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Sarcoma and the Spinal Column

Vincent Y. Wang, MD, PhD[a], Matt Potts, MD[b], Dean Chou, MD[a,*]

[a]*Department of Neurological Surgery, University of California, 505 Parnassus Avenue, Room M779, San Francisco, CA 94143-0112, USA*

[b]*Department of Surgery, University of California, 513 Parnassus Avenue, Med Sci 321, San Francisco, CA 94143-0470, USA*

Primary bone tumors of the spine are rare, representing only 2.8% to 13% of all bone tumors [1]. Sarcomas are malignant tumors of mesenchymal cell origin. The most common types of sarcoma seen in the spine are osteosarcoma, Ewing's sarcoma, and chondrosarcoma [1]. Because of their rarity, treatment experience of these tumors is limited. Treatment of primary spinal sarcomas has two main goals: to control local disease and to prevent metastasis. Surgical resection and radiation therapy, or a combination of both, are the first steps toward gaining local control. Chemotherapy is used for the treatment of any residual tumor and for the prevention of local recurrence. Chemotherapy is also the main treatment modality for metastatic disease. However, spinal sarcomas pose unique characteristics for treatment, especially given the anatomic constraints that can preclude a wide margin of excision. Furthermore, critical structures such as the spinal cord or abdominal organs also may limit the dose of radiation that can be used for treatment.

This article reviews some of the most common sarcomas, including osteosarcoma, retroperitoneal sarcoma, and Ewing's sarcoma. Chondrosarcoma is covered in another article elsewhere in this issue. The epidemiology, pathology, clinical presentation, and radiograph findings are discussed for each of these tumors, as are the surgical, chemotherapeutic, and radiation therapy for each tumor.

Osteosarcoma

Osteosarcoma is the most common sarcoma of the spine. It accounts for 3% to 15% of all the primary tumors of the spine but less than 3% of all sarcomas [1–3]. Osteosarcoma of the spine tends to occur in the fourth decade of life [4]. Spinal osteosarcoma is rare in the cervical spine, and it occurs at a proportionally higher frequency in the thoracic spine and the sacrum [2]. In one series, the sacrum was the primary site of origin in as many as 68% of the cases [2]. Most tumors arise from the posterior elements. One risk factor for osteosarcoma is Paget's disease. Paget's disease is relatively common and involves the spine in 50% of the cases [5]. Degeneration into osteosarcoma is a rare event, occurring in less than 1% of patients [5]. However, in some tumor registries, it accounts for as much as 50% of the osteosarcoma of the spine [1]. Spinal osteosarcoma in Paget's disease tends to occur in older patients, with a mean age of 67 [5].

Patients with spinal osteosarcoma usually present with pain [2,5]. Pain can either be axial back pain or radicular pain [2,5]. Neurologic symptoms such as weakness occur in more than half of the patients [5,6]. Bowel and bladder symptoms tend to occur late during the course of tumor progression. In most cases, there is a delay in diagnosis because the symptoms are nonspecific. The median delay in the Cooperative Osteosarcoma Study Group (COSS) trial, for example, was 5 months from the onset of symptoms [2].

Paget's disease

Paget's disease of the spine presents with pain and neurologic symptoms usually beyond the fourth decade. Imaging findings demonstrate

* Corresponding author.
E-mail address: choud@neurosurg.ucsf.edu (D. Chou).

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thickened cortical bone (giving a ''picture frame look'') and coarsened trabecular bone. Spinal involvement usually includes enlarged pedicles, lamina, and vertebral bodies with an ''ivory spine'' appearance. Diagnosis is aided by elevated serum alkaline phosphatase levels and marked increase uptake on bone scan. Long bones are commonly affected as well and present with pain and fracture.

Imaging

Imaging studies include plain radiographs, CT, and MRI. On plain radiographs, osteosarcomas can appear as either osteoblastic or osteosclerotic, although an osteoblastic appearance is more common, with a mineralization matrix being seen. An osteosclerotic appearance occurs in about 20% of the cases. Patients with spinal osteosarcoma can also have a normal X ray [2,5]. During recent years, CT scan and MRI have become more important in the evaluation of patients with spinal osteosarcoma. CT scans with reformatted images not only give excellent imaging of the bone, but they also allow for some evaluation of the soft tissue. Myelography has been used in the past to evaluate for spinal cord compression, but this has now been largely replaced by MRI. MRI evaluation yields great details of the soft tissue and also gives some information about the vasculature around the tumor (Fig. 1, Case 1). In addition to imaging of the spine, the metastatic workup may include chest radiograph, chest CT, or bone scans.

Histopathology

There are multiple histological subtypes of osteosarcomas. All osteosarcomas produce some osteoid matrix [7], and most have a medullary origin and are high grade [7]. Surface osteosarcomas tend to be lower grade but are rare in the spine [7]. Conventional osteosarcomas are the most common type, and production of osteoid matrix is a requirement for diagnosis [7]. Conventional osteosarcomas also produce other types of extracellular matrix and can be subclassified into osteoblastic, chondroblastic, and fibroblastic osteosarcoma [7]. The classification is based on the predominant type of extracellular matrix. Conventional osteosarcomas are high-grade tumors and contain significant amounts of anaplasia. Other types of histologies include telangiectatic osteosarcoma. Telangiectatic osteosarcoma resembles aneurysmal bone cysts and are composed mainly of multiple blood-filled sinusoids [7].

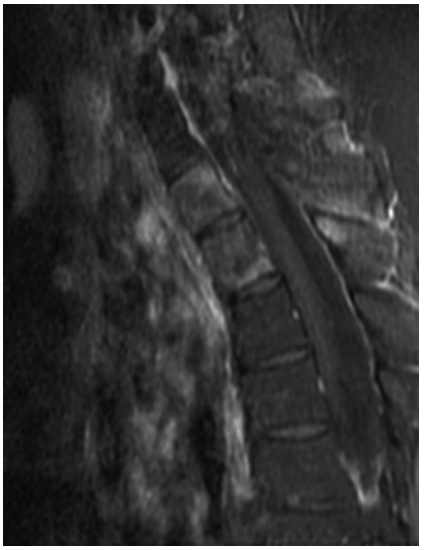

Fig. 1. Case 1: 22-year-old female with known history of osteosarcoma who was noted on bone scan to have increased uptake at both T1 and T2 vertebral bodies. Two-level spondylectomy undertaken. Sagittal T1-weighted MRI with gadolinium demonstrated both T1 and T2 vertebral body involvement.

Careful examination of the specimen should be performed to look for the presence of osteoid matrix [7]. Correlation with radiographic appearance on CT and MRI is useful as these imaging modalities may detect bone matrix. In the past, prognosis for telangiectatic osteosarcoma was worse than conventional osteosarcoma, although with modern chemotherapy regimens, telangiectatic osteosarcoma now carries a prognosis similar to conventional osteosarcoma [7].

Small-cell osteosarcoma is another rare variant of high-grade osteosarcoma, and it has round, hyperchromatic nuclei with little nuclear pleomorphism [7]. Small-cell osteosarcoma is positive for CD99, and it may be confused with primitive neuroectodermal tumor or Ewing's sarcoma [7]. In addition, translocation between chromosomes 11 and 22 has also been observed in small-cell osteosarcoma [7]. Spindle tumor cell types and the presence of osteoid matrix distinguish small-cell osteosarcoma from Ewing's sarcoma [7]. Epithelioid osteosarcoma tumor cells are poorly differentiated and may resemble a carcinoma [7]. Detection of osteoid matrix establishes the diagnosis of osteosarcoma [7]. If poorly differentiated cells are the predominant cell type in a bone tumor of a young patient, epithelioid osteosarcoma should be suspected and careful examination of

osteoid matrix must be performed. There are other variants of osteosarcoma such as giant-cell osteosarcoma or osteoblastoma-like osteosarcoma; however, these are rare histologic subtypes even in the extremities and even rarer in the spine. Surface osteosarcomas have epicenters located outside of the cortex of the underlying bone [7]. These tumors tend to be lower grade but are rare in the spine [7].

Treatment: surgical, radiation, and chemotherapy

Spinal osteosarcoma generally carries a poor prognosis. Compared with extremity osteosarcoma, patients with spinal sarcomas fare worse for several reasons [4]. The most important factor in the poorer prognosis is probably the inability to achieve adequate surgical margins [4]. In addition, the close proximity of the spinal cord also limits the radiation dose that can be delivered to the tumor bed. Decompressive laminectomy with biopsy was the mainstay of surgical intervention in the past [5,6]. Radiation therapy was used as the primary treatment in Shive's series, in which the median survival was only 10 months [6]. In cases of Paget's disease, the prognosis may be even worse due to the older age of the patients [5]. In Sharma's series of 13 patients with Paget's disease and spinal osteosarcoma, palliative radiotherapy was given to all patients, and they had a median survival of 4 months [5].

Recently, advances in surgical techniques have allowed for better excisions. Limited evidence suggests that wide margins or even marginal margins can significantly reduce the local recurrence rate, thus improving survival [2]. The development of en bloc techniques and earlier detection of disease may improve survival (Figs. 2–5, Case 1). However, there is no large surgical series that looks at the long-term outcome since the development of these techniques and early detection.

Development of effective chemotherapeutic protocols for spinal osteosarcoma has been based largely on trials for osteosarcoma of the extremities. On the basis of several large studies, four agents have been found to be effective against osteosarcomas (primarily in extremities)—methotrexate, cisplatin, doxorubicin, and ifosfamide—and current protocols use combinations of these agents. There is also some interest in the biologic immunostimulant agent muramyl tripeptide-phosphatidilehtanolamine (MTP-PE), which has been shown to have a beneficial interaction with ifosfamide. Addition of MTP-PE and ifosfamide to

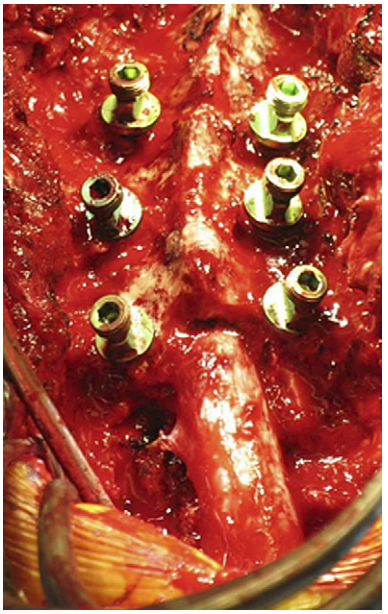

Fig. 2. Case 1: Posterior T1 2 elements removed en bloc.

a standard treatment of high-dose methotrexate, doxorubicin, and cisplatin increased the 3-year event-free survival rate in patients with osteosarcoma marginally from 71% to 78% [8]. Most of these patients had appendicular osteosarcoma, although 4% of the patients had axial or pelvic osteosarcoma [8].

Only one study of spinal osteosarcoma patients treated with current chemotherapy protocols has been published to date [2]. This retrospective review examined 22 patients treated with various osteosarcoma protocols from the COSS. The COSS protocols all included high-dose methotrexate and

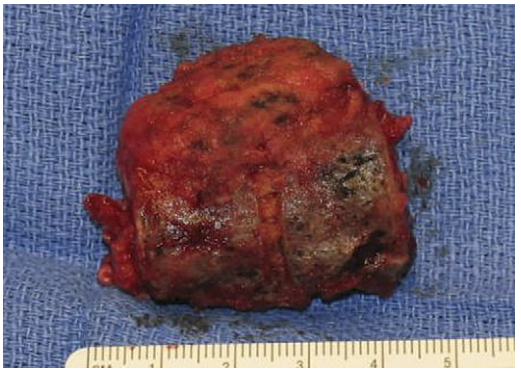

Fig. 3. Case 1: T1 and T2 vertebral bodies removed en bloc for total two-level spondylectomy.

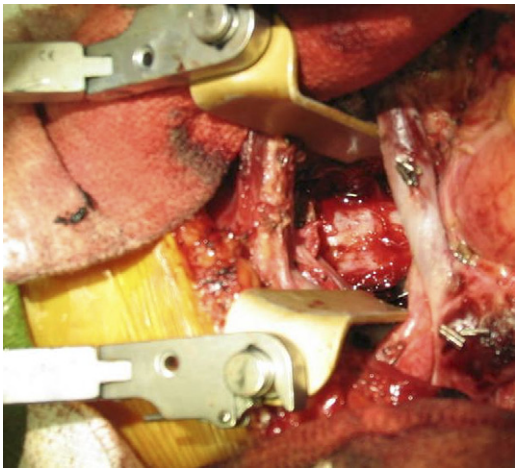

Fig. 4. Case 1: Dura visualized.

doxorubicin in various combinations with cisplatin, ifosfamide, bleomycin, cyclophosphamide, actinomycin D, and interferon α (which were changed over the 18 years of the study) [2]. Patients were also variably treated with surgical resection and radiotherapy. Median survival in this series was 22 months, with 3 patients surviving disease free for more than 6 years [2]. Although this study was too small to compare the various COSS protocols, it does begin to define the general effect of current chemotherapeutic agents on spinal osteosarcoma and serves as a basis for future studies.

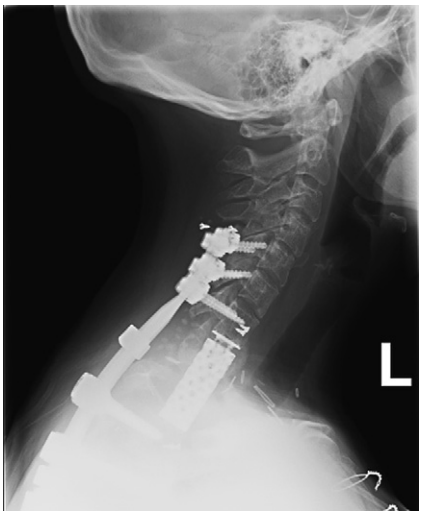

Fig. 5. Case1: Lateral radiograph showing final reconstruction.

More recently, an interdisciplinary approach that involves surgeons, oncologists, and radiation oncologists was used, and case reports of long-term survival, even with intralesional margin, have been reported. For example, in a case report, a 10-year-old child with osteosarcoma of the thoracic spine who underwent intralesional surgical resection; intraoperative brachytherapy; and external beam radiation followed by high-dose methotrexate, doxorubicin, cisplatin, ifosfamide, and MTP-PE demonstrated a greater than 5-year event-free survival [9].

Osteosarcomas are aggressive and relatively radioresistant tumors. Radiotherapy therefore has limited efficacy in their treatment [10]. Several series have suggested that radiation therapy can be used postoperatively after intralesional resection of osteosarcoma of spine and offered some benefit in local control [2,11]. Nevertheless, this benefit was limited with no definitive improvement of survival or outcome.

Ewing's sarcoma

Ewing's sarcoma occurs in the spine at a significantly lower rate than that of osteosarcoma. The spine is an uncommon primary site of Ewing's sarcoma, and only 3% to 15% of Ewing's sarcomas occur in the spine [12–14]. In general, Ewing's sarcoma can be separated into the sacral and nonsacral spine. Approximately 50% to 70% of Ewing's sarcomas are located in the sacral spine [12,13]. Distinguishing sacral Ewing's sarcoma from nonsacral Ewing's is important because of the difference in surgical management and survival [13,14]. In addition to the bony elements, extraskeletal Ewing's sarcoma can develop in the paravertebral muscles with invasion into the epidural space [15,16].

Spinal Ewing's sarcoma tends to present at young age, with the peak age of presentation at the second decade of life [14,17]. Pain is the primary symptom and occurs in virtually all the patients [12,14,17]. Neurologic deficits occur in 40% to 60% of the patients [12,14,17]. Bowel and bladder symptoms are rare and tend to occur late in the course of the tumor progression [17]. Frequently, there is a delay in establishment of diagnosis, and patients tend to have symptoms for 6 to 7 months before diagnosis is made [12,17].

Imaging

Unlike osteosarcoma, plain radiographs are rarely useful in diagnosing Ewing's sarcoma.

Changes on X ray usually do not occur until late in the course of the tumor. In most cases, a lytic lesion is observed [12,14]. Subtle findings, such as haziness of the endplates, may be the only changes observed. A completely negative radiograph can also be observed in the cases of Ewing's sarcoma [14]. CT and MRI have become the mainstays of diagnostic modalities to evaluate Ewing's sarcoma. CT is useful for the evaluation of bony elements, and MRI is useful to evaluate the soft tissue [12,17]. On MRI, Ewing's sarcoma is usually isodense or sometimes slightly hypodense on T1 and hyperdense on T2 [12,16]. Ewing's sarcomas tend to be enhanced with gadolinium [12,16]. In addition to CT and MRI, additional workup should include bone scans and chest X ray or chest CT to evaluate for metastasis.

Histopathology

Ewing's sarcoma consists of small, round cells with oval nuclei [14,18]. Recently, histologic diagnoses have become reliant on immunohistochemical stains. CD99 and Mic2 immunohistochemistry is commonly used for the diagnosis for Ewing's sarcoma [17,18]. Other additional stains that are useful includes periodic acid-Schiff stain for glycogen, as well as neuron-specific enolase, and S100 protein [14,18]. Translocation between chromosomes 11 and 22 is also seen in most cases of Ewing's sarcoma [16,17]. This translocation can be detected by cytogenetic methods [15].

Treatment: surgical, radiation, and chemotherapy

Currently, treatment of Ewing's sarcoma of the spine involves surgery, chemotherapy, and radiation therapy. Surgical interventions partly depend on the symptoms of presentation. If the patient presents with an acute neurologic deficit, prompt decompression is indicated. This can then be followed by chemotherapy, radiation therapy, or additional surgery. Note that kyphosis after laminectomy for decompression has occurred frequently, and stabilization with instrumentation should be considered [17]. In the case of the patient with full neurologic function or pain, a biopsy can be performed under CT guidance to establish the diagnosis. This can then be followed by chemotherapy to reduce the tumor size before surgical resection [14,18]. En bloc resection with spondylectomy should be used in these cases if possible [19,20]. It may improve local recurrence and, potentially, survival. However, data are sparse that show superior local control with en bloc spondylectomy versus intralesional resection when supplemented with chemotherapy.

Unlike with osteosarcomas, radiation therapy is almost a standard in the treatment of spinal Ewing's sarcoma owing to its sensitivity to radiation. Treatment protocols typically call for radiotherapy for unresectable lesions, intralesional or marginal resections, and poor histologic response to neoadjuvant chemotherapy [21]. Several recent studies of spinal Ewing's sarcomas included radiotherapy for their entire study population [17,22,23]. Typical radiation doses range from 45 to 55 Gy when combined with surgical resection to 55 to 65 Gy for radiotherapy alone, with fractionation regimens of 1.8 to 2 Gy daily or 1.6 Gy twice daily [24]. In pediatric patients, the field of radiation must either completely include or completely exclude a vertebral body to prevent asymmetric growth [24].

Schuck and colleagues [25] specifically examined the use of definitive radiotherapy in primary Ewing's sarcoma of the vertebrae. In this retrospective analysis of 116 patients, 75 received definitive radiotherapy as the only form of local treatment. Among those patients, local relapse occurred in 22.6%, a rate that is comparable to nonspinal Ewing's sarcoma patients treated with definite radiotherapy (26.3%) [21,25]. In this study, surgery with pre- or postoperative radiation does not appear to significantly improve local control, although it is unclear what margin was achieved in these cases [25].

Although the use of radiation therapy has become the mainstay of local therapy in spinal Ewing's sarcoma, it is likely that surgery will continue to have its role in local therapy of spinal Ewing's sarcoma. In extremities Ewing's sarcoma, it is clear that surgery with postoperative radiation provides the best local control and event-free survival [21]. With improved surgical techniques and possible combination with neoadjuvant chemotherapy/radiation therapy, en bloc resections of these tumors are more likely to be achieved in the future. Recently, radiosurgery has also been used for the treatment of Ewing's sarcoma of the spine, but long-term efficacy has not yet been borne out [26].

Ewing's sarcomas are also responsive to chemotherapy. The first large trial of spinal Ewing's sarcoma was the Intergroup Ewing's Sarcoma Study trial, which treated patients with radiation therapy as well as a combination of vincristine, actinomycin D, cyclophosphamide, and doxorubicin [13]. The results were excellent,

with a long-term survival rate of 86% and a 100% local control rate for patients with vertebral column Ewing's sarcoma. Patients with sacral Ewing's sarcoma, however, had a lower long-term survival rate—only 25%—and a 62% local control rate [13]. This lower survival rate was attributed to the more advanced disease at the time of diagnosis for sacral disease and limitations of radiation owing to adjacent organs in the pelvis [13]. This study served as the basis for most of the current protocols in practice, which employ combinations of vincristine, actinomycin D, cyclophosphamide, and doxorubicin [24].

Most of the current studies reported a 5-year survival rate for patients with spinal Ewing's sarcoma of around 48% to 58%. A recent retrospective series of 116 spinal Ewing's sarcoma patients treated under the Cooperative Ewing's Sarcoma Study and the European Intergroup Cooperative Ewing's Sarcoma Study protocols reported an overall survival at 5 years at 58% and a 47% 5-year event-free survival [25].

Ifosfamide and etoposide have been studied recently and may provide additional benefit in the treatment of Ewing's sarcoma in general, and these have been used in several trials that include spinal Ewing's sarcoma [21,27]. However, a direct comparison of addition of ifosfamide and etoposide with a standard regimen of vincristine, actinomycin D, and cyclophosphamide with or without doxorubicin did not appear to yield additional benefit [23].

Retroperitoneal soft-tissue sarcoma

Retroperitoneal sarcomas are tumors of connective tissues that are located in the retroperitoneal space. Approximately 15% of the 10,000 new cases of sarcoma will arise from the retroperitoneum [28]. Most of these tumors are managed by surgical oncologists. These tumors may involve multiple internal organs of the abdomen [29,30]. Given the close proximity of the retroperitoneal space to the spine, extension into the spine is possible. However, this is not observed frequently and has not been observed in many of the larger case series [29,30]. On the other hand, pelvic retroperitoneal sarcomas can compress the pelvic nerves, leading to radicular symptoms [31].

Pathology

Among the various types of sarcoma, liposarcoma is the most common type of retroperitoneal sarcoma, followed by leiomyosarcoma [29,30]. Liposarcoma makes up 40% to 50% of some of the case series [29,30]. Primary liposarcoma and leiomyosarcoma can also occur in the spine and paravertebral muscles (Figs. 6 and 7, Cases 2

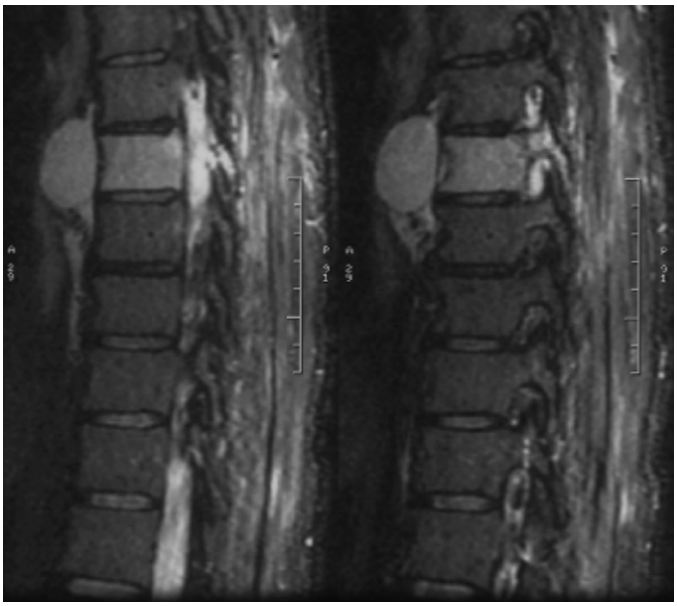

Fig. 6. Case 2: 57-year-old male who had liposarcoma resected from flank 9 years ago. Acutely presented out of state with neurologic deficit. Laminectomy performed; presented for oncologic resection. Sagittal T2-weighted MRI demonstrated T7 liposarcoma with paravertebral involvement.

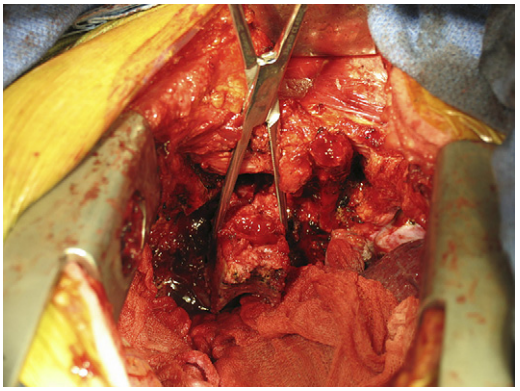

Fig. 7. Case 2: En bloc removal of T7 vertebral body after paravertebral soft tissue removed in one piece.

and 3) [32–36]. These tumors are rare and most are reported as cases series. There appear to be more case series of primary leiomyosarcoma of the spine than primary liposarcoma of the spine. Many of these tumors present with pain [35]; they can also present with fractures [33,36]. These tumors can also lead to weakness and acute deterioration of motor strength [34]. Workup usually consists of CT and MRI, which give good evaluation of the bony and soft-tissue details. Patients with previous radiation to the chest, abdomen, or pelvis, for childhood tumors such as Wilm's tumors, may have radiation-induced sarcoma originating from transformation of surrounding soft tissue such as muscle and fat years after initial therapy. These sarcomas tend to be aggressive and may grow extensively in the pleural or retroperitoneal spaces and involve the vertebral column (see Figs. 6 and 7).

Treatment: surgical, radiation, and chemotherapy

Surgery remains the mainstay of therapy. Approximately 70% of the cases undergo surgical

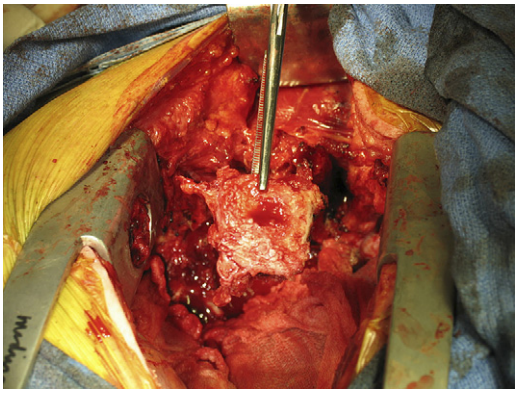

Fig. 8. Case 2: T7 vertebral body being removed.

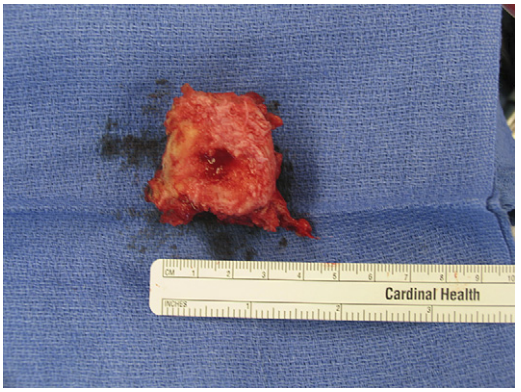

Fig. 9. Case 2: Specimen removed en bloc.

resection [28]. Obtaining an adequate margin is key to improving survival (Figs. 8–17, Cases 2 and 3). The 5-year survival rate ranges from 30% to 50% [29,30].

Adjuvant chemotherapy and radiation therapy have been used, but their clinical benefits have not been established yet [29,30]. Chemotherapy has been used only for those cases at high risk for metastases or unresectable or partially resected tumors. However, chemotherapy does not appear

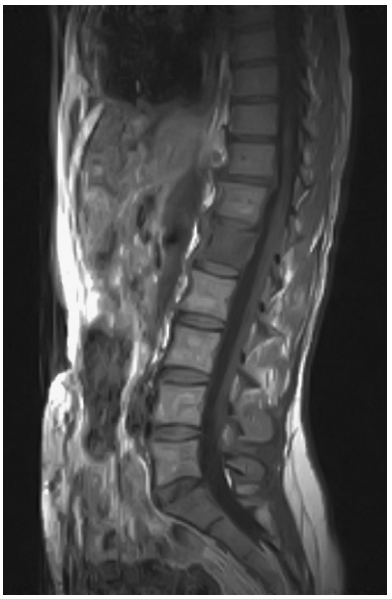

Fig. 10. Case 3: 58-year-old female with known history of leiomyosarcoma who presented with L1 signal changes on surveillance image (MRI obtained) and who wished to proceed with oncologic resection. Preoperative sagittal T1-weighted MRI without gadolinium. Lesion did not enhance with gadolinium administration.

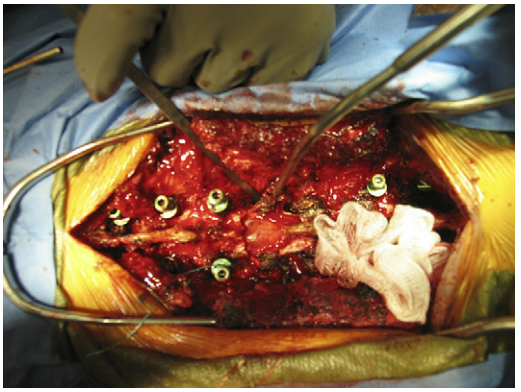

Fig. 11. Case 3: Posterior elements dissected free of all ligamentous attachments and pedicles amputated.

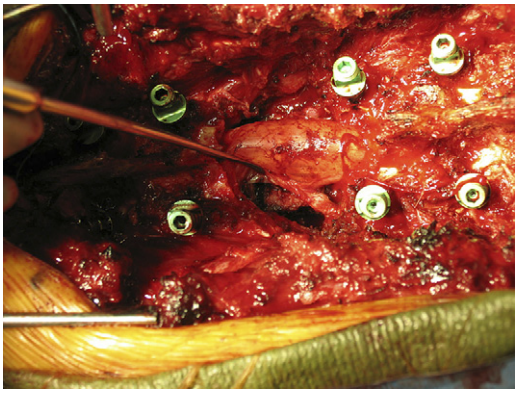

Fig. 13. Case 3: Aggressive discectomies and psoas dissection performed from a posterior approach. Emphasis of dissection placed contralateral to the intended side of approach for anterior resection.

to offer any significant benefit to unresectable tumors [37].

Recently, neoadjuvant chemotherapy has been combined temporally with radiotherapy to produce "chemo-radiation." Several phase I studies have demonstrated that this method is feasible for achieving local control in some patients with positive surgical margins [38]. This may also improve the probability of obtaining a wide margin of resection by reducing tumor burden.

Radiation therapy has also been studied for treatment of retroperitoneal soft-tissue sarcoma. Both brachytherapy and external beam radiation therapy are known to improve outcomes in patients with soft-tissue sarcomas of trunk and extremities [39,40]. Several studies of retroperitoneal sarcomas have also described some benefits from various forms of radiotherapy while used in conjunction with surgery. Tepper and colleagues demonstrated an association between long-term survival and high-dose (50–60 Gy) radiotherapy, whereas Heslin and colleagues reported that radiotherapy was the only factor that significantly reduced local recurrence in a prospective study of 198 patients with retroperitoneal sarcomas [41,42].

However, a recent retrospective review of 83 retroperitoneal sarcoma patients (including 60 with primary disease) treated with various forms of radiation demonstrated an overall disease-specific survival of 44% at a median follow-up of 47 months [43]. There was no effect of external beam therapy dose on outcome [43]. Likewise, the use of adjuvant intraoperative radiation therapy provided no benefits compared with external beam treatment alone [43]. This study suggests that the true effect of radiotherapy remains unclear.

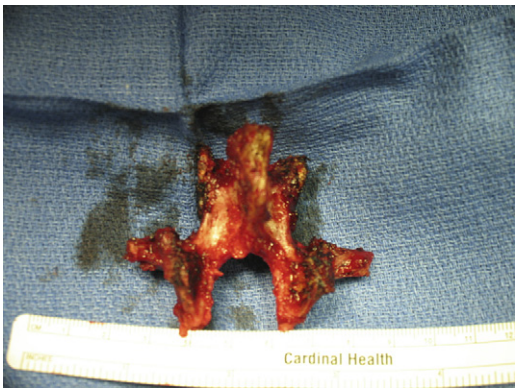

Fig. 12. Case 3: Posterior elements removed en bloc at pedicle level.

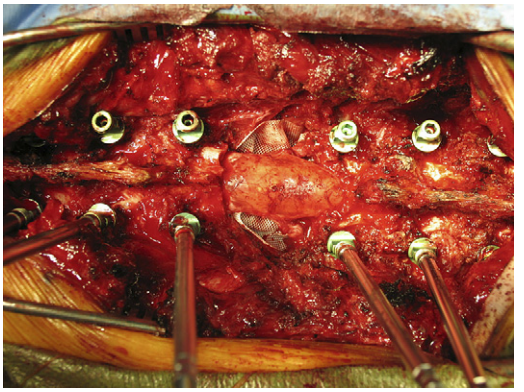

Fig. 14. Case 3: Protective barrier placed between the posterior longitudinal ligament and the thecal sac.

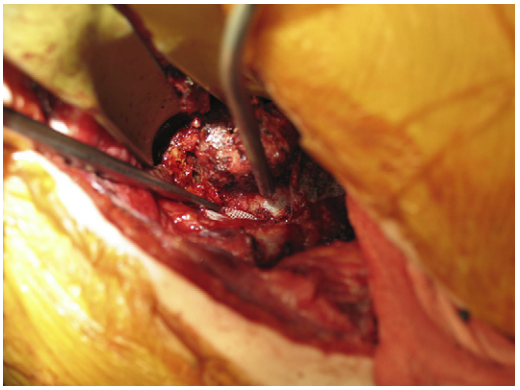

Fig. 15. Case 3: On anterior approach, vertebral body mobilized away from thecal sac after dissection performed. It can be removed en bloc.

Summary

Spinal sarcomas are rare tumors of the spine. Signs and symptoms of these tumors tend to be nonspecific. Delay of diagnosis is not uncommon. A high index of suspicion in young patients presenting with persistent back pain that is unresolved with conservative therapy should prompt further investigation, including CT or MRI of spine. Treatment of these tumors consists of surgery, radiation, and chemotherapy. Although surgery in the past mainly consisted of decompression and debulking, development of new surgical techniques such as en bloc resections may ultimately improve outcomes.

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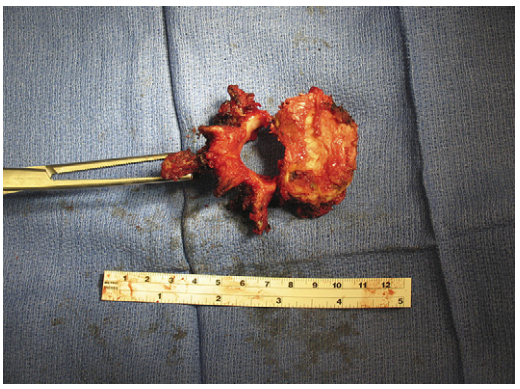

Fig. 16. Case 3: Anterior and posterior elements reconstructed, showing en bloc spondylectomy resection.

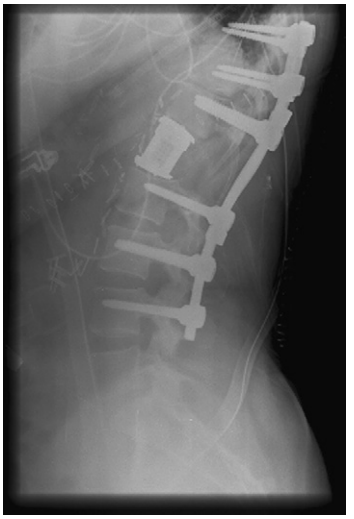

Fig. 17. Case 3: Postoperative X ray showing reconstruction.

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