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Case Report

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Traumatic Hemiparesis Associated with Type III Klippel-Feil Syndrome

Klippel-Feil Syndrome (KFS) is a complex congenital syndrome of osseous and visceral anomalies. It is mainly associated with multi-level cervical spine fusion with hypermobile normal segments. Therefore, a patient with KFS can be at risk of severe neurological symptoms even after a minor trauma. We report a patient with type III KFS who developed a hemiparesis after a minor trauma and was successfully managed with operation.

KEY WORDS : Hemiparesis · Klippel-Feil Syndrome · Trauma.

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INTRODUCTION

In 1912, Klippel and Feil first described a syndrome that consisted of a clinical triad of short neck, limitation in head and neck movements and low posterior hairline. Currently, this syndrome called Klippel-Feil syndrome (KFS) is referred to a congenital fusion of two or more vertebrae[7]. Multi-level fused cervical vertebrae may become symptomatic during the rapid growth of adolescence or in adult life by the entrapment of the brain and/or the spinal cord.

The classification of KFS is based on the site and extent of the cervical fusion[9]. Type I is applied to patients with an extensive cervical and upper thoracic spinal fusion. Type II refers to patients with one or two interspace fusions, often associated with hemivertebrae and occipitio-atlantal fusions. This type is considered most usual and asymptomatic. Type III is applied to individuals with both cervical and lower thoracic or lumbar fusions, which occur sporadically and are rarely reported.

We describe a patient of type III KFS who developed a hemiparesis after a minor trauma due to myelopathy at the cranio-cervical junction caused by multiple congenital anomalies such as assimilated atlas, basilar impression and multi-level fused cervical vertebrae. The patient was successfully managed with transoral anterior decompression and posterior decompression with occipito-cervical stabilization.

CASE REPORT

This 33-year-old man was admitted to the department of neurosurgery for the complaint of motor weakness in the right extremities. Two months before admission, he had a minor injury to the neck by slipping down at his workplace. Since that time, motor weakness had developed and progressed. There was no history of birth trauma, familial and genetic diseases.

On admission, we observed painful motion limitation of the neck with a lordotic stance, low hair line, and webbed neck (Fig. 1). Right-side muscle strength was determined to be 3/5 on the arm and 4/5 on the leg. Light touch and pin prick sensations were decreased below C3 bilaterally. Pathologic reflexes such as Babinski's sign, ankle clonus and Hoffman sign were also present.

Plain X-ray and CT scan of the cervical spine demonstrated an assimilation of atlas, basilar impression

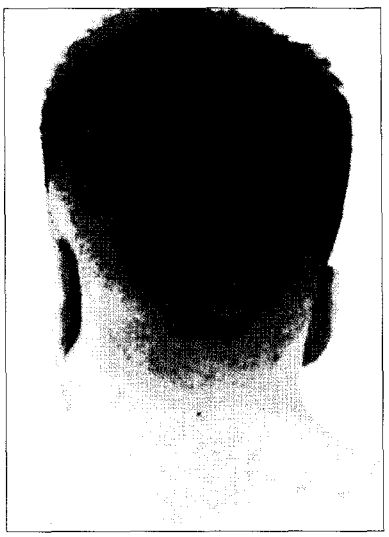

Fig. 1. Photography of the patient showing webbed neck and low posterior hair line.

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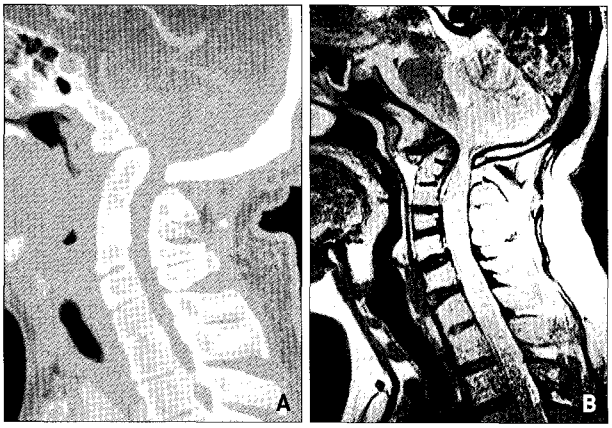

Fig. 2. A : Computed tomography sagittal reconstruction image showing assimilation of atlas, basilar impression and congenital fusion of C2-C4 vertebrae. B : T2 weighted sagittal magnetic resonance image demonstrating severely compressed spinal cord at cervicomedullry junction.

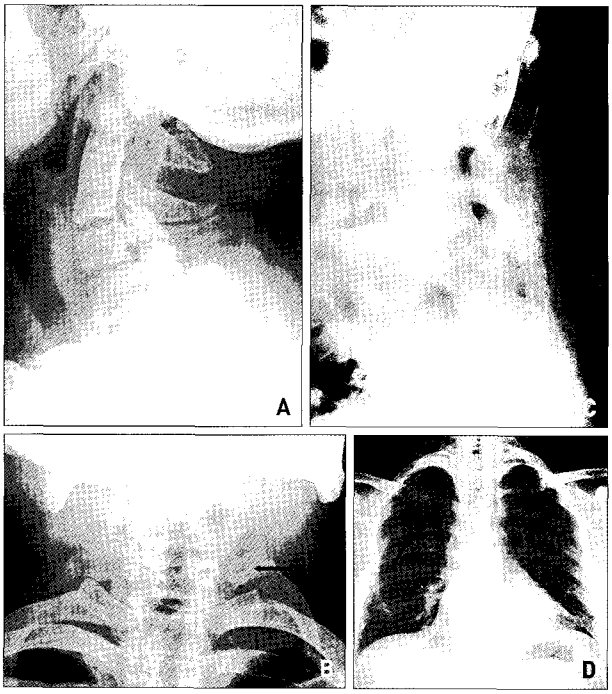

Fig. 3. Plain X-rays of patient A : Lateral view of cervical spine showing assimilation of atlas and congenital fusion of vertebrae from C2 to C4. B : AP view of cervical spine demonstrating cervical ribs (arrow) C : Lateral view of lumbar spine showing congenital fusion of lower lumbar vertebrae. D : Chest AP defining hypoplastic 5th rib (arrow).

and fusion of vertebral bodies from C2 to C4 (Fig. 2A, 3A). Sagittal MR image revealed that there was a severe canal stenosis at the cervical medullary junction and severely compressed spinal cord at the same level (Fig. 2B).

Chest X-ray revealed the 7th cervical ribs and the hypoplastic 5th rib on the right side (Fig. 3B, D). Lumbar spine X-ray revealed a fusion of vertebral bodies from L4 to S1 (Fig. 3C). CT scan of the abdomen showed left renal agenesis (Fig. 4).

Because the cord was compressed from the anterior and posterior aspect of the canal, we decided that decompression

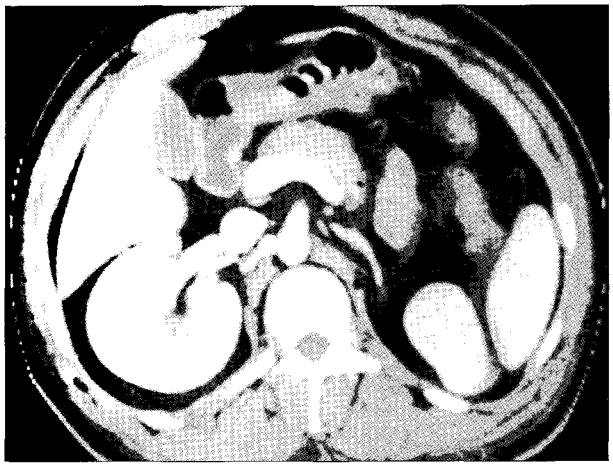

Fig. 4. Computed tomography scan of abdomen demonstrating a renal agenesis of left side.

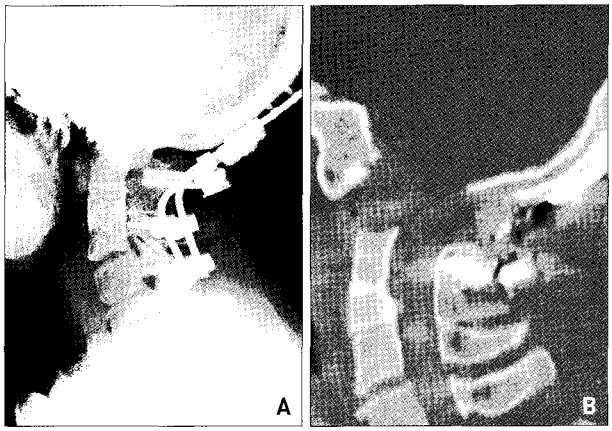

Fig. 5. Postoperative radiographic images revealing posterior occipito-cervical stabilization from occiput to C4 (A) and adequate decompression of cervicomedullary junction on computed tomography scan (B).

would be most effective through the anterior and posterior combined approach. The surgery was divided into two procedures. One was anterior decompression by transoral odontoidectomy and the other was decompressive suboccipital craniectomy and occipito-cervical fusion from occiput to C4 with additional instrumentation.

The first operation was performed in a supine position with neck extension by Mayfield head fixator under naso-tracheal general anesthesia. Transoral method in cooperation with the ENT team was then followed. Briefly, mouth was opened using a retractor and by incising a midline from the hard palate to the uvula base, nasopharynx was exposed. Then, dissection of retropharyngeal muscle to expose the odontoid process was followed and following odontoidectomy with a high speed drill, pulsation was checked by exposing anterior longitudinal ligament and dura.

Next, he was placed in concord prone position. Skin incision was made from occipital external protuberance to C5 levels followd by removal of the subocciput of foramen

magnum and the posterior arch of assimilated atlas. Two translaminar screws were then inserted into C2 lamina as previously described by Wright[12]. And, lateral mass screws were inserted, according to Anderson's method[1] at C3 and C4, inverted Y plates were posed in the midline with VERTEX system (Medtronic Sofamor Danek, Memphis, TN, USA). During the series of surgery, C-arm monitoring was used continuously. Finally, bone chips were placed around instruments with two iliac bone grafts. Two months after operation, his sensory change disappeared and motor weakness was improved to grade 4+/5 subjectively. Bone fusion was under progress in the last follow-up X-ray and his symptom improved (Fig. 5).

DISCUSSION

KFS is a complex syndrome of osseous and visceral anomalies. Although KFS includes the classic triad of short neck, namely limitation in neck movements and low posterior hairline, it is observed in less than 50% of patients[4] and the major feature of KFS is congenital fusion of two or more cervical vertebrae[7]. This syndrome appears to be a failure of the normal segmentation and fusion processes of the mesodermal somites, which occur between the third and eighth week of embryonic development[4]. Overdistension of the neural tube is also considered as another possible pathogenesis[5].

Several congenital abnormalities have been defined in relation with KFS. More frequently observed abnormalities are urogenital anomalies, deafness, scoliosis, Sprengle's deformity, hemi-vertebrae, spina bifida, and rib anomalies[10]. Our patient has cervical ribs, hypoplastic 5th rib, and unilateral renal agenesis, as well as multi-level fused cervical and lumbar spine[10]. Type III KFS is determined as type I or type II in association with lower thoracic or lumbar spine fusion. With these features, our case fits well with Type III of KFS.

KFS occurs in 1 of every 42,000 births, and 60% of the cases are female[2]. This syndrome can follow an autosomal recessive or dominant inheritance pattern with variable penetrations[2]. Therefore, although most cases of this syndrome occur sporadically, close evaluation of the immediate family is recommended.

In KFS, multiple fused vertebrae can alter normal spine kinematics. Mechanically increased stress by block vertebrae makes adjacent normal segments excessively mobile. Accordingly, the patient with KFS may be at a higher risk of spinal cord injury following a minor trauma due to the hypermobile spine. Strax and Baran reported two patients with KFS, which developed into tetraplegia after a minor trauma[11]. Elstar also noted a patient with KFS who showed quadriplegia after a slip-down injury[3]. In our patient, right side hemiparesis developed following slipping down. On radiological images, we could observe assimilated atlas and C2-C4 fused cervical vertebrae. Although there was no definite instability on dynamic X-ray, we believe that the serious clipping might happen between these two distinct conditions.

In our case, there was a marked canal stenosis at the cranio-cervical junction due to assimilated atlas and posteriorly displaced odontoid process. The decompression of the cervicomedullary junction was accomplished by transoral odontoidectomy via the anterior approach and suboccipital craniectomy via the posterior approach, consequently. We believe that the anterior decompression is insufficient in providing structural stability and decompression given the multi-level nature of the anomaly. Hence, we performed simultaneous posterior decompression and occipito-cervical stabilization. The key management principles in this case was spinal cord decompression, deformity correction, and immediate stabilization. We believe that only posterior occipito-cervical stabilization can provide stability and promote long-term sagittal, coronal, and rotational balance.

The incidence of congenital anomalies of the genitourinary tract is high in patients with KFS. Smith et al. reported that 25 out of 39 patients with KFS (64%) had significant genitourinary tract anomalies as defined by intravenous urogram and physical examination[10]. The unilateral renal agenesis is most common among the genitourinary-tract anomalies[8]. Our patient had a renal agenesis on the left side although it was not symptomatic. Other visceral anomalies related with KFS were congenital heart diseases such as ventricular septal defect[6]. Patients with KFS should also be informed of the medical complications of associated anomalies in other body systems.

Despite the extensive and complex spinal surgeries performed in our patient, both showed good outcome, defined as a functional improvement in his neurological condition.

CONCLUSION

We report one unusual case of type III KFS that was managed with operation successfully. Patients with KFS should be warned of the increased risk of spinal cord injury after minor trauma.

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