ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXV | No. 1 March 2021

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DOI: 10.33962/roneuro-2021-019

Romanian Neurosurgery (2021) XXXV (1): pp. 114-116
DOI: 10.33962/roneuro-2021-019
www.journals.lapub.co.uk/index.php/roneurosurgery

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

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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 manifes-tations and are typically prone to cervical cord injury after a minor fall or a major traumatic episode. (2, 5-8) 34 years old gentlemen, a plumber by profession presented with a history of slipped and fall about two stairs height while he was working.

Keywords
Klippel-Feil, cervical, spinal cord, cord contusion

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 gentlemen, a plumber by profession presented with 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 quadriparesis 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

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ISSN online 2344-4959
© Romanian Society of Neurosurgery

First published
March 2021 by
London Academic Publishing
www.lapub.co.uk

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 relive 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.

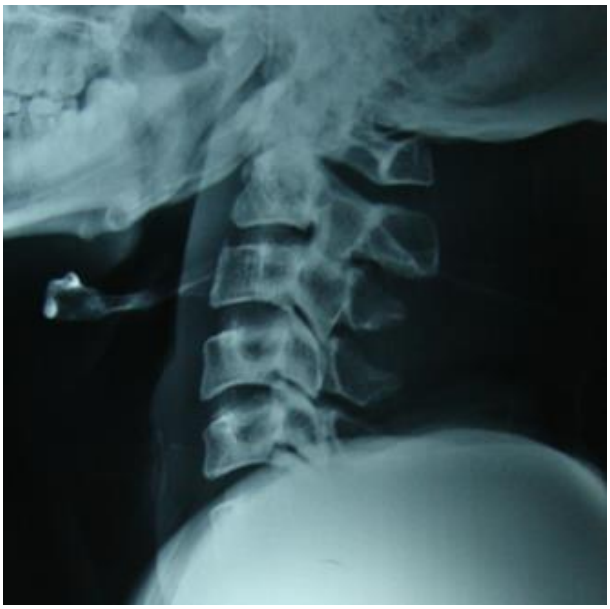

Figure 1. a - Lateral x-rays of the cervical spine showing fusion of the spinous processes of C2-3 vertebral body, also note the ossified posterior longitudinal ligament bridging the posterior border of the C2-3 vertebral bodies.

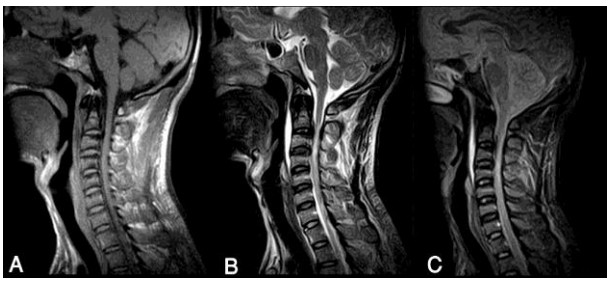

Figure 2. MRI cervical T1, T2 and FLAIR sagittal images showing calcified posterior longitudinal ligament behind C2-3-disc space with cord compression and cord signal changes.

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

1. Ulmer JL, Elster AD, Ginsberg LE, Williams DW, 3rd. Klippel-Feil syndrome: CT and MR of acquired and congenital abnormalities of cervical spine and cord. J Comput Assist Tomogr. 1993 Mar-Apr;17(2):215-24.
2. Guille JT, Miller A, Bowen JR, Forlin E, Caro PA. The natural history of Klippel-Feil syndrome: clinical, roentgenographic, and magnetic resonance imaging findings at adulthood. Journal of pediatric orthopedics. 1995;15(5):617.
3. Klippel M, Feil A. Un cas d'absence des vertebres cervicales. Avec cage thoracique remontant jusqu'a la base du crane (cage thoracique cervicale). Nouv Iconog Salpetriere. 1912;25:223-50.
4. Vaidyanathan S, Hughes PL, Soni BM, Singh G, Sett P. Klippel-Feil syndrome - the risk of cervical spinal cord injury: a case report. BMC Fam Pract. 2002 Apr 11;3:6.
5. Samartzis D, Kalluri P, Herman J, Lubicky JP, Shen FH.

Superior odontoid migration in the Klippel-Feil patient. Eur Spine J. 2007 Sep;16(9):1489-97.
6. Agrawal A, Badve AM, Swarnkar N, Sarda K. Disc prolapse and cord contusion in a case of Klippel-Feil syndrome following minor trauma. Indian J Orthop. 2009 Apr;43(2):210-2.
7. Karasick D, Schweitzer ME, Vaccaro AR. The traumatized cervical spine in Klippel-Feil syndrome: imaging features. AJR Am J Roentgenol. 1998 Jan;170(1):85-8.
8. Pizzutillo PD, Woods M, Nicholson L, MacEwen GD. Risk factors in Klippel-Feil syndrome. Spine (Phila Pa 1976). 1994 Sep 15;19(18):2110-6.
9. Adeleye AO, Akinyemi RO. Cervical Klippel-Feil syndrome predisposing an elderly African man to central cord myelopathy following minor trauma. Afr Health Sci. 2010 Sep;10(3):302-4.
10. Samartzis D, Lubicky JP, Herman J, Kalluri P, Shen FH. Symptomatic cervical disc herniation in a pediatric Klippel-Feil patient: the risk of neural injury associated with extensive congenitally fused vertebrae and a hypermobile segment. Spine (Phila Pa 1976). 2006 May 15;31(11):E335-8.
11. Strax TE, Baran E. Traumatic quadriplegia associated with Klippel-Feil syndrome: discussion and case reports. Arch Phys Med Rehabil. 1975 Aug;56(8):363-5.
12. Elster AD. Quadriplegia after minor trauma in the Klippel-Feil syndrome. A case report and review of the literature. J Bone Joint Surg Am. 1984 Dec;66(9):1473-4.
13. O'Donnel DP, Seupaul RA. Klippel-Feil syndrome. Am J Emerg Med. 2008 Feb;26(2):252 e1-2.
14. Matsumoto K, Wakahara K, Sumi H, Shimizu K. Central cord syndrome in patients with Klippel-Feil syndrome resulting from winter sports: report of 3 cases. Am J Sports Med. 2006 Oct;34(10):1685-9.
15. Shirasaki N, Okada K, Oka S. Cervical myelopathy in patients with congenital cervical block vertebrae. Eur Spine J. 1993 Jun;2(1):46-50.
16. Tracy MR, Dormans JP, Kusumi K. Klippel-Feil syndrome: clinical features and current understanding of etiology. Clin Orthop Relat Res. 2004 Jul(424):183-90.
17. Neroni M, Gazzeri R, Galarza M, Alfieri A. Intradural cervical disc herniation in a Klippel-Feil patient. Spine (Phila Pa 1976). 2007 Oct 1;32(21):E608-10.
18. Leung CH, Ma WK, Poon WS. Bryan artificial cervical disc arthroplasty in a patient with Klippel-Feil syndrome. Hong Kong Med J. 2007 Oct;13(5):399-402.
19. Yi S, Kim SH, Shin HC, Kim KN, Yoon DH. Cervical arthroplasty in a patient with Klippel-Feil syndrome. Acta Neurochir (Wien). 2007 Aug;149(8):805-9; discussion 9.