Case Report

Surgical Treatment of Klippel-Feil Syndrome with Cervical Spondylosis

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We report a case of Klippel-Feil syndrome. The patient was a 37-year-old man who developed progressive motor weakness and on C4-5 and C6-7 segments combined with severe cervical stenosis, basilar impression and C1 assimilation. He showed progressive quadriparesis and respiratory difficulty. He had combined congenital anomaly of right side facial palsy of peripheral type and right side sensorineural hearing loss. In May 2002, we performed, via transoral approach, anterior fusion with Harm's mesh and hydroxyapatite from the clivus to the C3. But, one month after the operation, Harm's mesh with hydroxyapatite slipped anteriorly due to non-union. So, we removed the mesh and reoperated with fibular bone graft from the clivus to the C3. Simultaneously, posterior approach was performed with on-lay autologous rib bone graft and wiring from the occiput to the C2,3,4. One year after the operation, his motor weakness has been gradually improved and there is minimal difficulty in self-respiration.

KEY WORDS : Klippel-Feil syndrome · Transoral approach · Congenital anomaly · Spine disease.

Introduction

Klippel-Feil syndrome is defined as congenital fusion of more than two cervical vertebrae and is believed to result from segmentation failure along the embryo's developing axis during 3rd to 8th gestational weeks. The classic clinical triad consists of low posterior hairline, short neck and neck motion limitation. Several associated anomalies like scoliosis, malformation of the renal, cardiovascular, central nervous and skeletal systems have also been reported. Klippel-Feil syndrome patients combined with cervical stenosis may have increased risk of spinal cord injury in spite of minor trauma as a result of hypermobility of various cervical segments[6]. Based on the literature, we report an experience of surgical treatment for Klippel-Feil syndrome with progressive motor weakness after minor trauma.

Case Report

A 37-year-old male was admitted with spastic quadriparesis and gait disturbance after slip down injury 2 years ago. On neurologic examination, he showed mild spastic quadriparesis(Grade IV). Congenitally, he showed Rt. side facial palsy of peripheral type and Rt. side sensorineural hearing loss. There was no combined anomaly in the genitourinary and cardiopulmonary systems. Radiologic examinations showed block vertebrae on C4-5 and C6-7 segments combined with severe cervical stenosis, basilar impression and C1 assimilation (Fig. 1). Cervical traction with Gardner-Wells tong was performed on the admission day. Two hours after the cervical traction with 5 pounds, he complained of severe neck pain and respiratory disturbance, and finally he developed res-

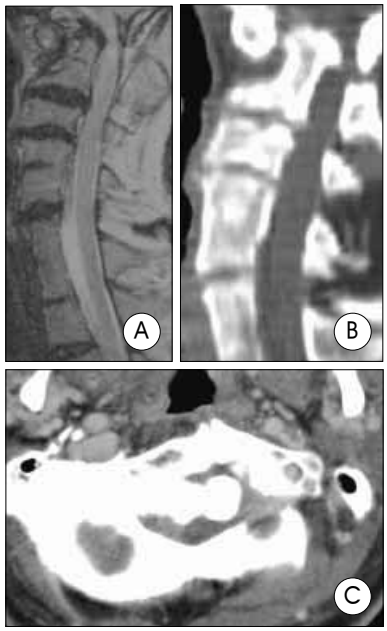

Fig. 1. A : Sagittal T2 weighted magnetic resonance reveals a compressed cord at the foramen magnum and C2/3 interspace, and also shows C4,5 and C6,7 block vertebrae. B : Saggital computed tomography shows basilar impression and C1 assimilation. C : Axial computed tomography shows chronic rotation anomaly and severe stenosis.

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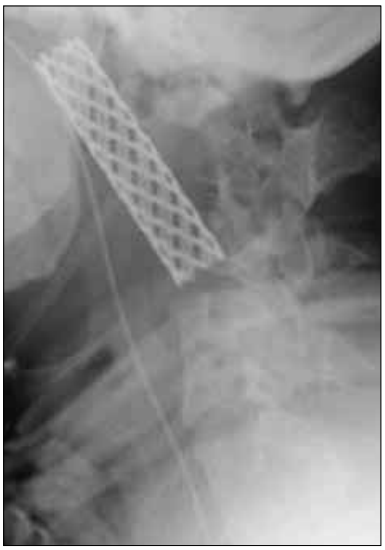

Fig. 2. One month later follow up plain lateral X-ray shows anterior slipping of the mesh.

piratory arrest. Emergency tracheostomy was done because of intubation failure due to severe anomaly in the oropharynx. His trachea deviated to the Rt. side and posteriorly. He was treated with steroid conservatively to reduce the cord swelling. His motor and respiratory function was recovered to the state before Gardner-Wells tong reduction. Although the risk was very high, we decided to perform the operation because he showed progressive myelopathy and respiratory difficulty due to severe stenosis in the craniocervical junction and upper cervical canal. Seven days before the operation, we applied Halovest because his craniocervical junction was severely stenotic and unstable, so cord compression and swelling happened only with the mild cervical traction. In May 2002, we performed, via transoral approach, anterior fusion with Harm's mesh and hydroxyapatite from the clivus to the C3. The operation was done in the state of Halovest application to stabilize the craniocervical junction. For transoral approach, we splitted the mandible and maxilla because he can not open the mouth enough to expose for transoral approach. One month after the operation, Harm's mesh slipped anteriorly due to non-union and infection(Fig. 2). So, we removed the mesh and reoperated with fibular bone graft from the clivus to the C3. Simultaneously, posterior approach was performed with on-lay autologous rib bone graft and wiring from the occiput to the C2,3,4 after C1 laminectomy(Fig. 3). After that, Halovest had been applied for 5 months. Immediate postoperative motor weakness was slightly worsened to Grade 3 as compared with the preoperative motor weakness of Grade 4 and he had to receive ventilator care due to reduced vital capacity and CO_2 retention. 3 months after the operation, the patient's motor weakness recovered to the preoperative state and ventilator weaning was possible. One year after the operation, his motor weakness has been gradually improved and there is minimal difficulty in self-respiration.

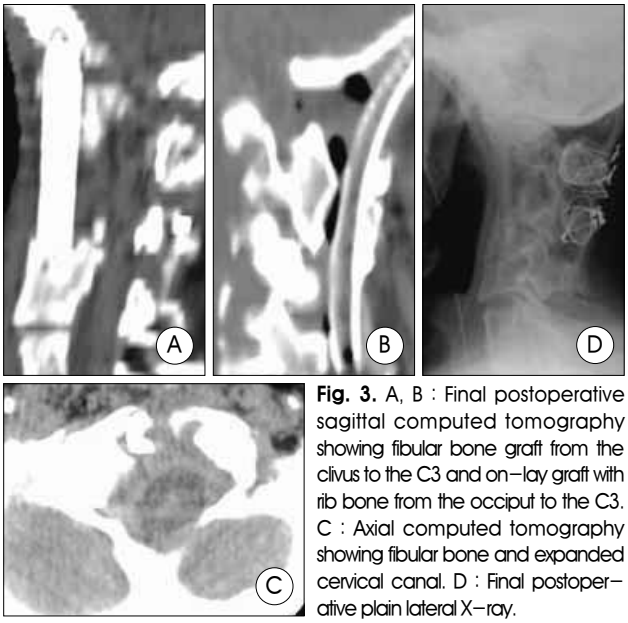

Fig. 3. A, B : Final postoperative sagittal computed tomography showing fibular bone graft from the clivus to the C3 and on-lay graft with rib bone from the occiput to the C3. C : Axial computed tomography showing fibular bone and expanded cervical canal. D : Final postoperative plain lateral X-ray.

Discussion

Klippel-Feil syndrome has an estimated incidence of 1:42,000 births with a slight preponderance of female(65%). Klippel-Feil syndrome patients combined with cervical stenosis may have increased risk of sustaining a transient neurologic deficit in spite of minor trauma[5]. This is probably related to the fused segments and the resultant altered mechanical force transfer that makes the adjacent non-fused segments excessively mobile[6]. When multiple block vertebrae are present, the normal segments may become hypermobile and be subjected to significantly increased stress. Potentially fatal subluxation may occur at these levels. Elster reported a 35-year-old male with Klippel-Feil syndrome who developed quadriplegia after only minor trauma[2]. Klippel-Feil syndrome patients, even those with minor cervical lesions, may have other less apparent or even occult defects in the genitourinary, nervous and cardiopulmonary systems, and even hearing impairment[1,4,7,8]. Many of these hidden anomalies may be more detrimental to the patient's general well-being than the obvious deformity of the neck. In the review by Hensinger and associates, a high incidence of related congenital anomalies was found, emphasizing that all patients with Klippel-Feil syndrome should be thoroughly investigated[4]. Radiographs will display only vertebral fusion. Minor vertebral anomalies may be associated with major hind brain anomalies. In such cases, magnetic resonance imaging is the ideal modality as it will also show degenerative disc diseases[3]. Treatment usually

includes regular follow up for the onset and progress of degenerative and neurological manifestations. Most respond to conservative treatment. Only a small percentage require judicious surgical stabilization. Kim et al. performed posterior craniocervical decompression and fusion in the Klippel-Feil syndrome patient with the cord compression at the foramen magnum level and the cervical instability[6]. In our case, we used Harm's mesh and hydroxyapatite. But one month later, graft site was infected and the mesh slipped anteriorly. We think that the cause of infection is foreign material, hydroxyapatite and oropharynx exposure. So, fibular bone graft was used in the next operation. To maintain the stability, we applied Halovest before, during and after the operation.

Conclusion

Klippel-Feil syndrome patients with cervical stenosis should be made aware of the potential for sustaining a neurologic deficit after minor trauma. Concerning the corrective operation for the severe anomaly in the cervical spine,

1) Concerning the general anesthesia, thorough preoperative evaluation should be made especially including the cardiopulmonary system. After that, appropriate approach method corresponding to the craniocervical junction anomaly should be performed.
2) Halovest apply is needed before, during and after the operation to maintain the stability of the patient who shows severe instability.
3) Mandiblectomy and maxillotomy is needed for the wide exposure in transoral approach, but should have a mind that the infection risk may be high.

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