Neuro-Oncology Practice
7(S1), i1–i4, 2020 | doi:10.1093/nop/npaa071

Multidisciplinary spine oncology care across the disease continuum

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Quote "If you would seek health, look first to the spine" – Socrates

Current State

Spine oncology encompasses both primary and metastatic tumors of the spinal cord and spinal column. Whereas primary tumors are very rare, metastatic tumors are very common. The prevalence of spine oncology is increasing secondary to an enlarging aged-population, greater medical awareness, and longer disease courses. Similarly, the complexity of spine oncology is increasing secondary to patient heterogeneity, expanded treatments, and improved outcomes. Modern stakeholders of spine oncology are increasing beyond the historical stakeholders of neurosurgery, orthopedic surgery, and radiation oncology, to now incorporate emergency medicine, primary care, neuro-oncology, medical oncology, interventional radiology, physical medicine and rehabilitation, palliative care, allied health professionals, and others. This modern multidisciplinary care has improved clinical outcomes, quality, satisfaction, and resource utilization.[1,2]

Challenges of the Current State

Despite the recent improvements of multidisciplinary care in spine oncology, substantial challenges limit them from being fully realized, including those listed in Table 1.[1,2]

Goals of This Supplement

We all want improved outcomes. Toward this goal, the *Neuro-Oncology Practice* (*NOP*) editorial team invited global multidisciplinary specialists who represent both the scope of patients we treat and the scope of providers who treat them. Author teams were asked to comprehensively review a specific aspect of the disease continuum: *Epidemiology, Tumor Types, Presentation, Initial Tumor-directed Management, Initial Symptom-directed Management, Subsequent Tumor-directed Management, Subsequent Symptom-directed Management, and Multidisciplinary Program Development and Resources for Stakeholders*. Anticipated outcomes of this supplement are listed in Table 2.[3–5] *NOP* and the authors welcome feedback, as well as correspondence toward future initiatives.

Initiatives of a Future State

Applying the opening quote from Socrates to spine oncology acknowledges that spine health begins with early seeking and prompt action. *NOP* and authors hope this primer serves as a beginning point for future initiatives, whether at the bench, bedside, classroom, ballot box, or pocketbook. Invest in local medical efforts and in professional societies that prioritize spine oncology. Contribute to the literature and teach those around you.

Thank you to the *NOP* editorial team; the senior editors of *NOP*, Susan Chang, MD, and Martin J.B. Taphoorn, MD, PhD, Oxford University Press; the authors; and our sponsor, BrainLAB.

Highlights Within Individual Manuscripts

The initial 3 manuscripts, by first authors Wewel, Kumar, and Fridley, focus on epidemiology, presentation, and diagnosis.

Epidemiology

Primary cord and column tumors are very rare (< 3% of all CNS tumors), can be either heritable or sporadic, and can be either benign or malignant. In contrast, metastatic cord and column tumors are very common (40-70+% of oncology patients develop during their lifetime). The skeletal system

Table 1. Current Challenges

Historical guidelines narrowly focused on specific treatments and episodes of care
Paucity of rigorously conducted clinical and outcomes research
Paucity of practical management guidelines
Paucity of insurance coverage and funding for the delivery of multidisciplinary care
Paucity of systematic education both for trainees and practicing providers
Paucity of resources and support for patients, caregivers, and providers
Paucity of prioritization by entities that drive the medical profession, including professional societies, pharma, industry, insurers, philanthropy, and governments

Table 2. Anticipated Outcomes of This Supplement

Primer for all stakeholders, including generalists, specialists, allied health professionals, patients, caregivers, trainees, researchers, pharma, administrators, and governments
Increased practical wisdom regarding early and accurate diagnosis, effective multimodality treatments, and multidisciplinary decision making
Increased prioritization of spine oncology research and training by the institutions, professional organizations, and funding mechanisms that support them
Increased advocacy for sufficient reimbursement of the true scope of multidisciplinary care
Increased attractiveness of spine oncology as a clinical and research career path

is the third most commonly metastatic site, following the lung and liver. Within the skeletal system, the spine is the most frequently involved (thoracic [60%-80%], lumbar [15%-30%], and cervical [< 10%]). Metastases are malignant, by definition, and spread via arterial, venous, cerebrospinal, or direct-extension mechanisms.

Anatomical Compartments Are Differentially Affected

Spine tumors can affect various compartments of the spinal cord or the spinal column and can be localized, regional, or diffuse. A deep understanding of spine anatomy is essential to safeguarding spine stability, formulating a differential diagnosis, obtaining and optimizing pathology, as well as predicting patterns of progression.

First, the authors review spine tumors based on whether they are primary or metastatic, benign or malignant, and hereditary or sporadic. *Primary spine tumors* can be either benign or malignant. *Benign primary spinal column tumors* are rare, often found incidentally, often require a biopsy, and are treated based on symptoms and morbidity risk. Examples include hemangiomas, osteoid osteoma, and osteoblastoma. *Malignant primary spinal column tumors* are even rarer than their benign counterparts, often found secondary to symptoms, and are treated in a highly individualized manner. Examples include chordomas, chondrosarcoma, Ewing sarcoma, osteosarcoma, plasma cell neoplasms, and lymphoma. *Metastatic tumors* are, *by definition, malignant*. They are very common and can dominantly affect the column, the cord, or both. Also, they can initially or subsequently involve the leptomeninges, cranial nerves, and cerebral spinal fluid. The most common solid tumor pathologies are lung, breast, prostate, and renal, and the most common liquid tumors are plasma cell neoplasms and lymphoma. They most commonly occur between the fourth to seventh decades of life and signify a symptomatic, shorted survival.

Second, the authors systematically review spine tumors based on the anatomical compartments they affect. Understanding the specific compartments involved is essential to safeguarding spine stability, formulating a differential diagnosis, coordinating multidisciplinary care, and optimizing outcomes. *Extradural, extramedullary tumors* represent 90% to 95% of malignant tumors. The overwhelming majority are skeletal metastases and carry a risk of epidural spinal cord compression (ESCC). *Intradural, extramedullary malignant tumors* represent only approximately 5% to 10% of malignant tumors and most commonly occur within the thoracic, lumbar, and cauda equina regions. They are "drop metastases" from intracranial deposits, including intracranial gliomas or metastases (usually breast and lung). *Intradural, extramedullary benign tumors* are most often primary and are most commonly either meningiomas or nerve sheath tumors. *Intradural intramedullary* tumors account for 20% of all intraspinal tumors in adults and can be primary (intraaxial), or metastatic. *Intradural, intramedullary primary tumors* are much more common. Ninety-five percent of these are ependymomas and astrocytomas. Importantly, they are very distinct from their intracranial counterparts

with respect to heredity, pathology, treatment, progression, and outcomes. *Intradural, intramedullary metastatic tumors* are extremely rare and typically occur in the cervical cord because of periventricular spread going down the fourth ventricle into the central canal. *Leptomeningeal carcinomatosis* is seen in 5% to 8% of solid tumors and 15% of liquid tumors. This usually presents later in the disease course and, at the time of diagnosis, averages a 2- to 4-month survival, regardless of treatment.

Presentation

The presentation of spinal cord and column tumors most commonly includes back pain (80%-95%), followed by motor dysfunction (35%-75%) and sensory dysfunction (20%), to include the emergency of ESCC. The authors detail how the evaluation of pain can localize a tumor, identify the mechanisms of pain, and determine the urgency and the modalities of treatment. They correlate pathology to patterns of presentation and patterns of progression. They point out that the worldwide prevalence of nononcologic pain can complicate and delay the diagnosis of spine tumors, thus urging patients and providers to maintain a high index of suspicion. Last, they provide guidelines for the evaluation and management of pain and other "red-flag" symptoms.

Diagnosis

Early and accurate diagnosis is strongly correlated with optimal management and thus to optimal outcomes. Diagnosis relies on a high index of suspicion, a detailed history and physical exam, and validated spine oncology assessments. The authors introduce the Neurologic assessments, Oncologic assessment, Mechanical assessments, and Systemic assessment (NOMS) framework as a tool for diagnosis.[6] They detail high-yield laboratory and imaging evaluations, as well as optimal pathology analyses. They emphasize the importance of a multidisciplinary approach from the moment a *potential* spine tumor presents, including the initial evaluation, formulation of the differential diagnosis, determination of the goals of care, obtainment of pathology, and coordination of subsequent treatment.

Initial Treatment

The next 2 manuscripts, by first authors Bilsky and Germano, focus on tumor-directed and symptoms-directed management at new diagnosis:

The primary treatment goal of spine oncology remains palliative, including optimizing symptoms, improving health-related quality of life, restoring neurologic function, safeguarding spinal stability, and achieving local tumor control. Yet an enlarging subset of patients are also achieving a secondary treatment goal: a modest improvement in survival. The authors detail how the greatest advancement toward these goals has been the incorporation of stereotactic body irradiation, followed by improvements both in surgery and interventional radiology, and followed, very modestly, by improvements in medicines. They reiterate the importance of the NOMS framework as a tool for diagnosis by facilitating and standardizing multidisciplinary decision making. They elaborate on it as a tool for management by integrating the most evidence-based technologies and treatments. They provide guidelines and references for validated pain scales, use of steroids, radiographic assessments, and spine stabilization techniques. They expand on the comprehensive management of pain by combining the modalities of radiation, surgery, neuromodulation, interventional procedures, medicines, rehabilitation, orthotics, psychotherapy, and others. Last, they highlight the importance of multidisciplinary care, including tumor conferences, treatment pathways, clinical trials, and education.

Subsequent Treatment

The next 2 manuscripts, by first authors Kotecha and Ruppert, focus on subsequent tumor-directed and symptoms-directed management.

Despite the aforementioned improvements in outcomes, the overwhelming majority of spine oncology patients will require subsequent treatment at recurrence/progression. Kotecha et al emphasize that the goals of care are the same as at initial presentation: optimizing symptoms, improving health-related quality of life, restoring neurologic function, safeguarding spinal stability, and achieving local tumor control. Yet, they also emphasize that patients require a much more individualized approach. Partly, this is secondary to the heterogeneity of a patient's health, tumor type, disease course, and goals of care. It is also partly secondary to the difficulty of matching patients to the trials that lead to available treatments. Accordingly, they point out the importance of assessing prior therapy (types, timelines, toxicity, and response), current pathology (if available), current staging, neurologic status, mechanical stability, overall performance status, estimated "all-cause" prognosis, and future oncologic treatment options. They detail published outcomes and practical considerations both for recurrent radiation and recurrent surgery. Last, they summarize multimodality guidelines, encourage referral to centers of excellence, and urge participation in clinical trials.

Rupert and Reilly provide a high-yield review of the most common symptoms encountered in spine oncology. They underscore that the anatomic locations and the mechanisms driving these symptoms codify the solutions to optimize them. They detail upper and lower motor neuron

dysfunction, sensory and autonomic dysfunction, abnormal tone and reflexes, bowel and bladder dysfunction, hemorrhage/thrombosis, integument and immune system dysfunction, neurocognitive dysfunction, and side effects to common treatments. They emphasize a multidisciplinary approach that includes various types of therapy and training for patients and caregivers. Last, they summarize a constellation of proven palliative care services and provide a robust list of resources for providers, patients, and caregivers.

Program Development and Resources

The last manuscript by senior author Benzil and colleagues focuses on multidisciplinary program development and resources for stakeholders:

The Benzil et al manuscript builds on many of the concepts introduced by prior authors. They detail evidenced-based justifications for distinct aspects of multidisciplinary care, including tumor conferences, multidisciplinary clinics, and overall program development. They demonstrate that multidisciplinary care improves outcomes, including clinical, quality, satisfaction, resource utilization, and reimbursement. At the patient level, benefits are seen in early and accurate diagnosis, consensus decision making, expansion of treatment options, and more access to clinical trials. At the provider and institutional levels, benefits are seen in work flow, training, collaboration, career satisfaction, scientific discovery, clinical trial accrual, insurance reimbursement, and resource accumulation. A construct for building and maintaining a multidisciplinary program is detailed. A robust list of resources for stakeholders is also included. Last, they emphasize the central role of palliative and hospice medicine across the disease continuum.

Conflict of interest statement. None declared.

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