

DEGENERATIVE CERVICAL MYELOPATHY IN A PATIENT WITH KLIPPEL-FEIL SYNDROME ACCOMPANIED BY CONGENITAL ANOMALIES OF THE PEDICLE AND LATERAL MASS: A CASE REPORT

 Besim Gülseren¹,  Atalay Başdemir¹,  İpek Gül Acartürk¹,  Hale Kayaer¹,  Ahmet Tolgay Akıncı²

¹Trakya University School of Medicine, Edirne, TÜRKİYE

²Trakya University School of Medicine, Department of Neurosurgery, Edirne, TÜRKİYE

ABSTRACT

Klippel-Feil syndrome is a rare congenital cervical fusion anomaly that may lead to degenerative changes and neurological symptoms later in life. Degenerative cervical myelopathy is progressive, and surgical treatment is recommended in moderate and severe cases. A 53-year-old man presented with progressive weakness, spastic gait, and neuropathic complaints. Neurological examination revealed spastic quadriparesis with hyperreflexia, ankle clonus, and bilateral Hoffmann sign positivity. The preoperative modified Japanese Orthopaedic Association score was 11, and the Nurick grade was three. Imaging demonstrated congenital C2-C3 fusion consistent with Klippel-Feil syndrome type II, multilevel cervical canal stenosis, and hypoplasia of the right C5 pedicle and lateral mass. C3-C4-C5 laminectomy was performed for decompression, and lateral mass screw and rod instrumentation was applied over C3-C6. Because the right C5 lateral mass was hypoplastic, no screw was placed at that site; the right-sided construct spanned C3, C4, and C6, and C5 was anchored on the left only. At 18 months the modified Japanese Orthopaedic Association score had improved to 16 and the Nurick grade to one, with no implant-related complications. Degenerative cervical myelopathy arising on a background of Klippel-Feil syndrome may require individualized surgical planning, because coexisting congenital anomalies can determine which fixation points are usable. Careful preoperative radiological assessment and posterior decompression with stabilization can provide satisfactory clinical outcomes.

Keywords: Klippel-Feil syndrome, spinal cord compression, spinal cord diseases, spinal stenosis, vertebral cervical fusion syndrome

INTRODUCTION

Klippel-Feil syndrome (KFS) is a rare spinal anomaly characterized by congenital fusion of the cervical vertebrae, which can present with neurological symptoms in adulthood in some patients. Congenital cervical fusion has been reported to be associated with increased mechanical stress and degenerative changes in adjacent segments. Degenerative cervical myelopathy (DCM) is one of the most common causes of non-traumatic spinal cord dysfunction in adults, and its progressive course calls for early diagnosis and appropriate surgical treatment (1, 2).

The clinical severity of DCM is commonly assessed using the modified Japanese Orthopaedic Association (mJOA) score and the Nurick grade. According to the mJOA classification, DCM

severity is categorized as mild (mJOA 15 or above), moderate (mJOA 12 to 14), or severe (mJOA below 12) (1). Preoperative Magnetic resonance imaging (MRI) parameters, particularly T2 intramedullary signal change, provide additional prognostic information about the outcome of surgery in patients with DCM (3).

Current clinical practice guidelines strongly recommend surgical treatment in patients with moderate and severe myelopathy (1). When multilevel decompression is required in the subaxial cervical spine, lateral mass screw fixation is the most widely used posterior fixation technique, as it provides immediate stabilization and reduces the risk of post-laminectomy kyphosis (4).



Address for Correspondence: Hale Kayaer, Trakya University School of Medicine, Edirne, TÜRKİYE

e-mail: halekayaer28@gmail.com

ORCID iD of the authors: BG: 0009-0004-4593-1770; AB: 0009-0007-9600-3693; İGA: 0009-0004-9683-8541; HK: 0009-0006-0017-2277; ATA: 0000-0002-9937-076X

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Surgical planning may be more demanding in patients with KFS, because congenital anomalies of the pedicle and lateral mass can occur alongside the fusion and become relevant once posterior instrumentation is considered. Congenital pedicle anomalies of the cervical spine include hypoplasia, aplasia, and dysplasia and may be accompanied by an abnormal vertebral artery course. Their prevalence in the general population remains unclear, but they have been documented in computed tomography (CT)-based morphometric studies and may influence screw trajectory planning (4). Lateral mass anomalies, such as hypoplasia or asymmetry, can further complicate fixation strategies. Both therefore call for detailed preoperative CT evaluation and careful selection of screw technique and fixation points before implants are placed (5, 6).

In this case report, we present the individualized surgical approach, the management process, and the clinical and radiological outcomes after posterior decompression and stabilization in a patient with DCM that developed on a background of KFS type II, with congenital anomalies of the C5 pedicle and lateral mass.

CASE REPORT

A 53-year-old man presented to our clinic with left-sided weakness and arm pain that had persisted for approximately eight years and had worsened in recent months. He had a 45 pack-year cigarette smoking history.

Physical examination revealed upper motor neuron findings: bilateral Hoffmann sign positivity, hyperactive deep tendon reflexes (+++/+++), ankle clonus, and increased muscle tone. Motor examination demonstrated asymmetric spastic quadriparesis, with strength graded 4+/5 on the right and 4/5 on the left in both the upper and lower extremities. The patient described dysesthetic complaints in the left hand and the medial aspect of the left leg. His gait was spastic, characterized by increased tone in the lower extremity extensor muscle groups (quadriceps and gastrocnemius-soleus complex), a stiff-legged pattern, reduced stride length, and circumduction, with scissoring observed during ambulation. The velocity-dependent hypertonicity on passive stretch, the ankle clonus, and this gait pattern together established the spasticity as being of upper motor neuron origin.

Preoperative neurological assessment showed an mJOA score of 11 and a Nurick grade of three. Functionally, the patient was able to button a shirt only with great difficulty, could manage stairs only with the aid of a handrail, had mild sensory loss in the hands, and reported mild to moderate difficulty with micturition. He remained independently ambulant but was unable to work at full capacity.

Plain radiographs of the cervical spine, including flexion, neutral, and extension lateral views, demonstrated congenital fusion of the C2 and C3 vertebral bodies with loss of the intervening disc space, consistent with KFS type II (Figure 1). Magnetic resonance

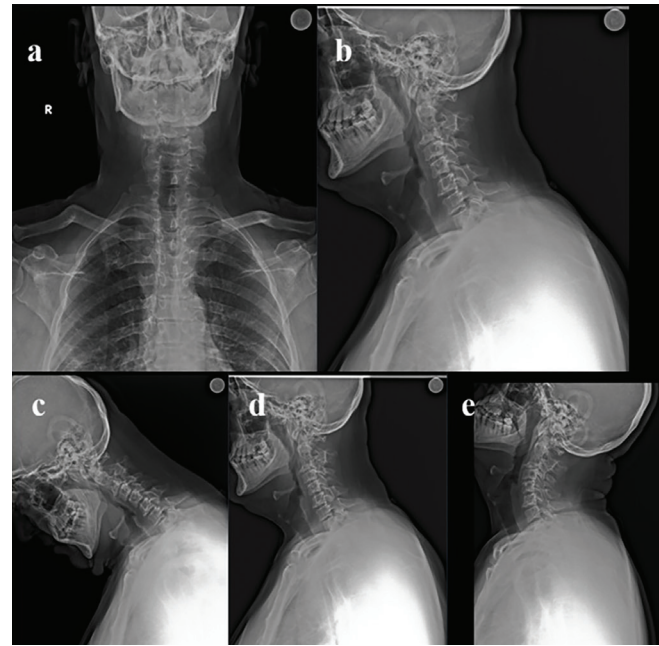


Figure 1: Preoperative plain radiographs of the cervical spine: (a) anteroposterior and (b) lateral views, and (c) flexion, (d) neutral, and (e) extension lateral views. The images reveal congenital fusion of the C2 and C3 vertebral bodies with loss of the intervening disc space, consistent with Klippel-Feil syndrome type II.

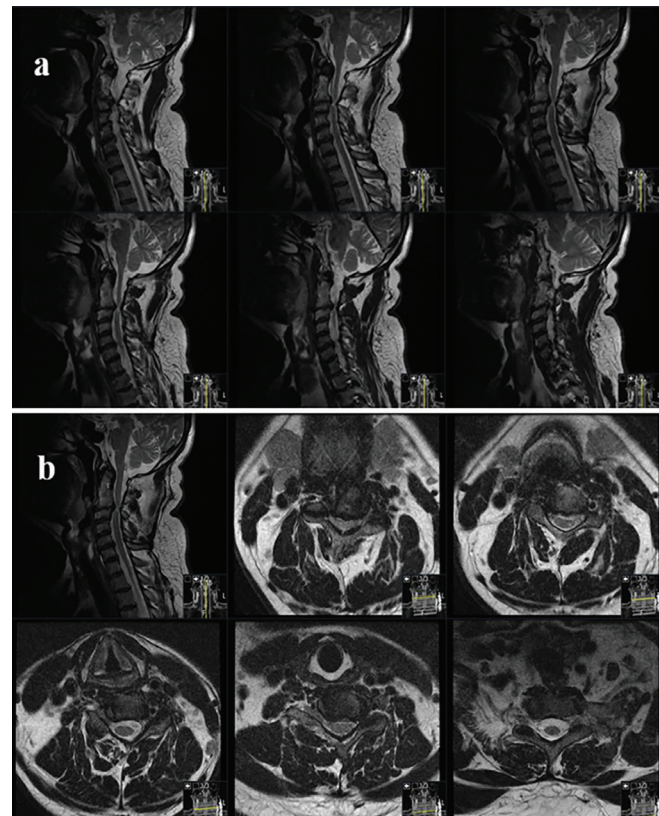


Figure 2: Preoperative T2-weighted MRI of the cervical spine: (a) sagittal images and (b) sagittal and axial images at the stenotic levels. The images reveal multilevel cervical canal stenosis with spinal cord compression, most prominent at the C3-C4 intervertebral disc level. Intramedullary T2 hyperintense signal change is present within the spinal cord at the level of maximum compression, suggesting myelopathic change. MRI: Magnetic resonance imaging.

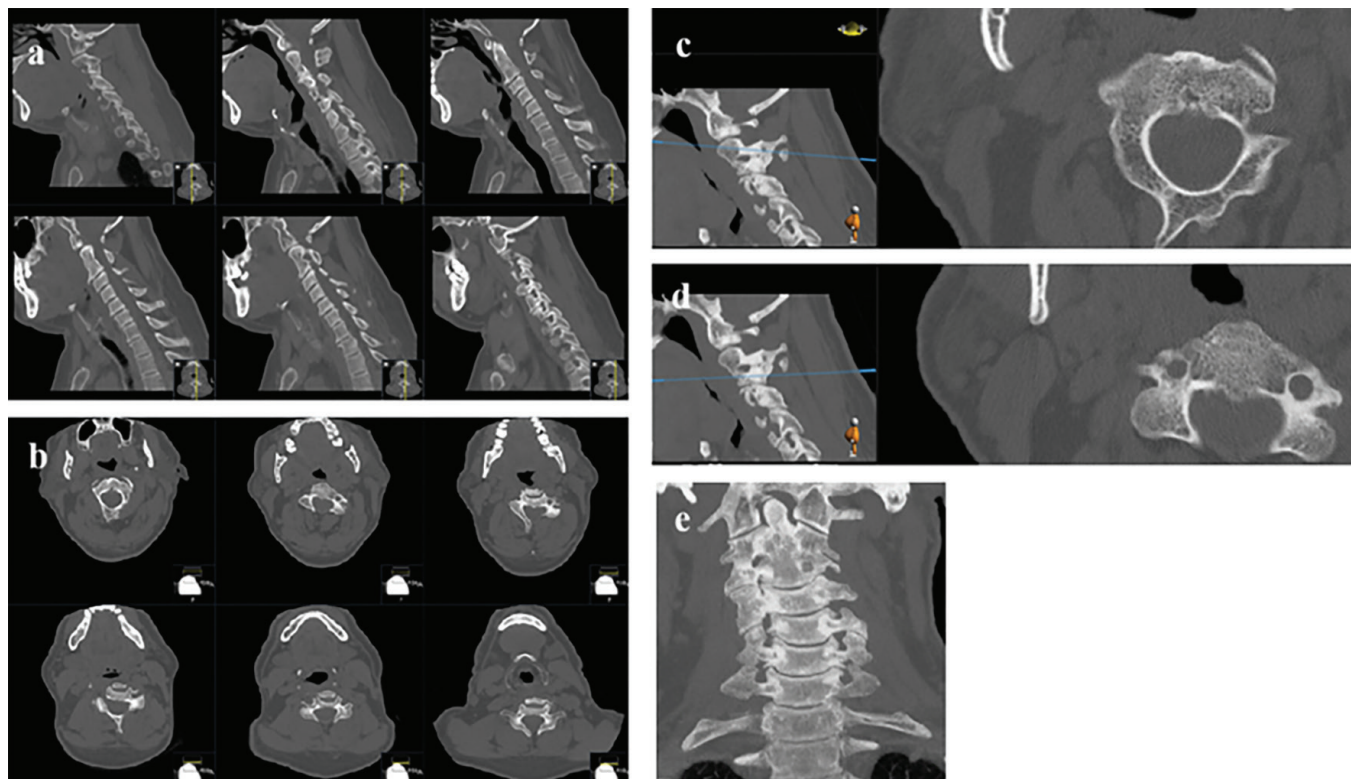


Figure 3: Preoperative CT images of the cervical spine: (a) sagittal reconstructions demonstrating congenital C2-C3 vertebral fusion with absence of the intervertebral disc and a continuous bony bridge, consistent with Klippel-Feil syndrome type II; (b) axial image at the C5 level revealing hypoplasia of the right pedicle and lateral mass; (c, d) axial images at the C3 and C4 pedicle levels, shown for comparison with the adjacent normal anatomy; (e) coronal reconstruction confirming the asymmetry of the right C5 lateral mass and pedicle compared with the left side. CT: Computed tomography.

cord compression, most prominent at the C3-C4 level, together with intramedullary T2 hyperintense signal change at the level of maximum compression (Figure 2). CT confirmed the continuous C2-C3 bony bridge and demonstrated hypoplasia of the right C5 pedicle and lateral mass, with clear asymmetry relative to the left side (Figure 3).

The patient's symptoms were predominantly left-sided, whereas the congenital bony anomaly was on the right. The two findings are not related. The clinical picture reflects the distribution of spinal cord compression rather than the side of the pedicle and lateral mass hypoplasia, which has no direct neural consequence in this patient and is relevant only to instrumentation.

Based on the clinical severity and the multilevel radiological findings, a single-stage posterior decompression and stabilization procedure was planned. The strategy comprised C3-C4-C5 laminectomy for spinal cord decompression and lateral mass screw fixation with rod instrumentation over the C3-C6 levels for stabilization.

Preoperative CT governed how that construct was built. The right C5 lateral mass and pedicle were hypoplastic to a degree that would not have provided adequate bone for safe screw purchase, and the reported association between congenital cervical pedicle anomalies and an anomalous vertebral artery course reinforced the decision not to instrument that site (5).

Screw placement at the right C5 was therefore abandoned at the planning stage rather than attempted and revised during surgery.

Using a posterior approach, C3-C4-C5 laminectomy and spinal cord decompression were performed. Lateral mass screws were then placed bilaterally at C3, C4, and C6, and on the left side only at C5, giving a seven-screw construct. The right-sided rod therefore spanned C3, C4, and C6, bridging the anomalous C5 level without an anchor, while the left-sided rod engaged all four levels. Extending the instrumentation caudally to C6, one level below the most caudal laminectomy, was a deliberate consequence of this asymmetry: it restored a bilateral distal anchor below the skipped level and prevented the right-sided construct from terminating on a single fixation point.

To promote posterolateral arthrodesis, the lateral masses and facet joints at each instrumented level were thoroughly decorticated using a high-speed burr, and locally harvested autogenous bone graft obtained from the laminectomy was applied over the decorticated surfaces (Figures 4, 5). There were no complications during surgery, and the early postoperative period was uneventful.

At the 18-month follow-up, marked neurological improvement was observed. Spasticity and motor weakness had regressed. The mJOA score improved to 16 and the Nurick grade to one.

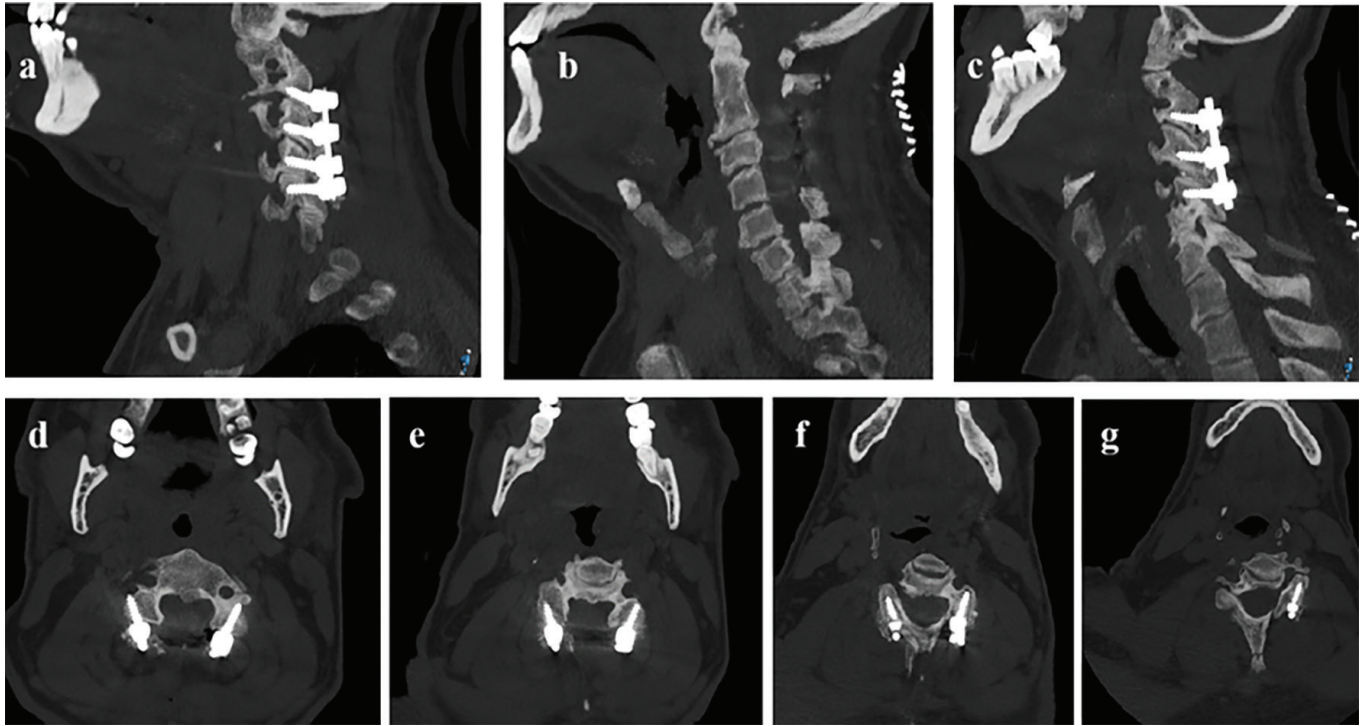


Figure 4: Early postoperative CT images of the cervical spine: (a, b, c) sagittal and (d, e, f, g) axial images. The images demonstrate satisfactory positioning of the lateral mass screws, placed bilaterally at C3, C4, and C6 and on the left side at C5, with the connecting rods in place. No screw was placed in the hypoplastic right C5 lateral mass. Adequate decompression following C3-C5 laminectomy is confirmed, with widening of the spinal canal and no evidence of hardware malposition or canal compromise.

CT: Computed tomography.

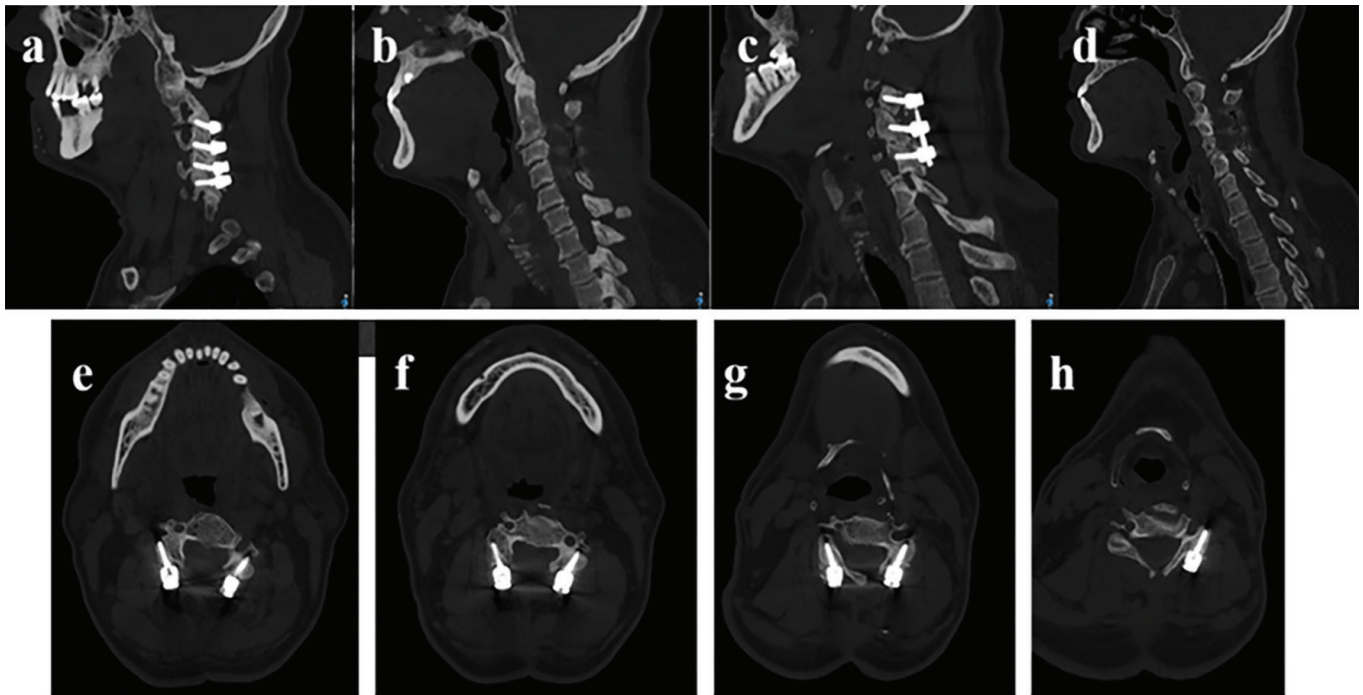


Figure 5: Six-month postoperative CT images of the cervical spine: (a, b, c, d) sagittal and (e, f, g, h) axial images. Compared with the early postoperative study, cervical alignment is maintained and hardware position is stable, with no evidence of screw loosening or rod displacement. The axial images confirm persistent canal decompression at the laminectomy levels. Early signs of posterolateral bony fusion are observed at the instrumented segments.

CT: Computed tomography.

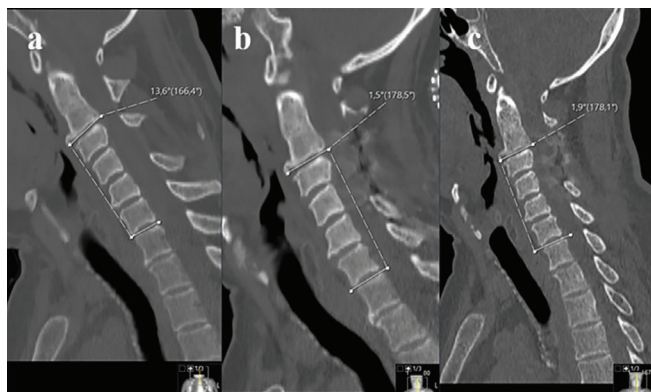


Figure 6: Cervical lordosis angles measured using the C3-C7 Cobb method on sagittal CT images at three time points: (a) preoperative, 13.6 degrees; (b) early postoperative, 1.5 degrees; and (c) six months postoperatively, 1.9 degrees. Lordosis decreased following decompression and instrumentation but showed no further decline between the early postoperative and six-month studies, and the instrumented segment did not progress to kyphosis. Because these angles were measured on supine CT rather than on standing lateral radiographs, they should be interpreted as a within-modality comparison.

CT: Computed tomography

The patient was able to button a shirt with only slight difficulty, walked unaided with a mild lack of stability, had no residual sensory loss in the hands, and reported normal micturition.

Imaging follow-up used two modalities at different time points. CT was obtained in the early postoperative period and at six months to assess implant position, canal decompression, and progression of fusion (Figures 4-6). MRI was obtained at 18 months and confirmed sustained decompression with partial resolution of the intramedullary T2 signal change (Figure 7). No implant-related complication was observed at any point during the 18-month period.

DISCUSSION

Neurological symptoms in patients with KFS may appear in adulthood and may be associated with degenerative changes developing in adjacent segments rather than at the fused level (7, 8). Nouri et al. (9) demonstrated, in a large multicenter cohort of patients with

DCM, that those with congenital cervical fusion exhibited degenerative changes preferentially affecting the segments adjacent to the fused levels, particularly at midcervical levels, supporting the role of adjacent segment pathomechanics in symptomatic KFS patients. Samartzis et al. (7) further characterized congenital fusion patterns in KFS, noting that Type II fusion patterns also carry a risk for developing clinically significant radiculopathic or myelopathic symptoms. Despite

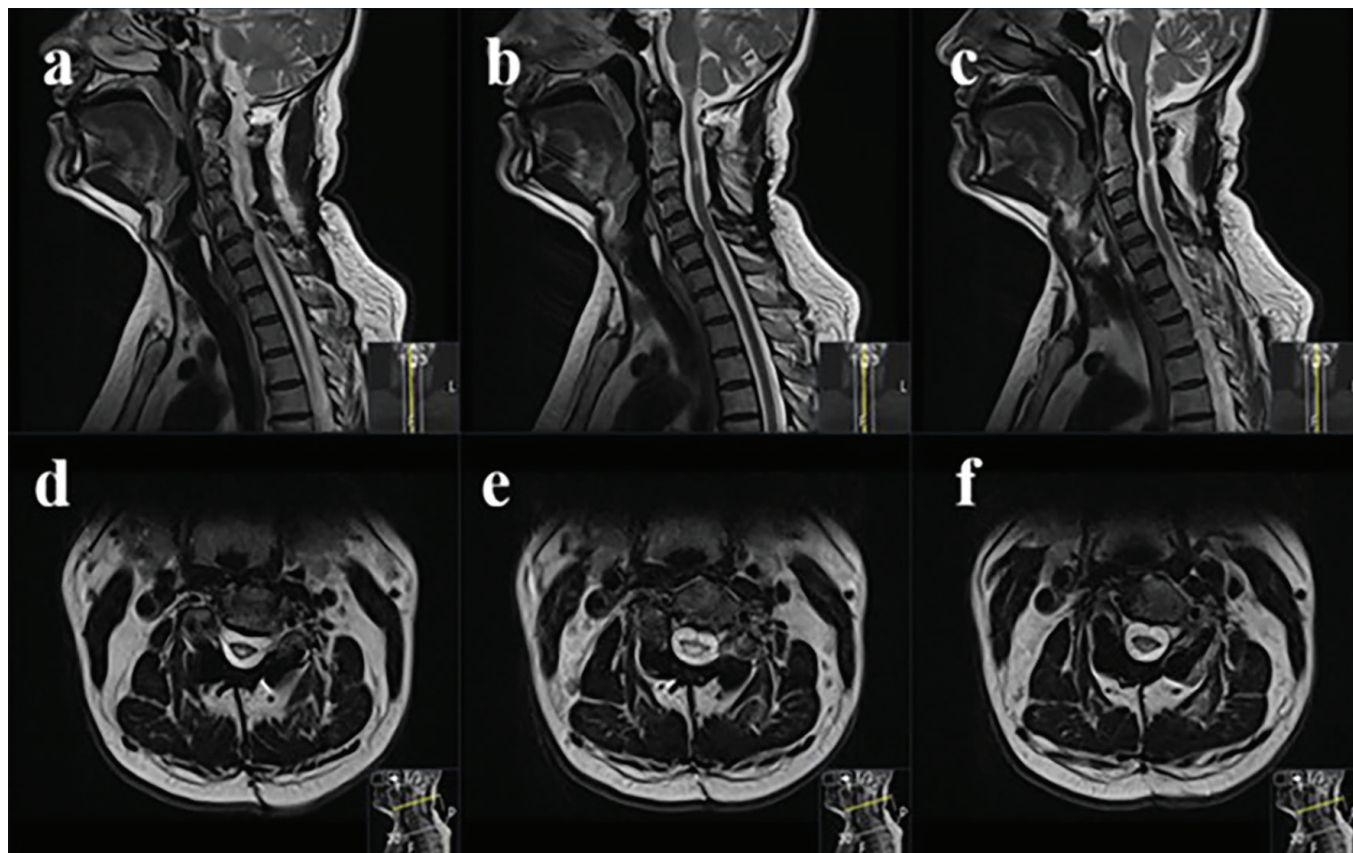


Figure 7: Eighteen-month postoperative T2-weighted MRI of the cervical spine: (a, b, c) sagittal and (d, e, f) axial images. The images reveal adequate spinal cord decompression with partial resolution of the previously observed T2 hyperintense signal change, suggesting recovery from myelopathic change. The sagittal images demonstrate maintained cervical alignment with no evidence of recurrent cord compression at the decompressed levels and no new major degenerative pathology at the adjacent segments.

MRI: Magnetic resonance imaging.

congenital fusion at C2-C3, spinal cord compression was maximal at C3-C4, the segment immediately caudal to the fusion, and extended across the subaxial levels below it.

Degenerative cervical myelopathy is a progressive disorder, and delayed treatment may lead to neurological deterioration. Current clinical practice guidelines strongly recommend surgical treatment in moderate and severe DCM (1). With a preoperative mJOA score of 11 and a Nurick grade of three, our patient fell clearly within that indication. The improvement to an mJOA score of 16 and a Nurick grade of one at 18 months objectively demonstrates the functional effectiveness of surgical treatment.

The technical difficulty in this case lay in the fixation rather than the decompression. Congenital anomalies of the pedicle and lateral mass are among the recognized challenges of posterior instrumentation in KFS, and they may render an intended fixation point unusable (5, 6, 10). Here the hypoplastic right C5 lateral mass was identified on preoperative CT, and the construct was designed around it. Three decisions followed from that single finding: the right C5 screw was omitted, C5 was anchored on the left side alone, and the instrumentation was extended caudally to C6, one level below the most caudal laminectomy, so that the right-sided rod regained a distal anchor and did not terminate on a solitary fixation point.

An asymmetric construct of this kind involves a trade-off. It reduces the number of fixation points on one side and is theoretically less rigid than a symmetric multilevel construct. We considered that trade-off preferable to placing a screw in bone unable to hold it, particularly given the reported association between congenital cervical pedicle anomalies and an anomalous vertebral artery course (5). The six-month CT showed stable hardware position without screw loosening or rod displacement, together with early posterolateral fusion at the instrumented segments, which suggests that the construct was adequate for this patient. In KFS, the preoperative CT therefore carries a second purpose beyond documenting the fusion, which is to establish which fixation points are usable.

The development of adjacent segment degeneration and instability in KFS remains controversial. Some early reports have suggested increased adjacent segment pathology in KFS patients (11), whereas more recent studies, including a large multicenter analysis by Nouri et al. (9), have demonstrated that congenital cervical fusion is associated with a higher prevalence of degenerative changes preferentially affecting midcervical levels, yet the risk of symptomatic adjacent segment disease is not uniformly elevated. Surgical decisions should therefore be individually tailored based on clinical findings and radiological evaluation. In this case, posterior decompression and stabilization were preferred due to multilevel canal stenosis and significant myelopathy.

Instrumentation accompanying posterior laminectomy has been shown to contribute to the maintenance of cervical stability and the prevention of post-laminectomy kyphosis, particularly in the presence of congenital anomalies. Kyphotic deformity is a well-documented consequence of multilevel cervical laminectomy

performed without supplementary fixation, and the addition of posterior instrumentation is therefore considered beneficial when multilevel decompression is performed, particularly in the presence of pre-existing instability (12-15). In the present case, cervical lordosis measured by the C3-C7 Cobb method decreased from 13.6 degrees preoperatively to 1.5 degrees in the early postoperative period. It showed no further decline by six months (1.9 degrees), and the instrumented segment did not progress to kyphosis. We therefore interpret the construct as having arrested, rather than prevented, the loss of lordosis that accompanied the decompression. No implant-related complications were observed during the 18-month follow-up period.

This report has several limitations. Sagittal alignment was assessed on supine CT rather than on standing lateral radiographs, which are the appropriate modality for measuring cervical Cobb angles. Postoperative standing radiographs were not available for this patient, so the reported angles should be read as a within-modality comparison and may not reflect standing alignment. The radiological follow-up is also uneven: CT extends to six months and MRI to 18 months, so the alignment data end at six months while the clinical and MRI follow-up extends to 18 months. Finally, the study is a single case, and the construct described here should not be generalized without further reports.

In summary, DCM developing on a background of KFS can be treated effectively by posterior decompression and instrumented stabilization. Coexisting congenital anomalies may determine which fixation points are available and should be identified on preoperative CT.

Ethics

Informed Consent: Written informed consent was obtained from the patient for this study.

Footnotes

Conflict of Interest: The authors declared no conflict of interest.

Author Contributions: Surgical and Medical Practices: A.T.A., Concept: B.G., A.B., İ.G.A., H.K., A.T.A., Design: B.G., A.B., İ.G.A., H.K., A.T.A., Data Collection or Processing: B.G., A.B., İ.G.A., H.K., A.T.A., Analysis and/or Interpretation: B.G., A.B., İ.G.A., H.K., A.T.A., Literature Search: B.G., A.B., İ.G.A., H.K., A.T.A., Writing: B.G., A.B., İ.G.A., H.K., A.T.A.

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