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L5 Corpectomy in a 12-Year-Old Owing to Rosai-Dorfman Disease: A Case Report

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ABSTRACT

Background: Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis typically presenting with massive, painless lymphadenopathy or subcutaneous masses. Extranodal involvement, particularly in the spine, is exceedingly rare and poses significant diagnostic and management challenges. Lumbar spinal RDD with vertebral collapse and neurological compromise is almost unheard of in the pediatric population, with no prior reports of surgical intervention requiring L5 corpectomy and circumferential fusion in an adolescent.

Case Description: We present a case of a 12-year-old girl with a 6-month history of worsening lower back pain unresponsive to conservative treatment. Imaging revealed collapse of the L5 vertebral body and a destructive, enhancing lesion encasing the neural elements, leading to severe canal and foraminal stenosis. Computed tomography-guided biopsy confirmed the diagnosis of RDD. A multidisciplinary surgical team implemented a 2-stage circumferential surgical plan. Stage 1 involved posterior decompression and posterior instrumentation from L3 to the pelvis using S2AI screws. A silastic barrier was placed to protect neural elements. Stage 2, performed via an anterior approach with vascular surgery assistance, involved complete corpectomy of L5 and reconstruction with an expandable cage spanning L4 to S1. Postoperative recovery was uneventful, and the patient remained neurologically intact. At 1-year follow-up, the patient demonstrated solid anterior and posterior fusion, maintained spinal alignment, and had returned to full activity.

Conclusion: This is the first reported case of pediatric RDD requiring an L5 corpectomy with circumferential spinal fusion. The case underscores the importance of early tissue diagnosis, individualized surgical planning, and interdisciplinary collaboration. Recognition of RDD as a potential etiology for pediatric spinal lesions is critical, particularly when imaging mimics malignancy. Our successful outcome supports the use of staged, tailored surgical approaches in managing rare spinal pathologies in children.

Case Report

Keywords: Rosai-Dorfman disease, L5 corpectomy, lumbar spine, pediatric spinal lesion

INTRODUCTION

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare, non-Langerhans cell histiocytic disorder characterized by the proliferation of histiocytes, specifically, sinusoidal macrophages, within lymph nodes and extranodal sites.[1] The exact cause remains unknown, although it is associated with viral infections (Epstein-Barr virus) and genetic mutations (*NRAS* mutations).[2,3] It has a demographic predilection for African American men younger than 20 years and presenting as a painless subcutaneous lesion.[4] Although RDD most commonly presents as painless subcutaneous masses, bone involvement, especially within the tibia and cranium, has been reported.[5] Involvement of the spine has also been reported, most commonly in the cervical and thoracic spine.[6]

With bone involvement, accurate diagnosis can be challenging, as radiographic findings vary and can mimic multiple disease processes.[7] Diagnosis is mainly histological, with the presence of Rosai-Dorfman cells, which demonstrate emperipolesis.[8] Although RDD most times has a self-limited course, in many cases, occasionally extranodal lesions can be rapidly progressive and lead to a poor prognosis.[6,9] In the present article, we present a case report of an atypical presentation of RDD in the lumbar spine, its treatment, and clinical outcomes.

CASE PRESENTATION

A 12-year-old girl presented with a 6-month history of lower back pain. Treatments included topical heating pads, physical therapy, and over-the-counter medications; however, the patient continued to have intractable pain. There was no history of lower extremity numbness, dysesthesias, bladder dysfunction, bowel incontinence, or shortness of breath. Initial laboratory workup for antinuclear antibodies, rheumatoid factor, and anticitrullinated peptide antibodies was negative.

Box 1. Case Highlights.

Key findings

- This is the first reported case of Rosai-Dorfman disease (RDD) requiring L5 corpectomy and circumferential spinal fusion in a pediatric patient. The patient presented with isolated lumbar spine involvement, vertebral body collapse, and severe spinal canal compromise. A 2-stage circumferential surgical approach allowed for gross total resection, decompression, and reconstruction, resulting in full neurological recovery and radiographic fusion at 1 year postoperatively.

What is known and what is new?

- RDD is a rare, non-Langerhans histiocytic disorder that typically presents with painless lymphadenopathy or subcutaneous masses. Spinal involvement is exceedingly rare, especially in the lumbar spine, and can mimic infections, malignancy, or inflammatory lesions on imaging.
- This case adds to the limited body of literature by detailing a successful surgical strategy in a pediatric patient with lumbar RDD and vertebral collapse. It underscores the importance of histological diagnosis, multidisciplinary surgical planning, and individualized treatment to achieve excellent clinical and radiographic outcomes.

What is the implication, and what should change now?

- Clinicians should maintain a high index of suspicion for RDD in pediatric patients presenting with destructive spinal lesions, even in the absence of systemic symptoms. Early biopsy and definitive histopathological diagnosis are critical for timely intervention. This case supports the use of personalized, staged surgical approaches in rare spinal tumors and highlights the role of 3D modeling and interdisciplinary collaboration in achieving optimal outcomes.

Serologically, there was no evidence of cytomegalovirus, human immunodeficiency virus, hepatitis B or C, *Borrelia burgdorferi*, or toxoplasmosis. Imaging, consisting of standing posteroanterior and lateral spine radiographs, magnetic resonance imaging, and computed tomography (CT) of the lumbosacral region, was obtained during the evaluation and is documented in Figure 1. After obtaining a positron emission tomography scan (Figure 2), the patient underwent a CT-guided biopsy of the lesion, as seen in Figure 3. A positron emission tomography scan was performed as part of the staging evaluation to assess for multifocal or systemic disease involvement. The study demonstrated intense hypermetabolic activity localized to the L5 lesion without evidence of additional metabolically active sites. The lesion was reported as a 6.8 × 5.7 × 1.4 cm aggregate of multiple tan-pink to tan-brown soft tissue and bone fragments. The diagnosis was RDD.

Surgical Plan

The surgical plan was developed jointly between the pediatric spine and oncology spine services. A 3-dimensional model of the lumbosacral spine (seen in Figure 4) was built for resection planning and intraoperative guidance. An en bloc resection was not technically feasible in this case because of the total collapse of the L5 vertebral body and the lack of encapsulation of the mass. A 2-stage plan was developed to allow for restoration of sagittal alignment, stabilization of the lumbosacral spine, safe decompression of neural elements, and complete mass resection.

Stage 1: The patient was positioned prone on a Jackson frame, and a posterior midline approach was completed, exposing L3 to the sacrum. Pedicle screw fixation was completed bilaterally at L3, L4, S1, and the ilium using the S2AI trajectory. The dorsal elements of L5 were resected from the L4 to L5 facet joints to the L5 to S1 facet joints along with any posterior RDD tumor. Resection was continued anteriorly, protecting and decompressing the L5 and S1 nerve roots. The anterior tumor was easily separated from the thecal sac and dura. Decompression was advanced laterally until the nerve roots were fully decompressed into the soft tissues. A silastic sheet was placed anterior to the thecal sac and L5 nerve roots, but dorsal to the residual vertebral body and tumor to protect the neural elements during the stage 2 anterior approach and resection. Decortication was carried out from L3 to S1, and rods and structural allograft were placed posteriorly along with demineralized bone putty and allograft.

Stage 2: Two days later, with the assistance of the vascular surgery team, an anterior approach was completed to the L5 body, allowing visualization from the inferior endplate of L4 to the superior endplate of S1, inferior to the bifurcation of the aorta. Discectomies were performed at L4-5 and L5-S1. The residual L5 body and tumor were resected, and the silastic sheet was visualized and removed. An expandable cage was placed from L4 to S1. Fluoroscopy confirmed appropriate cage placement and sagittal alignment from L4 to S1 (Figure 4C and D). Plain radiographs in the AP and lateral projections demonstrated achievement of adequate local and global alignment.

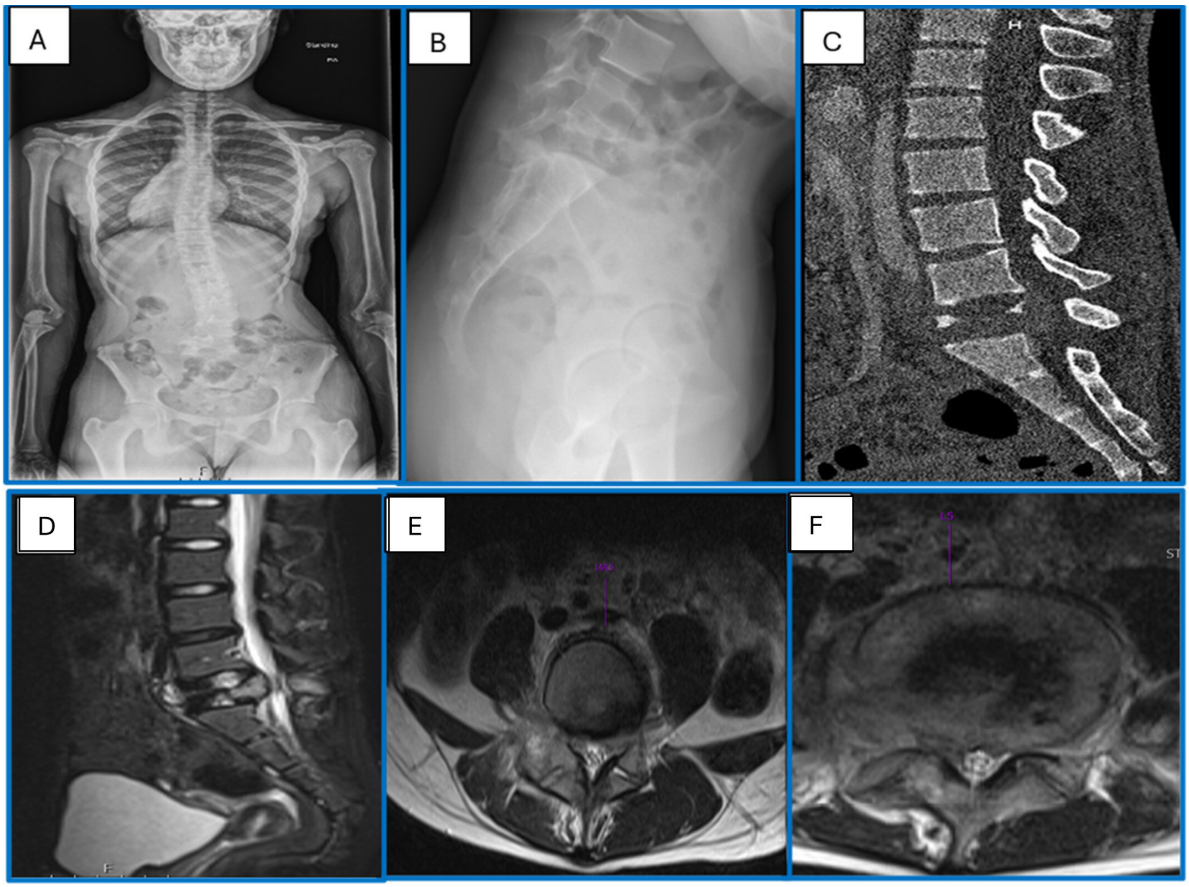

Figure 1. Standing scoliosis posteroanterior (A) and lateral radiographs demonstrate a pathological burst fracture (B) of L5 with height loss and focal kyphosis. Lumbar computed tomography scan (C) is shown. Lumbar magnetic resonance imaging (MRI) with and without contrast (D). There is a marrow replacing, enhancing soft tissue lesion of the L5 vertebral body resulting in severe compression and retropulsion into the spinal canal, causing severe spinal canal stenosis. Enhancement extends to involve the left pedicle and posterior elements as well as the right superior articular facet. Axial cut of a T2 MRI without contrast at the L4–L5 level (E) demonstrates an enhancing lesion that encased both L5 pedicles, more so the right pedicle. Axial L5 MRI (F) demonstrates the lesion encasing both exiting nerve roots and its extension into the extraforaminal space, resulting in severe bilateral neuroforaminal stenosis.

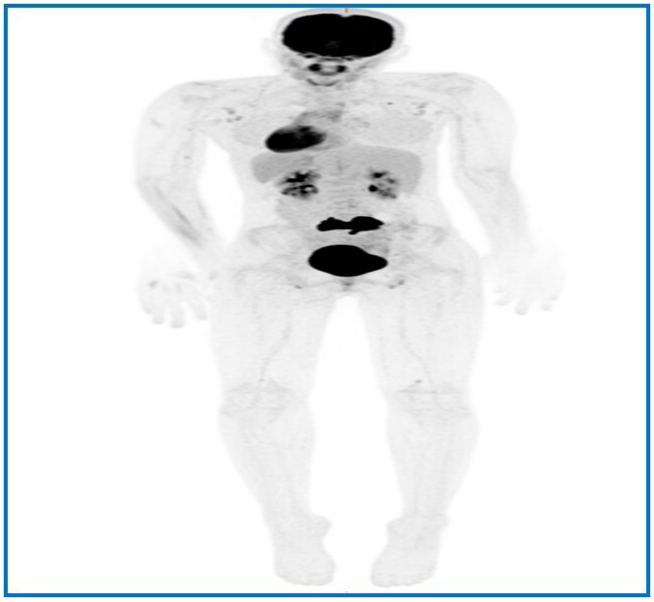

Figure 2. Positron emission tomography image displays intensely hypermetabolic soft tissue around the L5 burst fracture extending into L5–S1 neural foramina in keeping with the known diagnosis of Rosai-Dorfman disease.

Postoperatively, the patient remained neurologically intact, with motor and sensory function throughout L2 to S1 bilaterally. No new neurological deficits were observed. The patient was discharged 2 days after the second stage without any neurological deficits and was able to ambulate without assistance. At 3-month follow-up, she was able to participate in physiotherapy without bending or twisting. At ten months post-operatively, radiographs standing radiographs demonstrated restoration of alignment (Figure 5). At the 1-year follow-up, the patient was doing well without pain and resumed normal activities. A CT scan (Figure 3) demonstrated anatomic sagittal and coronal alignment and solid fusion anteriorly and posteriorly. The patient was placed on a 2-year course of trametinib, a MEK inhibitor used as targeted therapy in select histiocytic disorders.

DISCUSSION

RDD is an uncommon histiocytic disorder with a variable clinical presentation and a largely

self-limiting course.[10] While nodal involvement is most typical, extranodal manifestations, particularly in the spine, are exceedingly rare, accounting for fewer than 1% of RDD cases.[6] Spinal involvement presents a unique diagnostic and therapeutic challenge due to nonspecific clinical signs and the potential for neurological compromise. This case is of particular interest as it represents, to our knowledge, the first reported instance of RDD necessitating an L5 corpectomy and circumferential spinal fusion in a pediatric patient. The complex surgical management, multidisciplinary coordination, and favorable clinical outcome highlight the importance of early recognition and a tailored patient-specific approach in treating rare spinal pathologies like RDD.

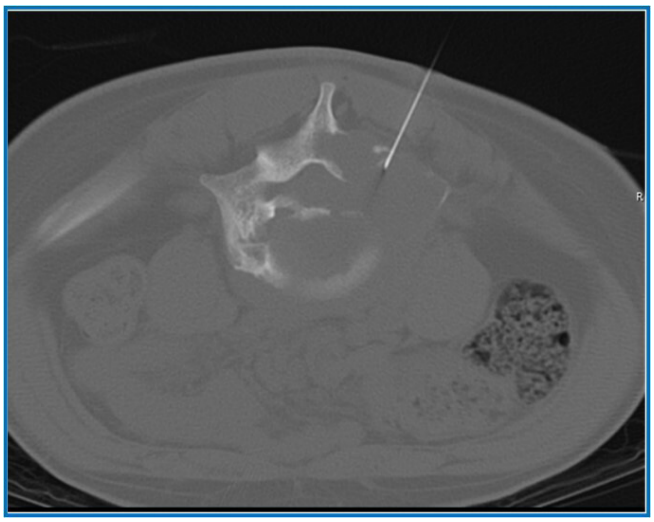

Figure 3. Interventional radiology computed tomography–guided deep bone biopsy demonstrates L5 bone lesion. Core specimens were then sent to surgical pathology and microbiology.

The differential diagnosis of a bone lesion in the spine of a pediatric patient includes various conditions, such as Langerhans cell histiocytosis, osteomyelitis, neoplasms (eg, Ewing sarcoma, osteosarcoma, etc), and infection (eg, tuberculosis, osteomyelitis). These entities often share overlapping clinical and radiological features, complicating diagnosis. In this case, CT-guided biopsy provided an early, definitive diagnosis. Histopathological evaluation revealed proliferative histiocytic disease with positive immunostaining for CD68, CD163, OCT2, S100, and cyclin D1/BCL1, consistent with RDD.[8]

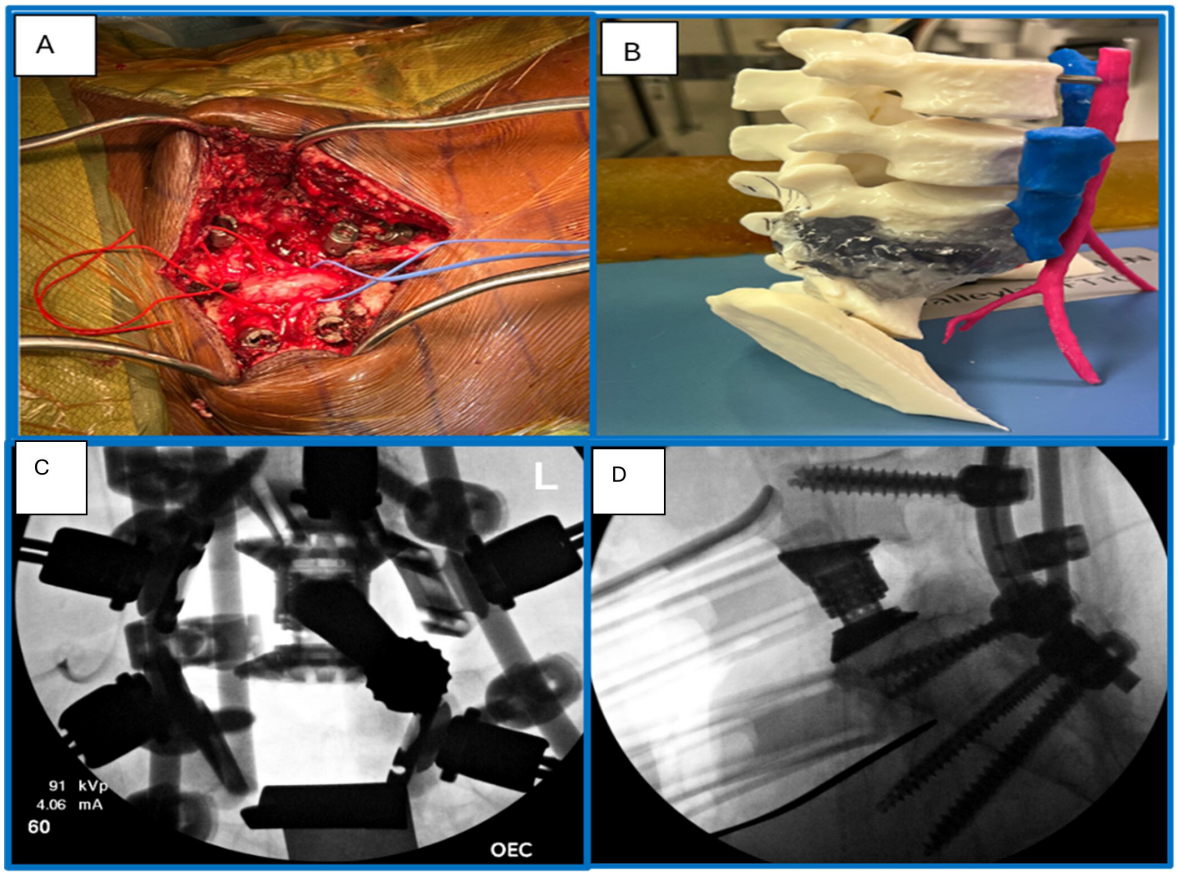

Figure 4. Intraoperative images of the posterior decompression and silastic sheath placement (A); 3D model of bone lesion (B); anteroposterior fluoroscopic view (C); and lateral fluoroscopic view (D) of expandable cage and posterior segmental instrumentation.

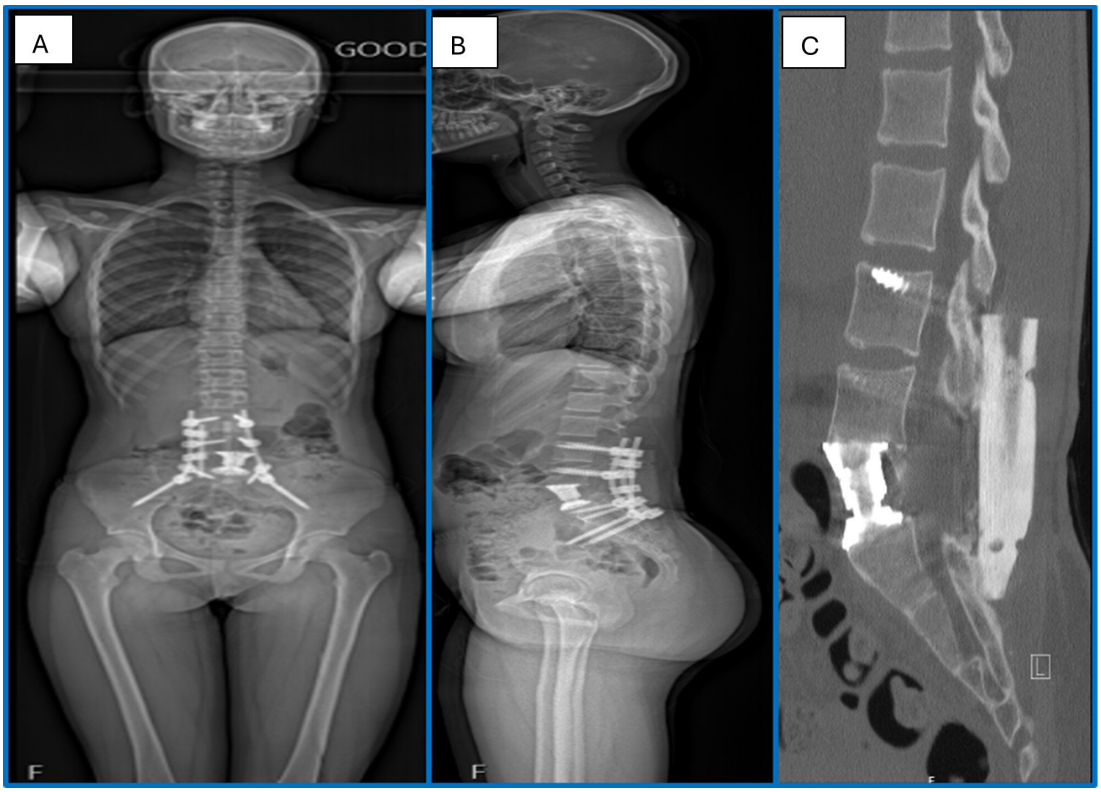

Figure 5. Ten months post-operative standing scoliosis x-ray images demonstrating restoration of alignment both coronally (A) and sagittally (B). Sagittal cuts of the computed tomography scan (C) demonstrate a solid fusion mass along the inside of the corpectomy cage and posteriorly over the strut allograft.

RDD generally has a benign course, most commonly presenting as painless lymphadenopathy or subcutaneous masses. Though not observed in our patient, systemic symptoms like fever, weight loss, and elevated inflammatory markers may occur.[10] Among extranodal sites, bone is a rare location, with spinal involvement being especially uncommon.[5,6] Awareness of its histological signature and atypical clinical behavior is essential for timely diagnosis and intervention.

Management strategies for RDD depend on disease location and severity. In 50% of cases, no medical treatment is needed as 50% of them resolve on their own, and of those treated conservatively, 82% do not relapse.[11] In the case of subcutaneous nodules, the most common therapeutic modality is surgical excision due to the low relapse rate. Immunosuppressive and chemotherapy agents such as methotrexate and 6-mercaptopurine have also been explored with modest effects.[12] Although systemic corticosteroids have been used with variable success, their benefits may be short-lived and are commonly used to treat fever and lymphadenopathy. While radiation therapy has been used in select cases, its efficacy remains limited, commonly resulting in persistent disease.[12]

Vertebral involvement with extradural extension, as seen in this case, is rare and sparsely documented in the literature.[4,6] Management of spinal RDD typically requires a multidisciplinary approach involving surgery, oncology, and radiology. Given the patient's progressive symptoms, extensive vertebral collapse, spinal canal compromise, and risk of neurological deterioration, surgical excision was deemed the optimal treatment. Gross total resection was performed in a 2-stage procedure with the assistance of a vascular surgery team. A 3D model of the lesion (Figure 4B) was used preoperatively to enhance surgical planning and improve intraoperative accuracy. The development of an individualized treatment strategy allowed safe tumor resection, decompression of neural elements, spinal stabilization, restoration of sagittal alignment, and ultimately, a favorable clinical outcome at 1 year postoperatively.

CONCLUSION

To the best of our knowledge, this is the first reported case of an adolescent patient with RDD with extensive vertebral column involvement at L5 with spinal canal compromise. Development of a detailed, personalized

staged surgical plan permitted gross total resection of the lesion, re-establishment of local, regional, and global spinal alignment, circumferential fusion, and no evidence of local or distant recurrence of the tumor.

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