Case Report

Cervical Klippel-Feil syndrome predisposing an elderly African man to central cord myelopathy following minor trauma

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Abstract

An otherwise-healthy, active 83-year-old Nigerian man developed reversible central cord myelopathy from a mild fall on a level surface. Cervical spine magnetic resonance imaging (MRI) revealed C5, 6, and 7 block vertebrae and marked disc extrusions only at the immediately adjoining upper and lower non-fused segments of the cervical spine. There was no spinal canal stenosis otherwise. We think that the unique presentation of this case of Klippel-Feil syndrome further supports the impression that following fusion (congenital or acquired) of one segment of the spinal column, hypermobility of the non-fused adjoining segments may strongly predispose to more disc extrusions.

Key Words: Klippel-Feil syndrome, Central cord syndrome, minor trauma

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Introduction

The Klippel-Feil syndrome is a multisystemic disorder in which there is congenital failure of cervical spine segmentation of varying degrees in association with congenital defects in other body systems like the cardiovascular, renal and so on[1]. It is thought to be due to faulty segmentation and resegmentation along the developing axis of the embryo during the 3 to 8 weeks of gestation[6]. Congenital cervical block vertebrae, or cervical spine synostosis, is the hallmark osseous element in this disorder and is sometimes referred to alone as Klippel-Feil syndrome[2,3] Though normally asymptomatic, it is known that there is an unusual occurrence, and extent, of myelopathy following minor trauma in people living with this anomaly[4]. Indeed, Strax and Baran (1975) were the first workers to suggest that patients with the Klippel-Feil syndrome might be at risk for sudden quadriplegia after minor trauma[2]. Other reports (5) later supported this view including the one on 32 patients aged 13 to 78 years by Shirasaki et al[4]. This latter study, amongst others, further showed that cervical myelopathy not only followed trivial trauma in people with Klippel-Feil syndrome, but also that the symptoms and signs of this myelopathy were usually attributed to spinal segments adjacent to the fusion. This report documents one such case in an 83-year-old Nigerian.

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Magnetic resonance imaging findings in the case of central cord syndrome from mild trauma in an elderly African who had background Klippel-Feil syndrome. Sagittal (A) T1W image and (B) T2W image showing the block vertebrae involving the C 5, 6, and 7 segments, adjoining discs extrusion, adequate thecal space away from the extruded discs even behind the block vertebrae.

Figure A

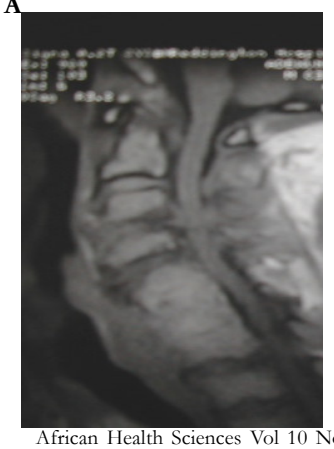

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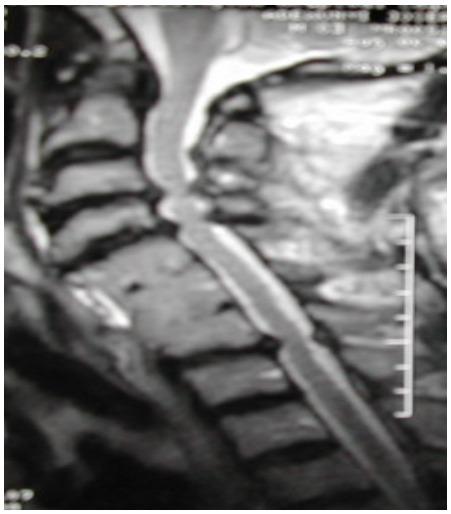
Figure B



Axial T2W images (C) at the level of C4/5 disc extrusion with markedly compromised thecal space and (D) at the level of the C5 vertebral body with well preserved thecal sac.

Figure C

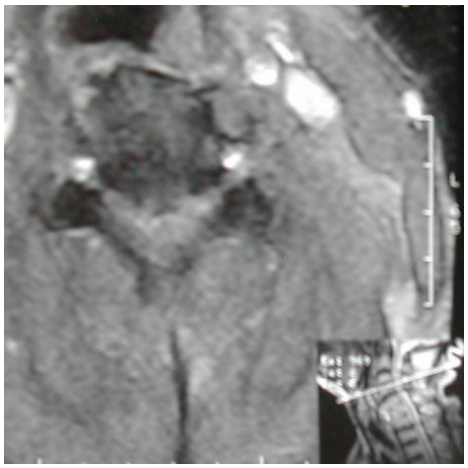
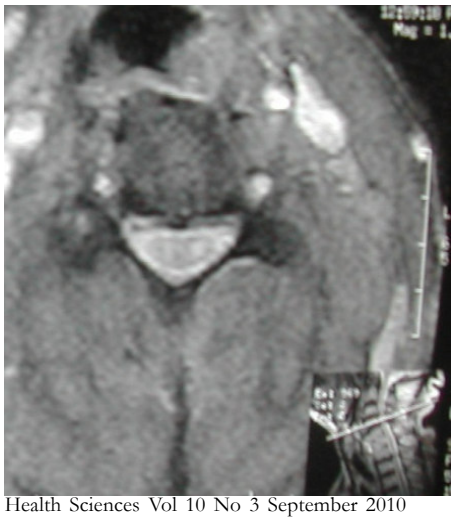


Figure D



Case Report

A previously healthy, active 83-year-old man presented in our clinic in July 2008 with a 3-week history of neck pain and weakness in all the four limbs. This was after a fall due to a slip on one foot whilst walking on a level surface. He landed on his fore-head, became completely paralysed in all 4 limbs and was bed bound for 3 days. All this while he had received in-patient medical attention which was not particularly directed at the cervical spine in a "private clinic". He denied history of sphincteric deficit. Power returned gradually; firstly to both lower limbs, and then more slowly to the upper limbs till the time of review in our unit.

His past medical history was not revealing of any other major co-morbidity. There was no history of hypertension, diabetes mellitus, or craniospinal surgery or trauma. There was no clinical evidence of urinary tract infection either. Preoperative electrocardiography and urine analysis amongst other operative workup revealed no significant abnormality.

Clinical examination revealed a generally healthy elderly man with normal general physical and neurological examination. He had difficulty walking even with a walker, appeared to be in some agony of neck pain but had his neck unsupported with any form of orthosis. There was a C3 hyper-reflexic quadriparesis worse in the upper limbs, the muscle power being 4/5 in all lower limb muscle groups, 4 minus in proximal and 3 in the distal upper limb muscle groups. The grip power was 3 and 2 on the left and right respectively. Sensory level was also C3 with only slight impairment down to the sacral dermatomes. The plantar response was absent bilaterally. Plain cervical spine roentgenologic study revealed only moderate spondylotic changes with no vertebral column malalignment.

We made a clinical diagnosis of post traumatic quadriparesis, probably a C3 central cord syndrome from background cervical spine spondylosis. Absence of fever or any other prodromal illness plus the absence of the typical distribution of sensory loss made this case to be unlikey to be due to Guillain-Barre syndrome. We therefore placed him on a hard, modified Philadelphia-type cervical collar and requested a cervical spine magnetic resonance imaging (MRI).

This revealed a Klippel-Feil (or block vertebrae) anomaly of the C5, 6 and 7, and posterior intervertebral disc extrusion of the adjoining C3/4, C4/5, and C7/ T1 interspaces worst at the C3/4 level. This is Type I Klippel-Feil anomaly using the classification of Klippel and Feil[3]. There was cord compression at the levels of the disc extrusions with

associated intramedullary signal changes worst at the C3/4 level. The rest of the thecal sac however appears well preserved even at the level of the block vertebrae. Neurophysiologic studies, including nerve conduction test and electromyography, could not be done in our practice setting.

Due to his good clinical status and his fairly advanced age, we chose to continue his management non-operatively on the neck collar, analgesia and physiotherapy. His neurological condition has continued to improve, 2 months now into his care. He has muscle power of at least 4/5 in all limbs' muscle groups including the left grip power; only on the right is his grip power only now 3/5.

Discussion

In February 2008, O'Donnel and Seupaul reported what they believed to be the first case of significant central cord impairment after moderate trauma in a patient with congenital cervical block vertebrae, or Klippel-Feil syndrome[6]. This was in a 19-year-old American. We here present another case of significant central cord myelopathy following minor trauma in a patient with block vertebrae C5, 6, and 7. This is in an 83-year-old Nigerian.

Some authorities believe that persons with the Klippel-Feil syndrome usually have associated spinal canal stenosis, and, hence, an increased likelihood to develop spinal cord injury even after minor trauma (7) whilst others hold contrary views[3]. Indeed, Strax and Baran (1975) were the first workers to suggest that patients with the Klippel-Feil syndrome might be at risk for sudden quadriplegia after minor trauma[2]. Other reports later supported this view including the one on 32 patients aged 13 to 78 years by Shirasaki et al[4]. However, this latter study, amongst others[2,3,5], further showed that the cervical myelopathy not only followed trivial trauma in people with Klippel-Feil syndrome, but also that the symptoms and signs of this myelopathy were usually attributed to spinal segments adjacent to the fusion. That is when trauma occurs it concentrates stress at the segment adjacent to fusion[4]. Magnetic resonance imaging, MRI, in some of their patients actually showed the cause of the cord lesions at the segments adjacent to the fusion to be the presence of cord compression by extradural causes like disc extrusion. The MRI showed the thecal sac to be well preserved in our case: not only in the region of the block vertebrae, but even at the levels of the vertebral bodies with the disc extrusion (figure 1 D). The thecal sac compromise and the cord injury appear therefore to be solely due to the extruded discs.

Central cord syndrome typically occurs in the elderly with preexisting cervical spine spondylosis and Matsumoto et al have reported 3 such cases in patients with Klippel-Feil syndrome following significant blunt force[8]. Our patient's central cord myelopathy occurred only after minor trauma. Although this event itself could not be said to be improbable in an 83-year-old man, we think the background block vertebrae and their after-effects on the adjoining vertebral segments (that is, disc extrusion from the hypermobility) greatly predisposed this man to this cord injury[2,3,4]. What is more, the series of Shirasaki et al, (1993) also included patients as old as 78 years.

Conclusion

There are a few clinical practice lessons in the unique presentation of this case. Firstly we think that the unique presentation of this case of Klippel-Feil syndrome further supports the impression that following fusion (congenital or acquired) of one segment of the spinal column, hypermobility of the non-fused adjoining segments may strongly predispose to more disc extrusions. Secondly, a high index of suspicion should be maintained always for more serious intraspinal lesions when severe limb weakness follows minor body trauma in any patient of any age group. Such should prompt quick neurology or neurosurgical referral for thorough neurologic evaluation and spinal computerized imaging.

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