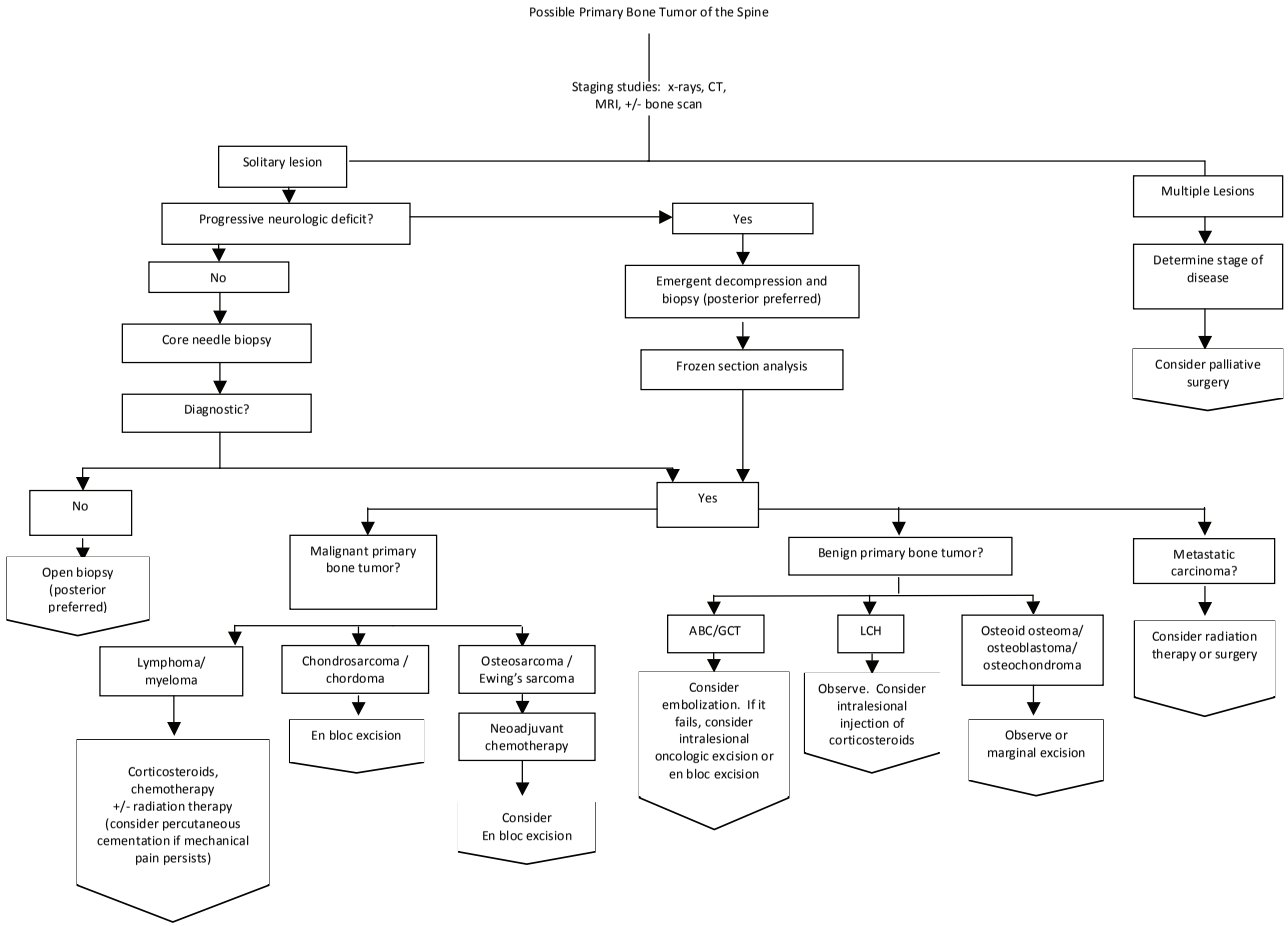
IX Spinal Tumors

Algorithm


Possible Primary Bone Tumor of the Spine
Staging studies: x-rays, CT, MRI, +/- bone scan
Solitary lesion
Progressive neurologic deficit?
Yes
No
Emergent decompression and biopsy (posterior preferred)
Core needle biopsy
Frozen section analysis
Diagnostic?
Multiple Lesions
Determine stage of disease
Consider palliative surgery
No
Open biopsy (posterior preferred)
Yes
Malignant primary bone tumor?
Benign primary bone tumor?
Metastatic carcinoma?
Lymphoma/ myeloma
Chondrosarcoma / chordoma
Osteosarcoma / Ewing's sarcoma
ABC/GCT
LCH
Osteoid osteoma/ osteoblastoma/ osteochondroma
Consider radiation therapy or surgery
Corticosteroids, chemotherapy +/- radiation therapy (consider percutaneous cementation if mechanical pain persists)
En bloc excision
Neoadjuvant chemotherapy
Consider En bloc excision
Consider embolization. If it fails, consider intralesional oncologic excision or en bloc excision
Observe. Consider intralesional injection of corticosteroids
Observe or marginal excision

46

Primary Spinal Tumors

Rex A. W. Marco

Primary bone tumors are those originating in bone (rather than metastasizing to bone). The treatment of benign and malignant primary spinal column tumors is diverse and complex. Most of these tumors present as a solitary lesion. The age, site, clinical presentation, and cell type must be considered prior to initiating treatment.

Classification

There are benign and malignant primary bone tumors. The neoplastic tissue is usually of mesodermal (bone, cartilage, fibrous, vessels, notochord) origin (**Table 46.1**).

Table 46.1 Classification of Benign and Malignant Primary Bone Tumors

Tissue of Origin	Benign Tumor	Malignant Tumor
Fibrous	Fibrous dysplasia Desmoplastic fibroma	Malignant fibrous histiocytoma
Cartilage	Osteochondroma	Chondrosarcoma
Bone	Osteoid osteoma Osteoblastoma	Osteosarcoma
Hematopoietic		Plasmacytoma Lymphoma
Vascular	Hemangioma	
Notochord	Ecchordosis	Chordoma
Unknown	Giant cell tumor Aneurysmal bone cyst Langerhans cell histiocytosis	Ewing sarcoma

Workup

History

The clinical presentation of primary bone tumors varies. A thorough history and physical examination will help direct the most appropriate care. Signs and symptoms of the tumor include persistent back pain, difficulty maintaining balance, wide-based gait, fatigue, bowel or bladder incontinence, paresthesias, and weakness of the extremities.

Physical Examination

A thorough physical examination should be performed in all patients with a solitary spine lesion, because the lesion may represent a metastasic deposit from a palpable organ such as the breast, prostate, rectum, testes, or thyroid. Moreover, sacral chordomas are often palpable on rectal examination.

Spinal Imaging

Conventional radiographs are obtained to evaluate the level of the lesion, the local anatomy, and the overall spinal alignment. A bone scan is helpful to confirm the solitary nature of the tumor. Computed tomography (CT) defines bony architecture and demonstrates characteristic findings of hemangiomas, osteoid osteomas, and osteochondromas. Magnetic resonance imaging (MRI) provides additional information about soft-tissue extension and neural involvement.

Special Diagnostic Tests

The precise diagnosis of the tumor is often dependent on obtaining tissue for pathologic analysis. A biopsy should be performed and thoroughly analyzed prior to proceeding with definitive care unless the patient has a progressive neurologic deficit due to spinal cord compression. However, most patients do not present with a progressive neurologic deficit even in the presence of marked spinal cord compression. Initiation of corticosteroids usually stabilizes the patient's neurologic function and provides some pain control in the short term. However, a biopsy should ideally be obtained prior to initiating steroids because corticosteroids have an oncolytic effect on myeloma and lymphoma, which might impair the diagnostic workup. Suboptimal technique in performing the biopsy may decrease the ability to perform complete excision of the tumor. A core needle biopsy rather than an open biopsy is preferred because a core needle biopsy minimizes contamination of the biopsy tract compared with an open biopsy. Minimizing contamination preserves surgical options, which include en bloc excisions. If the core needle biopsy is not diagnostic, then an open biopsy is indicated.

An emergent decompression with concomitant frozen section analysis is warranted in the rare individual presenting with a severe acute neurological in-

jury or progressive neurological deficit. A posterior biopsy and decompression procedure preserves more surgical options and can function as the first stage of an en bloc excision if deemed necessary. If the pathologist suspects a primary malignant bone tumor, then the thecal sac should be decompressed and an experienced spine tumor surgeon with expertise in en bloc excisions should be consulted as soon as possible. Minimizing local contamination with tumor cells is essential to decrease the likelihood of local recurrence and preserve treatment options. Decompression, tumor removal, and definitive stabilization are considered if carcinoma is identified on the frozen-section analysis.

Treatment

En bloc excisions of benign and malignant primary bone tumors of the spine can prolong survival and decrease the incidence of local recurrence more than intralesional excisions can. Although many symptomatic benign primary bone tumors can be successfully treated with an oncologic intralesional excision (complete removal of pathologic and reactive tissue), an en bloc marginal excision should be considered for patients with benign or malignant tumors isolated to the distal sacrum (below S2), the vertebral body, or the posterior arch of the vertebra. En bloc excisions of benign tumors involving the proximal sacrum or pedicle of the vertebral body may not be warranted because of the morbidity associated with these complex resections. Benign tumors involving the pedicle can be completely removed with an en bloc marginal excision combined with an intralesional excision of the tumor within the pedicle. En bloc excisions of tumors involving the proximal sacrum or vertebral body and pedicle are usually reserved for patients with malignant primary bone tumors.

Maintaining spinal stability by obtaining a solid fusion is especially important when treating patients with aggressive benign (giant cell tumor and osteoblastoma) and low-grade malignant (chordoma and low-grade chondrosarcoma) primary bone tumors, because these tumors have a relatively high incidence of local recurrence. A local recurrence in the setting of a solid fusion is a less challenging situation than a local recurrence in the presence of spinal instability. Appropriate oncologic excisions, combined with meticulous fusion techniques and biomechanically sound reconstructions, improve oncologic and functional outcomes by decreasing the incidence of local recurrence and increasing the likelihood of obtaining a stable spine fusion.

Conventional radiation therapy is not usually advised for the treatment of benign or malignant primary bone tumors. Most benign primary bone tumors are successfully treated with nonoperative measures or an appropriate oncologic excision. The potential benefit of decreasing the incidence of local recurrence thus does not usually outweigh the risk of malignant transformation or damage to the surrounding tissue following radiation therapy. Conversely, radiation therapy is not used for most malignant primary bone tumors because many of these tumors (chondrosarcoma, chordoma, and osteosarcoma) are relatively radioresistant. Ewing sarcoma and osteosarcoma are generally best treated with neoadjuvant chemotherapy followed by an en bloc excision for definitive local control. However, radiation therapy may decrease the incidence

of local recurrence in patients with Ewing sarcoma with residual microscopic disease following an oncologic excision with contamination. Additionally, stereotactic radiation therapy has been used to treat primary spinal sarcomas that have been deemed unresectable. Lymphoma is usually treated with chemotherapy alone, but radiation therapy is a useful adjuvant when necessary. Radiation therapy is also useful for patients with plasmacytoma and myeloma.

Complications

Incomplete resection of a lesion is associated with a high rate of recurrence for certain primary bone tumors. Other surgical complications, including infection, bleeding, failed fusion, and neurologic injury, are seen when surgical reconstruction is undertaken.

Outcome

The outcome is dependent on the type of tumor and the adequacy of resection.

Suggested Readings

Bergh P, Kindblom LG, Gunterberg B, Remotti F, Ryd W, Meis-Kindblom JM. Prognostic factors in chordoma of the sacrum and mobile spine: a study of 39 patients. Cancer 2000;88(9):2122–2134

This study reports the improved local control and survival of patients with sacral or mobile spine chordoma treated with en bloc excisions compared to intralesional excisions.

Boriani S, Bandiera S, Biagini R, et al. Chordoma of the mobile spine: fifty years of experience. Spine 2006;31(4):493–503

Forty-eight patients with chordoma involving the mobile spine were evaluated. Fourteen of these patients who received radiation alone, intralesional excision, or a combination had a local recurrence and died. Intralesional, extracapsular excision combined with radiation therapy resulted in local recurrence in 12 of 16 patients (75%), whereas only 6 of 18 patients (33%) who underwent en bloc excision had a local recurrence.

Boriani S, Biagini R, De Iure F, et al. En bloc resections of bone tumors of the thoracolumbar spine. A preliminary report on 29 patients. Spine 1996;21(16):1927–1931

The Weinstein-Boriani-Biagini (WBB) surgical staging system is discussed. This system divides the vertebra into 12 radiating zones in clockwise order from 1 to 12. This system allowed the authors to treat 29 patients with an en bloc resection uniformly. No local recurrences were reported.

Boriani S, De Iure F, Bandiera S, et al. Chondrosarcoma of the mobile spine: report on 22 cases. Spine 2000;25(7):804–812

These authors concluded that en bloc excision, with wide or marginal histologic margins, is the suggested management for chondrosarcoma of the spine. Three local recurrences occurred in 12 patients who had an en bloc excision compared to 10 recurrences in 10 patients treated with an intralesional excision. All patients who

had an en bloc excision were alive at last follow-up, while 8 of 10 patients who had an intralesional excision had died.

Boriani S, De Iure F, Campanacci L, et al. Aneurysmal bone cyst of the mobile spine: report on 41 cases. Spine 2001;26(1):27–35

These authors concluded that selective arterial embolization may be the first treatment option for aneurysmal bone cysts affecting the spinal column because 75% (3/4) of patients were cured with embolization. Moreover, embolization did not affect subsequent surgical treatment options.

Delauche-Cavallier MC, Laredo JD, Wybier M, et al. Solitary plasmacytoma of the spine. Long-term clinical course. Cancer 1988;61(8):1707–1714

Nineteen patients with plasmacytoma of the spine were treated with chemotherapy and radiation therapy. They reported an expected 85% 10 year-survival. Dissemination or local recurrence was observed in 13 of 19 patients.

Garg S, Mehta S, Dormans JP. Langerhans cell histiocytosis of the spine in children. Long-term follow-up. J Bone Joint Surg Am 2004;86-A(8):1740–1750

Twenty-six children with biopsy-proven Langerhans cell histiocytosis involving the spine were evaluated. Patients with solitary lesions were treated symptomatically with a spinal orthosis and followed clinically and radiographically. Two (8%) patients required surgical treatment for progressive deformity. These authors concluded that aggressive surgical management is usually not indicated for these patients.

Kawahara N, Tomita K, Murakami H, Demura S, Yoshioka K, Kato S. Total en bloc spondylectomy of the lower lumbar spine: a surgical techniques of combined posterior-anterior approach. Spine 2011;36(1):74–82

Ten patients with a spinal tumor of the lower lumbar spine underwent total en bloc spondylectomy (TES) by combined posterior-anterior approach. The authors found that the lumbar nerves were preserved by the combined posterior-anterior approach for spinal tumors of L4 or L5.

Lin PP, Guzel VB, Moura MF, et al. Long-term follow-up of patients with giant cell tumor of the sacrum treated with selective arterial embolization. Cancer 2002;95(6):1317–1325

The authors describe a reasonable alternative to surgical intervention of large giant cell tumors involving the sacrum or spinal column. They conclude that embolization should be considered in the treatment of this difficult disease because the risk-to-benefit ratio of surgical intervention with or without radiation therapy is probably higher than that of embolization.

Mankin HJ, Lange TA, Spanier SS. The hazards of biopsy in patients with malignant primary bone and soft-tissue tumors. J Bone Joint Surg Am 1982;64(8):1121–1127

These authors showed that biopsy-related problems occurred three to five times more often when the biopsy was performed at a referring institution rather than the treating center. The optimal treatment plan was altered in 18% (60/329 patients) of patients, and the prognosis and outcome were adversely affected in 8.5% (28/329) of patients. These authors concluded that the surgeon and the institution should be prepared to perform accurate diagnostic studies and to proceed with the appropriate definitive treatment or refer these patients to a treating center prior to biopsy.

Marco RAW, Gentry JB, Rhines LD, et al. Ewing's sarcoma of the mobile spine. Spine 2005;30(7): 769–773

This study evaluates a homogeneous group of patients with Ewing sarcoma of the mobile spine who were treated with multiagent chemotherapy combined with radiation therapy for definitive local control. These patients demonstrated improved survival rates compared to patients treated with radiation therapy alone. However, these patients had a high risk of local recurrence and postlaminectomy kyphosis. These authors concluded that current spinal resection and reconstruction techniques may lead to improved oncologic and clinical outcomes.

Marco RA, An HS. Chapter 32. In: McLain RF, Benzel E, eds. Cancer in the Spine (Handbook of Comprehensive Care): Complications, Anticipation and Management. Totowa, NJ: Humana Press; 2006:169–197

This is an excellent comprehensive review of the potential pitfalls involved with spine tumor surgery of the cervical, thoracic, lumbar, and sacral spine.

Rao G, Ha CS, Chakrabarti I, Feiz-Erfan I, Mendel E, Rhines LD. Multiple myeloma of the cervical spine: treatment strategies for pain and spinal instability. J Neurosurg Spine 2006;5(2): 140–145

The authors report the results of radiotherapy and surgical treatment of patients with myeloma involving the cervical spine. The authors suggest that external-beam radiation can effectively treat most patients with clinical or radiographically documented instability.

Schoenfeld AJ, Hornicek FJ, Pedlow FX, et al. Osteosarcoma of the spine: experience in 26 patients treated at the Massachusetts General Hospital. Spine J 2010;10(8):708–714

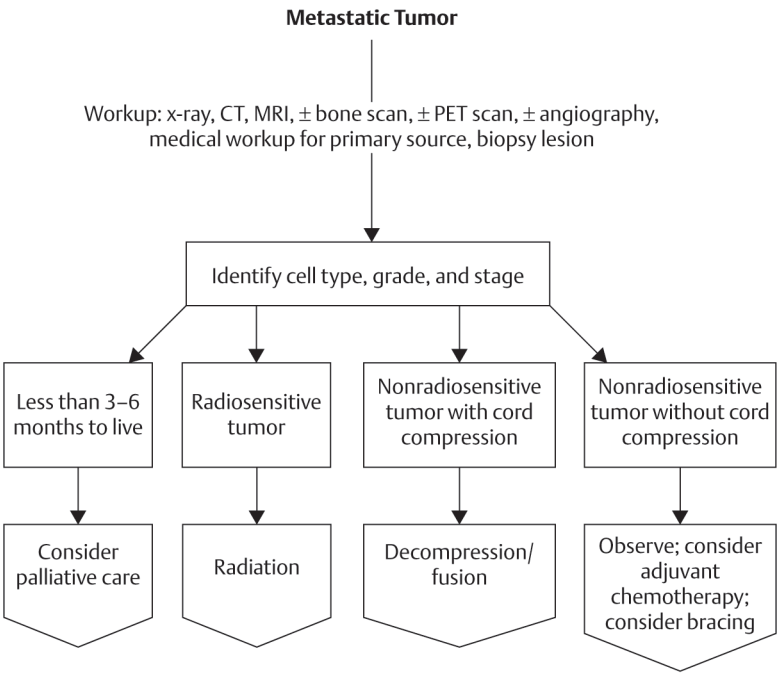
The authors confirm a poor prognosis for patients with osteosarcoma of the spine. They find that a combination of therapies, including surgery, chemotherapy, and high-dose radiation, achieve adequate short-term survival, but the 5-year mortality rate remains high.

Sundaresan N, Schmidek H, Schiller A, Rosenthal D, Eds. Tumors of the Spine: Diagnosis and Clinical Management. Philadelphia, PA: WB Saunders; 1990

This is an excellent review of the evaluation and treatment of benign and malignant primary bone tumors.

Tomita K, Kawahara N, Baba H, Tsuchiya H, Fujita T, Toribatake Y. Total en bloc spondylectomy. A new surgical technique for primary malignant vertebral tumors. Spine 1997;22(3): 324–333

These authors provide a step-by-step description of their en bloc spondylectomy technique.



CT, computed tomography; MRI, magnetic resonance imaging; PET, positron emission tomography.

Algorithm

47

Metastatic Spinal Tumors

Paul Kraemer and Rick C. Sasso

The spine is the most common site of skeletal metastatic disease. Spinal metastases account for the majority of spinal tumors encountered by spine surgeons. Breast carcinoma is the most common primary in women (**Fig. 47.1A,B**), while lung and prostate carcinoma are the most common primary sources in men. Renal, thyroid, and gastrointestinal tumors are also commonly seen, but in lesser frequency then the former mentioned tumors. Breast, prostate, and renal metastases are more likely to be seen by spine surgeons because of the longer relative survival compared with lung and gastrointestinal carcinoma.

Workup

The most common first symptom of spinal metastasis is mechanical pain, often well localized, insidious in onset, positional, and severe enough to disturb sleep. Patients may also present with various neurologic symptoms ranging from mild sensory or motor radiculopathy to complete paralysis due to neurologic compression, either directly from excessive tumor load or through instability of a pathologic fracture. Spinal metastasis may be the presenting complaint in a patient previously unaware of having a malignancy. The history should also address constitutional symptoms such as fever, chills, malaise, and weight loss as well as malignancy in the patient history and family history.

Radiographic evaluation begins with plain films looking for tumor location, bony destruction, soft-tissue extension, and pathologic fracture. Classic radiographic signs such as the "winking owl" (missing pedicle) or vertebra plana are present only after significant bony destruction. Magnetic resonance imaging (MRI) allows excellent evaluation of the neurologic elements and soft-tissue tumor involvement. Key tumor characteristics such as density and vascularity are also readily seen on MRI. Computerized tomography (CT) scans are helpful in evaluating bony lesions, determining stability, and help in preoperative

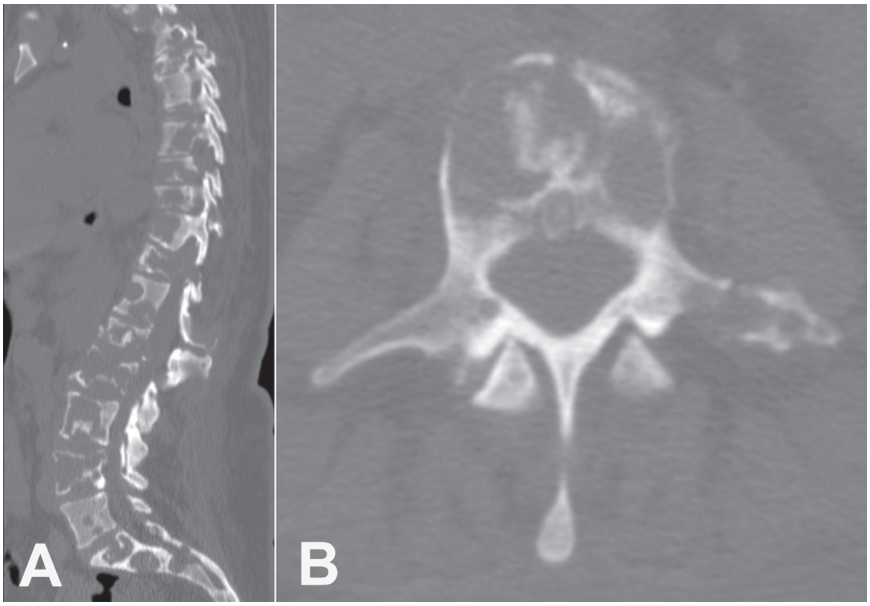


Fig. 47.1 CT of metastatic breast carcinoma of the vertebral column. **(A)** Sagittal. **(B)** Axial.

planning. Pathologic diagnosis should be obtained by biopsy, either CT-guided or open. Bone scans may be used as screening tools to identify other sites of metastasis, both in the spine and in the apical skeleton. Areas of metastasis identified outside of the spine, especially if they occur around the hips, should be imaged to evaluate for potential impending pathologic fracture.

For patients who present with metastatic lesions in the spine and who have not previously been diagnosed with cancer, a thorough examination should be performed. This often includes blood work, including complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), ionized calcium level, and serum and urine electrophoresis (SPEP and UPEP). Prostate-specific antigen (PSA) levels should be checked in men. CT scans of the chest, abdomen and pelvis should be obtained. Biopsy of either the spinal lesion or another lesion identified on imaging studies is necessary to make a definitive diagnosis.

Treatment

Goals of any intervention are improved quality of life via palliation of pain and prevention of neurologic worsening. The patient's overall health and prognosis must be taken into account when considering surgery or radiation, and close consultation with the medical oncologist, patient, and family should be the first step. Surgery should not be considered in patients who are hopelessly bedridden with an expected survival of less than six weeks.

Bracing is generally not considered except in terminal cases where no other options are reasonable, as there is no defined endpoint other than death. Surgical options should provide definitive stability without the need for adjunct bracing.

Vertebroplasty or kyphoplasty are useful for palliative treatment of select tumors wholly contained within a vertebral body. Indications for use are refractory pain without neurologic deficit. The posterior vertebral body wall must be intact to prevent extravasation of cement into the spinal canal.

Radiation therapy may be extremely useful for pain and tumor control in select pathologies, particularly germ cell and hematopoietic lines. Close coordination with the radiation oncologist is mandatory, as protocols change often, and variability in the direction of external beam may avoid irradiating a potential surgical approach.

Indications for surgical management include an isolated spinal lesion, pathologic fracture or deformity causing a neurologic deficit, or refractory pain and radioresistant tumors. The mainstay of treatment is intralesional excision and spinal reconstruction. Strong consideration should be given to "overfixing" with instrumentation, as immediate unbraced activity is necessary to maintain constitutional fitness, and solid osseus union may never be achieved in the face of an immunocompromised cancer patient. Anterior, posterior, and combined approaches all have a place in reconstructive efforts, largely depending on level, degree of instability, and surgeon preference. Multiple noncontiguous lesions may create a very difficult situation.

Patchels et al. in 2005 presented the only level I evidence comparing radiation to surgery. Exquisitely radiosensitive tumors were not studied and should be treated with radiation. Patients randomized to surgery had statistically better return of neurologic function, preservation of bowel and bladder function, relief of lower pain, and improved ambulation. Those who crossed over from radiation to surgery did not improve as much as those who were randomized to surgery, and the complication rate was higher. The study was stopped early by the review board because the results so heavily favored surgery over radiation. Though these results strongly favor surgical treatment of carcinoma and adenocarcinomas, a thorough discussion of risks and benefits of all options must be undertaken.

Outcome

Outcomes and prognosis for spinal metastasis vary based on several interrelated factors: primary tumor site, general health of the patient, number of extraspinal metastases, number of spinal metastases, metastatic involvement of internal organs, and severity of neurologic deficit on presentation.

Complications

Wound healing in cancer patients is often challenging. If radiotherapy is used as an adjunct to definite surgical stabilization, serious consideration should be given to deferring radiotherapy until after surgery to minimize complications of operating in an irradiated bed. Discussing the approach and incision with the radiation oncologist may help avoid unnecessary radiation to the incision.

Patients with diffuse metastatic disease may be systemically ill, cachectic, and malnourished. Aggressive nutritional support should be considered mandatory preoperatively as well as postoperatively and can help avoid or limit wound healing problems.

Suggested Readings

Abe E, Kobayashi T, Murai H, Suzuki T, Chiba M, Okuyama K. Total spondylectomy for primary malignant, aggressive benign, and solitary metastatic bone tumors of the thoracolumbar spine. J Spinal Disord 2001;14(3):237–246

Total spondylectomy was used to treat 14 patients with malignant or aggressive benign vertebral tumors. All patients had good pain relief and there were no serious complications. There were three local recurrences at 3.2 years.

Dudeney S, Lieberman IH, Reinhardt MK, Hussein M. Kyphoplasty in the treatment of osteolytic vertebral compression fractures as a result of multiple myeloma. J Clin Oncol 2002;20(9): 2382–2387

This article reports prospective evaluation of 55 kyphoplasties in 18 patients. Mean follow-up was 7.4 months. SF36 scores for bodily pain, physical function, vitality, and social function all significantly improved.

Fourney DR, Schomer DF, Nader R, et al. Percutaneous vertebroplasty and kyphoplasty for painful vertebral body fractures in cancer patients. J Neurosurg 2003;98(1, Suppl):21–30

Sixty-five vertebroplasties and 32 kyphoplasties were done in 56 patients with myeloma and primary malignant tumors. Median follow-up was 4.5 months with 84% complete pain relief.

Patchell RA, Tibbs PA, Regine WF, et al. Direct decompressive surgical resection in the treatment of spinal cord compression caused by metastatic cancer: a randomised trial. Lancet 2005; 366(9486):643–648

This landmark randomized controlled trial compared surgery to radiotherapy for metastatic lesions to the spine; it was ended early because significant benefits were seen for surgical patients.

Ryu S, Fang Yin F, Rock J, et al. Image-guided and intensity-modulated radiosurgery for patients with spinal metastasis. Cancer 2003;97(8):2013–2018

In this evaluation of 10 patients, most had significant pain relief within 2 to 4 weeks after treatment.

Sundaresan N, Rothman A, Manhart K, Kelliher K. Surgery for solitary metastases of the spine: rationale and results of treatment. Spine 2002;27(16):1802–1806

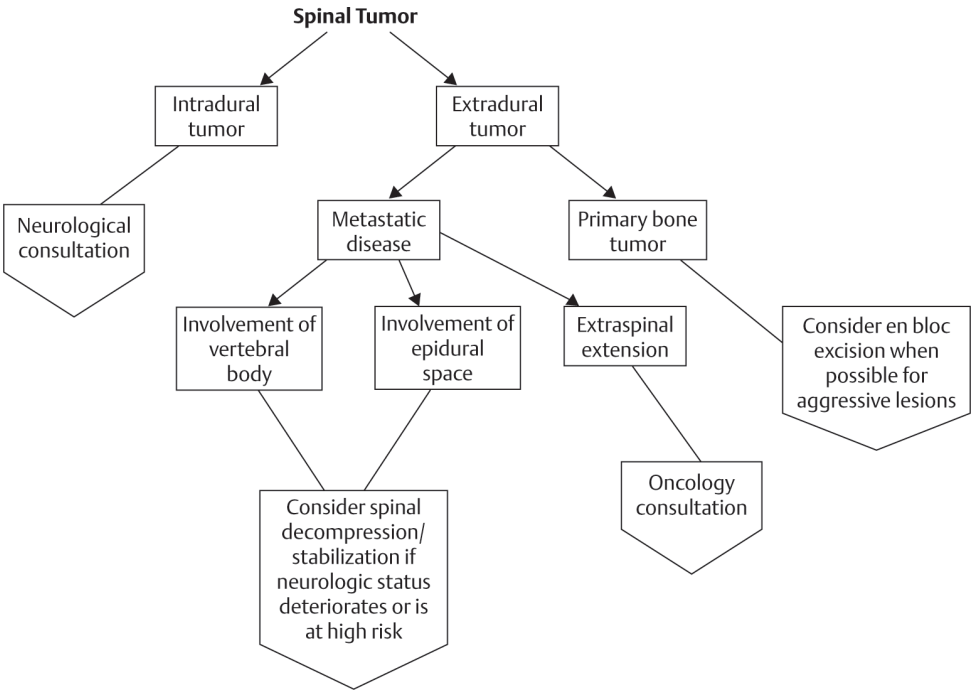
This is a retrospective review of 80 patients with solitary spinal metastasis from solid tumors. Median survival after surgery was 30 months. Surgical excision is recommended before radiotherapy to increase the chances of palliation and cure.

Wai EK, Finkelstein JA, Tangente RP, et al. Quality of life in surgical treatment of metastatic spine disease. Spine 2003;28(5):508–512

In this prospective evaluation of 25 patients undergoing surgery for spinal metastasis, the greatest improvement was with pain; however, improvements in constitutional symptoms were also seen.

Whyne CM, Hu SS, Lotz JC. Burst fracture in the metastatically involved spine: development, validation, and parametric analysis of a three-dimensional poroelastic finite-element model. Spine 2003;28(7):652–660

A finite-element study was undertaken to investigate features that contribute to burst fracture risk. The primary factors affecting fracture initiation were tumor size, magnitude of spinal loading, and bone density.


Spinal Tumor
Intradural tumor
Extradural tumor
Neurological consultation
Metastatic disease
Primary bone tumor
Involvement of vertebral body
Involvement of epidural space
Extraspinal extension
Consider en bloc excision when possible for aggressive lesions
Oncology consultation
Consider spinal decompression/ stabilization if neurologic status deteriorates or is at high risk

Algorithm

48

Classification, Staging, and Management of Spinal Tumors

Doniel Drazin, Noam Drazin, Amin Mirhadi, and Eli Baron

Tumors of the spine are relatively rare entities that affect only a small percentage of the population. They can, however, cause significant pain and neurologic compromise. Additionally, spinal metastasis, the most common spinal tumor, affects between 5 and 10% of cancer patients. In fact, ~50% of bony metastases involve the spine. The vertebral column is the most common site of spinal metastasis. The most common tumors that metastasize to bone include prostate, breast, lung, kidney, and thyroid.

Classification

The simplest classification for spinal tumors is based on the location within the spine. Tumors can be classified as involving the bony spinal column, extradural space, intradural extramedullary region, or intradural intramedullary. Magnetic resonance imaging (MRI) has greatly expanded the ability to place tumors in these categories.

Workup

The most common presenting symptom of spinal tumors is pain. Most patients with metastatic tumors present with back pain weeks to months prior to developing neurologic symptoms. Two pain patterns are typically seen in these patients: tumor-related pain and mechanical pain. Tumor-related pain typically occurs at night or in the early morning, improves with activity, and responds to administration of steroids. This pain may be caused by inflammatory mediators or be related to expansion of the tumor, causing stretching of the periosteum. Mechanical pain results from structural problems such as compression fractures. This pain is typically exacerbated with motion and does not respond to

steroids. Other extradural spinal tumors such as chordomas, osteoid osteoma, and aneurysmal bone cysts also typically present with pain. The pain associated with osteoid osteomas is exquisitely responsive to aspirin and nonsteroidal antiinflammatory drugs (NSAIDs).

Intradural extramedullary tumors such as spinal meningiomas (**Fig. 48.1**) and nerve sheath tumors also present with local pain. Patients with spinal nerve sheath tumors frequently have symptoms related to the affected nerve root (i.e., radiculopathy). Less often these tumors cause gait dysfunction, bowel and bladder dysfunction, and other symptoms of myelopathy.

Intradural intramedullary tumors such as spinal ependymomas and astrocytomas also tend to present with pain. Initially this is local, but it can also radiate elsewhere. Later-stage symptoms include myelopathy and, for filum terminale ependymomas, motor weakness and sphincter dysfunction.

Spinal Imaging

Magnetic resonance imaging (MRI), with and without gadolinium administration, of the whole spine is the imaging modality of choice for initial diagnosis of all spinal tumors. In the presence of known metastatic disease, it may be all

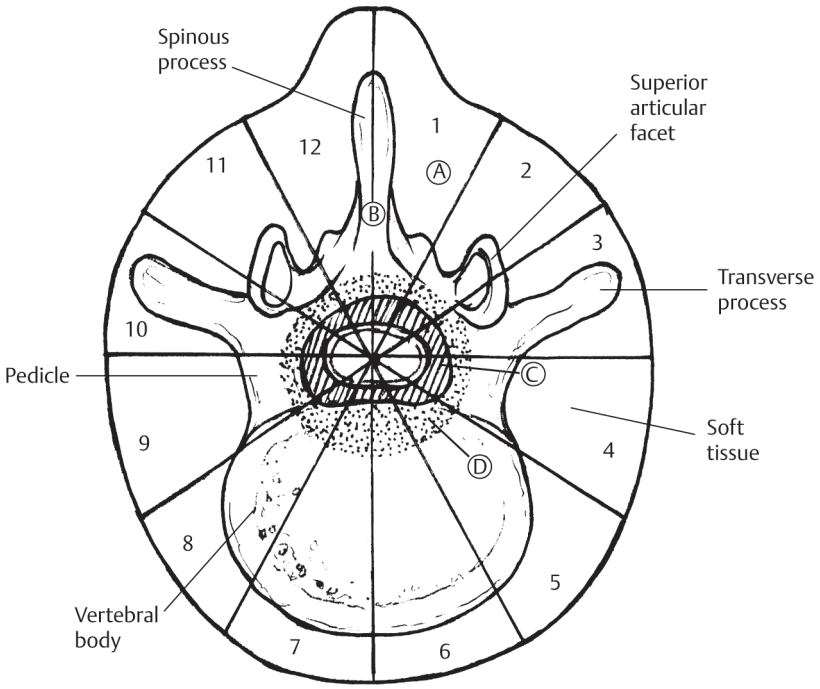


Fig. 48.1 Surgical staging. The location of the lesion using this scheme is useful for surgical planning prior to tumor excision.

that is required for workup. Consider myelography if other imaging modalities are contraindicated or inadequate. Other useful imaging modalities include nuclear medicine bone scans (technetium Tc 99) for workup of metastatic disease, computed tomography (CT), and plain radiographs for assessment of bony involvement for extradural lesions and assessment of spinal stability. Many primary bony tumors have distinct appearances on plain radiographs; thus, it is essential to obtain these imaging studies when bone involvement is suspected.

Though bone scans are more sensitive than plain radiographs in the detection of metastatic disease, they rely on osteoblastic reaction or bone deposition to detect metastatic disease. Thus, they may miss rapidly growing tumors and are insensitive for multiple myeloma and tumors of the bone marrow. Additionally, they are nonspecific and may also detect fractures, benign spinal tumors, and degenerative changes. Bone scans are most useful to screen the entire skeleton for evidence of metastatic disease. In cases of suspected metastatic disease, CT scan of the chest, abdomen, and pelvis may reveal a primary lesion. Needle biopsy is often useful to obtain an initial diagnosis in the case of extradural lesions.

Especially when considering surgery, angiography should be considered for preoperative embolization of vascular tumors (renal cell, thyroid, aneurysmal bone cysts, etc) to minimize operative blood loss.

Staging

Staging is commonly used for primary tumors of the spinal column such as sarcomas. Enneking proposed an oncologic staging system for musculoskeletal tumors that can be used for bony spinal column tumors. This system uses both radiologic and clinical data and presents two systems: one for benign tumors and the other for malignant tumors (**Table 48.1**).

Another scheme for surgical staging divides the transverse plane of the vertebra into 12 radiating zones, with five layers each (**Fig. 48.2A,B**). The tumor's longitudinal extent is calculated from the number of vertebral segments involved. MRI, CT, and even angiography may be useful in describing the transverse and longitudinal involvement of the tumor. This system may prove very useful in planning the surgical resection.

Treatment

Pharmacological and Minimally Invasive Management

The most feared aspect of a cancer diagnosis for most patients is the prospect of suffering from unrelieved pain. It has been reported that 50% of cancer patients receive inadequate analgesia. In patients with spinal metastases, the pain ranges from localized tenderness to progressive to severe and unremitting pain that may be nocturnal. It is important to offer analgesics in a stepwise order that allows for escalation of doses as well as adjuvant medications. The World Health Organization (WHO) outlined a three-step cancer pain relief ladder

Table 48.1 Enneking Staging System for Tumors of the Osseous Spine

Benign Tumors	
S1	Latent, inactive tumor: usually asymptomatic and bordered by a true capsule; often no management is required
S2	Slow, growing resulting in mild symptoms; thin capsule and reactive tissue; usually an intralesional excision can be performed with a low rate of recurrence
S3	Rapidly growing tumors where the capsule is very thin or absent; en bloc excision is usually appropriate management
Malignant Tumors	
IA	Low-grade malignant tumor confined to vertebrae
IB	Low-grade malignant tumor invading paravertebral compartments
IIA	High-grade malignant tumor
IIB	High-grade malignant tumor with infiltrating tumor spread beyond the cortical border with no gross destruction; often there is invasion of the epidural space
IIIA	High-grade malignant tumor with distant metastasis
IIIB	High-grade malignant tumor with infiltrating tumor spread beyond the cortical border with no gross destruction, with distant metastases

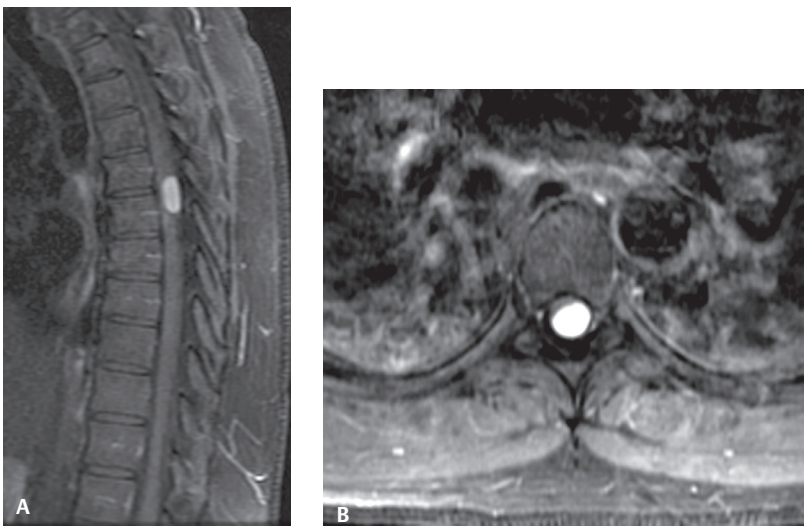


Fig. 48.2 MRI images, post contrast infusion, showing homogeneously enhancing intradural extramedullary meningioma compressing the spinal cord. **(A)** T1 sagittal. **(B)** Axial.

that has been shown to provide adequate analgesia to 90% of cancer patients. If the pain is mild, it is recommended to begin with NSAIDs. If pain persists or worsens, step 2 recommends weak opioids. If pain persists, strong opioids (e.g., morphine) should be started. For those patients who continue to have unremitting pain, a referral to a pain specialist should be considered. Additionally, a fourth 'interventional' step has been suggested that includes invasive procedures such as nerve blocks, spinal (epidural, intrathecal) infusions, and spinal cord stimulators.

Despite the use of all the aforementioned techniques for pain management, pain may persist. Some patients may be candidates for other neurosurgical analgesic techniques, namely radiofrequency ablation, vertebroplasty, or kyphoplasty. Radiofrequency ablation is a technique in which thermal energy is applied to the metastatic region, as guided by imaging. Under this high intensity of heat, tumor cells die immediately. Radiofrequency ablation is an option for patients in whom surgery is contraindicated, as it can be performed with local sedation. Initial studies have shown promising results, as a great majority of patients report a high degree of pain reduction that persists over time. While the accuracy provided by the imaging component, the immediacy of cell death, and the ability to use conscious sedation are the benefits to radiofrequency ablation, it is important to consider the proximity of the lesion to vulnerable structures and organs before applying this level of heat. As a guideline, it has been recommended that this technique be used only if the nidus is at least 1 cm from vital structures. There is also a rare, but reported, risk of local burning at the site of ablation.

Vertebroplasty is a procedure in which cement (typically polymethylmethacrylate, PMMA) is injected percutaneously into the vertebral body, guided by imaging, thereby solidifying and stabilizing vertebrae weakened by lesions and fractures. There is slightly more research on vertebroplasty than on radiofrequency ablation, and results (both with metastatic and with osteolytic lesions) show that a great majority of patients indicate a high degree of significant and lasting pain relief with this procedure. The pain relief is hypothesized to be a result of stabilizing the fractures, reducing mechanical forces, and/or killing nerve endings. In addition to the described benefits, it is also considered minimally invasive, making it accessible for a broad patient base. The main complication of this technique, however, is the possibility for PMMA to leak outside the vertebral body. PMMA extravasation is usually clinically insignificant but, rarely, has caused various deleterious secondary effects including pulmonary embolus. The rates of cement leakage vary widely, ranging from 2% to 73% per level, with little known definitively about what causes increased risk. There is also a correlation between the use of this technique and the development of new compression fractures in the vertebrae adjacent to the level treated with vertebroplasty.

In attempt to lessen the risk of cement leakage reported with vertebroplasty, the kyphoplasty procedure was developed. It varies from its predecessor in that a balloon is inserted into the vertebral body that, when inflated, decreases the fracture and, when deflated, creates a space to hold the cement. The balloon also enables the use of a higher-viscosity version of cement than PMMA, which, in combination with a lower injection pressure, decreases the risk for extravasation. Research shows a comparable level of pain relief to the high rate report-

ed for vertebroplasty (80–100% reporting decreased pain after the procedure). As intended, research shows that kyphoplasty is, in fact, associated with a lower rate of cement leakage. Additionally, one of the purported advantages of kyphoplasty, shown in cadaver studies, is a significantly increased height restoration (97%) as compared with vertebroplasty (30%). These important improvements have led some to believe that kyphoplasty should be the standard of care for patients with compression fractures. However, the procedure more often requires general anesthesia, takes longer, and may necessitate an overnight hospital stay. Randomized controlled trials, in time, will show whether the benefits of kyphoplasty over vertebroplasty outweigh these drawbacks.

Adjuvant medications are important part of cancer pain management, as they often enhance the efficacy of analgesics, alleviate concurrent symptoms, and address specific types of pain. Most commonly used adjuvants are corticosteroids and bisphosphonates (diphosphonates), while tricyclic antidepressants and anticonvulsants (gabapentin) have been used less frequently but appear to benefit some cancer patients with painful neuropathies.

Corticosteroids can help alleviate acute nerve compression, increased intracranial pressure, and soft-tissue infiltration. Unless contraindicated, most physicians will typically begin with at least 16 mg of dexamethasone and continue that dosage while treatment is being planned. If neurological function worsens, the dose is often increased temporarily. It is important to monitor blood glucose levels and side effects (proximal myopathy, psychosis, weight gain) in patients receiving corticosteroids. After surgery or the start of radiation therapy, the dose should be tapered gradually.

Bisphosphonates, such as clodronate, palmidronate, and zoledronic acid, have been used to treat bone pain and prevent skeletal complications of metastases. Based on several controlled trials, bisphosphonates provide a meaningful supportive but not life-prolonging benefit in patients with bone metastases. Specifically, patients with vertebral involvement from myeloma, breast cancer, and non-small-cell lung cancer may be offered biphosphonate to reduce pain and risk of vertebral fracture. Most experts agree that it takes at least 6 months of bisphosphate therapy before an effect is seen on skeletal morbidity outcomes. The potential benefits are still the topic of ongoing research; the risk of osteonecrosis of the jaw is a late toxicity has recently been recognized.

Surgery, Chemotherapy, and Radiation

Treatment depends on the cell type, grade, and degree of involvement of the spine. Common modalities include surgery, radiation, chemotherapy, or a combination of these. The extent and type of surgery depend on the goals of the procedure, with most surgery being performed for palliation or restoration of neurological function and only in rare cases curative. In general, most spinal tumors should be resected to “image complete” standards. The discussion below will assume complete or near complete surgical excision was possible.

It is helpful when discussing treatment options to classify tumors by location: intramedullary, intradural-extramedullary, and extradural.

Intradural tumors arise within the spinal cord itself and are most commonly ependymomas, astrocytomas, or metastases. Infiltrating tumors and malignant

astrocytomas cannot be resected and are best treated with radiation followed by chemotherapy. There is no currently proven role for chemotherapy in recurrent or metastatic spinal cord ependymomas. Additionally, the role of chemotherapy in spinal cord astrocytomas has yet to be defined. A literature review identifies only small observational studies in the mostly pediatric population. This scant research shows a limited number of chemotherapy regimens with some activity. Metastatic disease to the spinal cord is becoming more frequently recognized with increased use of MRI. Chemotherapy for metastatic disease is targeted to the tissue type of the primary tumor (e.g., breast or lung cancer). Another possible treatment option is focused radiation therapy. Historically, radiation has been given in a standard fractionated approach for both benign and aggressive tumors. This is mainly because the spinal cord has a limited tolerance to radiation dose. When the dose is exceeded, it can lead to demyelination, vascular injury, and subsequent paralysis. Radiation therapy, however, can be a very effective tool for both primary treatment as well as in the adjuvant setting. In the case of infiltrating tumors, usually these can be treated with standard fractionation up to 60 Gy (or at cord tolerance), but this dose may be adjusted depending on the aggressiveness and size of the tumor. Ependymomas are typically treated to 50 Gy and have up to 90% local control. Intradural extramedullary tumors arise from the dura but are outside the actual spinal cord. The most common types are meningiomas and nerve sheath tumors. The majority of intradural extramedullary tumors such as meningiomas and schwannomas are treated surgically via a standard posterior laminectomy. More ventrally located tumors may require facetectomy or a lateral extracavitary approach to the spine. Intradural intramedullary tumors are also usually approached via posterior laminectomy. Neurophysiologic monitoring including somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) is routinely used. Intraoperative ultrasound may be used to localize and determine the extent of tumor at surgery, as often the intramedullary tumor is not apparent by inspection of the surface of the cord. Usually a myelotomy is performed through the posterior midline septum, and a biopsy is taken during surgery. The intraoperative pathology can help to determine whether an aggressive resection is warranted. There is no defined role for chemotherapy in this tumor type. There are multiple clinical trials looking at extraaxial cranial meningiomas, but none currently looking at spinal meningiomas. With regards to radiation therapy, tumors in this location are much more amenable to treatment, especially with the utilization of more precise technology such as intensity modulated radiotherapy (IMRT), image-guided radiotherapy (IGRT), or stereotactic body radiotherapy (SBRT). Essentially, these types of treatment deliver precisely focused X-rays to the specific target and can avoid the spinal cord altogether, thus enabling dose escalation and better tumor control.

Extradural tumors are usually metastatic and arise most often in the vertebral bodies. Symptoms can be caused by spinal cord compression by epidural growth or direct intradural invasion. The most common primary tumors with metastatic disease to the spine are prostate cancer, breast cancer, and lung cancer. Most metastatic lesions in patients with limited life expectancy are amenable to radiation treatment alone. One study suggested that even in the face of epidural compression from metastatic disease, radiation was as effective as

decompressive laminectomy (without fusion) with regard to neurologic outcomes. Nevertheless, an aggressive surgical approach to metastatic disease is preferred in certain patients. Indications for surgical intervention in the presence of metastatic spinal involvement are listed in **Table 48.2**. Chemotherapeutic modalities in the treatment of extradural tumors are directed toward the primary tumor. Radiation therapy can be used as either primary or adjuvant treatment. However, whenever there is evidence of epidural cord compression, surgery should be considered first. Radiation therapy may help supplement the likelihood of local control, but it is significantly more effective when combined with tumor debulking, and can also improve functional outcomes.

Primary spinal tumors should be considered for en bloc resection. If a biopsy is performed, the tract should be placed in line with the future incision site for surgical resection of the tumor, so that the tract can be excised in one piece with the specimen.

Outcome

Spinal tumors are classified based on the anatomic compartment of the spine that they occupy. MRI is the initial imaging modality of choice for most spinal tumors and allows rapid classification based on tumor location. Primary tumors of the bony spinal column should be ontologically and surgically staged. A multidisciplinary approach including orthopedic surgery, neurosurgery, medical, and radiation oncology should be used to achieve optimal management of spinal neoplastic disease.

Table 48.2 Indications for Surgery in the Patient with Metastatic Disease

Radioresistant tumors (e.g., sarcomas, renal cell carcinoma)
Spinal instability
Pathologic fracture with bony cord compression
Circumferential epidural tumor with radioresistant lesion
Acute neurologic deterioration secondary to compressive disease with lack of improvement with steroids and unknown diagnosis
Post–radiation treatment with progressive neurologic symptoms or cord compression due to neural compression by tumor
Rule out residual tumor post–radiation therapy/chemotherapy for certain tumors (e.g., Ewing sarcoma, germ cell tumors)

Note: All patients undergoing evaluation for surgery for metastatic spinal lesions should undergo a full workup and assessment of systemic disease to be factored into any surgical decision making.

Suggested Readings

Aebi M. Spinal metastasis in the elderly. Eur Spine J 2003;12(Suppl 2):S202–S213

This article reviews spinal metastasis in the elderly including various pharmacologic, radiation, and surgical management strategies.

Bilsky MH, Lis E, Raizer J, Lee H, Boland P. The diagnosis and treatment of metastatic spinal tumor. Oncologist 1999;4(6):459–469

This work provides an excellent overview of spinal metastases and their presentation and treatment.

Boriani S, Weinstein JN, Biagini R. Primary bone tumors of the spine. Terminology and surgical staging. Spine 1997;22(9):1036–1044

This article reviews in detail oncologic and surgical staging of primary osseous spinal tumors.

Gilbert RW, Kim JH, Posner JB. Epidural spinal cord compression from metastatic tumor: diagnosis and treatment. Ann Neurol 1978;3(1):40–51

This classic article compares outcomes of patients undergoing laminectomy and radiation for epidural compressive spinal metastases with those of patients undergoing radiation alone. The authors find no differences in outcomes between the two groups.

Halpin RJ, Bendok BR, Liu JC. Minimally invasive treatments for spinal metastases: vertebroplasty, kyphoplasty, and radiofrequency ablation. J Support Oncol 2004;2(4):339–351, discussion 352–355

This article reviews the minimally invasive treatment options—vertebroplasty, kyphoplasty, and radiofrequency ablation—for spinal metastases.

Parsa AT, Lee J, Parney IF, Weinstein P, McCormick PC, Ames C. Spinal cord and intradural-extraparenchymal spinal tumors: current best care practices and strategies. J Neurooncol 2004; 69(1-3):291–318

This work reviews the presentation, diagnosis, and current management strategies with intramedullary and intradural extramedullary spinal tumors.

Ross JR, Saunders Y, Edmonds PM, Patel S, Broadley KE, Johnston SR. Systematic review of role of bisphosphonates on skeletal morbidity in metastatic cancer. BMJ 2003;327(7413):469

This review article provides evidence for the use of bisphosphonates to reduce skeletal morbidity in patients with bone metastases.

Sundaresan N, Boriani S, Rothman A, Holtzman R. Tumors of the osseous spine. J Neurooncol 2004;69(1-3):273–290

This article reviews primary osseous tumors of the spine, and approaches to classification, staging, and surgical strategies.

Van Goethem JW, van den Hauwe L, Ozsarlak O, De Schepper AM, Parizel PM. Spinal tumors. Eur J Radiol 2004;50(2):159–176

This work provides a detailed overview of the various spinal tumor types and their treatment.