Spontaneous rotational dislocation of the lumbar spine in type 1 neurofibromatosis
A case report and literature review

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1. Introduction
Neurofibromatosis (NF) is one of the most common autosomal dominant hereditary disorders, and is typically divided into 2 clinical forms.1,2 Type 1 neurofibromatosis (NF-1) is often associated with various musculoskeletal diseases, including spinal deformity, which was the most common skeletal manifestation of NF-1, affecting up to 64% of patients.3

A short-segmented, angulated, and rigid thoracic kyphoscoliosis is the most common spinal deformity observed;2,4 however, a spontaneous rotational dislocation of the lumbar spine in NF-1 is a rare entity.5,6 After carefully searching and reviewing the literature, we found that only 7 cases of thoracic spine had been reported in the past6–11 (Table 1). Most authors had advocated the use of circumferential fusion with anterior strut grafting from the concavity as the mode of treatment of this situation.

Here, we report a 51-year-old lady with spontaneous rotational dislocation of the L2 to L3. To the best of our knowledge, this is the 1st case of a dislocated lumbar spine reported with a solid circumferential fusion completed through posterior-only approach. We discuss the pathologic characteristics of this rare entity, report our experience on the management, and present a literature review.

2. Case description
2.1. Consent
The study was approved by the Medical Ethics Committee of Shandong Provincial Hospital affiliated to Shandong University. Informed consent was obtained in writing from the patient.

2.2. General information
A 51-year-old lady complaining of severe back pain was admitted to our center to be assessed for her disorder on August 7, 2017.
kyphosis of 31° sided scoliosis of 30° of the L2 to L3 with right-sided scoliosis of 40°

The anteroposterior radiograph revealed a rotational dislocation of 2.3°.

No abnormalities of bladder or bowel function were noted.

There was no tumor but a small bony protrusion of L3 vertebral body in the spinal canal without cord compression. A dural ectasia extending from lower thoracic spinal canal to sacral canal was noted (Fig. 2). The diagnosis of neurofibromatosis was ultimately verified by MRI-guided percutaneous biopsy.

### 2.3. Imaging characteristics

The anteroposterior radiograph revealed a rotational dislocation of the L2 to L3 with right-sided scoliosis of 40° at T12-L2 and left-sided scoliosis of 30° at L3-L5. The lateral radiographs showed kyphosis of 31° at L1-L4 (Fig. 1A, B). Axial computed tomography (CT) images revealed extremely thin pedicles and the classic double-vertebrae sign which is one mark of rotational dislocation of the spine (Fig. 1C, D). Reconstrucitive CT scan demonstrated the presence of dysplastic vertebral bodies and posterior elements (Fig. 1E, F). Magnetic resonance imaging (MRI) and enhanced MRI showed a large paravertebral mass on the left side at L2 level. There was no tumor but a small bony protrusion of L3 vertebral body in the spinal canal without cord compression. A dural ectasia extending from lower thoracic spinal canal to sacral canal was noted (Fig. 2). The diagnosis of neurofibromatosis was ultimately verified by MRI-guided percutaneous biopsy.

2.4. Therapeutic procedure

The patient was placed in skull-gravity traction at sitting position after admission, and the traction was increased gradually under close supervision. The initial traction weights were used 2 kg on the skull, which were finally up to 25 kg. During 4 weeks of traction, there were no neurologic deficit or severe complications observed in the patient. Radiographs obtained after traction showed the lumbar spine was slightly distracted and realized partial reduction. The improvement of kyphosis and scoliosis were also revealed in radiographs taken under the traction of maximum weights (Table 2).

Surgery was performed after 4 weeks of traction. Under general anesthesia, the patient was placed in a prone position. Somatosensory evoked potential (SEP) monitoring was used intraoperatively. Paravertebral muscles were detached using a subperiosteal dissection. Deficient posterior elements of spine were noted. Bilateral pedicle screws were inserted in the T12, L3, and L4. Another 2 pedicle screws were inserted in single vertebral body of L1 and L2 respectively because of contralateral defective pedicles. The inferior half of L2 lamina, the inferior articular process of L2, and the superior half of L3 lamina were removed. With the resection of ligament flavum, we exposed the spinal canal and the nerve roots. As much of the paravertebral neurofibromas as possible were removed. Discectomy and debridement of disc space were performed at L2-L3 disc for thorough release. Reduction of the dislocated vertebrae and realignment of the lumbar spine were achieved with the instrumented correction. Interbody fusion with polyetheretherketone (PEEK) cage and adequate autogenous iliac crest grafts were performed between L2 and L3 through posterior approach. Then posterolateral fusion was performed from T12 to L4. Large amount of milled autogenous and allogeneic bones were placed on both sides of the spinous process after decorticating the facets and laminas. Finally, we made artificial allograft bone plate to act as lamina. A cross-linkage was used to enhance the stability. SEP monitoring showed no neurologic complication during the operation.

2.5. Outcome and follow-up

Postoperatively the rehabilitation process was satisfactory and the patient was free of complaints. The VAS for back pain was
2 points. On radiographs the vertebral dislocation was reset and the lumbar lordosis was obtained (Fig. 3). She wore the Boston brace for 6 months. After a 13-month follow-up, she was completely asymptomatic and able to live a normal life. Radiographs and CT scans showed a good correction of the deformity and a solid bony fusion (Fig. 4). Late subsidence of the cage was not observed. No implant failure was found. However, there had been a slight loss of the initial correction. The loss may be due to the progression of neurofibromatosis instead of a nonunion of the fusion mass.

Table 2

| Stage          | Scoliotic angle, ° | Kyphotic angle, ° |
|----------------|--------------------|-------------------|
| Pretraction    | 40/30              | 31                |
| Posttraction   | 29/24              | 21                |
| Postoperation  | 28/18              | −18               |
| LFU (13 mo)    | 30/21              | −16               |

LFU = last follow-up, mo = months.
3. Discussion

According to the absence or presence of bone dystrophy, spinal deformities in NF-1 are generally classified into 2 basic types: nondystrophic and dystrophic. The presence of vertebral scalloping, rib penciling, defective pedicles, enlarged spinal canal, a short and angulated curve with severe vertebral rotation reflects vertebral dysplasia. Nondystrophic curvature has a more benign course, and its methods of management are very similar to idiopathic scoliosis. On the contrary, dystrophic curvature, characterized by pathologic morphologic deterioration, is very difficult to treat due to dysplastic vertebrae and remarkable osteoporosis.

Scoliosis is the most common form of dystrophic curvatures, whereas a spontaneous rotational dislocation of spine is a rather rare one. There is no recognized explanation for the cause or origin of the rotational dislocation, which may occur due to the rotatory instability for developmental defects of the posterior elements. On radiographs the deformity is characterized by short and angulated kyphosis at the junction of 2 lordoscoliotic curves. The apex vertebrae of the kyphosis on the lateral radiograph is in accordance with the junctional vertebrae of 2 scoliosis on the anteroposterior radiograph. The management of this type of deformity often produced disappointing results, because reconstructing a stable spine has practically technical difficulties and is correlated with high rates of implant failure and nonunion.

After carefully searching and reviewing the literature, we found that only 7 cases of surgically treated spontaneous dislocation of thoracic spine had been reported in the past. Most cases were associated with intraspinal pathology such as tumors, meningoceles, or dural ectasia. There were 4 cases existing neurologic deficits as reported. As seen in this case, there was an apparently rotatory glide of L2 to the lower right region of L3 with an extended dural ectasia. The deficient facets and lamina of L1-L3, along with the abnormal shape of L2 vertebral body because of partly compressed medulla, might leave it unstable enough to slowly dislocate during the spinal growth. Factors contributing to the absence of neurologic deficit might include dural ectasia and pathologic enlargement of bony vertebral canal. A small bony protrusion due to the hyperostogeny of L3 vertebral body after the erosive injury intruded into the canal but did not press on the spinal cord or nerve roots.

As for surgical procedure, combined anterior-posterior spinal fusion, which had more acceptable results by adding anterior strut grafts, had been recommended by many authors. Parisini et al reported 56 scoliosis patients with an overall fusion failure of 53% in patients who had posterior fusion alone compared to 23% who had an additional anterior fusion. Early circumbential fusion performed on patients with spontaneous spinal dislocations had also produced excellent outcomes and been considered standard treatment. The anterior strut-grafting from the concavity was recommended so that the graft undergoes compressive forces and conforms to biomechanical principle. With anterior support of the spine and posterior instrumented fusion, the reconstructed spine could achieve stability and solid bony fusion would be generally seen at follow-up. The difficulty of this procedure lies in the high technique requirement of anterior approach and vulnerability of reconstructed anterior column because of poor local bone stock for placing a graft.

In this case, we managed to achieve a good correction and solid circumbential fusion using posterior approach alone. After thorough release by means of posterior column osteotomy and disectomy, instrumented correction with pedicle screws and rods was performed to reduce the rotational dislocation and to restore the alignment in the coronal and sagittal plane. Good linear as well as positional alignment rather than in situ fusion can reduce the need for strut grafting technique. Interbody fusion was performed with the cage implanted from the concavity, where it is under compressive forces as those anterior strut grafts. When posterior corticotomy and posterolateral fusion were done, the circumbential fusion was achieved successfully through posterior-only approach. Although without fibular or costal strut grafts, the intervertebral cage and large amount of bone grafts could act as weight-bearing space fillers or struts. Surgical restoration of lumbar lordosis is significant to alleviate the tension of back muscles and to provide a conducive biomechanical environment for bony fusion to take place. Compared to combined operative approaches, single posterior approach is more well-controlled, less time-consuming, and brings fewer invasions to patients. However, the cross-segment strut-grafting from the concavity could provide better anterior support of the spine than this single cage and was less likely to result in pedicle screws loosening and pullout.

The role of preoperative short-term traction is important in the treatment of this patient: preoperative traction can improve the deformity and enable surgeons to evaluate the tolerability of spinal cord to tension by indirectly stretching the cord when the patient was still awake. In addition, a postoperative external support is essential to further protect the fusion mass and enhance the stability of spine. It is agreed that, compared to idiopathic scoliosis, bracing remains necessary to a dystrophic spine for a longer period. Six months of Boston brace was used in this case.

4. Conclusion

A spontaneous rotational dislocation of the lumbar spine secondary to neurofibromatosis is a rare entity. In view of the lack of clinical signs, one must be particularly aware of patients...
who have neurofibromatosis and unexplained back pain for early identification. Surgical stabilization should be considered urgently when the diagnosis is established by imageology and biopsy. Posterior instrumented reduction along with intervertebral-posteriorlateral fusion is a practical and promising surgical procedure with a good prognosis.

**Author contributions**

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