Top Ten Tips Palliative Care Clinicians Should Know About Spinal Tumors

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Abstract

Nearly 20% of cancer patients develop symptomatic spine metastases. Metastatic spine tumors are most commonly extradural tumors that grow quickly and often cause persistent pain, weakness, paresthesias, urinary/bowel dysfunction, and/or paralysis. Surgical intervention aims to achieve more effective pain management, preserve/restore neurological function, provide local tumor control, and stabilize the spinal column. The desired result of treatment is ultimately to improve a patient's quality of life. Neurosurgeons employ multiple decision frameworks and grading scales to assess the need and effectiveness of a variety of surgical interventions ranging from minimally to maximally invasive. Likewise, palliative care offers an array of treatment options that allows the best, individualized plan to be determined for a given patient. Therefore, crossfunctional collaboration between palliative care, radiation oncology, medical oncology, and neurosurgery is crucial both in the maximization of available treatment options and optimization of quality of life for patients.

Keywords: extradural tumors; intradural tumors; metastatic spinal tumors; neurosurgery; palliative treatment; spinal tumors

Introduction

PALLIATIVE CARE PHYSICIANS need to be familiar with the management of spine tumors as they commonly present in patients with cancer causing significant pain and disability. Metastatic lesions involving the spine can occur in up to 40% of cancer patients and ~15% of all primary central nervous system lesions arise from the spinal cord.¹ Spine tumors present in variable ways from insidious onset to more urgent neurological deficits and are treated using both conservative and aggressive means. The ultimate management paradigm depends on patient-specific factors, tumor-specific factors, and the clinical scenario. In this article, we review common questions and problems in spine tumor management pertinent to palliative care clinical practice.

Tip 1: All Tumors Are Not Created Equal: Intradural and Extradural Tumors Have Different Work-up and Management

Spine tumors are broken down into two major categories, extradural or intradural. It is helpful to know these delineations as they present in predictable ways and warrant different imaging tests and management approaches. Intradural

(within the dural sac covering the spinal cord and nerves) are further broken down into either intramedullary (within the spinal cord itself) versus extramedullary (outside the spinal cord but inside the dura).

Intradural tumors are divided into extramedullary and intramedullary. Extramedullary tumors are mostly benign and include nerve sheath tumors, meningiomas, and ependymomas, whereas intramedullary tumors include ependymomas, astrocytomas, or hemanangioblastomas. Intradural extramedullary tumors of the spine present with gradual neurological symptoms, such as axial back pain or radicular pain if they are compressing a nerve root. In the cervical or thoracic spine, they can present with “long-tract” or upper motor neuron signs, such as spasticity, unsteady gait, weakness, and hyperreflexia. Clinical features of intramedullary spinal cord tumors are variable and may present three to four years before diagnosis.² Abrupt symptoms are more likely due to a hemorrhage of this tumor type. Diagnosing an intradural tumor primarily occurs after MRI and a neurosurgical consult is appropriate to determine optimal management. In surgical cases, the goals of treatment for intradural tumors include biopsy for diagnosis, surgical debulking to relieve mass effect, and complete surgical resection. The extent of resection

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of an intramedullary tumor depends on the extent of tumor infiltration (i.e., whether there is a plane separating the tumor from the spinal cord), thus for some tumors complete resection is not always possible without incurring a significant neurological deficit. In cases of incomplete resection, post-operative radiation may be required.

Extradural tumors are most commonly metastatic and include tumors of the bony spine. Extradural metastatic tumors and primary bone tumors, such as chordomas, chondrosarcoma, Ewing's sarcoma, plasmacytoma, and multiple myeloma are considered as malignant neoplasms. Benign primary bone tumors include aneurysmal bone cysts and osteomas. Malignant or metastatic tumors typically show early symptoms within weeks to months.² Extradural disease grows quickly and causes both pain and compressive symptoms on the spinal cord and/or nerves. Compressive symptoms in the lumbar spine may be radicular with shooting pain down the leg or weakness. In the thoracic and cervical spine, weakness, gait instability, and bowel/bladder symptoms are more common. Because these tumors commonly arise from the bone, CT scan is appropriate to assess bony destruction from extradural tumors, whereas MRI can be used to evaluate the extent of epidural compression i.e., compression of the spinal cord or nerves. PET scan and X-rays also demonstrate the extent of bony invasion. Management of extradural metastasis depends on a variety of patient and tumor factors but can include surgical resection or stabilization and/or chemotherapy/radiation. Benign bony tumors can often be observed until they cause symptoms.^{2,3}

Tip 2: Refer to a Surgeon for Resection, Tissue Diagnosis, Intractable Pain, Instability, or Neurological Deficit

When oncologists or palliative care physicians evaluate a patient with a spine tumor, there are several reasons to call a surgeon.

Local tumor control/surgical resection

Any patient with a malignant bony tumor such as a chordoma should see a surgeon as resection portends the best

survival. Surgeons also consult on intradural tumors—the field shifted toward more aggressive surgical management of these tumors over the past 30 years.^{4–6} Metastatic and benign bony tumors do not all need surgery.

Tissue diagnosis

If a tissue diagnosis must be determined, and a biopsy is unable to be obtained from another source, surgical resection or open biopsy should be considered.

Pain or spinal instability

Oncologists and palliative care physicians can use symptoms to decide whether to refer a patient with metastatic spine disease or a benign primary bone tumor to a surgeon. The usual presenting symptom is pain. Tumor-related pain is typically located at the site of disease, dull, achy, and nocturnal. Mechanical pain from damaged vertebra and instability increases with standing or activity and improves when the patient lies down.⁷ Mechanical pain is a strong predictor of instability and warrants surgical consultation. Tumor-related medically refractory pain also should warrant surgical consultation.

Neurological deficit or epidural spinal cord compression

Ten to 20% of spinal metastasis patients will present with symptoms of epidural spinal cord compression (ESCC).⁸ Cord or nerve compression occurs direct tumor compression, retropulsed bone secondary to a pathologic fracture, or deformity related to vertebral body compromise. Symptoms can be divided into radicular (those that effect a particular nerve pattern with singular muscle group weakness, dermatomal numbness) and/or myelopathy (gait instability, spasticity, weakness, sensory loss, and bowel and bladder dysfunction).⁷ Quick onset symptoms of spinal cord compression are a neurological emergency and warrant stat imaging with a spinal MRI with and without contrast. While all tumors with epidural compression should be considered for surgical

FIG. 1. Types of tumor-related pain. Pain patterns depend on tumor location as well as extent of lesion.

Spinal Metastasis Decision-Making Framework Made Simple

FIG. 2. Summary of decision-making frameworks. Scoring systems to consider when making a referral.

consultation, those with symptom onset within 48 hours will benefit most from surgical decompression (Fig. 1).⁹

Tip 3: Neurosurgeons Utilize Diagnostic and Prognostic Scoring Systems to Stratify Surgical Decisions

Validated prognostic scoring systems and decision-making indices aid in assessment of a patient with a new spine tumor, and particularly for spinal metastases. One of the most commonly used treatment paradigms is the Neurologic, Oncologic, Mechanical, and Systemic (NOMS) decision framework, which provides a common language with which to best determine an individualized treatment plan for patients. NOMS evaluates the patient based on the presence or absence of a neurological deficit and/or myelopathy, oncological sensitivity to radiation, or other tumor-specific factors, the biomechanical stability of the spine, and the patient's systemic burden of disease and/or ability to tolerate surgical intervention.

Neurological status is best assessed by obtaining a thorough history and performing a physical examination. Radiographic parameters may also be assessed with the Bilsky ESCC grading scale, which helps to determine the degree of spinal cord compression using T2 axial-weighted MR images.¹⁰ Oncologic assessment is focused on tumor radio sensitivity and histology. Radio sensitivity is determined by the tumor response to external beam radiation therapy or stereotactic radiosurgery.¹¹ Mechanical stability can be evaluated using the Spinal Instability Neoplastic Score (SINS). This scoring system classifies patients into stable versus unstable using radiological and clinical components that include an assessment of pain.¹² Mechanical instability regardless of neurological status is an indication for stabilization and requires surgical consultation. Systemic status examines the patient's existing comorbidities, as well as the tumor histology and extent of metastatic disease. A patient's systemic burden of disease may be assessed using numerous prognostic scoring systems, including the Tokuhashi, Tomita, Modified Bauer, Katagiri, Linden, and SORG nomogram (Fig. 2).¹³

Tip 4: How to Make a Good Referral and What to Do "On the Ground" Before Referring: A Cheat Sheet

There are four main questions neurosurgeons ask after receiving a referral:

1. Do they have mechanical pain?
2. Is the tumor radiosensitive?
3. Do they have spinal cord compression with myelopathy?
4. What is their systemic burden of disease/prognosis?

The best surgical referrals come with an explanation of the neurological exam, onset of symptoms, prognosis, imaging findings, and tumor pathology. These can be summarized using the NOMS framework and ESCC grading scale or just simply explained. Neurological status is communicated through the neurological exam. All neurological exam findings should be reported (but the most important are muscle group strength, ability to ambulate, sensation, and bowel/bladder function), including the onset and duration of all deficits. If there are acute deficits it is important to make an emergent referral to neurosurgery. Information regarding tumor histology and radio sensitivity is important to assess the oncological status of a patient, and to direct appropriate management. Mechanical instability (SINS criteria) is suspected based on MRI, CT, and/or X-ray findings, but also based on intractable pain with activity (reclining is often a most comfortable position). Pertinent imaging information includes the location of the tumor and the number and location of metastases. Survival prognosis, staging workup by oncologist, and surgical risk stratification determine the patient's ability to tolerate and the likelihood of benefit from a surgical intervention.¹¹ Other useful considerations include the patient and family preferences regarding surgery.

Before referrals, a few prompt steps by the primary physician team can improve outcomes in patients with spinal cord compression and myelopathy symptoms:

1. A 10 mg intravenous bolus of dexamethasone followed by 16 mg daily (divided). This can temporarily

improve symptoms before definitive management and should be tapered as soon as possible to avoid complications. Close monitoring for hyperglycemia and concurrent use of proton-pump inhibitors help to avoid common steroid side effects.¹⁴

2. Medically manage pain.
3. Consider bracing for comfort and improved stability. A thoracolumbosacral orthosis can be used for low thoracic and lumbar lesions and a cervical thoracic orthosis for high thoracic lesions. The unmolded corset is more readily available and cost effective than the molded version and the molded version should not be ordered until recommended by a surgeon.

The above steps should ideally occur concurrently with referral to a spine surgeon. If a surgeon is unavailable, a referral to a tertiary care center should be made to discuss the possibility of patient transfer.

Tip 5: Minimally Invasive Treatment Options Provide Palliative Options for Pain Management

Minimally invasive procedures typically provide a palliative option for pain management in patients with spine tumors.

A brace may be used for patients that are potentially unstable (and/or do not immediately warrant surgical intervention) or for patients too sick to tolerate surgery but needing some degree of stabilization to improve their symptoms. Braces are not fully stabilizing and therefore patients that are appropriate surgical candidates typically receive internal fixation.

The advantage of minimally invasive spine tumor surgery is less tissue trauma, which is associated with less pain and less recovery time. Percutaneous pedicle screw instrumentation provides minimally invasive mechanical stabilization with less blood loss, improved cosmesis, decreased post-operative hospital length of stay, and decreased narcotic usage.¹⁵ Kyphoplasty and vertebroplasty are minimally invasive techniques used to treat painful bony lesions, specifically with vertebral body collapse. For a kyphoplasty, cement is percutaneously injected into the collapsed vertebral body after balloon dilation of the vertebral body. The procedure is contraindicated in patients with tumor extension into the posterior elements, significant epidural disease, or disruption of the posterior wall of the vertebral body cortex because cement could extravasate into the spinal canal and cause spinal cord compression.⁷ MRI is useful in the preoperative evaluation of these patients to evaluate the posterior elements, extent of epidural disease, and the degree of vertebral body height loss. Radiofrequency ablation, which uses a heat-generated current to destroy tumor tissue, can also be performed percutaneously and is coupled with kyphoplasty/vertebroplasty to treat metastatic disease. Minimally invasive surgical techniques do not always allow adequate decompression and therefore more invasive procedures can be utilized as well.

Radiation is the primary treatment option for radiosensitive tumors, such as lymphoma, multiple myeloma, small cell lung carcinoma, seminoma, neuroblastoma, and Ewing's sarcoma, but is also performed for the majority of tumor types after a surgical intervention. Additionally, radiotherapy alone is preferred for patients that cannot tolerate surgery or have a poor prognosis (less than three months).⁷ Despite the tumor-ablative effects of radiation treatment alone, patients may

warrant evaluation for resultant spinal instability (measured by SINS criteria) from 30 to 180 days post-radiation, particularly with single-fraction palliative radiation.¹⁶

Tip 6: Maximally Invasive Treatment Options Provide Tumor Control and Improve Outcomes in the Right Patients

The primary goals of spine tumor surgery are decompression of the neural elements, stabilization, local tumor control, treat medically refractory pain, and/or to obtain a tissue diagnosis. Patients with minimal comorbidities and better ambulation status preoperatively are shown to have better post-operative outcomes with surgery followed by radiation rather than radiation alone.¹⁷ Decompressive laminectomy involves the removal of the lamina at the level of the tumor dorsally to expand the spinal canal, providing more room for a compressed spinal cord. Laminectomy is often coupled with fusion, because removal of the posterior elements can lead to destabilization of the spine given the tumor typically alters the stability of the ventral portion of the spine. Results of decompressive laminectomy are better if internal fixation or "fusion" is performed. Fusions commonly span the index level of the tumor and typically extend 1 to 2 levels above and below for stabilization and/or correction of deformity. Circumferential decompression can address both the ventral and dorsal compression improving results, such as the ability to ambulate, as well as potentially providing better local tumor control.^{8,18}

Tip 7: Comprehensive Post-operative Care Improves Patient Outcomes

Recovery from spine tumor surgery is heavily influenced by any preoperative deficits. In our institutional experience, those without a debilitating deficit are discharged within one to seven days (shorter with minimally invasive surgery). On the other hand, patients with poor functional status often require inpatient rehabilitation. All spinal tumor patients benefit from a rehabilitation plan that incorporates their goals of care and treatment recommendation.¹⁹ Inpatient rehabilitation has positive effects on post-operative patients, such as improved mobility and bladder function.²⁰ Assessment of whether improvements in functional outcomes post-intervention are realized occur roughly three months post-operatively and/or approximately within one month to one year post-inpatient rehabilitation.²¹ When considering recovery from spine tumor surgery, one must consider the effects of cancer-related fatigue and mood disorders and provide support that complements the rehabilitation plan.²²

Possible complications that can occur during the first month post-surgery include deep vein thrombosis (DVT), infections, pain, and wound dehiscence. Early ambulation, as early as post-operative day zero, can decrease complications, improves outcomes, and is generally recommended for patients. Individual surgeon preference typically dictates when to restart DVT prophylaxis. At our institution, prophylactic low-molecular-weight heparin 24-hours post-operatively has not been associated with significant deleterious effects. For severe pain following surgery, patient-controlled analgesia may be indicated. Transition back to chronic pain medications or to oral dosing occurs as the patient recovers and before discharge. Wound care is individual surgeon-specific

and can include inspection of the wound, daily dressing changes, wedges to keep the patients from laying directly on the incision, and potentially wound vac placement. This is particularly important as the patient continues their rehabilitation plan, before and during radiation treatments. The goal, in general, is to keep a patient off of their wound and especially while in bed to minimize the risk of wound breakdown.

There are no guidelines for post-operative bracing, and some surgeons use bracing as an adjunct to externally immobilize the spine, prevent further collapse or kyphotic deformity, and improve fusion. Although the evidence for this is limited, surgeons often decide whether bracing is warranted based on intraoperative findings, such as bone quality. Braces are typically worn while the patient is upright and/or out of bed, although this is at the discretion of the individual surgeon.

Tip 8: Post-operative Plan: Expect Complications but Limit Them with Excellent Post-operative Care

Surgeons fear post-operative wound breakdown and infection because it often leads to readmission and potentially reoperation for patients. In a recent review of patients treated surgically for spine metastases, 56.6% were readmitted within one year and 13.8% in 30 days with 26.7% of the admissions related to surgical complications (predominantly wound complications).²³ Use of perioperative (both neoadjuvant and adjuvant) radiation therapy and chemotherapy correlate with rates of local wound complications. Age, gender, oncologic stage, type of tumor, and surgical approach also have been found to predict complications in certain cases.^{24,25} When balancing the severity of an individual patient's systemic illness and the potential improvement in quality of life derived from surgery, many consider the complication rate tolerable. A recent study of post-operative patients who underwent spinal deformity surgery showed that patient satisfaction did not correlate with post-operative complications.²⁶ The goal of complication rate reduction warrants interdisciplinary collaboration to improve quality of care. Factors that influence the rate of perioperative complications include: appropriate patient selection, careful wound care, consideration of plastic surgery consultation preoperatively for advanced wound closure, adequate nutrition, and planning of adjuvant therapy post-operatively to allow for wound healing.

Tip 9: Why Two Patients with the Same MRI Scan and Symptoms Get Different Treatments? Patient-Specific Factors Dictate Management Options

Neurosurgeons, palliative care physicians, and oncologists alike have seen patients with similar lesions on MRI scan, and even similar symptoms, get different treatments.

What do surgeons weigh when considering operating on a patient with a spinal metastasis? The use of prognostic indices and scoring systems has helped to decrease the variability in surgical decision making and appropriate surgical candidacy. Surgeons must weigh the perceived prognosis, often provided by the primary oncologist against the perception of surgical benefit, risk, and recovery. Many of these features are individualized to a given patient based on their

extent of disease, comorbidities, and age. The surgeon then discusses these findings with the patient, their family, the radiation oncologist, and the primary oncologist with the goal of making a multidisciplinary decision in the best interest of the patient's goals of care. While limited research exists as to how the decision-making process takes place, tumor boards and multidisciplinary discussions aid in communication regarding patients with newly diagnosed spine tumors.

Tip 10: Surgery for Spine Tumor Patients Can Mean Improved Quality of Life

Patchell's study in 2005, revolutionized the management of malignant, radioinsensitive spine tumors causing epidural compression. Coupled with subsequent studies, the Patchell study showed that patients managed with surgery and radiation had better outcomes, including the ability to ambulate post-surgery than patients managed with radiation alone.^{7,17} Of note, the patients included in this study had an expected post-intervention survival of three months or greater. Scales measuring motor function, such as the Frankel scale can demonstrate improvement post-operatively in appropriately selected patients.⁹ In general, post-surgical patients require less steroids and less narcotics than nonsurgically-treated patients after the perioperative period. The use of patient-centered outcome research in patients with malignant spine tumors will provide evidence to support the benefits of surgical intervention. Patient-driven metrics of surgical efficacy using Health-Related Quality-of-Life (HRQOL) Scores are currently being assessed by various groups with patients receiving surgery reporting improved and maintained quality of life compared with those nonsurgically managed.²⁷ Although surgery is invasive and can be associated with complications, it is important to note that even at the end of life, quality-of-life scores are high among surgical patients undergoing palliative procedures.

Conclusion

Nearly 20% of cancer patients develop symptomatic spine metastases.^{28,29} Metastatic spine tumors are most commonly extradural tumors that grow quickly and often cause persistent pain, weakness, paresthesia, urinary or bowel dysfunction, and paralysis.³⁰ These symptoms are indications for surgery and a prompt referral to neurosurgery may result in relief of pain as well as minimize loss of neurological function. The desired result of treatment includes tumor control, pain relief, preservation or improvement of neurological function, restoration of mechanical stability, and improvement of quality of life. Neurosurgeons employ multiple decision frameworks and grading scales to assess the need and effectiveness of a variety of surgical interventions ranging from minimally to maximally invasive. Likewise, palliative care offers an everevolving array of treatment options that make it imperative in every case to find the best, individualized plan. Crossdiscipline collaboration between palliative care and neurosurgery is crucial to maximize available treatment options and more importantly, quality of life for patients.

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