CASE REPORT

SURGICAL MANAGEMENT OF IRREDUCIBLE ATLANTO-AXIAL DISLOCATION WITH OS ODONTOIDEUM AND KLIPPEL-FEIL SYNDROME

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Klippel-Feil syndrome (KFS) is the congenital fusion of two or more cervical vertebrae which is often associated with various other abnormalities in the cervical spine. Involvement the upper cervical segments leads to atlanto-axial instability which manifests as progressive neurological symptoms due to compression on the spinal cord. These cases pose a surgical challenge due the abnormal and unique anatomy of each patient. A 37-year-old patient presented with neck pain and cervical myelopathy due to a posterior subluxation of C2-3 fused segment over C4-6 fused segment. The patient had an os odontoideum, incomplete posterior arch of C1, anomalous course of vertebral artery and C3 hemi-vertebra. The patient was successfully managed with transoral odontoidectomy and occipeto-cervical fusion. Irreducible atlanto-axial dislocation in a patient with an abnormal upper cervical spine anatomy may require transoral decompression followed by posterior fusion.

Keywords: Klippel-Feil syndrome; Atlantoaxial instability; AAD, Transoral surgery; Odontoidectomy

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INTRODUCTION

Klippel-Feil syndrome (KFS) was classically described in patients with a triad of short neck, low posterior hairline and restricted range of cervical mobility.[1] It is the result of normal segmentation failure in the cervical vertebrae during the early weeks of fetal development and frequently accompanies other cranial, cardiopulmonary and skeletal deformities.[2] Studies have found that less than half of these patients present with the classic triad.[3] Currently, patients with congenital fusion of any two of the seven cervical vertebrae are recognized as KFS.

Fusion of cervical vertebra at different levels leads to various degrees of instability in the cervical spine. As reported in a case series, C2-C3 fusion is frequently associated with atlanto-axial dislocation (AAD) and compression of cervical spinal cord.[4] These cases always pose a therapeutic challenge because of the abnormal anatomy of the spine. We present a 37-year-old patient with KFS presenting with cervical myelopathy due to with non-traumatic posterior subluxation of C2-3 fused segment over C4-6 fused segment. The patient had an os odontoideum, incomplete posterior arch of C1, anomalous course of vertebral artery and C3 hemi-vertebra.

CASE REPORT

The subject was a 37-year-old male, a security guard by occupation. The patient had switched many jobs mostly related to manual labor. He presented to our unit with a history of neck pain for the last 8 years that had increased in the last 6 months with new onset weakness in the right upper extremity, numbness in both upper extremities and gait disturbance. The patient had only used pain medications and had not sought specialist opinion. His main concern was being not able to do his job since worsening of these symptoms in the last 6 months.

On examination the patient had short neck and stature, low occipital hair line, mild scoliosis in dorsal spine and pectus excavatum. Neurological findings included unsteady gait, clumsiness of the hands and a positive Hoffman's sign. Other findings included fine-movement impairment, hyperreflexia, decreased sensation of bilateral forearms and hands more on the right side. The score according to the modified Japanese Orthopedic Association scoring system (JOA score) was 13.[5] Preoperative physical exam and routine investigations did not reveal any systemic abnormality.

CT cervical spine showed fusion of vertebral bodies and posterior elements of C2-C3 and C4-C5-C6. There was marked posterior displacement of C2-C3 fused vertebrae causing spinal canal narrowing. Additionally, the body of C3 was deficient on one side as well as the posterior arch of C1. An os odontoideum was visible on the CT scan. The C1-C2 facet joints were almost vertically inclined and C2-C3 fused segment had very thin pedicles and lateral masses. (Figure-1) Severe degenerative changes were visible at many levels. Extension radiographs did not show reduction of AAD. (Figure-2) MRI revealed severe cord compression with signal changes on T2 weighted images. (Figure 3)

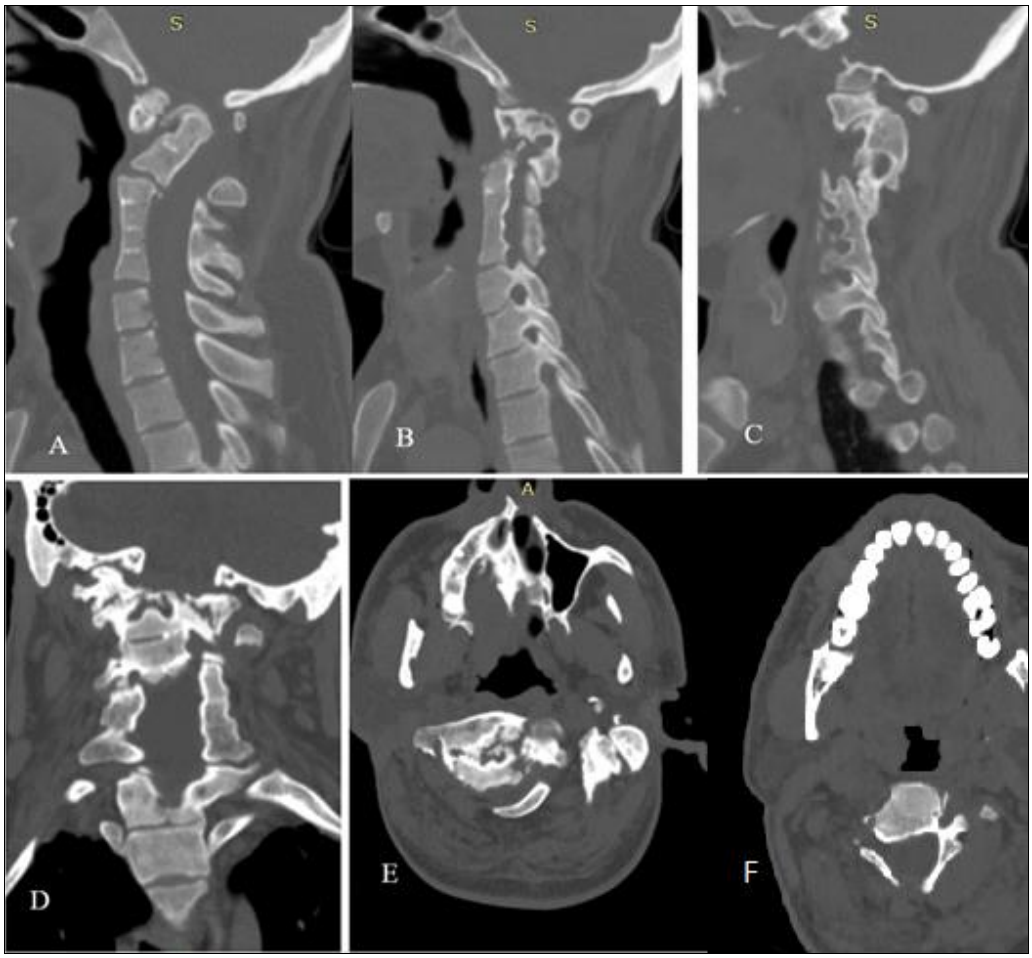

Figure 1: CT scan showing (A) posterior displacement of fused C2-C3 over C4-C6 segment. Os odontoideum is visible above anterior C1 arch. (B), (C) shows the degeneration and vertical orientation of right and left C1-C2 facet joints. (D), (E) shows coronal and axial view of C1-C2 anatomy. (F) shows thin C3 pedicles and lateral mass.

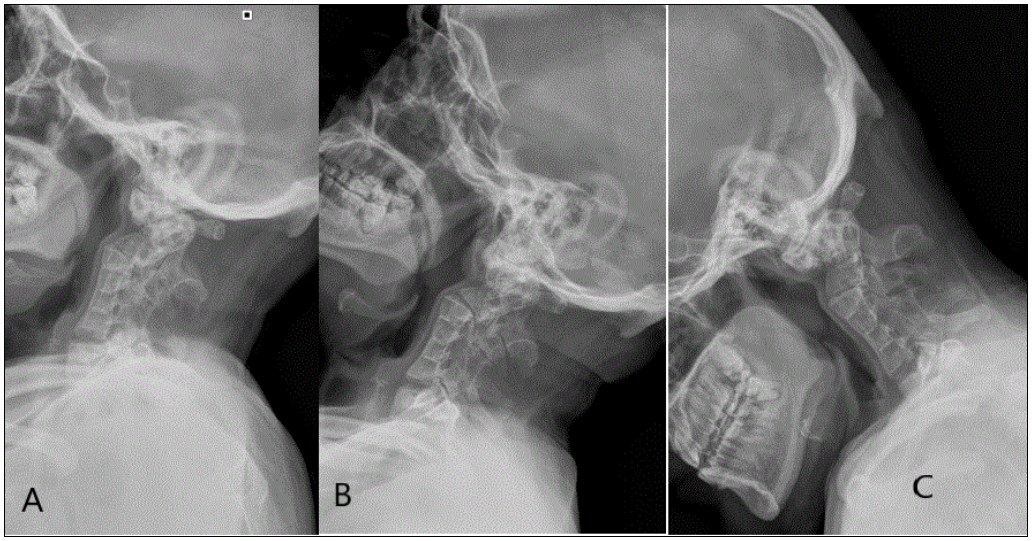

Figure-2: Lateral x-rays in (A) neutral, (B) extension and (C) flexion positions showing irreducibility of the AAD.
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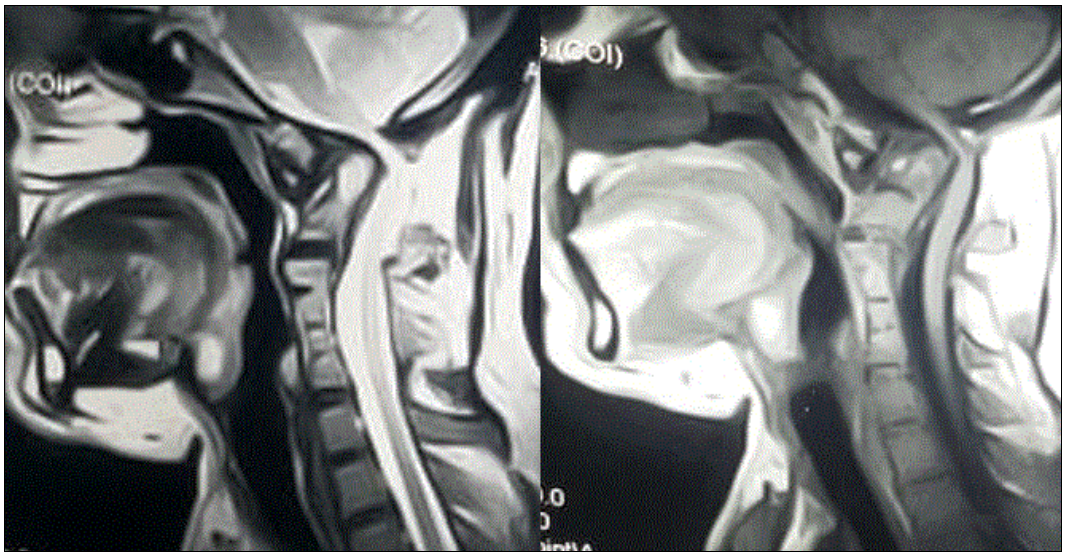

Figure-3: T2 and T1 weighted MRI shows significant compression of the spinal cord.

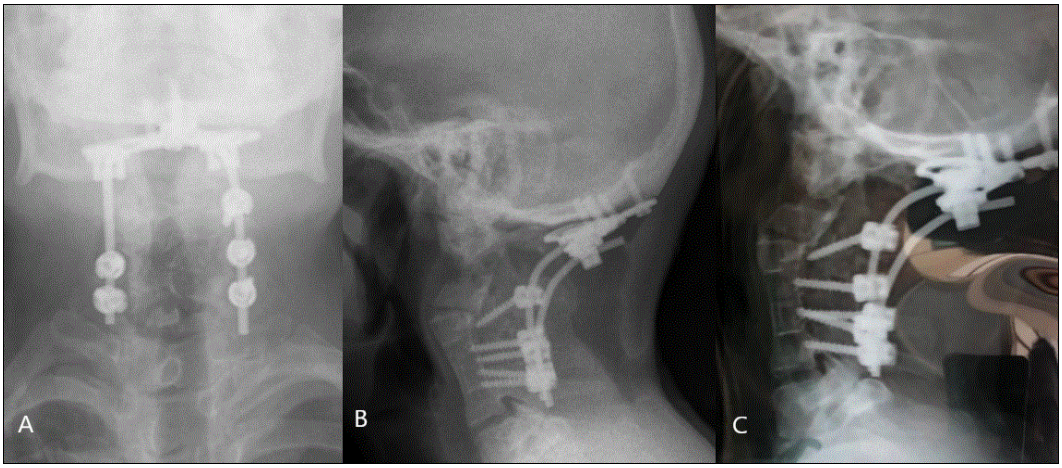

Figure-4: (A), (B) shows post-operative AP and lateral view of cervical spine. (C) radiograph taken at 3 months follow-up shows signs of fusion. Note: These x-rays have been photographed from films which may explain some visual distortion. The patients radiological and clinical status, however, was deemed satisfactory.

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Surgery was performed in two stages in the same setting. Initially the patient was positioned supine and intubated with fiberoptic guidance. A transoral odontoidectomy was performed along with C1 anterior arch removal, to relive the spinal cord. After adequate decompression the patient was shifted to prone position and occipetocervical fusion was performed. The construct consisted of an occipital plate, a single left sided reverse transarticular C2-C3 screw and lateral mass screws on C4 and C5. (Figure-4) A Nasogastric tube was left in place which was removed on third post-operative day. The patient was kept in neurosurgery HDU for three days. Daily examinations showed uneventful recovery.

A total follow-up of 6 months was completed. MJOA at final follow-up was 16. Follow-up x-rays showed adequate decompression maintained and early signs of fusion. (Figure-4)

DISCUSSION

Atlanto-axial dislocation is divided into traumatic, congenital and pathologic.[6] Atlanto-axial dislocation of congenital etiology is frequently accompanied by complex disorders of cranio-cervical junction such as KFS, occiput-C1 assimilation, abnormal dens and basilar invagination.[7] The presentation in this category of patients is usually with history of an insignificant trauma which may not be remembered by the patient like in this case report. Symptoms include neck pain, limitation of range of motion and long tract signs. No defined guidelines exist for the management of such patients and treatment is

individualized for each patient according to their symptoms, anatomy and degree of pathology. The overall principles of management are achieving adequate decompression and stability.[8,9]

The common cervical spine abnormalities associated with KFS are Atlas assimilation, C2-C3 fusion, contiguous or noncontiguous fusion of many levels, hemivertebra, basilar invagination, Arnold-Chiari malformation and syringomyelia.[4] The normal level adjacent to fused levels become hypermobile which results in early degeneration. Patients with atlas assimilation and/or C2-C3 fusion have a high rate of AAD. This can occur in anteroposterior plane noted by increase anterior atlanto-dental interval or vertical plane (basilar invagination) or a combination of both.[7] Anyone or combination of the above may usually present with neck pain and progressive compressive myelopathy. Our patient had fusion between C2-C3 and C4-C5-C6. This caused instability and degeneration at C2-C3 joint which finally led to subluxation of the C2-C3 segment over the lower fused block.

Klippel-Feil syndrome can have the involvement of cardiac, genitourinary and other systems. These need to be ruled out or managed before undertaking surgery. Fortunately, our patient had normal preoperative systemic examination and routine investigations. Due to the abnormal cervical spine anatomy of these patients, surgical intervention poses a challenge to the surgeon. The high rate of atlas assimilation associated with reduced volume makes the placement of screws difficult. Additionally, C2-C3 fusion leads to loss of normal bony landmarks. More than one third of C2-C3 fusion don't have suitable pedicle for screw placement.[10] In our patient none of C2 and C3 pedicles had enough diameter to hold screws. These bony abnormalities together with a very high rate of vertebral artery anomaly[11] makes surgical management extremely complex.

Atlanto-axial dislocation is divided into reducible, irreducible and non-reducible. Achievement of full alignment with extension or traction used to be the dividing line between two categories of treatment. Dislocations that reduced fully were managed with posterior fusion only and those which did not reduce required anterior decompression together with some form of posterior fusion.[6,12] Surgical management of AAD has changed significantly during the last two decades. Currently, the majority of both reducible and irreducible forms are managed via a direct posterior approach.[8,13] Anterior decompression, however, still has valid indications primarily as a salvage procedure in case of incomplete or non-reduction after posterior manipulation or in cases where posterior reduction only is not considered feasible or safe.[14,15] In our case, the vertical inclination of joints was anticipated to make facet distraction extremely difficult even with extensive joint remodeling. Additionally, the absence of a thick enough pedicle or lateral mass in the C2-C3 fused segment meant that posterior only approaches could not be useful. A decision was, therefore, made to perform transoral decompression and occipetocervical fusion.

Transoral, endoscopic trans-nasal or submandibular approaches are also used for removal of odontoid process and anterior arch of atlas for anterior decompression. In addition, anterior approach is used for odontoid release procedure in selected cases followed by anterior fixation via Transoral anterior reduction and fixation with plate (TARP) or anterior transarticular screw. Alternatively, a posterior fixation follows anterior release or anterior decompression.[9] The posterior construct is dictated by patient's anatomy in cases like occipetalization of atlas and aberrant vertebral artery and inadequate bony support for screws.

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