, hyper T2, hyper STIR, hypo Non-enhancing	Surgery	Good
Dermoid	Rare	Lumbar	T1, hypo T2, hyper FLAIR, hyper STIR, hypo DWI, bright Non-enhancing	Surgery	Good
Epidermoid	Rare	Lumbar	T1, hypo/hyper T2, hyper FLAIR, hyper STIR, hypo DWI, moderately bright Non-enhancing	Surgery	Good

17.3 Incidence and Etiology

17.3.1 Nerve Sheath Tumors

Nerve sheath tumors (NSTs) are categorized as neurofibromas or schwannomas. Although histological and immunohistochemistry studies support a common Schwann cell origin for both tumor types, each of these tumor types displays distinct clinicopathological characteristics during the formation of intradural, extramedullary spinal tumors and as such merits separate consideration.

NSTs account for about 23–25 % of intradural spinal cord tumors in adults and about 14 % in pediatric patients [15–17]. The peak incidence of NSTs is between the fourth through sixth decade of life, with no gender predilection [19]. Schwannomas are more common than neurofibromas and usually present as solitary tumors that occur proportionally throughout the spinal canal. Neurofibromas often show multiplicity, especially when associated with neurofibromatosis type 1.

The majority of nerve sheath tumors arise from a dorsal root although neurofibromas represent a higher proportion of ventral root tumors [20]. Most spinal NSTs (75–80 %) reside intradurally, but about 30 % of these tumors extend through the dural root sleeve as a dumbbell-shaped tumor with intradural and extradural components [20]; 10 % of spinal NSTs are located extramedullary and 1 % are located intramedullary. Intradural nerve sheath tumors most commonly affect the lumbosacral region, but cervical and thoracic tumors have been reported as well. The intramedullary NSTs are thought to arise from the perivascular nerve sheaths that accompany penetrating spinal cord vessels. Although NSTs are generally regarded as benign neoplasms, they can be malignant in a few cases, where they are designated the term malignant peripheral nerve sheath tumors (MPNSTs). 0.7 % of spinal NSTs are malignant, resulting in an exceedingly poor prognosis (median overall survival of ~22 months), irrespective of cranial or spinal location [18].

On imaging, NSTs have an isointense signal on T1-weighted images (T1WI) and a hyperintense signal on T2-weighted images (T2WI), with variable enhancement ranging from a homogeneous to a peripheral ring-like enhancement, after contrast administration.

Surgical resection is the primary treatment for NSTs, which is obtainable in most cases. Subtotal resection of these tumors might be an option when the tumor is attached to the spinal cord or when the tumor exhibits an extradural component closely associated with vital structures, such as the vertebral artery in the cervical region. Radiotherapy and chemotherapy are usually reserved for tumors that have malignant histological characteristics. Tumor recurrence is less than 5 % and might have a high association with subtotal tumor removal.

17.3.1.1 Schwannoma

Schwannomas are the most common intradural extramedullary (IDEM) spinal tumors, representing 30 % of such lesions and occurring at a rate of approximately 0.3–0.4 cases per 100 000 persons per year [18, 21]. They are benign tumors (WHO grade I), although malignant subtypes exist and usually arise from the dorsal

sensory roots [22, 23]. Patients usually present in the fourth through sixth decades. Topographically, spinal cord nerve sheath tumors are located in the upper cervical region (16%), cervical cord (31%), thoracic cord (22%), conus medullaris (7%), and cauda equina (24%) [15, 17, 24, 25].

The majority are solitary and sporadic; however, there is an association with NF2 [26]. Patients with NF2 often have multiple schwannomas and have high risk for malignant transformation [26]. The NF2 protein is thought to be a member of the ERM family of proteins, which is responsible for linking cytoskeletal components with proteins of the cell membrane that regulate cytoskeletal dynamics and cell-to-cell communication [15, 16]. Mutations in *NF2* may lead to the development of vestibular schwannomas (classically bilateral tumors of cranial nerve VIII), neurofibromas, ependymomas, gliomas, and meningiomas [9, 10, 15].

Schwannomas are either discovered incidentally or patients present with mild sensory symptoms consisting of shooting pain or paresthesias with nerve palpation; spontaneous pain can occur, but is uncommon.

On MRI, schwannomas appear as solid tumors in the dorsal sensory root region, with displacement of the spinal cord, conus medullaris, or filum terminale. Two thirds of schwannomas are slightly iso- to hypointense on T1WI and hyperintense on T2WI. Nearly all show intense enhancement following contrast administration. Enhancement patterns are homogeneous in 67%, mildly inhomogeneous in 10%, and heterogeneous with areas of intratumoral cystic degeneration in 22%. Peritumoral edema is present in 37%. Hemorrhage and calcification are not usually present [19, 23, 26].

Grossly, schwannomas appear as smooth globoid masses that do not enlarge the nerve but are suspended eccentrically from it with a discrete attachment. Histologically, they consist of elongated bipolar cells with fusiform, darkly staining nuclei arranged in a compact interlacing fascicles with a tendency toward palisade formation. Antoni A areas are more compact stellate-shaped cells while Antoni B areas are a loosely arranged pattern [25, 27, 28].

Especially in the elderly asymptomatic patient, schwannomas may be followed with serial imaging given their usual benign behavior. For symptomatic patients or radiographically enlarging tumors, maximal safe surgical resection is the most effective treatment modality. In most cases, schwannomas have an easily identified plane of dissection, and thus gross total resection (GTR) is the primary form of treatment [17, 25, 26, 29, 30]. Surgery is associated with minimal morbidity and provides symptomatic relief, and with complete surgical en bloc resection, local control rates of 90–100% have been reported. Adjuvant therapy is typically not recommended, and incompletely resected tumors should be followed with serial MR imaging given the benign growth and natural history of these tumors. Malignant schwannomas should be treated with postoperative radiotherapy, even if total resection was achieved. At present there are no compelling data to suggest a role for either chemotherapy or targeted therapy, notwithstanding recent reports of response of vestibular schwannomas to bevacizumab and epidermal growth factor receptor inhibitors. In elderly patients with poor functional reserve that are unable to undergo surgical resection, stereotactic radiosurgery is an option for these patients.

17.3.1.2 Neurofibroma

Neurofibromas are usually benign tumors (WHO grade I) that arise from peripheral nerves. Two types are recognized: general (solitary, circumscribed, or globular) and plexiform. Solitary neurofibromas are usually discretely localized, globular, or fusiform nodules. Plexiform neurofibromas are characterized by redundant loops of nerve fiber bundles and tumor tissue intermixed in a disorganized pattern that extends over multiple nerve roots [30–33]. Unlike Schwannomas, neurofibromas encase nerve roots rather than displacing them.

Neurofibromas appear to be more common in patients with neurofibromatosis type 1 (NF1), a condition that results from a mutation in the neurofibromin 1 gene located on chromosome 17q11 [8, 12, 14, 15, 25]. NF1 is thought to encode a protease involved in Ras-GTP phosphorylation, which reduces activation of downstream mitogen-activated protein kinases (MAPKs) involved in cell proliferation and survival [8, 12, 14, 15, 25]. NF1 mutations are associated with an increased risk for the development of malignant peripheral NSTs and of a set of diverse tumors, including carcinoid tumors, optic nerve gliomas, pheochromocytomas, and rhabdomyosarcomas [15, 16].

On imaging, spinal neurofibromas are often indistinguishable from schwannomas. They are most commonly fusiform in shape, unlike schwannomas, which tend to be characteristically round. They encase the nerve roots with fibers interwoven with nerve tissue, in contrast to schwannomas, which commonly displace the nerve root due to their asymmetric growth. They are generally hypointense on T1WI and hyperintense on T2WI, although a T2 hyperintense rim and central area of low signal may be seen. Moderate enhancement is usually seen post-contrast.

If asymptomatic, neurofibromas can be followed with serial MR imaging. Patients with symptomatic or enlarging solitary tumors should undergo surgical resection. GTR with minimal morbidity can be achieved. The clinical results following resection of a plexiform neurofibroma associated with NF1 are poor because GTR is rarely achieved. Plexiform neurofibromas may undergo malignant transformation (malignant peripheral nerve sheath tumor [MPNST]). Radiotherapy or chemotherapy is almost never employed for benign neurofibromas. Patients with NF1 may be at risk for malignant degeneration following radiotherapy [41]. Data on chemotherapy use is limited to MPNSTs and is usually Adriamycin based as with other soft tissue sarcomas.

17.4 Meningioma

Spinal meningiomas account for up to 50% of intradural spinal neoplasms and are the most common spinal tumors in adults [8–10, 13]. They usually arise from meningotheial arachnoid cap cells embedded in the dura near the root sleeve, which accounts for their predominant lateral position [25, 27]. They may also arise from pia or dural fibroblasts, probably as a result of their mesodermal origin [8–10, 13]. Although intradural meningiomas are usually solitary lesions, multiplicity might be encountered when these tumors are associated with neurofibromatosis type 2. Although meningiomas can develop in any age group, the preponderance occurs in

individuals between the fifth and seventh decades. The majority (~80%) of spinal cord meningioma patients are women, and 70–80% occur in the thoracic region [8–10, 13]. The female preponderance in the adult population is thought to be due to the effect of estrogen, although the exact mechanism remains unclear [15]. In men, spinal cord meningiomas are equally distributed between the cervical and thoracic cord. Overall, 15% of spinal cord meningiomas occur in the cervical spine, 81% in the thoracic spine, and 4% in the lumbar spine [18, 19, 26, 34].

The vast majority are WHO grade I lesions [18, 19, 26, 34]. Genetic predisposition (NF2) and prior exposure to ionizing radiation are the only definite risk factors [18, 19, 26, 34]. Similar histological subtypes are observed in both intracranial and spinal meningiomas, including meningothelial, metaplastic, psammomatous, transitional, atypical, and clear cell types. The psammomatous, meningothelial, and transitional subtypes are the most common meningiomas of the spine and, for reasons that are unknown, show a lower risk for recurrence than their intracranial counterparts [18, 19, 26, 34]. Multiple genes have been associated with spinal meningiomas – complete or partial loss of chromosome 22 and of its associated gene *NF2* along with loss of 1p, 9p, and 10q has all been implicated [29].

Meningiomas are either discovered incidentally or present commonly with back pain (70%), motor dysfunction (60%), sensory disturbance (40%), and incontinence (40%) [18, 20, 21, 35].

Radiographically, plain films are usually normal. Calcification is very rare and may be present in 1–5% of cases. Bone erosion is atypical. On MR scans, most appear isointense with the spinal cord on both T1WI and T2WI and lightly hyperintense on T2W/fluid-attenuated inversion recovery (FLAIR) images. Meningiomas display intense, homogeneous enhancement following contrast administration. Occasionally, densely calcified meningiomas are profoundly hypointense on MR and demonstrate minimal contrast enhancement [18, 20, 21, 35].

Surgery is the most effective treatment modality. Most intradural meningiomas are noninvasive, benign neoplasms, helping with gross total resection of the tumor.

In most cases, meningiomas have an easily identified plane of dissection, and thus GTR is the primary form of treatment [17, 25, 26, 29, 30]. With complete surgical en bloc resection, local control rates of 90–100% have been reported; however, gross total resection is not always achieved mostly due to tumor location and the need to preserve neurological function. The tumor recurrence rate with total or subtotal resection is between 3 and 7%. Atypical and anaplastic spinal meningiomas have a higher tumor recurrence rate though rarely metastasize. Radiotherapy could be considered after subtotal resection or recurrence of spinal meningiomas though data on its effectiveness remains sparse.

17.5 Ependymoma

About 40% of spinal canal ependymomas arise within the filum terminale. Although filum terminale ependymomas can develop in any age group, the majority occurs in individuals between the third and fifth decades [19, 26]. There appears to be a gender predilection as they tend to occur in men slightly more often than in women.

Myxopapillary ependymomas are by far the most common histologic type encountered in the filum terminale. They are WHO grade I lesions and histologically consist of papillary arrangement of cuboidal or columnar tumor cells surrounding a vascularized core of hyalinized and poorly cellular connective tissue [19, 26]. Mucinous changes undergone by tumor cells distinguish them from variants of ependymomas.

Patients typically present with radicular symptoms, lower extremity sensorimotor deficits, and/or sphincter dysfunction.

Myxopapillary ependymomas are typically benign, well-circumscribed tumors. Radiographically, they appear as a circumscribed mass with hypointense signal on T1WI and hyperintense signal on T2WI. They demonstrate homogeneous enhancement following contrast administration. Histologically, myxopapillary ependymomas display ependymal rosettes or perivascular pseudorosettes, with the characteristic deposition of myxoid material around blood vessels.

Surgery is the preferred treatment option. GTR of myxopapillary ependymomas is feasible if the nerve roots in the cauda equina are not entrapped within the tumor. In situations where the neural elements are entrapped within the tumor, maximal safe resection or subtotal resection is the preferred option in order to minimize surgical morbidity. Focal fractionated radiotherapy seems to be effective at improving neurological outcome and reducing tumor recurrence rate after subtotal tumor resection or piecemeal total excision. These tumors can seed to the spinal subarachnoid space, in which case broader field radiation is used, but this is uncommon. Although chemotherapy is sometimes started for recurrent or disseminated myxopapillary ependymomas, results are unconvincing.

17.6 Paranglioma

Spinal paragangliomas are rare heterogeneous tumors of neural crest origin commonly found in the cauda equina and filum terminale [30–32]. They are usually benign, nonfunctioning, sympathetic tumors and histologically resemble extra-adrenal paraganglia [30–32]. Parangliomas tend to occur in the fourth to fifth decades of life and show a male predominance. They appear to have a predilection for the cauda equina and lumbar regions, although intradural thoracic or cervical paragangliomas frequently occur.

Radiographically, they appear as well-circumscribed vascular tumors that are difficult to differentiate radiographically and clinically from filum terminale ependymomas [30–32]. They are hypointense to isointense on T1WI and hyperintense on T2WI. They characteristically produce a heterogeneous “salt and pepper” pattern after contrast administration. Hemorrhage and intratumoral vessels with flow voids are common features of this tumor [10, 19, 26]. Scanning using radiolabeled metaiodobenzylguanidine (mIBG), a noradrenaline analogue with uptake independent of catecholamine secretion, can allow visualization of paragangliomas. Histologically, paragangliomas display a highly vascularized tumor bed containing round and polygonal cells grouped in clusters called zellballen. Immunohistological methods to detect chromogranin and synaptophysin can be useful for diagnosis.

They are usually benign tumors and GTR is the preferred treatment. Although catecholamine-secreting spinal paragangliomas are uncommon, preoperative screening for a hyperadrenergic state is necessary, particularly in elderly patients, to prevent hypertensive crisis during tumor removal. Laboratory analysis may include fractionated plasma metanephrines and 24 h urine catecholamine and metanephrine tests. Where an elevation is found, a clonidine suppression test can be done. Recurrence rate after sub-total resection (STR) is less than 6% and does not appear to be reduced by concomitant radiotherapy or chemotherapy. Although iodine-131-labeled mIBG can slow progression and improve remission rate for metastatic paragangliomas, efficacy in primary intradural paragangliomas remains unknown.

17.7 Leptomeningeal Metastases

Leptomeningeal metastases are a common complication of cancer. These tumors are commonly the result of drop lesions from intracranial metastasis. They may also represent drop lesions from intracranial neoplasms such as gliomas and medulloblastomas. Metastatic spread from intracranial lesions can involve dissemination of neoplastic cells through the cerebrospinal fluid (CSF), and multiple lesions are common. They occur most frequently in the thoracolumbar or thoracic spine and tend to present as localized pain and spinal tenderness. Patients may also present with a radicular pain pattern; motor and sensory deficits are common. Sphincter dysfunction is present in about 33% of patients.

The diagnosis can be challenging; however, early diagnosis and aggressive treatment can prevent irreversible neurologic deficits. Diagnosis is usually established by the demonstration of malignant cells in the cerebrospinal fluid (CSF) or by the presence of enhancing tumor nodules on spinal MRI [31]. Leptomeningeal metastases may present with three different imaging patterns: (1) diffuse, thin, enhancing coating of the surface of the spinal cord and nerve roots; (2) multiple small enhancing nodules on the surface of the cord and/or nerve roots; and (3) as a single mass in the lowest part of the thecal sac [31]. Unenhanced T1-weighted images might be normal or demonstrate nodular lesions that are isointense to the spinal cord, whereas the contrast-enhanced images demonstrate significant enhancement.

Intradural extramedullary metastasis usually suggests advanced widespread progression of the systemic malignancy; aggressive surgical resection of lesions is usually not recommended. Treatment is, therefore, directed toward palliative and functional goals. Radiotherapy allows improvement in neurological function in a large population of patients. Adjuvant treatment with intravenous or oral corticosteroids can improve neurological function as well and provide symptomatic relief.

17.8 CNS Lipoma

Lipomas are congenital, benign tumors that are found infrequently in the intradural compartment. They arise from premature disjunction of the neural ectoderm from the cutaneous ectoderm and are frequently associated with spinal

dysraphisms. They are typically seen in young patients, however, and can be infrequently seen in older patients [15, 19, 26, 29]. They are typically slow growing tumors and become symptomatic because of the mass effect from their size. Intradural extramedullary lipomas are frequently located in the lower thoracic and lumbosacral regions.

Patients typically present with dysesthetic sensory changes, pain, gait abnormalities, paresis, and incontinence. Radiographically, they are hyperintense on T1- and T2-weighted images and show signal hypointensity on fat-suppressed or STIR images. These lesions are non-enhancing on post-contrast MRI. Microscopically, lipomas consist of mature adipose cells and connective tissue.

Surgery is the preferred treatment, although timing of surgery remains a controversial topic. Furthermore, the absence of a cleavage plane and the intermingling of neural and fibrofatty tissue at the periphery of the tumor make GTR very challenging. Intraoperative electrophysiological stimulation with evoked EMG monitoring is often used to allow differentiation between the functional spinal cord and the tumor. When complete or near total resection is achieved, there is a 90 % long-term progression-free survival at 16 years compared with only 35 % at 10 years after subtotal resection. Unfortunately, the improvement of neurologic symptoms post-operatively is not universal.

17.9 Hemangiopericytoma

Intradural extramedullary hemangiopericytomas are uncommon neoplasms of the CNS with uncertain histogenesis. They are hypercellular tumors with frequent mitoses and necrosis seen on microscopic examination. Hemangiopericytomas have an aggressive clinical course with high recurrence rates. Accordingly, surgical resection is usually needed. Because of the notable vascularity of these tumors, preoperative embolization is often attempted. Radiotherapy has proven effective in reducing recurrence of intracranial hemangiopericytomas, but there is insufficient evidence relating to spinal locations.

17.10 Epidermoid Cysts

Epidermoid cysts are uncommon lesions that comprise less than 1 % of all spinal tumors. They are more frequent in children and are usually congenital neoplasms arising from heterotopic ectodermal cell implantation into the neural tube early in embryonic development. They can be acquired and are considered a late complication of lumbar puncture. Lumbar punctures with nonstyleted needles have been implicated and are thought to introduce epidermal elements into the spinal canal that slowly grows, resulting in an intradural extramedullary neoplasm.

Patients typically present with radicular pain, motor deficits, gait abnormalities, and, infrequently, sphincter dysfunction. Radiographically, imaging findings are variable. These tumors are usually iso- to hyperintense compared to CSF on all sequences. There is usually minimum enhancement with gadolinium.

17.11 Dermoid Cysts

Intradural dermoid tumors are considered one of the congenital midline cystic tumors. Dermoids are thought to originate from epithelial inclusions within the neural groove during development. These tumors most commonly affect the lumbosacral region, with rare reports of thoracic involvement. Although these tumors usually present in the first two decades of life, they can infrequently present in elderly patients. On gross appearance, they may have skin appendages such as hair follicles and glandular tissue, which helps differentiate them from epidermoids. Radiographically, they have a variable imaging appearance though are hypointense to hyperintense signal on T1-weighted images and isointense to hyperintense signal on T2-weighted images. Dermoids weakly enhance after contrast administration.

Gross total resection is preferred when possible, but adhesion to neural tissue can prevent aggressive techniques. When STR is performed, emptying of the cystic contents and removal of a portion of the capsule are advised. Dissemination of the cystic contents spontaneously or during tumor removal might produce a granulomatous meningitis treatable with corticosteroids. Recurrence of resected intradural dermoid cysts is uncommon, and malignant transformation is uncommon. Adjuvant radiotherapy or chemotherapy for non-operable or malignant cases has not been thoroughly studied.

17.12 Principles of Treatment

The objectives of the surgical treatment for any spinal tumor must be clearly defined and discussed with the patient. In some cases, diagnosis may be the primary goal, and for many extradural tumors, this can be accomplished by image-guided biopsy, with relatively high diagnostic accuracy. IDEM tumors, however, require open exploration to obtain a biopsy safely for diagnostic certainty.

For most IDEM tumors and a number of primary spinal column tumors, lasting cure is the goal of surgery. Numerous studies have demonstrated that negative margins with en bloc resection of primary malignant tumors of the spine significantly decrease recurrence rates and prolong survival [9, 10, 15]. The surgical approach must be tailored to meet this marginal goal.

Conversely, for most metastatic lesions, symptomatic relief and palliation are the most common goals of surgical intervention; therefore, the effects of any selected treatment on the patient's quality of life must be carefully considered. In fact, for carefully selected patients with spinal metastases, surgical intervention may offer the best chance of improved quality of life.

In addition to the goals of (1) diagnosis, (2) tumor removal for local control or cure, (3) circumferential spinal cord decompression, and (4) symptomatic pain relief, any approach to spinal tumors must take into account the stability of the spinal segments involved. Where indicated, treatment options should incorporate arthrodesis, deformity correction, and fixation for levels that have been destabilized by the tumor or by the treatment itself.

17.13 Surgery in Elderly Patients

Traditional open decompressive operations for spinal tumors can carry up to a 30 % complication rate, including neurologic deterioration, severe medical complications, massive hemorrhage, wound infections and dehiscence, hardware complications, cerebrospinal fluid (CSF) leaks, and death. This is particularly relevant for elderly patients with poor functional reserve in whom surgery is being considered. Wound complications after open surgery can affect up to 40 % of patients.

Typical open surgical approaches also have significant drawbacks. First, subperiosteal dissection is required; therefore, denervation and devascularization of the paraspinal musculature ensue. This iatrogenic injury has been shown to lead to significant diminishment in postoperative axial muscle strength and performance. Second, in addition to sacrificing the bony and ligamentous portions of the posterior tension band, open laminectomy in the cervical spine injures the semispinalis capitis and cervicis muscles, which are thought to provide the primary force for maintained extension of the head and cervical spine. These untoward effects can produce iatrogenic sagittal plane destabilization that may lead to a progressive spinal deformity, most frequently seen in the cervical spine. Also known as postlaminectomy kyphosis, such deformity may occur in up to 10–40 % of adults after laminectomy and is most common after intradural tumor surgery. Such deformity has been shown to affect outcomes negatively.

17.14 Minimally Invasive Techniques

The motivation to develop minimally invasive strategies to treat spinal tumors in elderly patients is driven by the significant complication rates associated with established surgical approaches to neoplastic spinal disease. The factors that influence the decision to use minimally invasive techniques in the treatment of spinal tumors are the same as those affecting the decision to initiate surgical therapy in the first place, namely, life expectancy; health status of the patient; tumor type, location, and extent; symptomatology; prior therapies; and spinal stability. The indications for intervention are intractable pain, neurologic deficit, spinal deformity, need for diagnosis, and tumor cure or control. To that end, the primary objective of minimally invasive surgery is to reduce approach-related injury to normal spinal anatomy around the lesion of interest. Ultimately, this should translate into shorter operative times, reduced blood loss, shorter hospital stays, fewer complications, less postoperative pain, reduced medication use, decreased medical resource use, and faster recovery times.

17.15 Treatment of Intradural Extramedullary Tumors

Patients' preoperative neurologic status and tumor histopathology are the most important factors in determining long-term neurological and functional outcome after surgery for patients with IDEM spinal cord tumors. Surgical morbidity has

been significantly improved by advances in minimally invasive surgery, microsurgical techniques, and intraoperative electrophysiological monitoring.

GTR remains the primary goal of treatment for most types of IDEM tumors. Intraoperatively, the ability to identify a tumor–spinal cord plane has been a guiding principle for successful tumor resectability. Some tumors have a cleavage plane that facilitates the resection; however, other more infiltrative tumors lacking a tumor–normal spinal cord interface, such as lipomas, may not have such a plane, making complete resection impossible. When a plane of dissection is absent, resection can be associated with poor surgical outcomes despite electrophysiological monitoring during the procedure.

The treatment of recurrent IDEM tumors is largely based on tumor histology and remains controversial. In cases of subtotal resection, patients should be routinely followed by serial MRI for evidence of tumor progression, and radiation therapy is offered to patients with recurrence or enlarging tumors. In the rare cases where the tumor has infiltrated the spinal cord parenchyma, radical resection results in high morbidity. Therefore, conservative tumor removal or tissue diagnosis followed by radiation therapy is common practice. Re-resection is offered to patients with recurring IDEM tumors, as most are amenable to safe, complete removal with a low recurrence rate.

17.16 Radiosurgery

Stereotactic radiosurgery (SRS) has also been used in the treatment of intradural tumors [4]. SRS typically has been used to treat IDEM nerve sheath tumors and meningiomas but has also been applied to some hemangioblastomas, paragangliomas, and hemangiopericytomas [5, 8]. In 75–100% of benign IDEM tumors, growth arrest or a decrease in tumor size has been seen on follow-up imaging after SRS. In 2005, Bhatnagar and colleagues reported 45 cases of benign intradural tumors treated by SRS with the CyberKnife [36]. The median treatment time was 59 min. Tumor doses (80% isodose line) ranged from 9 to 31 Gy, with a median of 16 Gy [36]. Symptomatic improvement was seen in 78% of patients, and the local control rate with a median follow-up of 8 months was 96%. No toxicity from the treatments was observed. Interestingly, 42% of the lesions had undergone prior surgery, and 20% had received prior external beam radiation therapy (EBRT) [20, 27, 36]. Although these types of studies show promise for this modality in the treatment of benign intradural tumors, dosing has yet to be defined, the follow-up periods are far too short for lesions with benign histology, and comparative studies are lacking. For the time being, SRS of benign intradural tumors should likely be reserved for those patients who are unable to undergo more definitive surgical treatment or for whom other treatments have failed. SRS may be of particular benefit in patients with phakomatoses and multiple tumors, such as neurofibromatosis and von Hippel–Lindau disease.

Conclusion

Although rare, intradural spinal cord tumors should be an important consideration in the differential diagnosis of the elderly patients presenting with back or radicular pain associated with neurologic deficits. Radiographic assessment

combined with histological examination helps with the identification of the histogenesis of these tumors, which is vital when exploring surgical and adjuvant treatment options and patient management. Advances in minimally invasive and microsurgical techniques have facilitated complete resection and definitive treatment in some cases; however, for more infiltrative IDEM tumors, adjuvant radiotherapy may be required. Early diagnoses and aggressive definitive treatment, when possible, optimize the management of these tumors.

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