

Case Report

# Dystrophic thoracic spine dislocation associated with type-1 neurofibromatosis: Case report and rationale for treatment

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Abstract

The authors report a rare case of spontaneous dystrophic thoracic spine dislocation in a 14-year-old boy with neurofibromatosis type I (NF-1). Anteroposterior and lateral standing radiographs showed a dysplastic kyphoscoliotic deformity, with the thoracic kyphosis and scoliosis measuring 75° and 69°, respectively. Three-dimensional reconstruction after computed tomography demonstrated spondyloptosis at T5-T6 with overlapping of T5 over T6 and T7. The patient underwent circumferential fusion with anterior fibular strut grafting mechanically secured between the inferior and superior endplates of T5 and T7 followed by an instrumented posterior fusion from T2 to L1 and thoracoplasty. There was satisfactory resolution of the deformity with stabilization at the last follow-up evaluation.

**Key words:** Kyphoscoliosis, neurofibromatosis, scoliosis, spondyloptosis, thoracic spine

INTRODUCTION

Spinal deformities are the most frequent musculoskeletal manifestation in type-1 neurofibromatosis (NF-1), mainly affecting the thoracic region.<sup>[1]</sup> Despite the high variability in its reported incidence (ranging from 2 to 69%) studies carried out in an otherwise unselected cross-section of NF-1 patients had showed an overall prevalence of 10%.<sup>[1,2]</sup>

Spontaneous spinal column dislocation represents the rarest and most difficult-to-treat form of spinal deformity in patients

with NF-1. To date, only several cases have been reported and its optimal surgical treatment has not been clearly established [Table 1].

In this report a 14-year-old-boy who developed nontraumatic dystrophic thoracic spine dislocation associated with NF-1 is presented. There are also discussed relevant issues that must be taking in account during the planning and execution of its surgical management.

CASE REPORT

A 14-year-old adolescent boy, with known personal history of NF-1 was seen due to long-standing progressive thoracic hump in his back. The patient denied spinal pain, suboccipital headache, paresthesia, numbness, urinary retention, or other neurologic symptoms.

At the time of initial examination, he was found alert, oriented, and his cognitive status was unremarkable. There were detected several café-au-lait spots and subcutaneous

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neurofibromata spread over his body. A midthoracic kyphotic gibbus was found, which appeared to have some flexibility during trunk hyperextension. The curve was clinically well compensated.

The muscle strength during extension of the hip and knee joints was grade 5 for both sides. He did not have sensory loss and reflexes of lower extremities were not hyperactive. Babinski reflexes were negative.

Anteroposterior and lateral standing radiographs showed a dysplastic kyphoscoliotic deformity, with the thoracic kyphosis and scoliosis measuring 75° and 69°, respectively. Three-dimensional reconstruction after computed tomography demonstrated spondyloptosis at T5-T6 with overlapping of T5 over T6 and T7 [Figure 1].

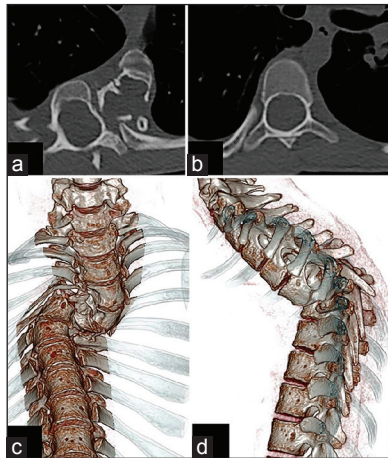
Magnetic resonance (MR) imaging showed a large dural ectasia extending from cervical spinal canal, at C7 level to the

lower thoracic spinal canal. There was no intraspinal tumor or abnormalities in the spinal cord [Figure 2].

The patient underwent circumferential fusion with anterior fibular strut grafting mechanically secured between the inferior and superior endplates of T5 and T7 followed by an instrumented posterior fusion from T2 to L1. A three-ribs thoracoplasty was carried out during the posterior stage of the treatment. The result was stable at the last follow-up evaluation [Figure 3].

## DISCUSSION

Spinal deformities in NF are generally classified into nondystrophic and dystrophic types, based on pathological morphological changes identified on imaging. Nondystrophic type resembles imaging patterns found in idiopathic scoliosis patients and account nearly 40% of all spinal deformities associated with NF-1.<sup>[2]</sup> The behavior of this type of curves



**Figure 1: Preoperative thin-cut computed tomography.** Axial cuts at level of spondyloptosis (a) and below (b) showing the dysplastic changes along the thoracic spine. Three-dimensional reconstruction after computed tomography; coronal reformation view (c) and lateral reformation view (d)

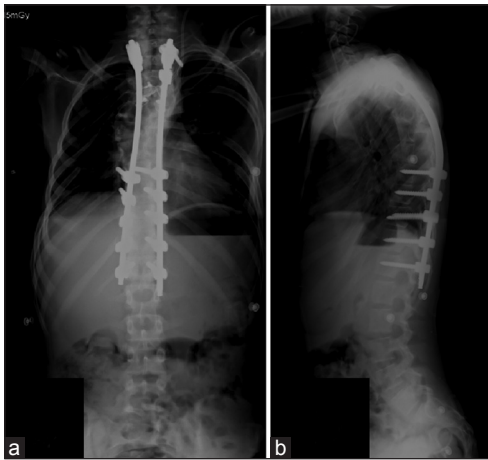


**Figure 2: Preoperative magnetic resonance imaging of the thoracic spine** shows the presence of extensive dural ectasia over the level of spondyloptosis (a) and below (b)

**Table 1: Case reports of surgically-treated spontaneous spondyloptosis in NF-I patients**

Author and year	Gender	Age (years)	Location	Neurologic deficit	Treatment
Rockower et al., 1982 <sup>[3]</sup>	Female	10	T4-T5	No	One-staged PF and AF; plaster body jacket for 9 months
Fasanelli et al., 1986 <sup>[4]</sup>	Male	10	NS	No	NS
Stone et al., 1987 <sup>[5]</sup>	Male	9	T1-T2	No	PF
Winter, 1991 <sup>[6]</sup>	Female	21	T9-T10	Thoracic myelopathy	Two-staged PF and AF (T3-T6), Risser cast for 6 months and plastic jacket for 6 months
Wong-Chung and Gillespie, 1991 <sup>[7]</sup>	Female	11	Lumbosacral	No	Hyperextension cast for 3 weeks, PLF, and postoperative casting for 3 months
Goffin and Grob, 1999 <sup>[8]</sup>	Female	41	C5-C7	Cervical myelopathy	Cervical traction, one-stage AF and PF
Hanna et al., 2002 <sup>[9]</sup>	Female	7	T6-T7	Paraparesis	Halo traction, PF followed by AF with structural grafting
Kim et al., 2007 <sup>[10]</sup>	Female	35	T5-T6	Progressive paraparesis	Halo-pelvic traction, PF (T2-T12) and AF with strut graft between T4 and T7
Melloh et al., 2013 <sup>[11]</sup>	Male	37	C6-C7	Cervical myelopathy	PF (C3-T9) and AF with vascularized fibula strut graft one year later

AF = Anterior fusion; PF = Posterior fusion; PLF = Posterolateral fusion; NS = Notstated, NF-I = Neurofibromatosis type I



**Figure 3:** Postoperative standing AP (a) and lateral radiographs (b) demonstrating a satisfactory correction and stabilization of the thoracic spine deformity. There is no progression of the residual deformity after 16 months. AP = Anteroposterior

is closely similar to patients with idiopathic scoliosis; and therefore, has been advocated to treat in a similar fashion.<sup>[1,2,12]</sup> On contrary, dystrophic curves requires a different approach, taking in account their aggressive behavior and highly deforming potential. These curves are characterized by several dysplastic changes, including vertebral scalloping, rib penciling or spindling of the transverse processes, vertebral wedging, intracanal rib head protrusion, foraminal enlargement, and/or dysplastic pedicles.<sup>[10,13]</sup> Those abnormalities may be intrinsic or associated with dural ectasia or dumbbell-shaped neurofibromas.<sup>[1]</sup>

The most frequent curves found in patients with NF-1-associated dystrophic deformities are scoliosis, kyphoscoliosis, and lordoscoliosis.<sup>[2]</sup> However, advanced dysplastic changes, particularly dural ectasia, may cause gross instability and bizarre spontaneous vertebral column dislocation.<sup>[3,4,8,9]</sup> This rare type of deformity represents the most severe and difficult to treat for in patients with NF-1, because reconstructive procedures are technically demanding and are associated with high rates of implant failure and nonunion. According to our comprehensive literature review, only nine cases of surgically-treated spontaneous vertebral column dislocation had been previously reported in the literature [Table 1].<sup>[3-9,11]</sup>

Due its rarity, the currently available literature is not able to offer strict guidelines for treatment of spontaneous vertebral column dislocation in patients with NF-1. However, several principles may be inferred from follow-up of patients treated for dystrophic curves. Firstly, circumferential fusion must be intended wherever possible, by means of anterior strut grafting and instrumented posterior fusion with a two-rod construct. In a large series of patients who underwent surgical correction of dystrophic spinal curves in NF-1, the failure incidence of the posterior instrumented fusion alone and of the planned anterior and posterior fusion after a mean follow-up period of 15 years was 53

and 23%, respectively.<sup>[14]</sup> However, achieving the anterior reconstruction is usually difficult because of inadequate local bone stock for positioning a graft because of dural ectasia. Another difficulty also must be faced during the implantation of pedicle screws and sublaminar or subpedicular hooks or wires because of thinning of pedicles and lamina.

Fibular or tibial strut grafts are the most used for anterior reconstruction in patients with thoracic dislocations associated to NF-1.<sup>[9,11]</sup> The graft can be secured between the inferior and superior endplates of two vertebral bodies aligned with the gravity line in the sagittal plane and more or less oblique in the coronal plane, as was accomplished in our case. The anterior strut-grafting must be done from the concavity, where it is put into an appropriate biomechanical situation, under compressive forces.<sup>[11]</sup>

Despite their magnitude, NF-1-associated spinal deformities usually do not origin spinal cord injury because dural ectasia also cause enlargement of the neural foramen and spinal canal. Therefore, attempts for reduction of spondyloptosis in absence of spinal cord dysfunction are deemed unnecessary and may be hazardous.<sup>[9]</sup> In those cases it had been advocated to correct the gibbus in a bayonet apposition (rather than anatomic reduction) and thoracoplasty, as was done in our case. On contrary, when the myelopathy is established, temporary stabilization with halo traction must be implemented during the initial workup.<sup>[9]</sup> This therapy must be followed by posterior correction of the deformity in a bayonet apposition and stabilization with a two-rod construct, anterior structural grafting, and an orthosis for 6 months.<sup>[8,9]</sup>

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