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Clinical Study

Treatment of dystrophic scoliosis in neurofibromatosis Type 1 with one-stage posterior pedicle screw technique

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Abstract

BACKGROUND CONTEXT: Corrective surgery for dystrophic scoliosis in neurofibromatosis Type 1 (NF-1) is challenging. There are various surgical methods, all with unsatisfactory outcomes.
PURPOSE: The purpose of the study was to evaluate the clinical outcomes of the treatment of dystrophic scoliosis in NF-1 with one-stage posterior pedicle screw approach.
STUDY DESIGN: This is a retrospective clinical study.
PATIENT SAMPLE: Sixteen patients with dystrophic scoliosis in NF-1 underwent one-stage posterior surgery with pedicle screw system.
OUTCOME MEASUREMENT: We used preoperative and postoperative whole-spine radiographs to determine coronal and sagittal Cobb angles (curve correction); distance between apex vertebra and central sacral vertical line (DAC), pelvic obliquity, and shoulder tilt (coronal balance improvement); and sagittal vertical axis and pelvic tilt angle (sagittal balance improvement). We assessed the fusion rate using fusion segment computed tomography scan.
METHODS: Patients underwent surgery with or without osteotomy according to spinal flexibility. Fusion segment selection method of fusion segments selection which mean fusing from one or two levels proximal to upper end vertebra to one or two levels distal to the lower end vertebra (EV+1 or 2) or stable vertebrae fusion. There were no study-specific conflict of interest–associated biases.
RESULTS: The average follow-up time was 40.9 months. Mean scoliosis and kyphosis improved from 83.2° to 27.6° and 58.5° to 26.8°, respectively; at the last follow-up, it was 30.4° and 27.4°, respectively. Mean DAC, pelvic obliquity, and shoulder tilt improved from 53.0 to 23.9, 8.1 to 4.9, and 9.8 to 7.5 mm, respectively. Sagittal vertical axis and pelvic tilt angle improved from −5.8 to 1.6 mm and 17.9° to −5.8°, respectively. During follow-up, mean coronal and sagittal correction losses were 2.8° and 0.7°, respectively. Two EV+1 or 2 patients had decompensation. No pseudoarthrosis was identified.
CONCLUSIONS: The one-stage posterior pedicle screw approach is safe and effective in the treatment of dystrophic scoliosis in NF-1. Posterior vertebral column resection is recommended if flexibility is less than 35%. Stable vertebrae fusing is recommended.

Keywords: Neurofibromatosis; Dystrophic scoliosis; One stage; Posterior instrumentation and fusion; Pedicle screw techniques; Posterior vetebral column resection

FDA device/drug status: Not applicable.

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EVIDENCE & METHODS

Context
Given the rarity of the condition, results following surgical treatment for dystrophic scoliosis in the setting of neurofibromatosis (NF)-I are not widely available in the literature. The authors present their experience with a single-stage posterior technique.

Contribution
This was a retrospective case series limited to 16 patients. Average follow-up exceeded three years post-surgery. Satisfactory outcomes were documented in most patients.

Implications
As a retrospective case series, this work is confounded by selection and indication bias, among others. While demonstrating that, in the authors' hands, the single-stage all-posterior technique is safe and effective for the treatment of dystrophic scoliosis in patients with NF-I, these findings are not necessarily translatable. Results may not be comparable in other surgical settings, among other surgical providers with different levels of experience and in a different set of patients.

—The Editors

Introduction

Neurofibromatosis (NF) is an autosomal dominant hereditary disease characterized by the abnormal proliferation of cells from the neural crest; it can occur in both the peripheral and the central nervous systems. Typically, NF is divided into two clinical types: Type 1 (NF-1), also termed von Recklinghausen disease or peripheral NF, and Type 2 (NF-2). Neurofibromatosis Type 1 is often associated with different orthopedic disorders, especially scoliosis. Scoliosis is the most common skeletal manifestation of NF-1; prevalence is between 10% [1] and 64% [2]. However, NF-1 accounts for just about 3% of all scoliosis cases [3]. Furthermore, scoliosis in NF-1 is usually classified into two basic types (nondystrophic and dystrophic) based on the natural history and curvature characteristics. The manifestations of nondystrophic curvature and its management are very similar to those of idiopathic scoliosis. Dystrophic curves often occur earlier than nondystrophic curves and have worse prognosis. Owing to its specific pathologic characteristics and process, decisions on the management strategy of scoliosis in NF-1 should depend on the curvature location, range, flexibility, and Cobb angle (coronal and sagittal planes). Hook-rod–based instruments and hybrid systems have been most frequently used to correct dystrophic scoliosis in NF-1 [4–8]. Most of these techniques require the combining of anterior and posterior procedures or a hybrid of screws and hooks. Occasionally, a multistage procedure is required. The shortcomings of these methods are apparent: longer treatment cycle, greater injury, more blood loss, and more complications. Literature on the surgical management of dystrophic scoliosis in NF-1 with the one-stage posterior segmental pedicle screw system is scant.

The purpose of this retrospective study was to report the clinical outcomes of treating dystrophic scoliosis in NF-1 with the one-stage posterior pedicle screw approach in 16 patients in our spinal surgery department.

Materials and methods

We reviewed all dystrophic scoliosis patients who presented with two or more diagnostic criteria of NF-1 and who were treated between 2001 and 2009 with the one-stage posterior segmental pedicle screw technique. Preoperative clinical examination included thorough neurologic evaluation and radiological examination. All patients underwent standard standing plain radiography of the whole spine (posterior-anterior and lateral positions) and side-bending radiograph examination. Coronal and sagittal curve measurements were made on the whole-spine radiograph images using the Cobb method, and we measured apex rotation according to the Nash-Moe method. We measured the distance between apex vertebra and central sacral vertical line (DAC), pelvic obliquity, shoulder tilt, sagittal vertical axis, and pelvic tilt angle to assess spinal balance. Spinal flexibility was evaluated using side-bending images. We also investigated fusion segments using computed tomography (CT) to assess both preoperative bone quality and severity of pedicle deformity (Fig. 1); seven patients underwent three-dimensional CT (3D-CT). Computed tomography was also used to help assess the fusion rate after surgery. We considered loss of correction of 10° or more as another indicator of nonunion or pseudoarthrosis [9–12]. After 2005, all patients underwent magnetic resonance imaging (n=13) to assess spinal cord deformities. As most dystrophic patients have severe thoracic deformities, all patients underwent pulmonary function testing and echocardiography to assess cardiopulmonary function and the presence of congenital cardiac malformations, respectively.

Operative techniques

All surgeries were performed by the corresponding author with somatosensory-evoked potential monitoring. Patients received general anesthesia and were placed in the prone position on prone frames. A posterior median incision was made along the spinous processes. Two methods of screw insertion and fusion segment strategies were applied. One was applied in nine patients from one or two levels proximal to the upper end vertebra to one or two levels distal to the lower end vertebra (EV+1 or 2); the remaining seven patients underwent fusion to stable vertebrae (SV). All patients were treated with one-stage posterior segmental pedicle screw instrumentation and spinal fusion. The