Two-level thoracic pedicle subtraction osteotomy for progressive post-laminectomy kyphotic deformity following resection of an unusual thoracolumbar intradural extramedullary tumor

Case report

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The authors report a case in which multilevel thoracic pedicle subtraction osteotomy (PSO) was performed to correct post-laminectomy kyphotic deformity in a 9-year-old boy presenting with worsening lower-extremity neurological deficits. Five years prior to presentation, the patient underwent multilevel thoracolumbar laminectomies for resection of an atypical teratoid/rhabdoid tumor (AT/RT), a rare lesion that typically occurs intracranially and has a poor prognosis, making this particular presentation unusual and the patient’s subsequent postoperative course remarkable. No fusion was undertaken at the time of resection, given the patient’s age and presumptive poor prognosis. Over the next 5 years, the patient developed progressive thoracolumbar kyphotic deformity, with a Cobb angle greater than 110°, despite bracing, and bilateral lower-extremity weakness requiring ankle-foot orthotics for continued ambulation due to progressive foot drop. Worsening gait and the onset of respiratory issues prompted surgical intervention. Multilevel thoracic PSO and thoracolumbar fusion were performed, resulting in improved lower-extremity function and correction of the kyphotic deformity to approximately 65°. This report outlines an unusual AT/RT presentation and postoperative course and also discusses literature related to PSO within the context of pediatric kyphotic deformity. The authors’ experience supports the use of multilevel PSO with fusion as a potential treatment option for significant pediatric thoracolumbar kyphotic deformity requiring surgical correction.

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KEY WORDS • pedicle subtraction osteotomy • kyphotic deformity • atypical teratoid/rhabdoid tumor • spine

Correction of spinal deformity in the pediatric population is a challenging surgical problem given limitations related to bone size and the need to accommodate future growth. While corrective surgery is often delayed to allow for additional growth as well as further development of bony structures to improve options for spinal instrumentation, certain conditions necessitate more urgent surgical intervention. Kyphosis is a condition characterized by abnormal sagittal alignment of the spine and is not often encountered in the pediatric population. While frequently seen in elderly persons as a consequence of aging, pediatric kyphotic deformity within the thoracolumbar region may be due to congenital, neuromuscular, or idiopathic etiologies. It may also be due to a post-laminectomy effect secondary to a previous procedure. Progressive deformity may lead to problems with ambulation due to sagittal imbalance and, if severe, neurological compromise. Initially, nonsurgical treatment options such as bracing are attempted; however, if these options fail to correct deformity, corrective surgery is required. In this article, we describe the use of multilevel PSO and segmental instrumentation in a pediatric patient to correct severe progressive thoracolumbar kyphosis resulting from previous multilevel laminectomies for resection of a relatively rare intradural extramedullary tumor.

Case Report

History and Presentation. This 9-year-old boy had progressive post-laminectomy thoracolumbar kyphosis following urgent surgery for a malignant spinal cord tumor 5 years previously. The patient was first referred to the neurosurgical service at our institution at the age of 4 years. At that time he had a 2-week history of constipation and had been unable to ambulate for several days.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
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On physical examination, he was found to have lower-extremity paraplegia. Imaging revealed an intradural examedullary mass lesion extending from approximately T-10 to L-1 (Fig. 1). Spinal axis and intracranial imaging was otherwise unremarkable for disease (Fig. 2). The patient underwent emergent T9–L3 laminectomies for subtotal tumor resection and was found to have an AT/RT. He recovered from his paraplegia to paraparesis requiring ankle-foot orthotics and crutches for ambulation. His lower-extremity strength varied between 2 to 4 out of 5 in his various muscle groups. He had no urinary or fecal incontinence. He subsequently received localized radiation therapy as well as multiple rounds of systemic and intrathecal chemotherapy. Over the next few years the patient did well, surviving various complications related to his tumor therapy, including neutropenia and bacteremia. He was followed in pediatric neurosurgery and orthopedic surgery clinics during this time, and despite bracing, development of progressive thoracolumbar kyphosis was observed. Measurements eventually indicated a deformity greater than 110° (Fig. 3). His gait worsened to the point of bilateral foot drop, and breathing difficulties developed due to the severity of his deformity. Given the progressive nature of the patient’s symptoms and deformity, surgical correction was undertaken.

Operation and Postoperative Course. The patient was placed prone on a Jackson table and positioned so that appropriate correction of the kyphotic deformity could be achieved. Neuromonitoring was used to evaluate somatosensory and motor evoked potentials; intraoperative electromyography was also performed. The previous incision was marked and reopened just off midline to avoid injury to the spinal cord. The posterior elements of T-7 through L-4 were exposed, and intraoperative fluoroscopy was used to confirm appropriate levels. The inferior facets were removed bilaterally from T-7 through T-10 and from L-2 through L-4, and polyaxial pedicle screws were placed. Reduction screws were used at L-2 and L-4. The first in a series of temporary rods contoured for the kyphosis was then placed. Pedicle subtraction osteotomy was performed first at T-12 and then at T-11. Using progressively straighter rods, a safe degree of correction was achieved. Intraoperative radiographs revealed correction to within the range of 50°–60°. Additional correction was limited by a reduction in motor evoked potential amplitude that occurred with continued kyphotic reduction, but the motor evoked potential amplitude returned to baseline with lessening of the reduction. Six-millimeter-diameter titanium rods were then sized, cut, shaped, and secured with setscrews. The reduction screws were broken off. Cortical cancellous autograft bone chips were placed along the length of the construct to promote arthrodesis. Following copious irrigation, members of the plastic surgery service performed a latissimus dorsi flap rotation to cover the surgical area and provide wound closure. The patient was at his neurological baseline following the procedure.

Over the next few days, members of the physical therapy staff worked with the patient, and he was able to ambulate with the assistance of a rolling walker, spinal arthrodesis, and ankle-foot orthotics. He was discharged home on postoperative Day 8. He continued to wear a total contact orthosis brace for approximately 1 year after surgery. In postoperative follow-up, he was able to stand erect and ambulate with a shuffling gait. His proximal lower-extremity strength was better than his distal strength. Follow-up imaging indicated a stable construct, with correction of his kyphosis to approximately 65° (Fig. 4). A PET study performed for oncological surveillance 18 months after the spinal deformity corrective surgery (approximately 5 years after initial surgery and diagnosis) did not show any evidence of tracer uptake within the previous surgical site or was systemically suggestive of recurrent or metastatic disease (Fig. 5).

Discussion

Our case report is unique in 2 respects. The first is the unusual tumor location and postoperative course; the second is the description of a 2-level thoracic PSO deformity-correction procedure in a pediatric patient. Atypical teratoid/rhabdoid tumors account for less than 5% of all pediatric CNS tumors and usually are seen in patients younger than 2–3 years of age. They are highly malignant pediatric embryonal CNS tumors (WHO Grade IV) that exhibit rhabdoid neoplastic cells with or without nonrhabdoid components. Medulloblastoma-like and/or primitive neuroectodermal tumor–like components are the most common nonrhabdoid elements. Atypical teratoid/rhabdoid tumors are associated with deletion or mutation of the SMARCB1 gene (also referred to as INI-1, hSNF5, or BAF47) on the long arm of chromosome 22 (22q11.2). Cytopathological analysis revealed a highly cellular tumor with a high degree of pleomorphism. Larger tumor cells showed ample eosinophilic cytoplasm and occasionally hyaline inclusion material. Nucleoli were also prominent in numerous tumor cells. There were frequent mitotic figures and areas of hemorrhagic necrosis with Ki 67 staining estimated at 47%. Focal inflammatory infiltrates were also observed. Staining was
positive for vimentin, cytokeratin, EMA, and SMA and negative for skeletal muscle markers including myogenin and myoD1, distinguishing it from rhabdomyosarcoma (Fig. 6). Immunohistochemical testing of tumor sections showed no evidence of INI-1 expression (Fig. 7). This tumor is most commonly found intracranially within the posterior fossa, and there are only rare reports of AT/RTs with a primary spinal cord origin. Even with radiation and chemotherapy, mean survival for patients with this lesion is typically less than 6 months; the 3-year overall survival rates are less than 25%, although one study suggests improved survival for those patients who are at least 3 years old at the time of diagnosis, while a second study indicates that early initiation of radiotherapy may benefit patients 3 years or younger.

In the present case, given the diagnosis and associated prognosis, spinal fusion was deferred. Our patient’s unexpected 5-year survival after initial diagnosis and treatment was a welcome surprise.

As previously mentioned, the surgical correction of complex spinal deformities in pediatric patients is a challenging endeavor. Prior to undertaking kyphosis-correction surgery in this population, it is important to appreciate that normal thoracic kyphosis progresses until the end of adolescence and ranges from 20° to 50°. In the developing spine, however, loss of the posterior ligamentous restraints is concerning because the remaining structural elements will continue to grow, allowing for rapid progression of the deformity. Thus, laminectomy and postsurgical radiation therapy in the developing thoracic spine carry an increased risk for progressive kyphosis. If the deformity remains flexible, bracing remains a good treatment option, with favorable outcomes achieved if the curve can be reduced to less than 50°; however, these outcomes relate to treatment of Scheuermann kyphosis, a congenital condition with pathophysiology significantly different from that of progressive post-laminectomy kyphosis. Bracing therapy may be required for up to a year or longer, although continued worsening of the deformity angle accompanied by the development of neurological, orthopedic, cardiac, and/or respiratory problems necessitates surgical correction.

Depending on the severity and flexibility of the deformity, a potential surgical approach involves a combined anterior-posterior fusion to achieve adequate sagittal correction, but the potential morbidity from pulmonary com-

**Fig. 2.** Representative midline sagittal T1-weighted postcontrast MR image demonstrating no evidence of intracranial disease to suggest potential drop metastasis etiology.

**Fig. 3.** Preoperative sagittal plain radiograph of the spinal axis showing significant thoracolumbar kyphotic deformity with a Cobb angle of approximately 110°.
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Complications associated with an anterior approach makes this surgical option less attractive. Given the morbidity associated with anterior surgical approaches as well as continued improvements in surgical technique, a study by Luhmann et al.9 indicated that the anterior component of a combined approach may not be necessary for correction of adolescent idiopathic scoliosis. With the severity of our patient’s deformity as well as his pulmonary issues, combined anterior-posterior fusion was rejected in favor of a posterior-only approach. Smith-Peterson osteotomy and PSO allow for approximately 10° and 30°–40° sagittal plane correction, respectively, for each spinal level at which the procedure is performed.7,11 With a sagittal deformity greater than 100°, a 2-level PSO was favored over a multilevel Smith-Peterson osteotomy to reduce the total number of vertebral bodies requiring osteotomies. The T-11 and T-12 levels were selected for PSO given that

Fig. 4. Postoperative sagittal plain radiograph of the spinal axis showing T-11 and T-12 PSO along with T7–10 and L2–4 screw-rod fusion. The Cobb angle approximates 65°, with significant improvement of thoracolumbar kyphotic deformity.

Fig. 5. A PET scan obtained 18 months after spinal deformity correction (approximately 5 years after the initial surgery and diagnosis) showing no evidence of tracer uptake within the surgical site or systemically to suggest tumor recurrence.
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these levels were the sites of relative kyphosis and maximum deformity.\textsuperscript{7} Instrumentation was extended 4 levels cephalad and caudad to ensure adequate fixation and reduce the likelihood for potential junctional kyphosis.

There has been limited discussion of pediatric PSO in the literature,\textsuperscript{1,6} presumably because surgery for deformity correction is typically deferred until after adolescence to allow for adequate bone growth and maturation. Unfortunately, our patient’s neurological deficits and progressive respiratory difficulty did not allow for continued nonsurgical management. While numerous studies have examined adult outcomes following PSO, only Bakaloudis et al.\textsuperscript{1} has reviewed single-level thoracic PSO outcomes in pediatric deformity, and their study showed generally favorable results. Academic discussion may also be limited by the relative rarity with which complex spinal corrective surgery is performed in the pediatric population. Pedicle subtraction osteotomy is a technically demanding procedure and may result in complications such as neurological deficits, deep wound infection, and significant intraoperative blood loss, although a posterior approach does entail less risk of damage to major blood vessels than an anterior approach.\textsuperscript{7} At one point during our procedure, neuromonitoring signals were diminished while the spine was cantilevered but returned with lessening of the correction. Cheh et al.\textsuperscript{4} reviewed this phenomenon, proposed etiologies for signal alteration including vascular insufficiency and/or overcorrection of the deformity, and developed an algorithm to address this event intraoperatively. Our patient did not suffer any neurological deficits attributed to temporary neuromonitoring signal change and had no postoperative complications.

Conclusions

Pediatric spinal deformity correction remains a complex and challenging neurosurgical discipline. The ability to correct thoracic spinal kyphosis regardless of its etiology allows surgeons to help patients avoid potentially debilitating neurological and/or orthopedic conditions and, hopefully, to achieve a level of functional independence. Rigorous preoperative evaluation, surgical planning, and intraoperative neuromonitoring are essential to promote favorable outcomes. Continued progress in surgical instrumentation and technique will likely increase the utility of PSO and afford pediatric patients suffering from severe spinal deformity an opportunity for a more normal childhood.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kelley. Acquisition of data: Kelley, Johnson. Analysis and interpretation of data: Kelley, Johnson, Vortmeyer. Drafting the article: Kelley. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Kelley.

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