Ossified longitudinal ligament causing same level cord contusion in a case of Klippel-Feil syndrome

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Amit Agrawal
Professor of Neurosurgery, Department of Neurosurgery, Narayna Medical College Hospital, Nellore, (AP), INDIA

ABSTRACT
Klippel-Feil syndrome (KFS) is a congenital fusion of two or more cervical vertebrae due to faulty segmentation of the vertebral axis during gestation. (1-5) These patients present with a constellation of manifestations and are typically prone to cervical cord injury after a minor fall or a major traumatic episode. (2, 5-8) 34 years old gentleman, a plumber by profession presented with a history of slipped and fall about two stairs height while he was working.

LETTER TO THE EDITOR
Dear Sir,

Klippel-Feil syndrome (KFS) is a congenital fusion of two or more cervical vertebrae due to faulty segmentation of the vertebral axis during gestation. (1-5) These patients present with a constellation of manifestations and are typically prone to cervical cord injury after a minor fall or a major traumatic episode. (2, 5-8) 34 years old gentleman, a plumber by profession presented with a history of slipped and fall about two stairs height while he was working. Following that he had transient loss of consciousness when he regained the consciousness, he realized that he was not able to move his all four limbs. He also could not pass urine. There was no history of vomiting, ENT bleed or convulsion. At the time of admission, he was conscious, oriented to time place and person. His general examination was normal. He had breathing difficulty and the respiration was of diaphragmatic type. Chest wall expansion was absent. Single breath count was 5. Neurologically he was conscious, alert and oriented. Cranial nerves were normal. Motor system examination revealed quadripareisis with muscle power grade 3/5 in shoulders, 2/5 at elbow, 1/5 at wrist and absent grip in both upper limbs. All the deep tendon jerks were sluggish. The patient was catheterized as he was not able to pass urine. All the sensations were reduced below D8 level. Bilateral planters were extensor. The gag and palate reflexes were normal. X-ray films of the
cervical spine showed fusion of the spinous processes of cervical vertebrae at C2 and C3 vertebrae with ossification of posterior longitudinal ligament at the same level (Figure-1). Magnetic resonance imaging (MRI) of the cervical spine showed congenital fusion of the same vertebral spinous processes and in addition there was ossified posterior longitudinal ligament with canal stenosis and cord contusion (Figure-2). The patient received injection Methylprednisolone as per the standard protocol and planned for surgical decompression at the earliest. The patient underwent posterior approach and C2 and C3 laminectomy to relieve the compression of the cord. The patient made gradual recovery. At follow-up the power in all four limbs was improved to grade 4/5.

In cases of Klippel-Feil syndrome fusion of one segment of the spinal column causes hypermobility of the non-fused adjoining segments thus cervical spine is unable to compensate for excessive flexion, extension, rotation and lateral bending and predisposing disc prolapse at adjacent levels and neurological deterioration following even minor trauma. (2, 4, 7-13) Central cord syndrome typically described in the elderly patients with preexisting cervical spine spondylosis, only few reports discuss in patients with Klippel-Feil syndrome with cord compression at adjacent level. (9, 11, 14, 15) In contrary to many other cases where the patients with Klippel-Feil anomaly presented with adjacent level disc prolapsed and instability, the present case was unique as he developed spinal cord injury due to canal compromise at the level of used segment because of ossified posterior longitudinal ligament leading to canal stenosis. X-ray cervical spine is useful to understand the bony anomalies, however as in present case the MRI will better delineate the normal as well pathological spinal cord anatomy including disc prolapsed and cord contusion. (6, 9) Treatment regimens depend on the severity of symptoms and the segmental instability or cord compression and vary from modification of activities to extensive spinal surgery including microsurgical removal of the herniated disc with instrumentation. (6, 10, 16-19) However, in presented case the extensive ossification in the posterior ligament was cause of canal compromise and cord compression and, as there was no instability, we opted for posterior decompression alone.

REFERENCES
5. Samartzis D, Kalluri P, Herman J, Lubicky JP, Shen FH.
Amit Agrawal


