Total En Bloc Spondylectomy of L3 Vertebra for Histiocytic Sarcoma

Bin Lin, MD; Zhi-wen Chen, MD; Ning Wang, MD; Zhi-min Guo, MD; Hui Liu, MD; Ming Zeng, MD

Abstract

This article describes a rare malignant spinal tumor successfully treated with total en bloc spondylectomy via a posterior approach. The purpose of this study was to emphasize the occurrence of primary histiocytic sarcoma in the lumbar spine. Histiocytic sarcomas are rare, malignant neoplasms of the lymphohematopoietic system that usually occur in the skin, lymph nodes, and intestinal tracts. Primary spinal column histiocytic sarcoma is rare. To the authors’ knowledge, no reports have been published of treating this tumor with total en bloc spondylectomy.

A 27-year-old woman presented with a 2-month history of intermittent low back pain and right lower extremity pain. Magnetic resonance imaging and computed tomography (CT) revealed a lumbar vertebra tumor. Positron emission tomography/CT showed focal accumulation in the tumor site. The patient was diagnosed with a histiocytic sarcoma based on biopsy findings and underwent total en bloc spondylectomy of L3 and reconstruction via a posterior approach. The patient maintained normal neurologic function, and the pain was lessened. No major complications occurred. No radiotherapy or chemotherapy was administered postoperatively, and no local tumor recurrence or distant metastases existed at 2-year follow-up.

The diagnosis of histiocytic sarcoma relies predominantly on the verification of histiocytic lineage and the exclusion of other, poorly differentiated, large-cell malignancies by immunohistochemical stain. Total en bloc spondylectomy at L3 via a posterior approach can be performed safely and is an important approach in the treatment of selected spinal tumors.

Figure: Photographs of the gross view of the anterior (A) and posterior (B) elements of the L3 vertebrae after total en bloc spondylectomy.
Histiocytic sarcomas are rare lymphohematopoietic malignant neoplasms composed of tumor cells showing morphologic and immunophenotypic features of mature tissue histiocytes. The malignancy is a proliferation of histiocytes (tissue macrophages), which are characterized by positive expression of the macrophage-associated antigen cluster of differentiation (CD)68, negative expression of the T-cell-associated antigen CD1a, negative expression of the dendritic cell-associated antigens CD21 and CD35, and lack of Birbeck granules on electron microscopy. This tumor usually occurs in the lymph nodes, skin, and intestinal tract. Primary spinal column histiocytic sarcomas are rare. This article describes an unusual case of histiocytic sarcoma in the L3 vertebra treated with total en bloc spondylectomy and L1-L5 segmental fusion via a posterior approach. No neurovascular structures were sacrificed.

Case Report

A 27-year-old woman presented with a 2-month history of low back pain and right lower-extremity pain. On examination, she had isolated right quadriceps muscle motor weakness (Medical Research Council Grade 4). Right knee deep tendon reflex was diminished. Laboratory tests showed normal blood counts, liver and renal functions, and tumor markers.

Radiographs showed an osteogenic tumor with a clear border. Computed tomography (CT) images showed a well-demarcated tumor in the L3 vertebral body. The tumor showed homogeneous high-signal intensity on a T1-weighted magnetic resonance imaging (MRI) and heterogeneous low- and high-signal intensities on T2-weighted MRI (Figure 1). The tumor extended into the paravertebral tissue and anterior epidural space (C).

Surgical Technique

The patient was placed in the prone position, and the posterior elements of L1-L5 were exposed via a standard midline posterior approach. The posterior route biopsy tract was isolated and resected. An intraoperative anteroposterior radiograph with markers confirmed the appropriate levels. Titanium pedicle screws (Wego Ortho, Weihai, China) were placed at L1, L2, L4, and L5. Posteriorly, wide dissection was performed along and underneath the L3 transverse processes bilaterally. After laminectomy of the lower half of the L2 lamina using a high-speed diamond burr (Medtronic, Minneapolis, Minnesota), a Gigli saw (Wego Ortho, Weihai, China) was introduced into the vertebral foramen and pulled out through the adjacent intervertebral foramen. After the pedicles were cut, the entire posterior element of L3 was removed in 1 piece by transection of the L2-L3 and L3-L4 facet capsules. An electric cautery knife (Devel, Beijing, China) was inserted into the affected pedicle to coagulate the tumor tissue inside the pedicle to prevent tumor cell contamination. After en bloc laminectomy, the cut surfaces of the pedicles were sealed with bone wax to

Figure 1: Sagittal T1-weighted magnetic resonance image (MRI) (A) and low-signal coronal T2-weighted MRI (B) showing a tumor with homogeneous high-signal intensities. Axial T2-weighted MRI revealing the tumor affecting the L3 vertebral body and right pedicle and extending into the paravertebral tissue and anterior epidural space (C).

Figure 2: Photographs of the gross view of the anterior (A) and posterior (B) elements of the L3 vertebrae showing total en bloc spondylectomy.

Figure 3: Anteroposterior (A) and lateral (B) postoperative radiographs showing L3 laminectomy, corpectomy, and placement of an expandable anterior cage between L2-L4 and L1-L5 posterior fusion using pedicle screws, respectively.
The tumor cells had abundant clear and eosinophilic cytoplasm, and the nuclei were grooved, folded, or lobulated. Distinct, small nucleoli existed. Binuclear or polynuclear cells occasionally existed. Acidophilic granulocytes and occasional lymphocytes existed (hematoxylin–eosin stain, ×100 objective) (A). Positive immunohistochemical staining for cluster of differentiation 68 (immunoperoxidase stain, ×100 objective) (B).

The anterior spinal column was reconstructed by implantation of a titanium mesh cage (Stryker, Kalamazoo, Michigan) filled with allograft bone (Xin Kang Chen Medical Technology Development Co., Ltd., Beijing, China). The fluoroscopic image showed excellent position, and the opposite segmental rod was attached to the pedicle screws. The L1-L2 and L4-L5 facets were subsequently destroyed, and allograft bone graft was used for posterolateral fusion (Figure 3). The L3 vertebra was completely removed (Figures 2-4), and the wound was closed. Operative time was 6 hours, and blood loss volume was 1900 mL. The wound healed uneventfully.

Postoperative Management
The patient had intact sensorimotor function of the lower extremities postoperatively. Two weeks postoperatively, the patient was allowed off of bed rest and walked with a thoracolumbosacral orthosis. The patient and her family refused chemotherapy and radiotherapy. At 2-month follow-up, the patient was able to walk with no restriction, had no pain, and had normal neurological function. No local tumors or distant metastases recurred, and MRI (Figure 6) and whole-body PET/CT (Figure 7) revealed no disease at 2-year follow-up.

Histological Findings
Histological examination of the sagittal sections showed that the disks were not involved and that the portion of the tumor involving the right pedicle was surrounded. The axial sections revealed that the right paravertebral and epidural portions of the tumor were covered with a pseudocapsule. The margin of excision was marginal or more than marginal, except at the right paravertebral and right pedicle, where it was intrasional.

Histopathologically, the tumor cells had abundant clear and eosinophilic cytoplasm. The nuclei were grooved, folded, or lobulated. Small distinct nucleoli were present. Binuclear or polynuclear cells occasionally existed. Focal necrosis existed, but mitotic figures were rare. Scattered acidophilic granulocytes and occasional lymphocytes existed between these cells (Figure 5).

Immunohistochemically, the tumor cells were positive for CD68, leukocyte common antigen, vimentin, CD43, CD45RO, S100 protein, and were negative for cytokeratin, epithelial membrane antigen, CD34, synaptophysin, smooth muscle antigen, des, human melanoma black 45, kappa, Lambda, CD15, CD30, alkaline, CD20, CD79a, CD3, CD57, CD99, glial fibrillary acidic protein, and myeloperoxidase. The Ki-67 (MIB-1) index was 20%. These morphologic and immunohistochemical features are typical of histiocytic sarcoma (Figure 5).

Discussion
Histiocytic sarcomas are rare malignancies, representing <1% of all non-Hodgkin’s lymphomas, which demonstrates morphologic and immunopheno-
Morphologically, malignant cells have large eccentrically placed oval nuclei with vesicular chromatin and a prominent single irregular nucleolus. The nucleus may appear grooved. Cytoplasm is abundant and eosinophilic and may be foamy or vacuolated. Large multinucleated tumor cells and multiple nucleoli may also exist. Ultrastructural features of the neoplastic cells include abundant cytoplasm, with several lysosomes. Birbeck granules and cellular junctions are essentially absent. The tumor cytology and architecture is not particularly unique; therefore, immunohistotypic and molecular studies are essential for diagnosis.

The immunohistochemical profile shows expression of the lysosome-associated (macrophage or histiocytic) markers CD68 (in 100% of cases) and lysozyme (in 94% of cases),\textsuperscript{9} CD45, CD45RO, and human leukocyte antigen-disease resistance are usually positive in histiocytic sarcoma. The Ki-67 index varies from 10% to 90% of tumor cells. In addition, they were negative for molecules related to B-cells, T-cells, accessory or dendritic cells, myeloid cells, epithelial cells, and melanocytes. Focal reactivity for the S100 protein (typically expressed in Langerhans` cell tumors) can exist in normal macrophages and histiocytic sarcoma. Because of the monocytic origin of histiocytes, monoblastic leukemia should be excluded.\textsuperscript{10} Recently, CD163, a hemoglobin scavenger receptor, has been recognized as a new macrophage-related differentiation marker, which is more specific than the conventional histiocyte-related molecules, such as CD68 and lysozyme.\textsuperscript{7}

Histiocytic sarcoma has a poor prognosis because of its aggressive nature and high proliferative rate, extranodal spread, and poor response to therapy, although the clinical presentation and involvement site vary.\textsuperscript{5,9} Radical surgery with wide surgical margins at an early stage of the disease are beneficial for those with histiocytic sarcoma to prevent continuous dissemination from the primary tumor site.

Enneking et al\textsuperscript{11} examined the process of the local spread of primary bone and soft tissue tumors of the extremities and proposed "the concept of compartment and anatomic barriers." In a histological study of specimens resected at total en bloc spondylectomy in the spine, Fujita et al\textsuperscript{12} determined the pattern of local spread of spine tumors and identified barrier tissues to the spread of a tumor. Although en bloc resection with marginal or wide margins is a common procedure in malignant tumors of the extremities, it is difficult to achieve in spinal tumors due to anatomic reasons. Conventional piecemeal intralosional resection with curettage is more common, even in primary malignant spinal tumors or solitary spinal metastases. After Lièvre et al\textsuperscript{13} performed the first total spondylectomy by piecemeal resection of a giant cell tumor involving L4 in 1966, several reports and techniques have been described for total spondylectomies for reducing local recurrence of vertebral tumors.\textsuperscript{14-17}

However, most reported procedures were based on intracapsular curettage or

**Figure 6:** Magnetic resonance imaging revealing no local tumor recurrence at 2-year follow-up.

**Figure 7:** Coronal (A), sagittal (B), axial (C), and coronal (D) positron emission tomography/computed tomography of the whole body showing no local tumor recurrence or distant metastases at 2-year follow-up.
piecemeal resection of malignant neoplasms. Total spondylectomy can result in tumor cell contamination, massive bleeding that is difficult to control, and incomplete resection.17,18 To avoid this mishap, Tomita et al18,19 modified and refined the techniques to achieve an en bloc resection of the entire vertebra via a posterior approach using a T-saw (modified Gigli saw) and reported good results for metastatic tumors and primary malignant vertebral tumors.20 Total spondylectomies with en bloc tumor excision seem to promote more complete tumor resection than piecemeal total spondylectomy, with good bleeding control, decreased local recurrence, and possibly improved survival. More recently, Abe et al,21 Matsuda et al,22 and Melcher et al23 described total en bloc spondylectomy for reducing the local recurrence of vertebral tumors, with excellent clinical results.

Total en bloc spondylectomy for the treatment of disease localized in the lower 3 lumbar vertebrae has rarely been reported. This region’s unique anatomy makes total en bloc spondylectomy particularly challenging, especially posterior total en bloc spondylectomy. The nerve roots are important for motor function of the legs. Although sacrificing the nerve root above L2 does not cause serious neurologic deficits, sacrifice of the L3 nerve root does.21 The ilopsoas muscle is bilateral and must be dissected. The vertebral bodies are larger than they are elsewhere, and the iliac vessels are close to them bilaterally and are frequently adherent. In addition, the ureters, located just laterally, are at risk for injury at the lower lumbar levels. Therefore, tumor resection at this level via a posterior approach can be difficult.

According to Tomita et al,18,19 the ipsilateral lateral margin and contralateral pedicle should be cut if 1 pedicle is involved. Because the L3 vertebral body is larger than the vertebral body of the rostral side, the risk for nerve root injury is high when it is rolled out around the dural tube between the nerve roots. Furthermore, if the right lamina is cut, limitations exist to the lateral exposure of the right L3 nerve root. The narrow pedicles was the ideal site because the nerve roots and cord can be freed with no trauma, and the intralesional cut surface area and the risk for contamination can be minimized. Therefore, it was possible to obtain a wide surgical margin in the posterior elements, a marginal margin in the spinal canal, and an intralesional margin in the pedicles. Total L3 en bloc spondylectomy was accomplished via a posterior approach, and no significant sensory or motor deficits occurred. This case illustrates that posterior total en bloc spondylectomy at L3 can be performed safely and should be recognized as an important approach in the treatment of selected cases of spinal tumors involving L3.

**References**