Surgical Treatment of Congenital Kyphosis Associated With Progressive Spastic Paralysis in an Adult Patient
—Case Report—

Hiroshi NOMURA*, Kazumasa TERADA*,**, Nobuo KOBARA*,**, Kiyoshi MIYAZAKI*,**, Michitaka YUASA*, Dai MURATA*, and Hisaaki MIYAHARA*,**

*Department of Orthopedic Surgery and **Clinical Research Institute, National Kyushu Medical Center, Fukuoka

Abstract

A 38-year-old man presented with untreated congenital kyphosis associated with progressive spastic gait. To prevent progression of the spastic paralysis, rigid correction of the severe spinal deformity arising from the congenital kyphosis was performed by one-stage posterior closing-wedge osteotomy, without occurrence of neurological complications. Progression of the paralysis has not been identified for 30 months after the operation and a slight improvement in gait was recognized. The current case is categorized as type I deformity of congenital kyphosis in the upper thoracic spine, which is normally treated surgically before the adolescent growth phase begins.

Key words: congenital kyphosis, closing-wedge osteotomy, single posterior approach, segmental spinal instrumentation, spastic paralysis

Introduction

Congenital kyphosis is an extremely rare deformity of the spine which, if left untreated, may result in strict angular deformity associated with severe spinal compression myelopathy. Accordingly, early surgical treatment is critical for pediatric patients with this condition.3,4,9–11) We treated an adult patient with congenital kyphosis associated with progressive spastic paralysis of the lower extremities.

Case Report

A 38-year-old man presented with a 3-year history of progressive gait disturbance and numbness of the bilateral lower extremities. No episodes of gait disturbance had been noted during the adolescent period of growth, although kyphotic deformity of the thoracic spine had been recognized. Gait disturbance with numbness in the lower extremities began from age 35 years with no episode of trauma. The patient then developed progressive paraparesis of the lower limbs requiring the use of two sticks to aid walking since age 36 years. The gait disturbance became markedly worse during the 2-month period prior to our initial consultation.

Neurological examination revealed hyperactive deep tendon reflex in the bilateral lower extremities. Gait was patently spastic, and obvious bilateral ankle clonus and moderate muscular weakness of the lower limbs were observed. Babinski reflex was easily evoked. Urinary frequency was slightly increased. Sensory disturbance was not prominent. Frontal radiography detected no scoliotic deformity of the spine, but lateral radiography exposed severe local kyphosis of the middle column of the thoracic vertebrae (Fig. 1). The sagittal Cobb angle was 75° between the upper endplate of T-7 and the lower endplate of T-9 (Fig. 1 right). In addition, the an-
Fig. 1 Preoperative radiographs of the patient aged 38 years showing congenital kyphosis. Left: Frontal radiograph showing no scoliotic deformity. Right: Lateral radiograph demonstrating severe local kyphosis of the middle column of the thoracic vertebrae. Note that the sagittal Cobb angle is 75° between the upper endplate of T-7 and the lower endplate of T-9.

terior and middle columns of the T-8 vertebral body were not visualized. Three-dimensional computed tomography (CT), sagittal CT reconstructed imaging, and postmyelography CT of the thoracic spine revealed the bilateral T-8 quarter vertebrae attached to the bilateral T-8 rib bones (Fig. 2A–G). Magnetic resonance imaging and postmyelography CT showed that the extreme narrowing of the spinal cord at the apex in the kyphosis was due not only to sagittal compression from the sharply angulated kyphosis, but also to axial compression from the bilateral posterior quarter vertebrae (Fig. 2H, I). Because of these findings, we performed a laminectomy and closing-wedge osteotomy by correction with segmental spinal instrumentation via a single posterior approach (Fig. 3).

Blood for autotransfusion (800 ml) was prepared before the operation. The patient was placed in the prone position on a four-poster frame. A midline longitudinal incision was made over the spinous processes from T-3 to T-11. After retraction of the paravertebral muscles, the operative field was extended laterally above the rib heads for a distance of approximately 5 cm along the proximal part of the ribs between T-4 and T-10. The posterior elements, involving the bilateral T-8 quarter vertebrae, were identified (Fig. 4A). The bilateral ribs of T-7, T-8, and T-9 were transected approximately 3 cm lateral to the costotransverse joint.

Under epidural-epidural spinal cord-evoked potential control, a laminectomy from T-6 to T-9 was performed. During this procedure, there was enough workspace on the proximal side of the dorsal surface of the ventrally-shifted spinal cord. Massive fatty tissue around the dorsal surface of the theca was resected and both tent-like heap and sandglass-like constriction of the dural sac were exposed at the corner of the apex in the kyphotic thoracic spine. To create some space beyond the compressed spinal cord, osteotomy of the apex was carried out using an airtome attached with a cutting burr, Luer’s bone rongeur, and curette. Then, a wedge-shaped osteotomy was carefully performed between the caudal side of T-7 and the rostral side of T-9 using a thin chisel, preserving the anterior tip of the vertebral body and the anterior longitudinal ligament. The intervertebral disc was also removed. Consequently, the dural tube at the apex was circumferentially decompressed.

After the wedge-shaped osteotomy, hooks and pedicle screws were placed segmentally from T-4 to T-11 (Fig. 4B). A bent rod was positioned vertically, then the compression force was loaded using a compressor to close the wedge-shaped osteotomy gap with the hinge of the remaining anterior tip of the vertebral body. During this step, spinal cord-evoked potential monitoring recorded no abnormal electrical waves although the dural tube was gradually shortened. A second rod was also positioned using adequate compression force. The double Cotrel-Dubousset M8-rods were connected with a low-profile crosslink (Fig. 4C). The spinal instrumentation was set to be immobile, providing extreme rigidity.

No neurological deterioration was evident after surgery. After the operation, the sagittal Cobb angle was corrected from 75° to 38° (Fig. 5). Because gait was still unstable, the patient was allowed to begin walking 6 weeks after surgery. From 2 weeks until 4 months after surgery, a hard thoracolumbosacral brace was applied. Bony union was confirmed by tomography 4 months after surgery, and then the hard corset was changed to a soft one. From 6 months after surgery, the numbness in the lower extremities had almost completely disappeared and
the ankle clonus was apparently improved compared with the preoperative condition. Sagittal CT reconstructed imaging and CT revealed favorable bony union and circumferential decompression of the spinal cord at the level of the apex 15 months after the operation (Fig. 6). Progression of the spastic paralysis has not been identified for 30 months after the operation. Gait disturbance has recovered slightly, although the patient is still unable to move independently.

Discussion

Congenital kyphosis is classified into three types.1,2) Type I deformity, which is defined as congenital absence of the vertebral bodies, is the most likely to result in neurological complications. In particular, deformities in the upper thoracic spine usually result in neurological problems.11) The current case was categorized as type I deformity of congenital kyphosis in the upper thoracic spine associated with severe spastic paralysis. Conservative treatment by bracing is ineffective for congenital kyphosis, so surgical treatment is recommended before the adolescent growth phase begins.3,4,6-11) However, the current patient had not received any clinical treatment until age 38 years. As a result, the natural development of the kyphotic deformity was structurally complete and the spastic paralysis continued to progress. Based on the various image findings, we suggest that the origin of the current paralysis was associated with both sagittal compression from the sharply angulated kyphosis and axial compression from the bilateral posterior quarter vertebrae. Consequently, we decided that circumferential decompression of the spinal cord might prevent further progression of the spastic paralysis, and selected a combination of spinal correction of the structural kyphosis and removal of the bilateral quarter vertebrae.

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Fig. 3 Diagram of the lateral view of the thoracic spine at the left side from T-6 to T-10. The area of the wedge osteotomy is indicated by the oblique lines. Left T-8 quarter vertebra is shown by an asterisk.

Fig. 4 Photographs of closing-wedge osteotomy. A: Posterior aspects of the fusion range were exposed laterally above the rib heads for a distance of approximately 5 cm along the proximal part of the ribs. B: After wedge-shaped osteotomy between the caudal side of T-7 and the rostral side of T-9 with the segmental placement of Cotrel-Dubousset (CD) M8-hooks and pedicle screws. Note that the dural tube on the apex is circumferentially decompressed. C: After the positioning of double CD M8-rods which were connected with a low-profile crosslink.

Anterior fusion is indicated in type I deformities if the angle of kyphosis is greater than 50 degrees.11) Recently, with the development of many spinal instrumentation systems, closing-wedge osteotomy via a single posterior approach has been recommended for correction of kyphotic deformity for three reasons.1–8) First, this operative method enables one-stage rigid correction of the spinal deformity using segmental spinal instrumentation instead of the former two-stage procedure of combined anterior and posterior fusion. Thus, operative stress, including postoperative management, is greatly reduced. Second, since the closing-wedge osteotomy results in the contact of two wide cancellous surfaces, fusion rates are very high.3) Third, circumferential decompression of the spinal cord can be performed during closing-wedge osteotomy. For these reasons, we chose to carry out closing-wedge osteotomy in the current case.

Prior to the closing-wedge osteotomy, a laminectomy must be performed to provide a direct view of the theca. This can be performed safely because there is enough working space behind the narrowing spinal cord following the anterior shift of the spinal cord, thus allowing safe handling of the circumferential decompression of the spinal cord. In particular, the surgeon needs to pay attention to the neurological complications associated with the shortening of the expanding dural tube following closure of the wedge-shaped osteotomy gap. Such neurological defects may also be caused by loss of the bilateral segmental blood supply to the spinal cord following ligation of the radicular artery during circumferential decompression of the spinal cord.1,5) During this procedure, the spinal cord-evoked

Fig. 5 Frontal (left) and lateral radiographs (right) 3 months after the operation. Note that the sagittal Cobb angle after the correction is 38° between the upper endplate of T-7 and the lower endplate of T-9 (right).
Fig. 6  A, B: Sagittal computed tomography (CT) reconstructed images of the middle column of the thoracic spine at the left side (A) and the median (B) 15 months after the operation showing satisfactory bony union between T-7 and T-9 and the decompressed spinal cord at the level of the apex.  C: CT scan at the level of the apex showing adequate circumferential decompression of the spinal cord.

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References


Address reprint requests to: K. Terada, M.D., Department of Orthopedic Surgery, National Kyushu Medical Center, 1–8–1 Jigyouthama, Chuo–ku, Fukuoka 810–8563, Japan.
 e-mail: kterada@qmed.hosp.go.jp