L2 Chordoma in an 11-Year-Old Girl

Asheesh Bedi, MD, Ramesh C. Srinivasan, MD, Laurel C. Blakemore, MD, and Gregory P. Graziano, MD

Chordomas are rare malignant bone tumors that account for only 1% to 4% of all primary bone tumors. Originating from embryonic remnants of the primitive notochord, these lesions arise in the midline of the axial skeleton.[1-4] Chordomas are predominantly found in the sacrococcygeal and clival (skull base) regions and typically affect adults in the fifth to seventh decade of life. Prognosis for these radioresistant lesions is highly dependent on complete resection with proven, tumor-free surgical margins.[5-9]

The unusual case of an L2 chordoma in a pediatric patient managed with en bloc vertebral resection and T12–L3 segmental fusion via a posterior approach is presented. A computer image guidance system was used to assist with pedicle screw placement and spinal instrumentation.[10,11]

Case Report

An 11-year-old girl was brought to her primary physician for the complaint of intermittent abdominal pain over 1 year. Her mother reported that the girl had frequent (weekly) episodes of generalized abdominal pain without any clear precipitants. These episodes were often accompanied by nausea and diarrhea, but no emesis, melena, or blood per rectum was noted. The patient's past medical history was remarkable for premature birth at 24 weeks complicated by necrotizing enterocolitis requiring multiple abdominal surgeries as an infant. Review of systems was significant for intermittent mild headaches with vertigo and infrequent "fainting" spells over the preceding 6 months. The patient denied seizures, focal neurologic deficits, back pain, and visual deficits. Computed tomography (CT) and magnetic resonance imaging (MRI) scans of the head (obtained by the primary physician) showed no abnormalities. Given the patient's history of complex abdominal surgery, she was referred to gastroenterology specialists for further evaluation of symptoms.

An abdominal CT scan taken to assess for inflammatory bowel disease incidentally identified an enhancing lesion within the L2 vertebral body. Plain radiographs of the lumbar spine showed a lytic, sclerotic lesion involving the posterior cortex of the L2 vertebral body (Figure 1). No compression deformity or canal extension was noted. A bone scan revealed no significant abnormalities or increased activity at the L2 level. An MRI scan of the lumbar spine showed a contrast-enhancing lesion in the posterior two thirds of the L2 vertebral body with slight distortion of the posterior cortex and extension into the left pedicle (Figure 2). Musculoskeletal CT scan obtained per image-guided software protocol (StealthStation Surgical Navigation Technology; Medtronic-Sofamor-Danek, Louisville, Colo) confirmed the presence of a large, lytic lesion affecting the posterior L2 vertebral body and anterior left pedicle. Multiple punctate areas of calcification were noted within the lesion, and erosion of the posterior cortex of the L2 body was appreciated (Figure 3). Given the aggressive appearance of the lesion, image-guided biopsy was performed; it yielded physaliporous cells consistent with chordoma. The diagnosis was discussed extensively with

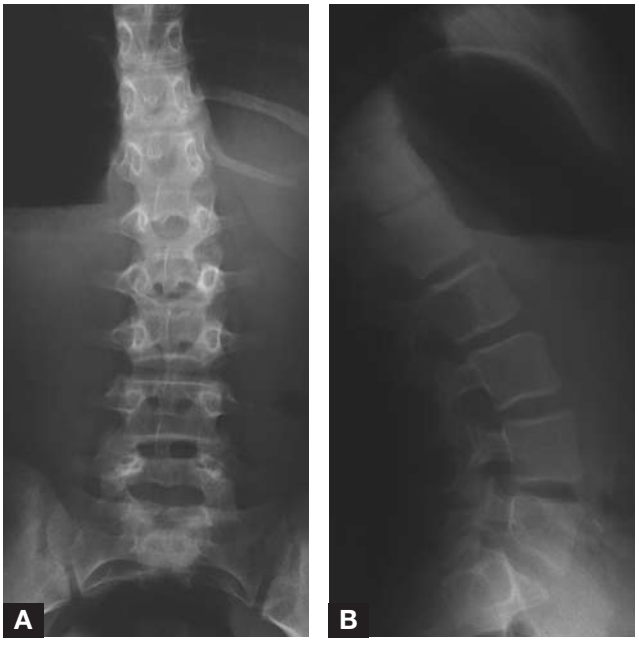

Figure 1. Anteroposterior (A) and lateral (B) radiographs of lumbar spine show lytic, sclerotic lesion within posterior L2 vertebral body.

Dr. Bedi is Fellow, Sports Medicine and Shoulder Surgery, Hospital of Special Surgery, New York, New York.
Dr. Srinivasan is Resident Physician, Department of Orthopaedic Surgery, University of Michigan Health System, Ann Arbor, Michigan.
Dr. Blakemore is Chief, Division of Orthopaedic Surgery, Children's National Medical Center, Washington, DC.
Dr. Graziano is Associate Professor and Chief, Section of Spine Surgery, Department of Orthopaedic Surgery, University of Michigan Health System, Ann Arbor, Michigan.

Address correspondence to: Gregory P. Graziano, MD, Section of Spine Surgery, Department of Orthopaedic Surgery, Taubman Center 2912, University of Michigan Health System, 1500 E Medical Center Dr, Ann Arbor, MI 48105 (tel, 734-936-7299; fax, 734-936-8611; e-mail, graziano@umich.edu).

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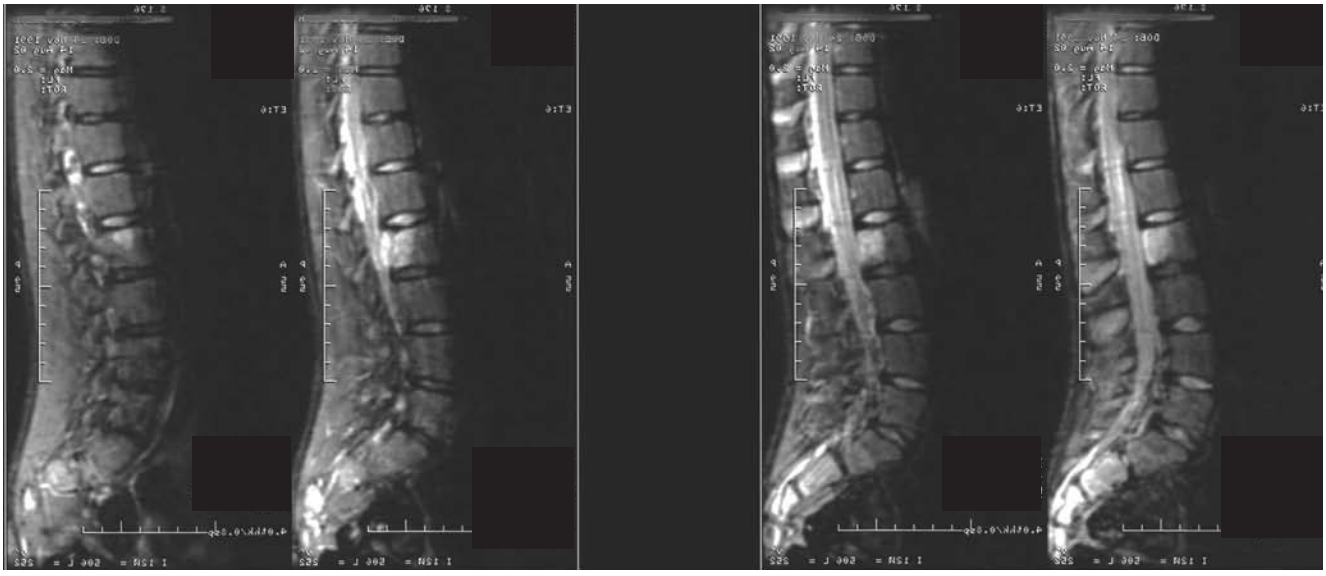

Figure 2. Sagittal T_2-weighted magnetic resonance imaging scan shows enhancing lesion in posterior L2 vertebral body with distortion of posterior cortex and extension into left pedicle.

the patient and family, and recommendation was made for en bloc resection of the tumor and image-guided spinal instrumentation via a posterior approach.

One year after initial presentation, the patient underwent surgery. She was positioned prone on the Jackson spinal frame, and somatosensory evoked potential (SSEP) monitoring was instituted. The posterior elements of T12–L3 were exposed via a standard midline posterior approach.[10,11] The posterior route biopsy tract was carefully isolated and resected. An intraoperative anteroposterior x-ray with markers confirmed appropriate levels. Titanium pedicle screws were placed with StealthStation guidance at the T12, L1, and L3 levels without complication.[10,11] Wide dissection was carried out along and underneath the L2 transverse processes bilaterally. The L1–L2 and L2–L3 foramina were identified and cleared of soft tissue to allow for easy insertion of dural separators. The L2 pedicles were transected with a Kerrison rongeur below the transverse processes. After the pedicles were cut, the laminae were carefully removed by transection of the L1–L2 and L2–L3 facet capsules. The cut surfaces of the pedicles were sealed with bone wax to decrease bleeding and risk for tumor cell contamination. This provided excellent visualization of the L2 nerve roots and thecal sac. Circumferential extraperiosteal dissection was completed at the L1–L2 disc level and extended distally to the L2–L3 disc. Two Gigli saws were passed anterior to the L2 vertebral body and were used to cut through the L1–L2 and L2–L3 discs without complication. Ribbon and Scoville rib retractors protected the dural sac, nerve roots, and blood vessels during passage of the saw. One rod was then placed to stabilize the spine. The L2 vertebral body was subsequently rotated out to the left and removed en toto (Figure 4). Residual disc material on the L1 and L3 vertebral bodies was removed with curettes. A femoral strut allograft packed with Healos osteoconductive matrix (DePuy Spine, Raynham, Mass) soaked in

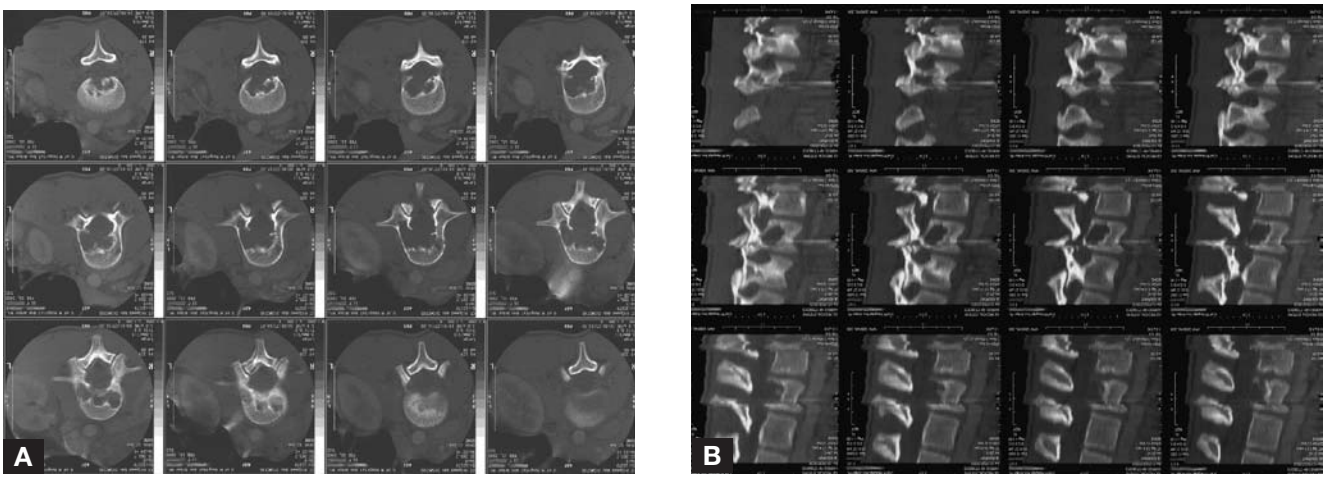

Figure 3. Thin-section axial (A) and sagittal (B) computed tomography scans show large, lytic lesion affecting posterior L2 vertebral body and left pedicle. Erosion of posterior wall is appreciated. Multiple areas of punctuate calcification within lesion are noted.

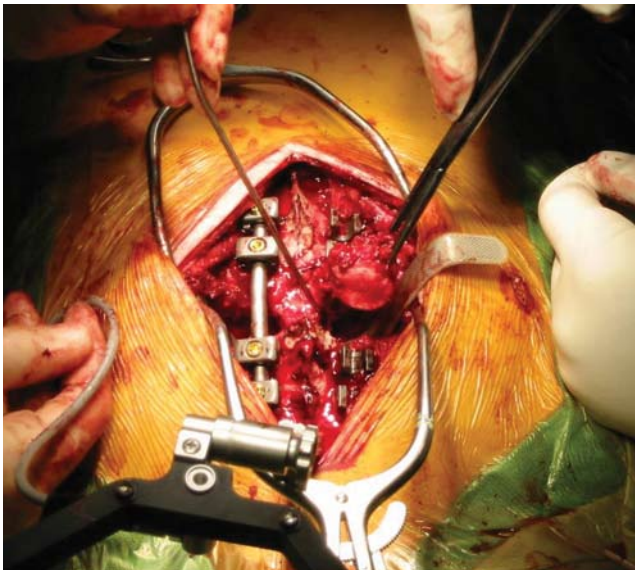

Figure 4. Intraoperative photograph shows technique of unilateral spinal column stabilization and en bloc corpectomy of L2 body.

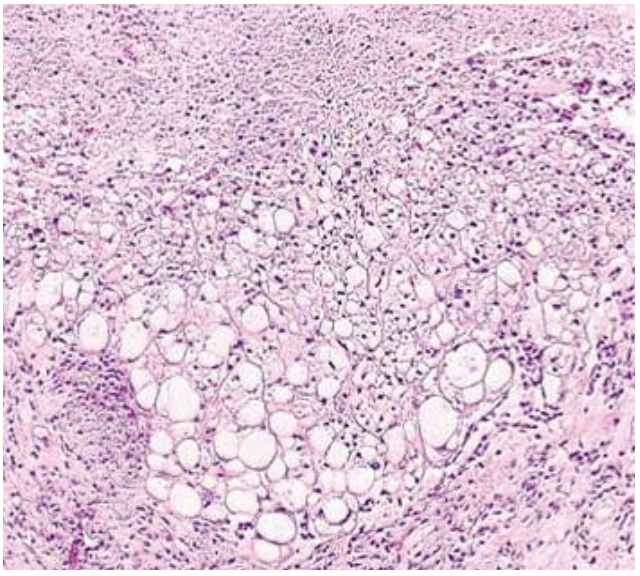

Figure 5. Hematoxylin-eosin–stained section of surgical specimen shows lobules of physaliporous cells separated by fibrous septa.

bone marrow aspirate was rotated into position around the dural sac. The opposite segmental rod was attached to the pedicle screws, and compression was applied bilaterally across the anterior strut graft. In situ rod benders were used to restore physiologic lumbar lordosis. T12–L1 facets were subsequently destroyed, and autogenous bone graft, bone marrow aspirate, and Healos osteoconductive matrix were positioned for posterolateral fusion. The wounds were copiously irrigated, and closure of the muscular fasciae and dermal layers was completed in standard fashion. No SSEP signal abnormalities were noted throughout the procedure, and the patient had intact sensorimotor function of the lower extremities after surgery.

Radiographs showed excellent position and locking of the anterior strut graft as well as restoration of lumbar lordosis with posterior instrumentation. Specimen analysis revealed a 2×1.5×1-cm cystic lesion within the L2 vertebral body. Histologic analysis revealed lobules of physaliporous cells separated by fibrous sheets consistent with the diagnosis of chordoma (Figure 5). In the horizontal plane, marginal negative margins along the L2 body edges were obtained. An intralesional margin was expected and confirmed at the left pedicle. In the sagittal plane, the upper and lower edges of the L2 body demonstrated wide negative margins. The laminae and spinous process had wide negative margins in both planes.

After surgery, the patient was fit with a thoracolumbosacral orthosis (TLSO) and mobilized out of bed by postoperative day 4. Recovery was marked by postoperative ileus and mild pancreatitis, which resolved with bowel rest and nasogastric decompression. The patient subsequently resumed a regular diet and was discharged without complication.

The patient, followed regularly in our clinic after surgery, continued to do well. She was weaned from the TLSO and discontinued using it 3 months after surgery. She was pain-free and had no focal neurologic deficits. She resumed noncontact sports at school and initiated gentle back flexion-extension exercises. Compared with thin-cut CT scan, radiographs are somewhat limited in their ability to show fusion and incorporation of large allografts in the lumbar spine. Although radiographs obtained at the patient's most recent visit (12 months after surgery) did not convincingly show allograft incorporation inferiorly or bone formation posterolaterally, significant bony callus was appreciated with preservation of sagittal alignment and hardware integrity (Figure 6). Clinical examination and chest radiographs showed no metastatic disease.

Discussion

Accounting for only 1% to 4% of primary osseous lesions, chordomas are rare bone tumors arising from remnants of the

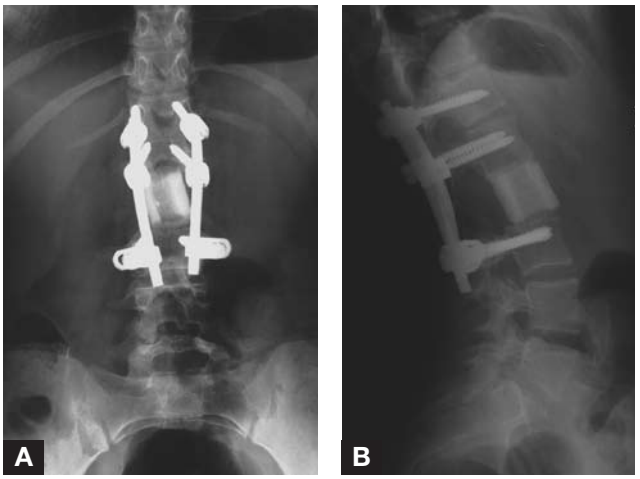

Figure 6. Anteroposterior (A) and lateral (B) radiographs of lumbar spine at 1 year follow-up show incorporation of graft and preserved sagittal alignment with T12–L3 segmental posterior fusion.

embryologic notochord. They typically arise in adults with a 2-to-1 male-to-female predominance. Eighty-five percent of chordomas are localized to the skull base and sacrococcygeal regions, and 50% of all sacral tumors are chordomas. Infrequently, chordomas arise in the cervical, thoracic, or lumbar spine; involvement of the cervical spine has been most commonly reported.[1-4,6,9,12-14] In these instances, isolated vertebral involvement with sparing of the adjacent discs is typical. Osseous destruction progresses from the body to the posterior elements via the pedicles, with progressive risk for vertebral body fracture and collapse.[15] Epidural extension of the tumor is not uncommon.[3,6,7,16] Metastatic disease is found in approximately 7% to 14% of patients. Spread typically occurs to regional lymph nodes, lungs, brain, and abdominal viscera and is associated with a poor prognosis. For patients with sacrococcygeal chordomas, mean duration of survival after symptom onset is 5.7 years, despite treatment.[2-6,8]

Clinical Presentation. The clinical presentation of chordomas varies widely and is highly dependent on tumor location. The insidious growth of these lesions often results in a significant delay to diagnosis. Clival lesions are benign, but local growth and invasion of critical adjacent structures can precipitate cranial nerve palsies, facial pain, and ataxia. Sacrococcygeal and vertebral lesions initially present with focal back pain.[2,3] Constipation, urinary retention, or radicular symptoms are suggestive of tumor extension into the spinal canal or foramina. It is unclear whether our patient's abdominal pain resulted from the occult malignancy or from the history of complex abdominal surgery. Lack of intraoperative cord or root compression is more suggestive of primary gastrointestinal pathology.

Imaging Findings. Chordomas appear as expansile, lytic lesions on plain radiographs. Intralesional calcification and trabeculation are common.[1-3,13] These findings, however, are neither highly sensitive nor highly specific for chordoma. Bone scans are similarly unreliable and often fail to identify the lesion, as in our patient's case.[3,17,18] MRI and CT are both extremely helpful and have complementary roles in diagnosing and managing chordomas. MRI accurately evaluates extension of the tumor and involvement of neural roots and the thecal sac.[17] CT defines the degree of bony destruction, stability of the vertebral column, and pattern of intralesional calcification.[18] The aggressive appearance of our patient's lesion on CT called for image-guided biopsy, which led to prompt diagnosis of the malignancy.

Pathology Findings. Chordomas grossly appear as soft, lobulated masses with a surrounding fibrous pseudocapsule. They often track along nerve roots or into the epidural space and encase critical structures, making complete resection without neurologic injury difficult.[1,3-8,13] Lobules of physaliporous cells separated by fibrous septa is a pathognomonic histologic finding.[1,3] These cells are defined by eosinophilic cytoplasm and prominent mucin-filled vacuoles and are identical to those seen in the embryonic notochord (Figure 4).

Management

Successful treatment of chordomas is difficult. Extension and invasion of local neurovascular structures complicate attempts at curative resection. Local recurrence is frequent, and metastasis, though uncommon, may occur in 7% to 14% of cases.[1,2,5,8] Complete excision remains the only chance for curative treatment, as the role of primary chemotherapy or radiation remains unclear. En bloc resection with proven tumor-free surgical margins is one of only a few interventions that have been correlated with a favorable prognosis for disease-free survival.[5-9,12-14] Boriani and colleagues[6] reported on the treatment and outcome in 21 cases of chordomas in the mobile spine. Radiation therapy was not effective in eradicating the tumor, even when accompanied by intralesional or debulking surgery. En bloc excision, even when marginal, has yielded the most favorable outcomes.[6,7,18] Biagini and colleagues[15] recently reported 4 years of disease-free survival after en bloc vertebrectomy and dural resection for an L1 chordoma with extensive dural infiltration. Adjuvant radiation therapy may improve the prognosis when marginal excision of the tumor is achieved.[3,5-9,12-15]

"...intralesional curettage and failure to biopsy the lesion could have catastrophic consequences on the ultimate prognosis for tumor-free survival."

Our patient's case highlights several important principles. It is critical for a clinician to maintain a high level of suspicion for chordoma despite unusual axial location or atypical patient demographics. The age and sex of our patient and the location of the lesion at the L2 level are not characteristic of chordomas. Aneurysmal bone cyst, hemangioma, osteoblastoma, or fibrous dysplasia may be higher on the differential diagnosis in the pediatric population. Empiric management with intralesional curettage and failure to biopsy the lesion could have catastrophic consequences on the ultimate prognosis for tumor-free survival.

In addition, en bloc excision with negative surgical margins is one of the controllable factors associated with improved survival and reduced rate of local recurrence of chordomas.[5-8,12-15] Conventionally, piecemeal intralesional resection with curettage was used, given the difficulty of the surgical approach and the proximity of the lesion to the aorta. Magerl and Coscia[19] described a total posterior vertebrectomy technique with a piecemeal resection technique for spinal metastatic lesions. This procedure, however, is associated with significant potential for local

tumor cell contamination and incomplete resection with intralesional margins.[19,20] Tomita and colleagues[20,21] subsequently described the total en bloc spondylectomy (TES) technique, in which bilateral pediculotomy allows for posterior en bloc laminectomy and en bloc corpectomy, facilitating tumor mass excision with wide or marginal margins. Encasement of the spinal cord within the ring of the vertebra necessitates bilateral ring transection for excision without neurologic compromise. The narrow pedicles are often an ideal site, as the nerve roots and cord can be protected, and the intralesional cut surface area and the risk for contamination can be minimized.[20,21]

The TES technique, used in our patient's case, allowed for complete vertebrectomy of L2 with negative margins and subsequent segmental fusion without complication. The potentially increased morbidity associated with an anterior exposure was avoided.[3,19-22] Furthermore, use of a computer image-guidance system allowed for secure pedicle screw fixation and maintenance of a stable spine in spite of vertebral resection in this pediatric patient. She had no postoperative complications and remained disease-free according to surveillance imaging studies. Use of this approach extends beyond primary tumors to include isolated vertebrectomy for metastatic disease or infection.[3,7,20,21] Boriani and colleagues[7] reported on 29 cases of en bloc resections for aggressive primary lesions or solitary metastases with no local recurrences or iatrogenic neurologic complications.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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