Successfully Treated Traumatic Dislocation of a Thoracic Vertebra Caused by Minor Trauma in a Patient with Neurofibromatosis Type I: A Case Report and Literature Review

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Introduction

Although a high incidence of spinal deformity is reported in patients with neurofibromatosis type I (NF-I),¹⁻³ the true prevalence of spinal deformity remains unknown. Spinal decompensation in neurofibromatosis is generally classified into non-dystrophic and dystrophic types based on the absence or presence of skeletal deformities during plain radiographic evaluation.¹⁻³ Dystrophic changes may cause destabilization of the vertebrae, occasionally leading to spontaneous subluxation or dislocation⁴⁻⁶ and indicating vulnerability to extrinsic forces, subsequently leading to spinal cord injury.¹⁻³ However, a small number of spinal cord injuries has been reported to be expected even for patients with NF-1 in severe neurological condition after acute spinal cord injuries.

Keywords: Neurofibromatosis type I, dystrophic type, spinal cord injury, minor trauma

Case Report

A 29-year-old woman slipped on the floor of her office and sustained minor injuries to her buttocks. Immediately after the injury, she complained of low back pain, motor weakness, and numbness in her lower limbs. She was admitted to the hospital where she worked at. Her past medical history was not contributory, but her mother was confirmed to have NF-1. She denied all CT or MRI examinations in the past. On physical examination, many “café-au-lait” spots and subcutaneous masses were discovered throughout her trunk. She was confirmed to have NF-1. Urinary incontinence was observed. All of her vital signs were normal. Manual muscle testing revealed that she scored no trace in her leg muscles, and normal in her upper limbs. She showed incomplete sensory loss below her lower belly with sparing sacral segments. These were consistent with the American Spinal Injury Association Impairment Scale (AIS) B.¹² Routine blood tests were normal. Emergency neuro-radiological workup was performed. A CT scan showed a significant vertebral dislocation at T11/T12 with no dislocation of bilateral facet joints, and scalloping of the vertebral body margins from T9 to L1, as well as widening of the spinal canal (Figs. 1A–1C). MRI revealed a spinal medullary with high intensity associated with the vertebral dislocation, including prevertebral hemorrhage and a narrowing of the spinal canal (Figs. 1D and 1E). A T2-weighted coronal image clearly visualized dural ectasia from T9 to L1, as well as level and lateral meningocele, indicating a considerable enlargement of the neural foramina (Fig. 1F). Each pedicle was shown as very thin, bony structures and no pedicle was visualized from T7 to L1 on the right (Fig. 2). Vertebral body fractures, spindling of the transverse process, rib penciling and rib head dislocations were not present. There was no tumor in and around the vertebral column. It was noted that there was no scoliosis or kyphosis.

In order to reposition the dislocated vertebral body and stabilize the spinal column, an emergency operation was undertaken. But after consideration of the risks and benefits, a posterior spinal fusion surgery was carried out instead 40 hours after the injury. After the introduction of general anesthesia, transcranial motor-evoked potentials and somatosensory-evoked potentials could not be detected and so could not be used to measure damage during the surgery. So, the patient was meticulously placed in a prone position. A posterior spinal fusion surgery was performed for this case by using conventional rods and screws, as pedicle screw placement.

The authors reported a rare case of young women with neurofibromatosis type I (NF-I) who were successfully treated from the traumatic dislocation of a thoracic vertebra caused by a simple fall, and the relevant literature was reviewed. Due to various spinal dystrophic changes, the conventional posterior spinal fusion surgery was modified for the treatment. Spinal deformity is a common feature of NF-1, and a dystrophic lesion, like dural ectasia, provokes weakness in spinal structural. Unexpectedly, only seven similar cases were found. The review suggested that it is mandatory to thoroughly examine the spine in patients with NF-1, and that a good outcome can be expected even for patients with NF-1 in severe neurological condition after acute spinal cord injuries.
was impossible for these pedicle abnormalities, and the bone was grafted. Through a posterior midline approach from T6 down to L4, conventional laminectomy was performed on T11 and T12, taking great care not to apply unnecessary force. The screws (CD HORIZON SOLERA Spinal System, Medtronic, Memphis, TN, USA) were placed into the left L2 pedicle as well as both sides of the L3 and L4 pedicles. Since the pedicles above L1 appeared inappropriate for pedicle screw placement due to their brittleness or disappearance, hook systems (CD HORIZON SOLERA Spinal System, Medtronic) were applied to T6 through T9. Two metal rods 6 mm in diameter were used for rigid stabilization. Fortunately, reduction could be achieved using a rod-compression maneuver among the anchors under intraoperative radiographic control. Sublaminar tape (ultra-high molecular weight polyethylene tape, Nesplon, Alfresa, Inc., Osaka) was utilized for reinforcing rigidness (Figs. 3A and 3B). After decortication of the posterior bony element of the spine, plentiful bone
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A graft substitute from the iliac crest was added to stimulate fusion.

Postoperative course was uneventful (Figs. 3C and 3D). A postoperative CT scan showed successful correction of the spinal dislocation (Fig. 3E). She wore a rigid spinal brace for 8 months. In the following week, signs of recovery in her lower limb movement were found. Her rehabilitation program at the gym was started 4 weeks after the surgery. Fortunately, 3 months later, she became to stand with the aid of a rehabilitation walker. Her neurological condition improved day by day. At present, she can walk without any aid, and she has recovered completely from sensory and urinary disturbances. A CT scan was taken 1 year after the surgery showed no worsening in spinal alignment or corresponding bony structures (Fig. 3F).

Discussion

NF-1 is an autosomal, dominant, inherited genetic disorder with a multitude of manifestations potentially involving any organ system affecting approximately 1 in 4000 persons.\(^1\)\(^{-}\)\(^3\) Classically, it is manifested as abnormalities of the skin, nervous system, bone, and soft tissues.\(^1\)\(^{-}\)\(^3\) The skeletal system is the most frequently involved mesodermal derivative. Spinal deformity is the most commonly observed, but its incidence was reportedly ranged from 2% to 69% of NF-1 patients.\(^1\)\(^{-}\)\(^3\) Akbarnia et al.\(^1\) believe that 10% is representative of the true prevalence of spinal deformity. Recent studies using MRI suggested a much higher incidence. Spinal deformities are usually classified into non-dystrophic and dystrophic types based on the absence or presence of skeletal deformities on plain radiographic evaluation.\(^1\)\(^{-}\)\(^3\) Dystrophic features include vertebral scalloping (posterior, lateral, or anterior), rib penciling or spindling of the transverse processes, wedging of one or more vertebral bodies, a short curve with significant apical rotation, foraminal enlargement, defective pedicles, and associated paraspinal masses.\(^1\)\(^{-}\)\(^5\)\(^{-}\)\(^10\) These changes often occur in sectioned vertebral columns.\(^7\)\(^{-}\)\(^13\) Introduction of the recent sophisticated neuro-imaging studies has pointed out that patients with the radiographically labeled non-dystrophic type were found to have significant dystrophic changes. Dystrophic changes are thought to be either intrinsic in origin or associated with intraspinal anomalies, namely abnormalities of the dura mater, such as dural ectasia, or dumbbell neurofibromas extending through the intervertebral foramina and causing foraminal enlargement. These dystrophic changes may be progressive.\(^2\)\(^{-}\)\(^3\) The patient presented here was thought to have several dystrophic changes associated with dural ectasia that was mostly localized to the lower thoracic area. We should know that, no matter what the classification is, NF-1 may cause destabilization of the vertebrae, occasionally leading to spontaneous subluxation or dislocation\(^5\)\(^{-}\)\(^9\) and indicating vulnerability for minor extrinsic forces, subsequently leading to spinal cord injury.

Reported cases of NF-1 with spinal cord injury caused by minor trauma are summarized in Table 1.\(^1\)\(^{,}\)\(^6\)\(^{-}\)\(^10\)\(^11\) Despite a meticulous review of the literature, only seven cases, including the present case, were found. Considering the expected incidence of NF-1 with spinal deformity, this small
### Table 1  Summary of all reported cases of neurofibromatosis type I with spinal cord injury caused by minor trauma

<table>
<thead>
<tr>
<th>Author</th>
<th>Age/Sex</th>
<th>Examination of the spine</th>
<th>Mechanism of injury</th>
<th>Location</th>
<th>Symptoms</th>
<th>Dysplastic changes</th>
<th>Findings</th>
<th>Operation</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rockower S, et al.</td>
<td>10/F</td>
<td>-</td>
<td>Fall</td>
<td>T4/5</td>
<td>Back pain</td>
<td>Vertebral scalloping</td>
<td>Subluxation</td>
<td>Anterior and posterior fusion</td>
<td>GR</td>
</tr>
<tr>
<td></td>
<td>10/M</td>
<td>-</td>
<td>Fall</td>
<td>C6/7</td>
<td>Quadripareis</td>
<td>-</td>
<td>Dislocation</td>
<td>-</td>
<td>GR</td>
</tr>
<tr>
<td>Ferner RE, et al</td>
<td>71/M</td>
<td>-</td>
<td>Traffic accident</td>
<td>C1/2</td>
<td>Quadripareis</td>
<td>Dural ectasia</td>
<td>Atlanto-axial subluxation</td>
<td>Transoral decompression and posterior stabilization</td>
<td>D</td>
</tr>
<tr>
<td>Lam KS, et al.</td>
<td>21/F</td>
<td>-</td>
<td>Slip, occiput</td>
<td>C3/4</td>
<td>Neck pain, dysoesthesia, paraesthesia</td>
<td>Vertebral scalloping, dural ectasia, paravertebral soft tissue mass</td>
<td>Subluxation with a 40° kyphosis</td>
<td>Anterior C3/4 disectomy and fusion, C2-6 posterior cervical fusion</td>
<td>GR</td>
</tr>
<tr>
<td></td>
<td>40/M</td>
<td>-</td>
<td>Traffic accident</td>
<td>C1/2, C5/6</td>
<td>Bilateral C5 root dysoesthesia, myelopathic gait</td>
<td>-</td>
<td>Intradural extramedullary mass</td>
<td>Resection, fixation from occiput to T2</td>
<td>GR</td>
</tr>
<tr>
<td>Sakamoto H, et al.</td>
<td>12/M</td>
<td>+</td>
<td>Fall</td>
<td>T7</td>
<td>Paralysis, ischuria</td>
<td>Vertebral scalloping, transverse process spindling, paravertebral soft tissue mass, spinal rotation</td>
<td>40° scoliosis and 76° kyphosis</td>
<td>Resection and posterior fusion from T2 to L3, anterior fusion from T5 to T9</td>
<td>GR</td>
</tr>
<tr>
<td>Our case</td>
<td>29/F</td>
<td>-</td>
<td>Slip</td>
<td>T11/12</td>
<td>Paralysis</td>
<td>Vertebral scalloping, dural ectasia, intervertebral foraminal enlargement, dysplastic pedicles</td>
<td>Dislocation</td>
<td>Decompression and posterior fusion from T6 to L4</td>
<td>GR</td>
</tr>
</tbody>
</table>

**NF-1**: neurofibromatosis type I.

number is said to be incredible. There must be many unreported cases. The recorded patients were generally young, with the exception of one patient, and all had no preexisting symptoms related to NF-1. Consequently, no spinal survey was carried out in these patients. Unrecognized spinal deformities related to NF-1 can have serious consequences. Therefore, it is mandatory to thoroughly examine the spine in patients with NF-1. It was noted that six out of seven patients had a good outcome; only one patient died from complications 2 weeks after the surgery, despite the favorable neurological recovery. The important suggestion obtained from this review appears to be that, even in severe neurological condition after acute spinal cord injuries, a good outcome can be expected in patients with NF-1.

Stabilization of the vertebral column can be easily achieved through posterior fixation. In cases with no severe kyphosis or scoliosis due to NF-1, posterior fixation alone can be attempted.\(^2,8,14\) In progressive deformities, however, anterior fixation is performed additionally, and a combined anterior and posterior fixation should be carried out to ensure solid fusion.\(^2,8,9\) In the present case, there was no severe kyphosis and scoliosis. There were dural ectasia and lateral meningoceles in the surrounding dislocation. The dura in that area is extremely thin and fragile.\(^9,13\) Anterior fixation concerns the inevitable dural injury and uncontrollable leakage of cerebrospinal fluid. Therefore, we decided that only posterior fixation and anterior fixation was not feasible for this case. When a posterior fixation was to be carried out, many authors recommend the use of pedicular screws. However, because of the pedicular shape associated with dural ectasia, the use of hooks and rods for posterior fixation is recommended.\(^14–16\) However, the dural ectasia may cause erosion of the vertebral column and posterior fixation alone may not be mechanically viable and may be subsequently proved inadequate through breakage. Spinal fusion of dystrophic type in NF-1 is fraught with difficulties. Although
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this case is successful deformity correction and fusion, long-term follow-up is required.

Informed Consent
The patient has consented to submission of this case report to the journal.

Conflicts of Interest Disclosure
The authors report no conflict of interest concerning the materials and methods used in this study or the findings specified in this article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

References

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