Fracture-Dislocation of the Cervical Spine Secondary to Low-Impact Trauma in a Patient with Klippel-Feil Syndrome: A Case Report

Yusuke Dodo, Yoshifumi Kudo, Koji Ishikawa, Ryo Yamamura, Haruka Emori, Hiroshi Maruyama, Akira Matsuoka, Katsunori Inagaki and Tomoaki Toyone

Department of Orthopaedic Surgery, Showa University School of Medicine, Tokyo, Japan

Keywords:
Klippel-Feil syndrome (KFS), Fracture-dislocation, Cervical spine, Low impact trauma

Spine Surg Relat Res 2020; 4(1): 84-86
dx.doi.org/10.22603/ssrr.2019-0007

Klippel-Feil syndrome (KFS) is characterized by abnormal fusion of two or more vertebra in the cervical spine, which creates the characteristic appearance of a short neck with resulting facial asymmetry, low hairline, and limited neck mobility. Recently, a Meox-1-deficient medaka was developed as a KFS model, leading to developments in the understanding of this congenital disease[1]. Here, we report a case of cervical spine fracture-dislocation in a patient with KFS secondary to low-impact trauma, which ultimately required cervical posterior decompression and fusion surgery.

A 38-year-old female with a history of scoliosis and cleft lip and palate was transferred to our facility with severe neck pain, left shoulder paralysis, and numbness of the left thumb and index finger. The symptoms occurred during a yoga exercise, when she flexed her neck while putting her weight on her forehead. She suddenly heard a crunching sound with the onset of symptoms.

Physical examination indicated severe posterior midline neck tenderness with limited range of motion. On further evaluation, she did not have a low hairline, dysphagia, or ataxia. Radiography and non-contrast computed tomography (CT) of the cervical spine showed a C4/5 fracture-dislocation (Fig. 1, 2). CT showed evidence of congenital fusion of C5 through T1, which could have been a predisposing factor to the patient's injury. Magnetic resonance imaging of the cervical spine revealed C5 nerve root compression, which explained her left upper extremity paralysis (Fig. 3).

Based on these findings, the main concern was cervical spine nerve compression due to the dislocation and it was decided to perform urgent surgery using the posterior approach. In the operating room, she underwent bilateral facet joint resection and open reduction with partial laminectomy of C4/5. After checking the C5 nerve root, fusion surgery was performed from C3 to T1. Owing to the anomalous shape of the vertebrae, the operative repair was technically challenging and standard pedicle screws could not be used. We instead used anchoring devices for the anomaly part of vertebrae, including lamina screws and hooks, to ensure stability. During the inpatient recovery period, her neck pain was greatly relieved from the first postoperative day, and she was able to start ambulation with a neck collar. Additionally, her neurological deficits, including left shoulder paralysis and numbness of the left thumb and index finger, fully recovered. Postoperatively, she was doing well, and follow-up performed at 1 year revealed bone union at the neck fusion site.

KFS is characterized by congenital union of the cervical vertebrae, which can predispose patients to adjacent vertebral body instability[2]. Mishima K et al. previously reported on a patient with KFS with a fracture through fused cervical segments following trauma[3]. Additionally, there have been other case reports on spinal cord injury or other complications in patients with KFS[4,5]. However, to our knowledge, this is the first case report describing a patient with KFS requiring instrumentation surgery for cervical fracture-dislocation.

The potential instability of C4/5 in this case is attributable to the abnormal vertebral union between C5 and T1, which resulted in cervical dislocation with only slight force. Spine

Corresponding author: Tomoaki Toyone, tomotomot2001@gmail.com
Received: March 31, 2019, Accepted: June 11, 2019, Advance Publication: August 16, 2019

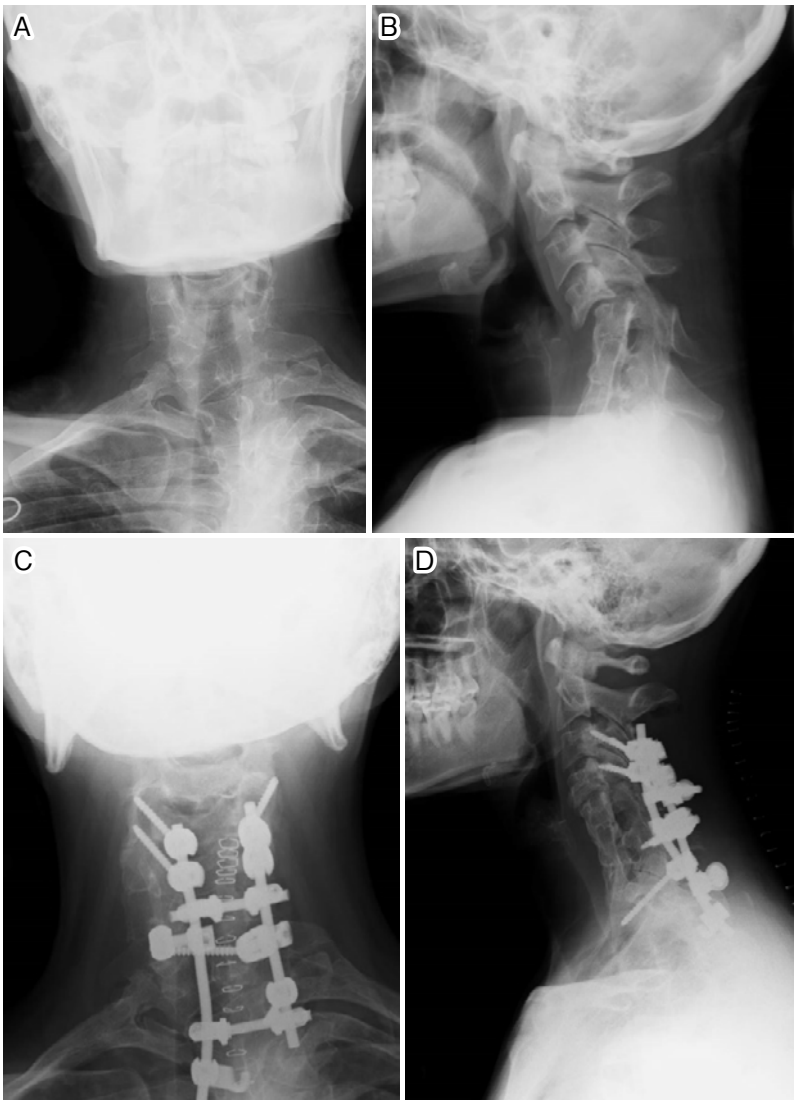

Figure 1. Preoperative Cervical Spine Radiography
A: Anteroposterior view; B: Lateral view
C4/5 shows anterior dislocation, and C5-7 shows vertebral body fusion
C: Anteroposterior view, D: Lateral view

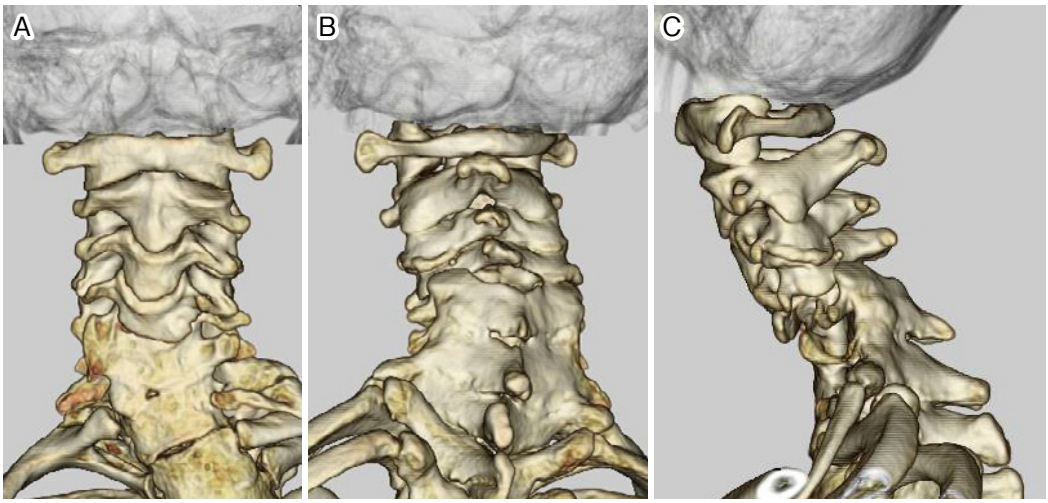

Figure 2. Three-dimensional Computed Tomography (3DCT)
A. C5-7 shows vertebral body fusion on 3D-CT (anterior view)
B. C5-T1 shows abnormal vertebral union on 3D-CT (posterior view)
C. Cervical vertebrae dislocated fracture is seen at C4/5 (sagittal view)

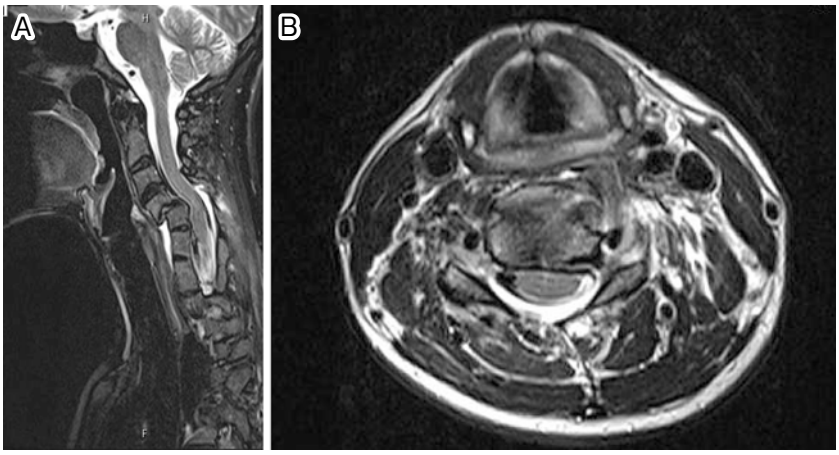

Figure 3. Preoperative Cervical Spine Magnetic Resonance Imaging (MRI)
A: MRI shows C4/5 anterior dislocation on sagittal view
B: MRI shows left C5 nerve root compression on axial view

surgeons should have a high index of suspicion for severe injury with a history of low-impact trauma in the setting of vertebral anomalies, as in the present case. Intraoperatively, care should be taken when there are differences in vertebral shape, which could complicate the technical aspect of screw insertion. A wide variety of instrumentation devices, such as lamina screws and hooks, might be effective. Moreover, although not utilized in this case, navigation system might be helpful in surgical management.

In conclusion, we reported a rare case of cervical spine fracture-dislocation in a patient with KFS secondary to low-impact trauma, which was successfully treated with cervical posterior decompression and fusion surgery.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Author Contributions: Yusuke Dodo wrote and prepared the manuscript, and all the authors participated in creating the study design. All authors have read, reviewed, and approved the article.

References

1. Dauer MVP, Currie PD, Berger J. Skeletal malformations of Meox 1-deficient zebrafish resemble human Klippel-Feil syndrome. J Anat. 2018;233(6):687-95.
2. Menger RP, Notarianni C. Klippel Feil syndrome. StatPearls [Internet]. 2019 Jan. [cited 2019 Jan 21]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK493157/
3. Mishima K, Itoi A, Sugita M, et al. A case of fracture through fused cervical segments following trauma in a patient with Klippel-Feil syndrome. J Emerg Trauma Shock. 2016;9(2):85-6.
4. Ogihara N, Takahashi J, Hirabayashi H, et al. Surgical treatment of Klippel-Feil syndrome with basilar invagination. Eur Spine J. 2013;22(Suppl 3):S380-7.
5. Futane S, Salunke P. Klippel-Feil syndrome with atlanto-axial dislocation, anomalous vertebral artery, dextrocardia and situs inversus. Clin Neurol Neurosurg. 2013;115(10):2304-6.