CASE REPORT

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THE SURGICAL TREATMENT METHOD FOR AN ADULT POSTTRAUMATIC THORACOLUMBAR KYPHOSIS PATIENT WITH OSTEOGENESIS IMPERFECTA

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ABSTRACT

Osteogenesis imperfecta (OI) is an inheritable bone disorder characterized by osseous fragility and ligamentous laxity. It is sometimes difficult to obtain bone union in patients with OI. The purpose of this report is to present a rare case of posttraumatic kyphosis due to a L1 burst fracture in a patient with OI, and to discuss how to treat it to achieve an adequate correction and circumferential fusion. The patient was a 29-year-old man with OI (Sillence type-IA) who had sustained an L1 fracture when he dived head first into a river. After 3 months of conservative therapy with a body cast, he showed disability at work because of his persistent low back pain and fatigue in his whole back. He showed no neurological disorder. Diagnostic imaging revealed localized kyphotic deformity at L1. Therefore, lumbar lordosis and thoracic kyphosis worsened. Anterior release and fusion, and posterior fusion were conducted. Three months after surgical treatment, circumferential fusion was obtained. His low back pain and fatigue in the whole back disappeared, and he could resume work without any difficulty. From the bone union standpoint, the surgical strategy for spinal correction in OI patients is still controversial because of the intractableness of bone union and fragility of the bone itself. The authors achieved circumferential union using anterior fusion and posterior fusion, in which wide bone bed is available owing to spared posterior elements of the spinal column.

Key Words: Osteogenesis imperfecta, posttraumatic kyphosis, thoracolumbar, surgical treatment

INTRODUCTION

Osteogenesis imperfecta (OI) is an inheritable bone disorder caused by abnormal synthesis of type-I collagen[1)]. It is characterized by osseous fragility and ligamentous laxity, and classified into four types (type I, mild; II, lethal; III, severe; IV, moderate), based on clinical, radiographic, and genetic characteristics and bone fragility[2, 3)]. The prevalence of OI is rare (1/5,000–10,000)[1-3)]. Therefore, there are few case reports of surgery for OI, and an optimal operative strategy has not yet been found[4-6)]. The authors had the opportunity to treat a patient with OI, who had an injured L1 fracture that afterward worsened into symptomatic kyphotic deformity without any

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neurological deficit. Because of the intractableness of bone union caused by the abnormal bone metabolism associated with OI, the surgical strategy for correcting this kyphosis is controversial. Eventually, a preferable outcome was obtained after anterior-posterior surgery with instrumentation, without any decompression.

There has been no report to date on the correction of adult posttraumatic kyphosis in a patient with OI. Therefore, the authors considered that reporting this case would be worthwhile, with its particular emphasis on the advantages of circumferential bone union.

CASE REPORT

History and Presentation

The patient was a 29-year-old man with OI (Sillence type-IA) who had sustained an L1 fracture when he dived head first into a river. After 3 months of conservative therapy with a body cast in another clinic, he showed disability at work because of his persistent low back pain and fatigue in his whole back. Then, he was referred to our institution.

Neurologic examinations upon admission were entirely normal. In his childhood and school days, he had suffered from fractures 5 times in the upper extremities, which were treated conservatively and did not show any functional disorder. He also had an episode of an abdominal injury (hepatic rupture and perforation of sigmoid colon) due to 2 separate traffic accidents in the past, in which open abdominal operations were required. His mother and cousin were also patients with OI.

Examinations

On examination, the findings from routine laboratory tests were within normal limits. His height was 164 cm and his weight was 55 kg. His dentine appeared normal, but showed blue sclera. Severe kyphotic deformity was observed when sitting, while the trunk was symmetric. There was no hearing disorder.

The spinal radiographs revealed osteopenia of the vertebral bodies and the presence of severe thoraco-lumbar kyphosis. On the coronal plane (Figure 1A), the C7 plumb line was 19 mm right of the center sacral vertical line (CSVL). On the sagittal plane (Figure 1B), the Cobb angle of the upper curve from T2 to T10 was 4 degrees, whereas that of the lower curve from L2 to S1 was 29 degrees. The C7 plumb line was +18 mm, whereas localized kyphosis from T12 to L2 showed 36.4 degrees in neutral position (Figure 2B), 41.1 degrees in flexion (Figure 2C), and 30.1 degrees in extension (Figure 2D). Although computed tomography (CT) showed approximately 40% spinal canal occupation due to retropulsed bony fragment (Figure 3A and B), cerebro-spinal fluid space was wide enough in the axial and sagittal magnetic resonance images. The Japanese Orthopaedic Association (JOA) scoring system of lumbar spine (maximum score 29 points) for him was 19, and the Japanese Orthopaedic Association Back Pain Evaluation Questionnaire (JOABPEQ) implied considerable difficulties in daily life.

Operation

The patient then underwent anterior-posterior surgery with instrumentation and both auto iliac bone graft and allograft, extending from T12 to L2. Instrumentations used in the anterior approach were a mesh cage with auto bone graft from the iliac bone and rib bone, and in the posterior approach, they were a mono-axial pedicle screw with a diameter of 6.5 mm and a laminar hook with a decortication and allograft (Figure 4A and B). In the operation, T12 and L1 were completely fused. Then, anterior release was first conducted from the left chest wall anterior

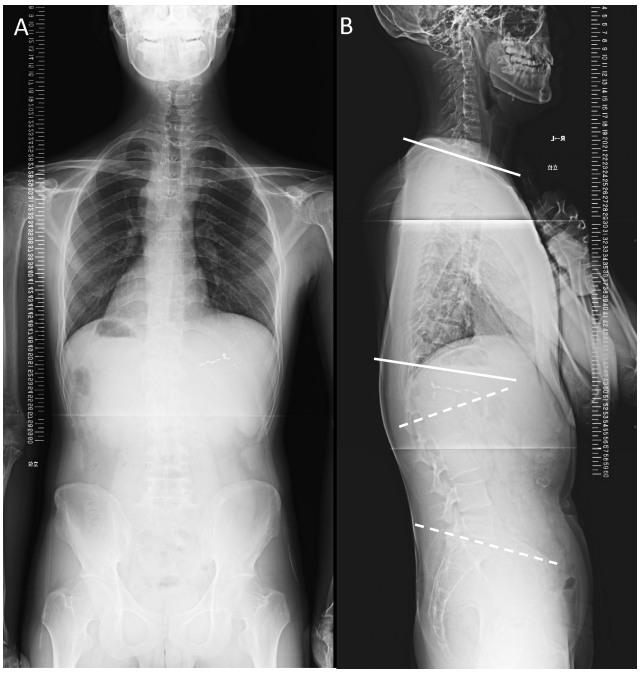

Fig. 1 Preoperative radiographs of whole spine in frontal **A** and lateral view **B**.

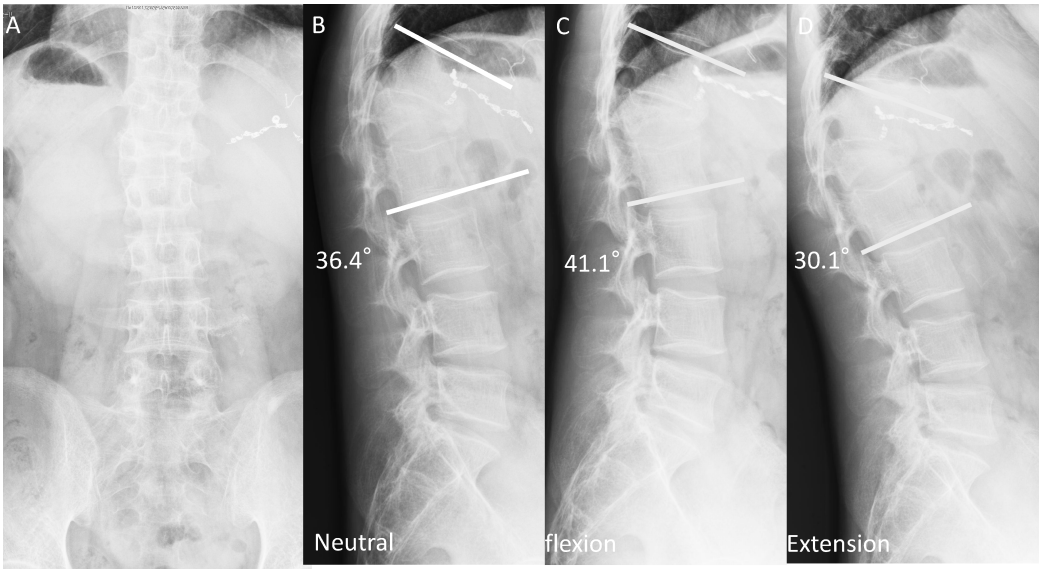

Fig. 2 Preoperative radiographs of lumbar spine in frontal **A** and lateral view **B-D**. In the lateral view, images in flexion position **B**, neutral position **C**, and extension position **D** are shown.

approach with a resection of the 11th rib. After releasing the anterior fusion mass from the left side of the spinal column to the right side, spinal kyphosis was released and manipulated to a

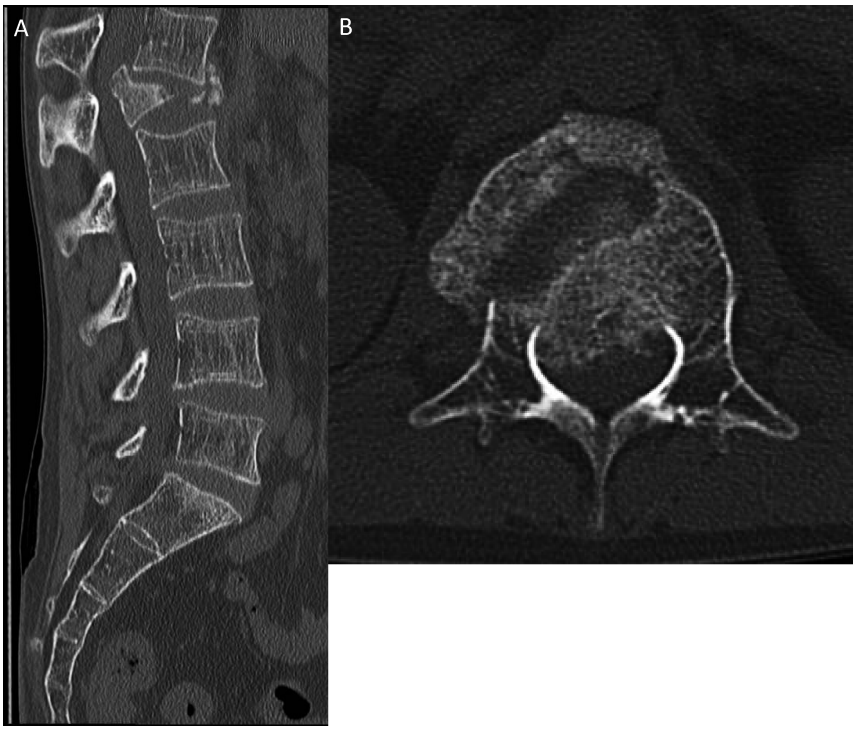

Fig. 3 Preoperative reconstructed sagittal image **A** and axial image **B** of computed tomography.

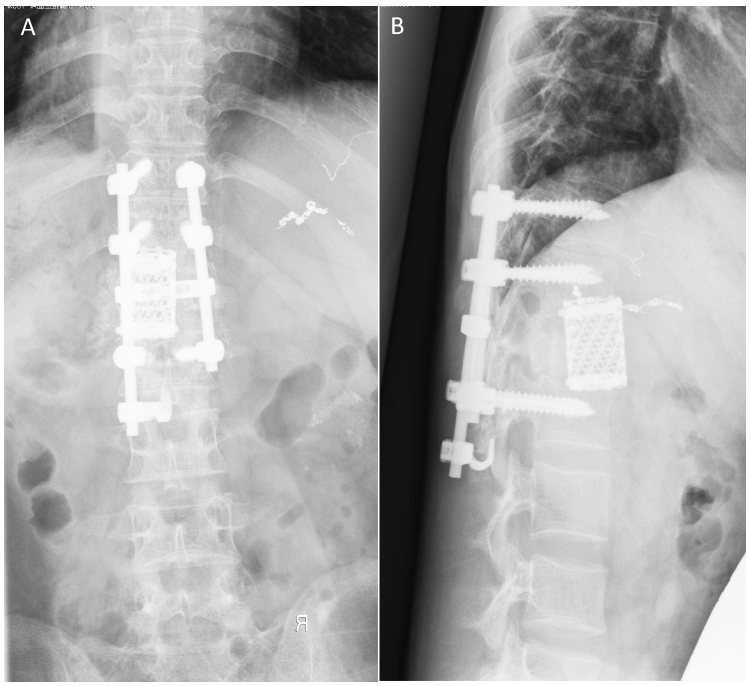

Fig. 4 Postoperative radiographs of lumbar spine in frontal **A** and lateral view **B**.

nearly straight alignment under a radio-fluoroscopy image. Next, a mesh cage with the height of 42 mm was set up, in which an auto bone graft from the iliac and rib bone was filled up. Because neuromonitoring with motor and sensory-evoked potential did not show any abnormal change throughout this anterior procedure, anterior decompression was not conducted. After completing the anterior procedure, posterior fixation with pedicle screw was performed. Because the bone quality of the vertebra was found to be poor, laminar hooking was added. After the decortication of laminar and facet joint fusion from T12 to L2 with allograft, the operation was completed. Blood loss was estimated to be approximately 900 ml, and the operation time was 372 minutes.

Postoperative course

Postoperatively, the patient recovered well without any complication, and his height increased to 166 cm. Low back pain and fatigue, which made him unable to work, diminished 1 month after the operation. The spine radiographs showed that on the coronal plane, the C7 plumb line changed from 19 mm right to 8 mm (Figure 5A). On the sagittal plane, compensatory change of upper thoracic curve (pre-op; 4 degrees, post-op; 18 degrees) and lower thoracolumbar curve (pre-op; 29 degrees, post-op; 38 degrees) were both remedied accompanied with the correction of localized kyphosis from T12 to L2 itself (pre-op; 36.4 degrees, post-op; 8 degrees), and the C7 plumb line was +21 mm, which implied the global sagittal balance was considered to be preferable (Figure 5B). Fusion T12 to L2 was obtained on the CT sagittal reconstruction view and lateral functional radiograph 2 years after the surgery (Figure 5C). The postoperative JOA

Fig. 5 Postoperative radiographs of whole spine in frontal **A** and lateral view **B**. Postoperative reconstructed sagittal image of CT shows bone fusion inside the mesh cage with auto bone graft **C**. Postoperative axial CT image shows that canal stenosis due to fracture fragment has been improved **D**.

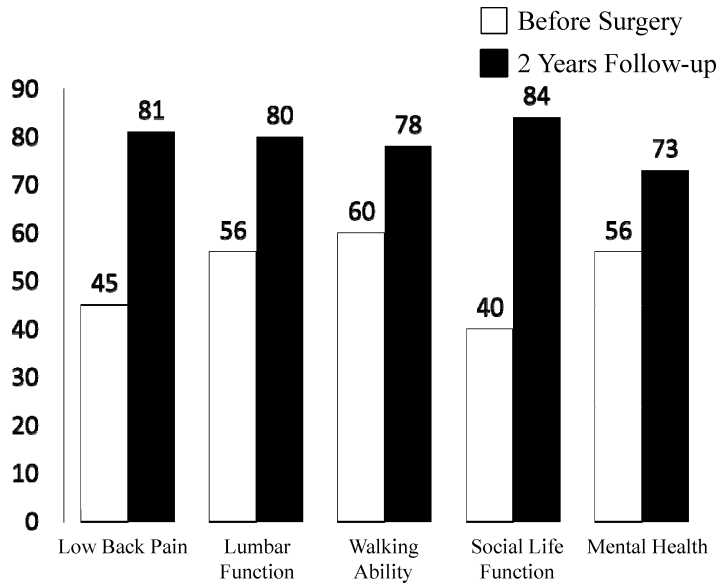

Fig. 6 Graph showing improvement in JOABPEQ scores. In 5 categories (low-back pain, lumbar function, walking ability, social life function, and mental health) the 2-year follow-up scores were higher than the preoperative scores.

scorering system of lumbar spine was 27 points, and also JOABPEQ scores showed improvement in any aspect of difficulties because of the low back pain (Figure 6). The patient appeared well after 2 years of follow-up. He was fully informed that his data would be submitted for publication, and he gave his consent.

DISCUSSION

Posttraumatic kyphosis can occur in patients after conservative therapy or inadequate surgical treatment, and may impact the conus or cauda equine[7]. It can be treated with both conservative and surgical treatment; however, when inadequately treated, there is a tendency for the injured vertebra to gradually collapse because of the direct force on the anterior and middle column of the spine. In addition, posterior muscle strength becomes weaker year by year, thus worsening the collapse[8]. Patients treated conservatively often experience chronic back pain because of the gradual increase of kyphosis, which causes further damage to the posterior soft tissues. A recent study indicated that among posttraumatic kyphosis patients, 94% have continuous back or lower extremity pain, and 46% will have worsening of the kyphosis[9]. Therefore, the patient in this report required surgical treatment even though he was a patient with OI.

Because of the difficulty in establishing bone union in OI patients, treating their spinal compression fractures is thought to be difficult and complicated, and there are few reports on vertebroplasty or kyphoplasty for OI spinal fractures[10, 11]. However, to our knowledge, there has been no detailed report on an OI patient with posttraumatic thoracolumbar kyphosis.

Generally, the problematic points in surgery for OI were associated with bone fragility in both the laminar and vertebral body, which sometimes led to instrumentation failure afterward, excessive blood loss during and after the operation, and late pseudoarthritis or non-union[4-6]. In

treating this case with symptomatic rigid kyphosis, the authors needed to obtain both correction and fusion, considering several of the issues described above. Then, the authors considered vertebral column resection (VCR)[12, 13], pedicle subtraction osteotomy (PSO)[14-16], and anterior-posterior surgery[17] as surgical options which could achieve an adequate correction for kyphosis. Although the capability of each surgical method for correcting kyphosis in this case was thought to be adequate[12-17], the greatest difference among them was the utility of bone bed of posterior elements. VCR and PSO could treat this kyphosis with only one posterior incision, which was an advantage for patients with past abdominal injury. However, those methods invariably cut the posterior bone continuity, which is thought to be a disadvantage in respect of utility of posterior bone bed. On one hand, anterior and posterior fusion requires two different incisions, which was thought to be a serious disadvantage for the patient with 2 abdominal surgeries in the past. But this method could use both anterior and posterior bone beds, which could reduce the risk for postoperative non-union to a minimum. The authors made bone union after the operation a matter of highest priority, and selected anterior and posterior surgery. Moreover, allograft[18] as well as auto iliac bone graft was adopted to avoid the lack of bone stock. Eventually, both an anterior and posterior, namely a circumferential union, was obtained 3 months after the operation, and the fused segment (T12-L2) did not show any mobility in the functional X-ray images. The authors achieved their goal of circumferential bone union.

In conclusion, the authors conducted anterior and posterior fusion with allograft and iliac bone graft for a symptomatic thoracolumbar kyphosis which occurred in an adult patient with OI, and circumferential bone union and drastic improvement of symptoms were obtained 3 months after the surgery. This method was considered to be the optimal surgery for achieving an adequate correction of kyphosis and appropriate fusion.

DISCLAIMER

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