Surgical correction of severe cervical kyphosis in patients with neurofibromatosis Type 1

Report of 3 cases

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Severe cervical kyphosis requiring surgical treatment is rare in patients with neurofibromatosis Type 1 (NF1). When it occurs, however, dystrophic changes in the vertebrae make surgical correction and fusion of the deformity extremely difficult.

The authors report on 3 cases of severe cervical kyphosis associated with NF1 that were successfully treated with combined anterior and posterior correction and fusion. All patients underwent halo-gravity traction for approximately 1 month prior to surgery to correct the deformity gradually. Posterior correction and fusion were performed with segmental spinal instrumentation consisting of lateral mass screws, lamina screws, pedicle screws, and polyethylene tape for sublaminar wiring. Anterior spinal fusion was performed using a fibula strut to induce solid bone fusion. All patients used a halo vest for postoperative external fixation.

Preoperative CT scans showed dystrophic cervical spine changes, and MR images demonstrated extensive neurofibromas outside the cervical spine in all 3 patients. The preoperative kyphotic angles were as follows: Case 1, 140°; Case 2, 81°; and Case 3, 72°; after halo-gravity traction, the kyphosis angles improved to 50°, 55°, and 51°, respectively; and after surgery, they were 50°, 15°, and 27°, respectively. Solid bone union was observed in all patients at the latest follow-up. All three patients experienced postoperative complications consisting of superficial infection, severe pneumonia, and partial dislocation of the distal fibula graft after removing the halo vest, in one patient each.

Although dystrophic cervical vertebral changes in these patients with NF1 complicated the correction of severe cervical kyphosis, the use of preoperative halo-gravity traction, a combination of spinal instrumentations, an anterior strut bone graft, and postoperative halo-vest fixation made it possible to correct the kyphosis, maintain the correction, and achieve solid bone fusion.

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Key Words • cervical kyphosis • neurofibromatosis Type 1 • deformity • dystrophic vertebra

Neurofibromatosis 1 (NF1) is a multisystemic disease presenting with major symptoms of neurofibromas and café-au-lait spots. Although spinal deformity in NF1, first reported in 1918 by Gould, is a common manifestation, with a reported incidence between 10% and 69%, the thoracic spine is usually involved. Cervical spine deformities requiring surgery are rare.

Characteristic dystrophic changes found with spinal deformity in NF1 include scalloping of the posterior vertebral margins, spinal canal widening, enlarged neural foramina, defective pedicles, and spindling of the transverse processes. These changes complicate the placement of stable anchors in the vertebrae, which are essential for effective surgical correction. Posterior fusion involving wire loops between the spinous processes or lateral mass plates, along with anterior fusion with a strut bone graft, has been reported, but most procedures resulted in suboptimal correction of the cervical deformities. Recently developed spinal instrumentation, such as pedicle screws,
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lamina screws, and lateral mass screws, and their use in combination, have improved outcomes for surgical correction of severe cervical spinal deformities. We report on 3 patients with severe cervical kyphosis associated with NF1 who underwent correction and fusion in which a combination of modern spinal instrumentations was used.

Case Reports

Three patients with severe cervical kyphosis associated with NF1 underwent combined anterior and posterior correction and fusion surgeries to correct the spinal deformities. The ethics committee of Keio University School of Medicine approved this study.

Case 1

This 20-year-old man presented with a 2-month history of deteriorating neck pain and upper-extremity numbness (Table 1). His preoperative function was reflected by a JOA score of 16. Radiography showed severe cervical kyphosis of 140° at C3–C7 (Fig. 1A, Table 2). Magnetic resonance imaging demonstrated extensive neurofibroma growth around the cervical spine. Computed tomography revealed extensive dystrophic changes in the cervical spine. Halo-gravity traction for 1 month prior to surgery gradually corrected the kyphosis from 140° to 50° (Fig. 1B). The patient’s numbness in the upper extremities worsened when the traction weight reached 8 kg, and the weight was decreased to 7 kg. The patient underwent same-day combined posterior and anterior surgery. The posterior correction from C-2 to T-4 was performed by placing pedicle screws at C-2 and T1–4, lateral mass screws at C-3 and C-4, and polyethylene tape at C5–7. A fibula strut bone was then grafted anteriorly from C-3 to T-2 to reinforce the bone fusion. The decompression of the spinal cord was performed only via the posterior approach. The distal end of the strut bone was fixed at the T-2 vertebral body with a cortical screw. The total operative time was 675 minutes, and the EBL was 2870 ml. The kyphosis was corrected from 81° to 55°. The surgery was conducted in 2 stages. A posterior correction from C-2 to L-3 was performed by placing pedicle screws at C-2. In the middle and lower thoracic spine and the lumbar spine, lateral mass screws were placed at C-3, C-4, and C-6. Lamina screws were placed in the upper thoracic spine, where severe dystrophic changes made the use of pedicle screws impossible. Two months after the first operation, anterior C3–7 fusion was performed using a fibular strut bone. The distal end of the strut bone was fixed at the C-7 vertebral body using a cortical screw. The total operative time was 675 minutes, and the EBL was 1413 ml. The kyphosis was corrected from 81° to 15° (Fig. 2 B and C). After surgery, the patient developed pneumonia and was treated successfully with intravenous antibiotics. A halo vest was applied for 1 month. Three years after surgery, radiography showed a solid bone union and no significant loss of correction.

Case 2

This 10-year-old girl presented with progressive cervical kyphosis after resection of a spinal cord tumor without neurological deficit (Fig. 2, Table 1). Radiography showed cervical kyphosis of 81° between C-5 and C-7 (Fig. 2A), as well as scoliosis of the thoracic spine (Table 2). Magnetic resonance imaging demonstrated extensive neurofibroma growth around the cervical spine. Computed tomography revealed severe dystrophic vertebral changes. Prior to the surgery, the patient was placed in halo-gravity traction with a weight of 5 kg for 1 month, which corrected the cervical kyphosis from 81° to 55°. The surgery was conducted in 2 stages. A posterior correction from C-2 to L-3 was performed by placing pedicle screws at C-2. In the middle and lower thoracic spine and the lumbar spine, lateral mass screws were placed at C-3, C-4, and C-6. Lamina screws were placed in the upper thoracic spine, where severe dystrophic changes made the use of pedicle screws impossible. Two months after the first operation, anterior C3–7 fusion was performed using a fibular strut bone. The distal end of the strut bone was fixed at the C-7 vertebral body using a cortical screw. The total operative time was 675 minutes, and the EBL was 1413 ml. The kyphosis was corrected from 81° to 15° (Fig. 2 B and C). After surgery, the patient developed pneumonia and was treated successfully with intravenous antibiotics. A halo vest was applied for 1 month. Three years after surgery, radiography showed a solid bone union and no significant loss of correction.

Case 3

This 19-year-old woman presented with gait disturbance and a sensory deficit and clumsiness in both hands. A huge neurofibroma was located at the right side of her neck (Fig. 3). Her preoperative function was reflected by a JOA score of 11. Radiography showed an anterior C-2 subluxation resulting in an angular kyphosis of 72° at C2–5 (Fig. 4). Computed tomography revealed severe dystrophic changes in the cervical vertebrae. Magnetic resonance imaging demonstrated a huge paraspinal tumor and an intradural tumor at C2–3. The patient was placed in halo-gravity traction with a weight of 4 kg for 1 month prior to surgery, which corrected the kyphosis from 72° to 51°. The patient underwent surgery in 2 stages. At the first

TABLE 1: Summary of characteristics in patients with severe cervical kyphosis and NF1

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yrs), Sex</th>
<th>Chief Complaint</th>
<th>Ant/Pst Approach</th>
<th>Op Time (mins)</th>
<th>EBL (ml)</th>
<th>Fused Area</th>
<th>Complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20, M</td>
<td>neck pain &amp; numbness of arms</td>
<td>same day</td>
<td>625</td>
<td>2870</td>
<td>C3–T2</td>
<td>superficial infection</td>
</tr>
<tr>
<td>2</td>
<td>10, F</td>
<td>progressive cervical kyphosis after resection of spinal cord tumor</td>
<td>staged</td>
<td>675†</td>
<td>1414†</td>
<td>C2–L3</td>
<td>pneumonia</td>
</tr>
<tr>
<td>3</td>
<td>19, F</td>
<td>gait disturbance due to cervical myelopathy</td>
<td>staged</td>
<td>893†</td>
<td>3300†</td>
<td>Oc–T3</td>
<td>partial dislocation of graft bone after removal of halo vest.</td>
</tr>
</tbody>
</table>

* Ant = anterior; Oc = occiput; Pst = posterior.
† Total value of first and second surgeries.
stage, posterior correction and occiput–T3 fusion were performed using occipital screws, with pedicle screws being placed at C-2, C-4, C-5, and T1–3 and lateral mass screws being placed at C-4, C-5, and C-6. The spinal cord tumor was resected at this time. Three weeks after the first surgery, a fibula strut bone was grafted at C2–5, and the distal end of the strut bone was fixed at C-5 with a bioabsorbable cortical screw. The total operative time was 412 minutes, and the total EBL was 2860 ml. The kyphosis was corrected from 72° to 27°. A halo vest was applied for 1 month after surgery. After the halo vest was removed, the grafted bone became partially dislodged at the distal end (Fig. 5). Because the bone was still partially in contact with the C-5 vertebra, the patient was placed in a Philadelphia collar and carefully observed. Postoperatively her preoperative disability mostly resolved and her JOA score improved from 11 to 16. Four years after the surgery, the patient was free from her preoperative neurological deficits and could walk without any assistance. Solid bone fusion was documented, and the cervical spine correction was maintained.

Discussion

Several factors complicate the treatment of severe cervical kyphosis associated with NF1, including the following: 1) a potentially high risk of SCI during the correction, 2) coexisting paraspinal and spinal cord tumors, 3) difficulties in placing stable anchors in dystrophic vertebrae, and 4) difficulty in obtaining solid bone fusion.

Kyphosis Correction and SCI

The risk for SCI is from the increased compressive forces placed on the apex of the kyphosis during correction. To achieve a gradual and safe correction, each pa-

![Fig. 1. Case 1. Radiographs obtained in a 20-year-old man with deteriorating neck pain and numbness of the upper extremities. A preoperative cervical kyphosis of 140° at C3–7 (A) was corrected to 50° after halo-gravity traction (B). The kyphosis correction was maintained at 50° after surgery (C).](image)

![Fig. 2. Case 2. Radiographs acquired in a 10-year-old girl with progressive cervical kyphosis after a spinal cord tumor resection. A preoperative cervical kyphosis of 81° at C5–7 (A) was corrected to 15° after surgery (B and C).](image)
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Patient was placed in halo-gravity traction prior to correction surgery. Traction started with a weight of 2 kg, and the weight was gradually increased day by day while we carefully observed the patient for neurological deficits. Rinella and colleagues\(^1\) have reported that the maximum traction force for spinal kyphosis or scoliosis should be 33%–50% of the patient’s body weight. However, in the case of cervical kyphosis associated with NF1, we recommend smaller traction forces because of the fragility of the cervical spine due to dystrophic changes. In the case of rigid cervical kyphosis, Wu et al.\(^3\) have recommended anterior release of the cervical spine prior to initiating traction. There is the possibility that patients with NF1 may have an aneurysm at the vertebral arteries.\(^11\) Thus, before starting traction, the patient should be evaluated by MR angiography or contrast-enhanced CT because traction carries a potential risk of aneurysmal rupture.\(^11\) In the present study, the traction weight was increased to a final weight of 4–7 kg, and the kyphosis was corrected to safe preoperative levels.

**Table 2: Kyphotic angle of the cervical spine**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Kyphotic Angle (°)</th>
<th>Flexibility (%)</th>
<th>After Halo-Gravity</th>
<th>Final Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preop</td>
<td>Extension</td>
<td></td>
<td>Traction</td>
</tr>
<tr>
<td>1</td>
<td>140</td>
<td>125</td>
<td>11</td>
<td>50</td>
</tr>
<tr>
<td>2</td>
<td>81</td>
<td>78</td>
<td>4</td>
<td>55</td>
</tr>
<tr>
<td>3</td>
<td>72</td>
<td>56</td>
<td>22</td>
<td>51</td>
</tr>
</tbody>
</table>

**Fig. 3.** Case 3. Studies obtained in a 19-year-old woman with gait disturbance and clumsiness and sensory disturbance in both hands. A huge neurofibroma was located in the right side of the neck (left). The patient was placed in preoperative halo-gravity traction with a weight of 4 kg for 1 month (right).

**Fig. 4.** Case 3. Left: Preoperative radiograph shows a C-2 subluxation resulting in an angular kyphosis of 72° at C2–5. Right: Postoperative radiograph demonstrating that the kyphosis was corrected to 27°.

**Coexisting Paraspinal and Spinal Cord Tumors**

Magnetic resonance imaging should be used to detect coexisting paraspinal and spinal cord tumors, since paraspinal tumors often cause massive hemorrhaging and spinal cord tumors increase the risk of SCI during corrective surgery. Patients should also be observed for postoperative hematoma, because hematomas often cause respiratory problems by compressing the airways.\(^29\) Preoperative tumor embolization has been reported to decrease hemorrhage from neurofibromas,\(^27\) although it was not used in our 3 patients. If a spinal cord tumor exists in the spinal canal, we recommend tumor excision before correction of the kyphosis to prevent SCI, even if the patient had no neurological deficit.

**The Difficulty of Placing Stable Anchors**

It is often extremely difficult to place stable anchors where there are severe dystrophic changes in the cervical spine.\(^7,18,28\) Without stable anchors, the correction will eventually be suboptimal. A morphological evaluation of the cervical vertebrae should be performed by CT prior to surgery, to determine the best type of spinal instrumentation for each vertebra. Although pedicle screws have superior biomechanical stability,\(^15–17\) they are rarely usable in cervical vertebrae with dystrophic pedicle changes. Instead, a combination of lateral mass screws, lamina screws, and high polymer polyethylene tape was used in our cases.

**Difficulty of Obtaining Rigid Bone Fusion**

Most previous investigators have recommended combined anterior and posterior correction and fusion surgeries.\(^5,6,12,21,25,30,32,33\) A very few have recommended anterior fusion alone\(^13\) or posterior fusion alone.\(^19\) In all 3 of our patients, dystrophic changes in the laminae and lateral
The surgical treatment of severe cervical kyphosis in NF1 is complicated by dystrophic changes in the cervical vertebrae that make rigid fixation and sufficient bone fusion challenging. Preoperative halo-gravity traction and the combined use of applicable spinal instrumentations, anterior strut bone grafts, and postoperative halo vest fixation made it possible to correct the kyphosis, maintain the correction, and achieve solid bone fusion.

Conclusion

The surgical treatment of severe cervical kyphosis in NF1 is complicated by dystrophic changes in the cervical vertebrae that make rigid fixation and sufficient bone fusion challenging. Preoperative halo-gravity traction and the combined use of applicable spinal instrumentations, anterior strut bone grafts, and postoperative halo vest fixation made it possible to correct the kyphosis, maintain the correction, and achieve solid bone fusion.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Acquisition of data: Kawabata, Watanabe, Hosogane, Ishii, Nakamura. Analysis and interpretation of data: Watanabe, Ishii, Nakamura. Drafting the article: Kawabata, Watanabe. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Matsumoto. Study supervision: Matsumoto, Watanabe, Toyama.

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