Dear Editor,

The hallmark feature of Klippel-Feil syndrome (KPS) is congenitally fused cervical segments as a result of improper segmentation along the growing axis of the embryo. Patients may have neurological deterioration either spontaneously or even after minor trauma. Instability at the junction of the fused and mobile segments has been thought to predispose to neurological symptoms.

We provide the second case concerning a fracture of fused bones in a patient with KFS.

An 80-year-old male was admitted to our hospital following a fall accident which had occurred while he was carrying a log. On arrival, he was in a comatose state (Glasgow Coma scale score - 7) so that the evaluation of his paralysis was initially difficult. He developed respiratory failure with abdominal breathing requiring intubation and ventilation. A computed tomography scan showed an isolated fracture through C5 to C7 fused cervical segments with Os odontoideum at C2 and a unilateral hypoplastic kidney. Magnetic resonance imaging showed two high-intensity areas at C4 and C6 of the cervical spinal cord [Figure 1]. After he underwent anterior and posterior interbody fusion, he had grade 3/5 power in both upper limbs and grade 1/5 power in both lower limbs with sensory loss below C5. He required a temporary pace-maker for treatment of severe bradycardia that resulted in a few episodes of cardiac arrest. After treatment for the respiratory failure and neurogenic shock, he remained tetraparesis and was transferred to other hospital for rehabilitation.

This is the second reported case of a fracture through a Klippel-Feil fused segment resulting in neurological injury. This case also had os odontoideum and a unilateral hypoplastic kidney. According to a classification system for the KFS proposed by Samartzis et al., three broad categories have been described, Type 1, Single congenitally fused segment; Type 2, multiple noncontiguous segments; and Type 3, multiple contiguous segments. Al-Tamimi et al. and our case would be classified as Type 3 which is associated with a higher incidence and risk of neurological deterioration. Even the fractures occurred within one or two levels from the previous fused segment, congenital fused cervical segments are also at risk of fracture following trauma. Patients diagnosed with KPS should be counseled about contact sports and working environment as even minor cervical trauma can have disastrous consequences.

The os odontoideum may occur due to the presence of transitory intervertebral disk or as a result of increased movements at the time of ossification of the dens. As the presence of fusion of cervical and/or other vertebrae along with the os odontoideum strongly pointed toward KFS, the os odontoideum in our case was judged as complication of KFS.

There is high incidence of congenital anomalies of the genito-urinary tract in patients with KPS. A unilateral renal agenesis is the most common. Accordingly, the unilateral hypoplastic kidney in our case might be complication of KFS.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Kentaro Mishima, Akira Itoi¹, Manabu Sugita², Youichi Yanagawa
Mishima K, Itoi A, Sugita M, Yanagawa Y. A case of fracture through fused cervical segments following trauma in a patient with Klippel-Feil syndrome. J Emerg Trauma Shock 2016;9:85-6.

Access this article online

Quick Response Code:   Website:   www.onlinejets.org
DOI:   10.4103/0974-2700.179455

How to cite this article: Mishima K, Itoi A, Sugita M, Yanagawa Y. A case of fracture through fused cervical segments following trauma in a patient with Klippel-Feil syndrome. J Emerg Trauma Shock 2016;9:85-6.

Received: 23.11.15. Accepted: 17.12.15.

REFERENCES

1. Al-Tamimi YZ, Sinha P, Ivanov M, Robson C, Goomany A, Timothy J. Fracture through fused cervical segments following trauma in a patient with Klippel-Feil syndrome. Br J Neurosurg 2014;28:408-10.

2. Samartzis DD, Herman J, Lubicky JP, Shen FH. Classification of congenitally fused cervical patterns in Klippel-Feil patients: epidemiology and role in the development of cervical spine-related symptoms. Spine (Phila Pa 1976) 2006;31:E798-804.

3. Mac Millan M, Stauffer ES. Traumatic instability in the previously fused cervical spine. J Spinal Disord 1991;4:449-54.

4. Begum S, Roy T, Ghosal Sen T, Gupta I. A rare presentation of os odontoideum with multiple vertebral fusion in type III Klippel-Feil syndrome (KFS) – A case report. J Clin Diagn Res 2014;8:AD03-5.

5. 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;3:6.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.