Rotational dislocation of the thoracolumbar spine

Case report and review of the literature.

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✓ The authors report a rare case of rotational dislocation of the thoracolumbar spine in a 12-year-old girl with neurofibromatosis Type 1. The patient had progressive spinal kyphoscoliosis with acute-onset paraplegia. She was treated with corrective traction preoperatively, followed by spinal decompression and circumferential spinal fusion without instrumentation. She had complete neurological recovery after a solid fusion of her spine.

KEY WORDS • kyphoscoliosis • neurofibromatosis Type 1 • spinal dislocation • spinal fusion • traction

Rotational dislocation of the spine is a rare disorder first described by Duval-Beaupere and Dubousset in 1972.¹ The common site of dislocation occurs at the thoracic spine, and secondarily at the thoracolumbar junction. The etiology of rotational dislocation includes NF1, diastrophic dysplasia, congenital myopathy, metaphyseal chondrodysplasia, Gorham disease, congenital defects such as butterfly vertebrae, and others.²,³ The clinical features of this disorder include progressive spinal deformity with or without neurological deficit. Deformity as seen on radiological images is characterized by short, sharp angled kyphosis at the junction of two lordoscoliotic curves.¹ The diagnosis of rotational dislocation is confirmed by noting the apex of the kyphosis on a lateral radiograph, which should correspond with the junction of two scoliotic curves on a frontal radiograph. Dubousset recommends initial partial correction of the deformity before surgery using controlled traction under close supervision, followed by early circumferential fusion using anterior strut grafting and posterior fusion with instrumentation.¹ We report a case of rotational dislocation of the thoracolumbar spine with progressive neurological deficit that required surgical correction with fusion resulting in a successful outcome.

Abbreviations used in this paper: CT = computed tomography; NF1 = neurofibromatosis Type 1.

Case Report

This 12-year-old girl with a history of NF1 presented with spinal deformity, back pain, and lower-limb weakness. Although the spinal deformity was present in the patient for many years, progressive lower-limb weakness had been present only for 1 year. She complained of back pain and worsening neurological deficit in the 2 months before presentation. At presentation the patient was unable to walk and was wheelchair bound. On examination, she had café-au-lait spots on her trunk and arms. The patient had Medical Research Council Grade 3 strength in all muscles of the right lower limb and Grade 4 strength in all muscles of the left lower limb, with classical upper motor neuron signs. Both upper limbs were neurologically intact. There was no sensory deficit and anal tone was normal. Routine blood tests were normal. Her anteroposterior radiograph showed right-sided scoliosis of 48° at T8–11 and left-sided scoliosis of 54° at T12–L3. Lateral radiographs showed kyphosis of 82° at T9–L1 (Fig. 1 left). Results from an oblique radiograph suggested a significant rotation and displacement of the T11–12 vertebrae (Fig. 1 right). Electrodiagnostic studies revealed no peripheral neuropathy, myopathy, or root lesion. A plain myelogram showed a complete block at T11–12. A myelographic CT scan revealed the characteristic double-vertebrae sign and cord compression at T11–L1 (Fig. 2). Magnetic resonance imaging findings were unequivocal with no intracanal mass.
The patient was initially placed in halo bilateral femoral traction in the supine position, and traction was gradually increased under close supervision. The final maximum traction weights used were 12 kg on the head and 6 kg on each femur. After approximately 3 weeks of traction, her neurological status improved resulting in increased muscle power and decreased clonus in both lower limbs. Radiographs obtained during traction showed improvement of kyphosis to 45°, right-sided thoracic scoliosis to 38°, and left-sided thoracolumbar scoliosis to 45° (Fig. 3).

After a total of 4 weeks in traction, the patient was scheduled for anterior and posterior spinal fusion. The patient was placed in the lateral decubitus position with a removable bolster at the apex of the curve. A lower thoracotomy was performed through the concavity of the inferior scoliotic curve. After careful dissection, the apex was identified at the T11–12 junction. All the tethering segmental vessels were ligated and divided. An autogenous fibular graft was harvested from the left leg and was cut into three struts measuring 7, 5, and 3 cm. These strut grafts were inserted in a palisade fashion starting with the spanning of a short gap at the level of the kyphotic apex. The bolster positioned under the apex was removed in order to lock the grafts into position. The space between the fibular grafts was then filled with rib grafts. Posterior decompression was followed by posterior fusion. Harvested autologous iliac crest grafts were placed on both sides of the spinous process after decorticates the facets and the transverse processes. Posterior instrumentation could not be placed because of poor bone quality, thinned pedicular diameter, and dystrophic posterior elements. Therefore, the patient underwent anterior spinal fusion and posterior in situ fusion followed by the placement of a halo pelvic cast. A plaster jacket was applied extending down to the pelvis, leaving the abdominal area free. The halo was connected to the plaster jacket by incorporating four vertical rods into the plaster. At the 3-month follow up, the patient was walking independently with crutches and had normal power in both lower limbs. The halo plaster cast was worn for a total of 12 months.

Although final x-ray images (Fig. 4) obtained 3 years postoperatively showed some loss of the initial correction (kyphosis to 50°, right-sided scoliosis to 40°, and left-
sided scoliosis to 50˚), the patient had complete neurological recovery and no back pain (Table 1).

**Discussion**

Spinal deformity in NF1 has dystrophic features. Rotational dislocation could occur due to deficient posterior elements in the spine. The deformity progresses slowly, leading to a rotation at the junction of two curves. There is a high risk of spontaneous dislocation in patients with NF1 and dural ectasia.

Progression of spinal deformity may lead to neurological deficit. The natural history of this type of deformity is relentless progression of kyphosis associated with myelopathy. Minor trauma at this stage may lead to complete paraplegia with or without a fracture. Thus, an expedited evaluation at presentation followed by surgical correction is required to prevent the consequences of progressive spinal deformity.

Use of plain radiography is the gold standard for assisting in the diagnosis of rotational dislocation of the spine. Rotational dislocation differs from congenital dislocation, in which one could find a definitive indicator of the dislocation. A definitive indicator is evident in the cervical spine in NF1 due to defects in the posterior elements, but may be difficult to recognize in the thoracolumbar spine.

To understand the deformity, further diagnostic tests such as CT or magnetic resonance imaging are required before making the decision to use surgical correction. The double-vertebrae sign noted on CT scans is a hallmark of rotational dislocation of the spine, as evident in this case.

An early age of onset, a high Cobb angle, and scoliotic curves with more than 11˚ apical vertebral rotation with

![Fig. 3. Left: Anteroposterior radiograph obtained 4 weeks after traction showing partial correction of the rotational deformity. The right-sided scoliotic curve angle was 38˚; the left-sided curve angle was 45˚. Right: Lateral radiograph demonstrating improvement in the kyphotic angle to 45˚.](image1)

![Fig. 4. Left: Anteroposterior radiograph obtained 3 years after surgical correction showing solid fusion of the spine. The right-sided scoliotic curve angle was 40˚ and the left-sided was 50˚. Right: Oblique radiograph obtained 3 years after spinal fusion.](image2)
The specific risk factors for the progression of deformity in NF1. The specific risk factors for rotational dislocation are not noted in the literature. Dias et al. postulated that embryonic buckling at the 4th and 6th embryonic weeks—producing segmental translation at a single level—results in congenital dislocation of the spine. We hypothesize that in NF1 the developmental defects in the posterior elements of the spine, along with abnormal shapes of the vertebrae, may create an imbalance in which the rotation may take place along an axis. The center of rotation of this axis may lie in the vertebral body cranial to the apex. The cranial lordoscoliotic curve may displace on the caudal curve at the apex as the child grows. The increasing forces that develop across the apex may further displace and rotate the vertebrae, causing progressive neurological deficit.

It is recommended that early circumferential arthrodesis is performed to achieve a stable spine. Anterior strut grafting from the concavity is recommended so that the graft undergoes compressive forces. Arthrodesis from the convex side is not recommended because it decreases the structural resistance of the spine. Decompression of the spinal cord is indicated for patients who have a neurological deficit of recent onset or a progressive neurological deficit. When correcting the deformity through a combined approach in the present case, we performed decompression of the spine and an anterior strut grafting from the concave side. An association of decreased bone mineral density with NF1 is well recognized. The pathogenesis of the osseous manifestations in NF1 may involve impaired development of the skeletal system and impaired maintenance of bone structure. Posterior instrumentation could not be placed in the present case due to the presence of structural weakness in the bone. The soft bone of the dystrophic vertebrae often undermine the placement of implants in NF1.

Conclusions

Rotational dislocation of the spine is a rare pathology, which may cause progressive neurological deficit in children with NF1. This pathology has a potential risk of severe kyphoscoliosis, which requires early identification, urgent surgical correction, and circumferential fusion. The present case is unique because after spinal decompression and fusion, we could not perform any instrumentation due to deficient posterior spinal elements and poor bone quality. Nevertheless, this patient underwent a successful fusion with a complete recovery of her neurological deficit.

References


TABLE 1
Pretraction, posttraction, and follow-up values of the magnitude of the radiological curve angle*  
<table>
<thead>
<tr>
<th>Stage</th>
<th>Scoliotic Angle (L/R) (˚)</th>
<th>Kyphotic Angle (˚)</th>
</tr>
</thead>
<tbody>
<tr>
<td>pretraction</td>
<td>54/48</td>
<td>82</td>
</tr>
<tr>
<td>posttraction (4 wks)</td>
<td>45/38</td>
<td>45</td>
</tr>
<tr>
<td>FU (3 yrs)</td>
<td>50/40</td>
<td>50</td>
</tr>
</tbody>
</table>

* FU = follow up.