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Adult Congenital Lumbar Kyphosis Requiring Anteroposterior Correction and Fusion: A Case Report with 32-Year Follow-Up

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Abstract

Congenital pure kyphosis due to failure of vertebral body segmentation is a relatively rare entity, and surgical intervention is infrequent compared to that for failure of vertebral body formation [1] [2]. There are very few reports of long-term follow-up of surgical treatment in patients with congenital pure kyphosis, and all the reported cases were diagnosed as failure of formation and had an age at the time of surgery of less than 18 years. It is important for orthopedic surgeons to follow the postoperative course of rare cases over 30 years. Here, we present a surgically treated case with ultra-long term follow-up of a 50-year-old patient with congenital pure kyphosis of the lumbar spine. Imaging of the lumbar spine showed six vertebrae and an unsegmented bar at L3-4 causing a pure kyphosis of 54˚. The wedge-shaped block vertebra had 4 pedicles with the neural foramen between the pedicles without concomitant disc space, with compensatory thoracic hypokyphosis and lower lumbar hyperlordosis. One-stage correction and fusion surgery using anterior opening and posterior closing osteotomy was successfully performed. Both clinical and radiographic results were excellent and have been maintained for over 30 years postoperatively. The basic principle in the surgical treatment of adult spinal deformity is to achieve and maintain a good global sagittal balance over time. This case reaffirms the importance of spinopelvic harmony.

Keywords

Adult Congenital Kyphosis, Anterior Posterior Spinal Fusion, Failure of Vertebral Body Segmentation, Long-Term Follow-Up, Spinopelvic Harmony

1. Introduction

Congenital spinal malformations that cause progressive spinal deformity, such as

congenital scoliosis and kyphoscoliosis, are not uncommon, with a reported prevalence of 0.13 - 0.5/1000 live births. However, congenital kyphosis of the lumbar spine caused by failure of vertebral body segmentation is a less frequent deformity compared to kyphosis or kyphoscoliosis due to failure of formation. Most cases occur in childhood and surgical treatment is not mandatory because paraplegia is infrequent and the severity of this type of deformity is relatively mild, unlike in patients with failure of formation. Here, we present a neglected case of lumbar congenital pure kyphosis in an adult that resulted from failure of segmentation. A successful one-stage combined anteroposterior osteotomy and deformity correction was performed at age 50. In the follow-up period of over 30 years to date, the patient has maintained a stable and well-balanced spinopelvic sagittal alignment, resulting in a good clinical outcome. We conducted a retrospective evaluation of this case, using modern sagittal spinopelvic parameters for adult spinal deformities.

2. Case Presentation

2.1. Patient

A 50-year-old male visited our hospital for the first time with severe low back pain and gait disturbance. His past medical history included nephroptosis. He had noticed a lumbar hump since childhood, but he had not sought medical care because he was asymptomatic for low back pain and paralysis.

2.2. History of Present Illness

Four years prior to his first visit, the patient had low back pain and walking difficulty, and had undergone surgery at another hospital. He told us that his symptoms had worsened after the surgery, and severe low back pain had appeared 6 months before he visited our hospital. Conservative treatment was used, but the patient gradually found it difficult to maintain a standing position, even for five minutes. Thus, he was referred to our hospital in 1991.

2.3. Status at Admission

A scar of approximately 10 cm from the previous surgery was observed at the midline caudal of the lumbar hump. Lumbar kyphosis was present with a vertex at the conjoined vertebra. Muscle strength and sensation of the lower extremities were both preserved, suggesting that the neurologic status was normal.

2.4. Imaging Findings

No imaging results were available to document progression of the deformity prior to the first visit, but plain X-ray radiographs showed several abnormal findings. There were 13 thoracic and 6 lumbar vertebrae, including an L3-4 unsegmented bar, causing pure kyphosis in the lumbar spine. The wedge shape block vertebra had 4 pedicles with the neural foramen between the pedicles without concomitant disc space. L3-4 kyphosis of 54˚, compensatory thoracic

hypokyphosis, and lumbar hyperlordosis were observed. A laminectomised L6 vertebra in conjunction with 22% retrospondylolisthesis was also present. The sagittal L1-S1 Cobb angle, which indicates lumbar lordosis (LL), was 5˚. Pelvic parameters were not available because femoral heads were invisible due to poor resolution of the old X-ray film (Figure 1). Lumbar spine MRI revealed no disc space of the block vertebra without spinal canal stenosis (Figure 2). Based on these findings, the patient was diagnosed with congenital kyphosis of the lumbar spine due to failure of L3-4 vertebral body segmentation.

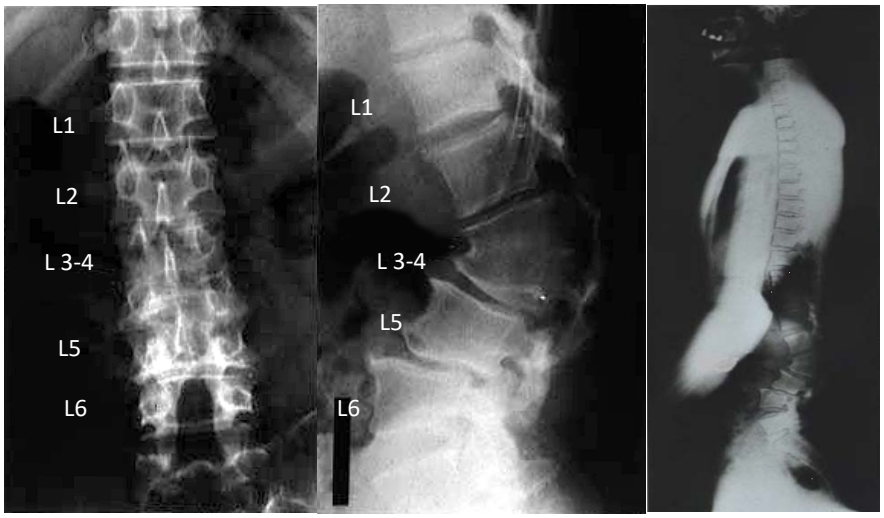

Figure 1. Plain X-ray radiographs before surgery. (a) Anteroposterior view of six lumbar vertebrae, with L6 laminectomised. (b) Lateral view of a L3-4 unsegmented bar with 54˚ kyphosis and retrospondylolisthesis of L5. (c) Full-length lateral standing view showing lumbar lordosis of 5˚.

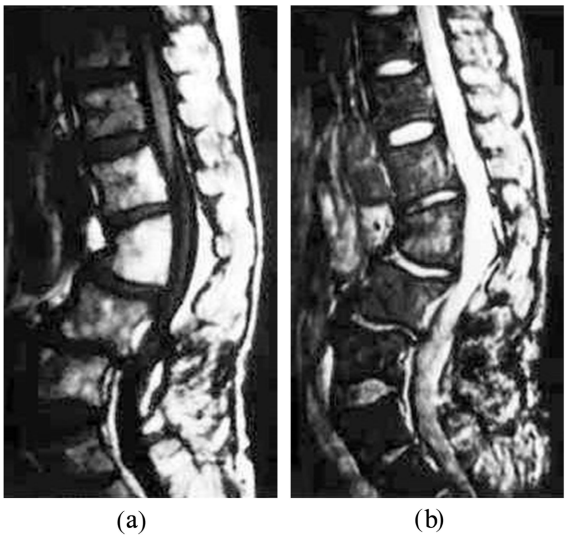

Figure 2. Sagittal view of lumbar spine MRI before surgery. (a) T1-weighted image. (b) T2-weighted image. Disc space of the block vertebra showed a complete defect. No spinal canal stenosis is present.

2.5. Surgical Procedure and Findings

One-stage correction surgery using anterior opening and posterior closing osteotomy was planned. A conventional midline posterior skin incision was made. The spinous process, laminae, and facet joints of the L3-4 apex were all fused, and there was no interlaminar space between L3-4. Posterior wedge osteotomy was performed using the Smith-Peterson osteotomy technique in the apical region of the kyphotic deformity, followed by application of Cotrel-Dubousset (CD) compression hooks (Sofamor, France) to the cranial (L2) and caudal (L4) laminae bilaterally, after which the wound was temporarily closed. The patient was then rolled to the right decubitus position, and an extrapleural-retroperitoneal approach was used to expose the anterior portion of the lumbar spine. Vertebral osteotomy of the L3-4 body was performed at the level of the bisected line parallel to the horizontal plane. Kostuik-Harrington (KH) anterior screws (Zimmer, IN, USA) were inserted in the L1, L2, L5 and L6 vertebral bodies, all intervertebral discs between L1 and L6 were removed, and dual KH distraction rods were set to correct the kyphosis. A distraction force on the KH rods and a compression force on the CD rods were simultaneously loaded, and anterior opening and posterior closing osteotomy were completed. Anterior strut grafts were added using autologous rib and tricortical iliac bone for all instrumented levels (**Figure 3**). The operative time was 577 min and the estimated blood loss was 2246 ml. The procedure was essentially the same as that described by Kostuik *et al.* [3]. A pedicle screw system was not available at the time of the surgery.

2.6. Postoperative Course

The patient was mobilized one week after surgery in a body cast that was worn for 3 months, followed by a thoracolumbosacral orthosis for 3 months. On postoperative imaging at 6 months after surgery, the LL had increased from 7˚ to 28˚,

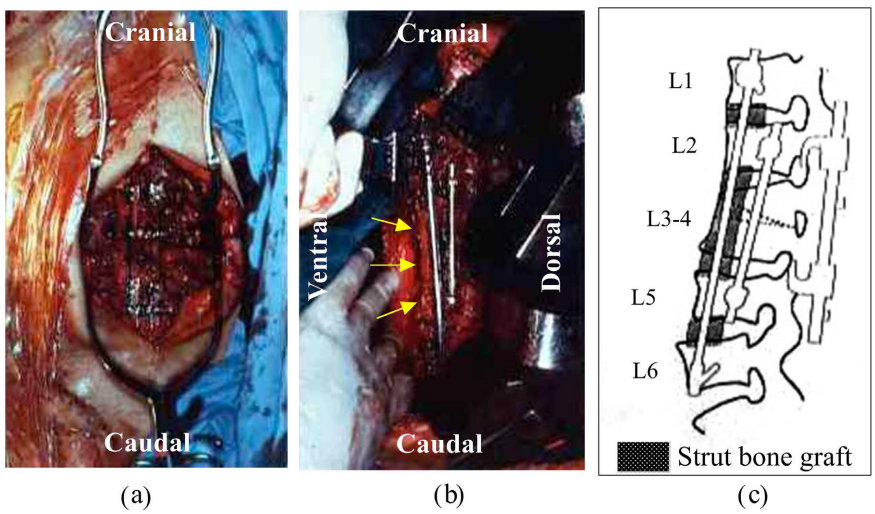

Figure 3. Operative field after one stage anterior opening and posterior closing osteotomy and bone grafting. (a) Posterior view. (b) Anterior view. The yellow arrow indicates autologous strut bone. (c) Schematic drawing of the postoperative view.

showing favorable realignment of the lumbar spine (Figure 4). Severe low back pain and gait disturbance were diminished without any neurological complications. However, at 8 months after surgery, the patient complained of irritation in his back. He underwent posterior device removal for a prominent CD instrument 11 months postoperatively, when fusion was evident radiographically. Thereafter, the chief complaints were resolved, and an excellent surgical outcome has been maintained for a long period. The most recent follow-up was in 2023 when the patient was 82 years old, 32 years after surgery. His clinical condition at this time included mild low back pain, no neurological symptoms, and an Oswestry disability index of 23%. On radiographs, the only motion segment of the lumbar spine at the L6-S disc space was well preserved. There was no proximal or distal adjacent segment kyphosis, and global spine lateral view radiographs indicated pelvic incidence (PI): 28˚, LL: 23˚, PI-LL: 5˚, pelvic tilt (PT): 14˚, and sagittal vertical axis (SVA): 10 mm (Figure 5). Adjusting those results to modern SRS-Schwab criteria [4], the coronal curve type was N and the sagittal modifiers (PI-LL, global alignment, and PT) were all 0. The patient has maintained the reconstructed spinopelvic alignment for 32 years after surgery.

3. Discussion

Congenital kyphosis due to failure of vertebral segmentation is an uncommon spinal deformity. In 1973, Winter *et al.* [1] radiographically classified this deformity into three types: type I, anterior failure of vertebral body formation; type II, anterior failure of vertebral body segmentation; and type III, mixture of failure of formation and segmentation based on the pathologic embryology. These types of deformities are less common compared to congenital scoliosis and are potentially more critical [1]. A type I deformity produces a sharp angular kyphosis

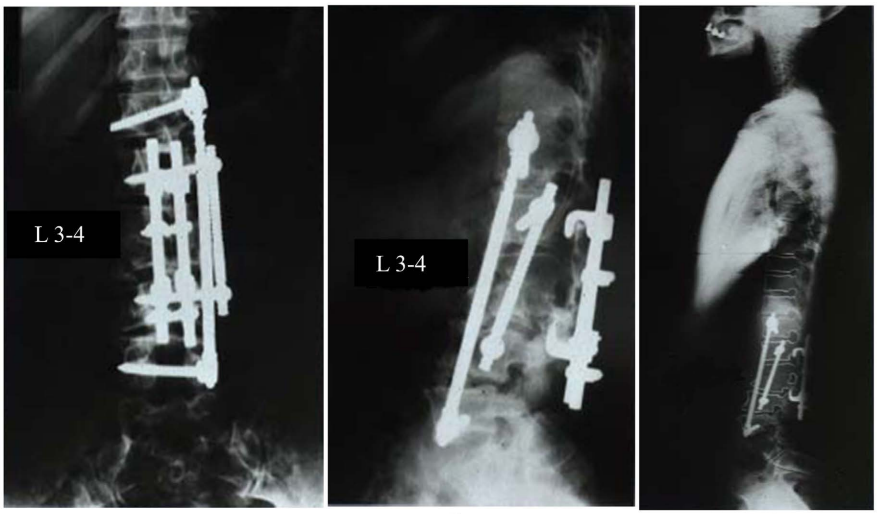

Figure 4. Plain X-ray radiographs at six months after surgery. (a) Anteroposterior view. (b) Lateral view. (c) Full-length lateral standing view. Lumbar lordosis was restored to 28˚.

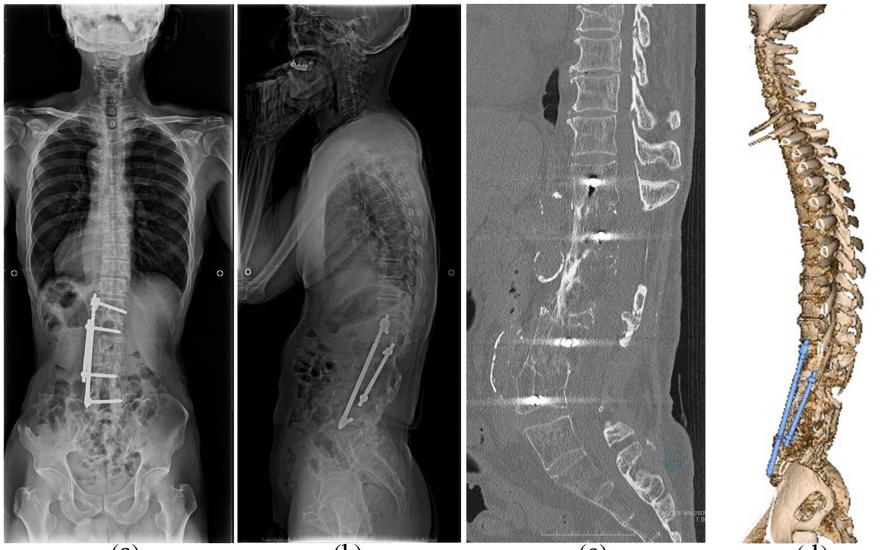

Figure 5. Plain X-ray radiographs and CT at 32 years after surgery. (a) Full-length postero-anterior view. (b) Lateral view showing pelvic incidence—lumbar lordosis of 5˚. (c) Sagittal reconstruction CT showed solid fusion. (d) Lateral view of 3D-CT reconstruction.

that tends to progress to spinal cord compression and paraplegia, and for which conservative treatment is generally ineffective. In contrast, a type II deformity is much less common than type I, is mostly observed at the thoracic and thoracolumbar levels, and does not involve neurologic complications because it produces a smooth kyphosis [1] [2]. The current case is categorized as a type II deformity arising at the mid-lumbar level. There are very few reports with over 10-year follow-up of surgical treatment in patients with this disease, and all the reported cases were diagnosed as type I and had an age at the time of surgery of under 18 years old [5] [6] [7] [8]. To the best of our knowledge, there have been no reported cases of ultra-long follow-up after one stage anteroposterior corrective surgery for lumbar type II pure kyphosis in adult.

McMaster and Singh suggested that progression of type II pure kyphosis is relatively slow and has a low risk of paraplegia [9]. Thus, a type II deformity does not necessarily require immediate surgical treatment unless it is severe enough to require correction or has a smaller curve that shows signs of progression while under observation [10]. Our patient had a significant deformity that was neglected, leading to low back pain with aging and degenerative changes. Considering the diagnosis before the initial surgery, the complaint of low back pain and gait disturbance could have led to a misdiagnosis of lumbar spinal canal stenosis and treatment with wide laminectomy, which might accelerate disc degeneration and L6 retrospondylolisthesis. We speculate that loss of lumbar posterior structures such as the spinous process, supraspinous ligament, and back muscle injuries accelerated disc degeneration and aggravated low back pain before the first visit to our hospital. Mayfield *et al.* [11] found a frequency of 38% for low back pain requiring treatment in a large series of type II cases, with the pain related to compensatory lumbar hyperlordosis caudal to the block ver-

tebra. Remarkably, these authors predicted 40 years ago that patients with such a type II deformity treated with observation only would develop significant deformity and lumbar hyperlordosis, and that low back pain may be a problem during adulthood. The current case is an absolute match to this prediction.

Many innovative surgical techniques, rigid spinal instruments, and comprehensive clinicoradiographic evaluation systems for diagnosis and treatment of spinal deformity have been introduced since 1991 [12] [13] [14] [15], while aging of society has increased the number of patients with various types of adult spinal deformities. The classic surgical procedure performed for this patient is highly invasive and is unlikely to be used in the era of new surgical instrumentation and osteotomy techniques. A similar case today would undergo pedicle subtraction osteotomy (PSO) or Grade 4 osteotomy with pedicle screw fixation by a less invasive, single posterior approach [14] [16]. Because this case has several anomalous lumbar vertebrae, the alignment values cannot be accurately compared to the normal SRS-Schwab classification. However, the key principle of obtaining a "balanced spine" with maintenance of ideal sagittal balance for a long period has not changed in surgical treatment of adult spinal deformity, and our case reaffirms the importance of "spinopelvic harmony".

4. Conclusion

To the best of our knowledge, this case presents the longest follow-up report to date of single-stage anterior and posterior correction and fusion for type II adult congenital kyphosis. The clinical and radiographic outcomes were excellent, with a well-maintained achieved correction for 32 years after the surgery.

Declarations

Consent to Patient

The patient was informed that data from the case would be submitted for publication and gave his consent.

Conflicts of Interest

The authors declare that there is nothing to disclose regarding the publication of this manuscript.

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