

Surgical Management of a Klippel-Feil Patient with Basilar Invagination and an Intramedullary lipoma: A Case Report

Klippel-Feil Sendromu ile Birlikte Görülen Baziler Invajinasyon ve İntramedüller Lipomlu Olguda Cerrahi Tedavi: Olgu Sunumu

ABSTRACT

Patients with Klippel-Feil syndrome (KFS) and associated craniocervical junction abnormalities are at high risk for the development of progressive neurologic deficits. A 60-year-old male patient with Klippel-Feil syndrome was admitted to the emergency room with an 8-year history of a progressive spastic quadraparesis which began with numbness in the right lower extremity. Neurologic status of the patient improved significantly following posterior decompression and stabilization. Dramatic resolution of his myelopathy with the initial procedure led to postponing the second procedure. Early post-operative improvement in neurologic function after the initial procedure may be misleading. For this reason, the two-staged ventral and dorsal decompression was suggested for the restoration and preservation of neurologic function.

KEY WORDS: Klippel-Feil Syndrome, lipoma, basilar invagination, surgical management

ÖZ

Klippel-Feil sendromu ile birlikte görülebilen kranioservikal bileşke anomalileri ilerleyici nörolojik defisit gelişmesinde büyük risk taşır. Klippel-Feil sendromlu 60 yaşındaki erkek hasta 8 yıllık spastik kuadriparezi hikayesi ile birlikte sağ alt ekstremitede yeni başlayan uyuşukluk şikayeti ile acil servise başvurdu. Hastaya uygulanan posterior dekompresyon ve stabilizasyon operasyonundan sonra nörolojik tablosunda belirgin düzelme gözlemlendi. Erken postoperatif dönemde nörolojik tablodaki düzelme yanıltıcı olabilir. Bu nedenle, bu olgularda nörolojik fonksiyonları korumak ve yeni nörolojik defisit gelişimini önlemek için, iki basamaklı anterior ve posterior cerrahi girişim önerilmektedir.

ANAHTAR SÖZCÜKLER: Klippel-Feil sendromu, lipom, cerrahi tedavi

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INTRODUCTION

Patients with Klippel-Feil syndrome (KFS) often have associated craniocervical junction abnormalities and other visceral anomalies. These patients seem to be at higher risk for the development of progressive neurologic deficits. Abnormalities at the occipitoatlantal junction, spinal canal stenosis, scoliosis and cervical instability may cause neurologic deficits in patients with KFS (3, 10). A review of the literature does not generate a uniform clear-cut strategy for the surgical management of these patients (10). We present such a case and suggest both anterior and posterior decompression with posterior stabilization for the restoration and preservation of neurologic function.

CASE REPORT

Presentation: This 60-year-old wheelchair-bound male presented to the emergency room with an 8-year history of a progressive spastic quadraparesis which began with numbness in the right lower extremity.

Examination: On neurologic examination, the patient had profound sensory loss to light touch, pin-prick, and temperature in all four extremities. His motor strength was graded 2/5 in the lower extremity and right upper extremity. The left upper extremity was held flexed against the chest with marked muscular atrophy. Bowel and bladder function, and lower cranial nerve function were normal. The remainder of his physical examination revealed a low-lying hair line, low set ears, a wide, webbed neck, torticollis, and a left Sprengle's deformity. Additionally, he had an abnormal airway, cardiomegaly, and he stated he was born with only one kidney.

Craniocervical imaging revealed profound basilar invagination, a Chiari I malformation, Klippel-Feil fusion of the cervical spine, and a compressive intramedullary lesion of the cord at the C3 level (Figure 1). Anterior and posterior impression on the cord was seen at the cervical-medullary junction. There was marked kyphoscoliosis of the thoracolumbar spine.

Surgery and Post-operative course: The patient was placed on the solumedrol protocol and a two-staged procedure was planned. After placement of a tracheostomy and percutaneous gastrostomy, a suboccipital craniectomy, C1-C4 laminectomy, subtotal resection of the C3 intramedullary lipoma and duroplasty were performed. His left scapula,

which reached the level of the C2 and C3 spinous processes, was subtotally resected. Additionally, an occipital-cervical fusion to the C6 level was performed utilizing a bent Stinmen pin and Songer cables as well as an autologous bone graft in anticipation of a second stage anterior procedure.

Immediately post-operatively, his spasticity, tone and strength were dramatically improved but his sensory symptoms improved only marginally. It was decided to delay the anterior procedure because of the marked improvement in his neurologic findings. Post-operative sagittal reconstructed CT imaging showed a capacious upper cervical canal, but considerable kinking of the cervical-medullary junction remained (Figure 2). On the 10th postoperative day the patient was transferred to a rehabilitation hospital. During the second week of rehabilitation, the patient began to develop recurrent hypertonicity in the lower extremities. Within a few days he was profoundly quadraparetic. Lower cranial nerves, bowel and bladder function remained normal. Follow-up CT imaging disclosed no acute changes from the immediate post-operative studies and axial CT showed the odontoid process embedded within the parenchymal tissue (Figure 3). He underwent replacement of his tracheostomy, and was taken to surgery for a transoral resection of his odontoid, lower clivus, and anterior arch of C1 utilizing frameless stereotactic technique (Figure 4).



Figure 1: Post-Gadolinium sagittal T1 MRI of the craniocervical junction showing fusion of the C3 and C4 vertebral bodies, atlantooccipital fusion, basilar invagination, Chiari I malformation, cervicomedullary kinking and a C3 intraspinal enhancing mass.



Figure 2: Post-operative reconstructed sagittal CT image of the craniocervical junction after the first procedure. There is a capacious upper cervical canal, but the odontoid process continues to invaginate the cervicomedullary junction.

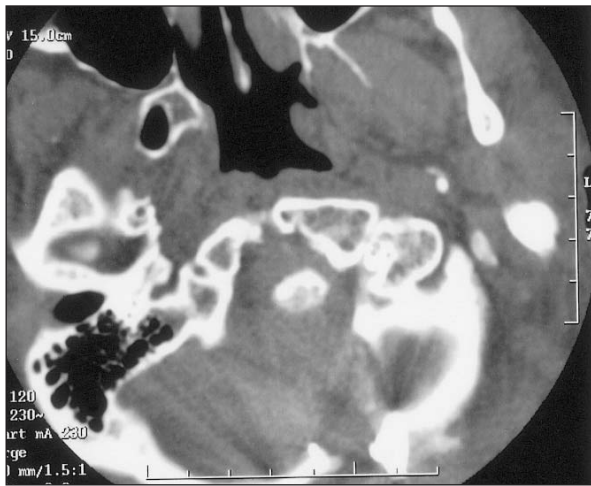


Figure 3: Post-operative axial CT of the craniocervical junction after the first procedure showing the odontoid process embedded within the parenchymal tissue.



Figure 4: Post-operative axial CT after the second procedure showing the extent of the resection of the odontoid process. The cervicomedullary junction has been decompressed both anteriorly and posteriorly after this second procedure.

The patient had significant improvement in his motor function after the second procedure. Lower cranial nerves function remained normal but bowel and bladder function remained unchanged.

DISCUSSION

Klippel and Feil (6) first described the classic triad of short neck, low posterior hairline, and severe restriction of motion of the neck because of complete fusion of cervical vertebrae; a syndrome which now bears their name. According to their classification system this consists of a one- or two-interspace fusion associated with hemivertebrae and occipitoatlantal fusion (2, 7). The patient we present had a Type II Klippel-Feil abnormality. This syndrome can cause neurological symptoms which are quadriplegia, radiculopathy and myelopathy (9). Neurological deterioration may be caused by spondylosis or hypermobility of unfused segments, which result in mechanical pressure on the spinal cord or nerve roots; by congenital malformations of the spinal cord; or by vascular disorders (9, 11). Visceral congenital anomalies are associated with the syndrome (4). The presence of an intraspinal lipoma, as seen in our patient, has rarely been reported in association with the KFS (1, 12, 13).

Hensinger and Maceven (5) described three potentially unstable cervical fusion patterns in patients with KFS that could lead to neurologic compromise and stated that this compromise is more likely related to other anomalies at the occipitoatlantal junction than to the abnormal fusion patterns. Such abnormalities include basilar invagination and the Chiari I malformation. Nagib et al (10) reviewed their series of 21 KFS patients with respect to identifying those at high risk for neurologic compromise. One of the patients in this series, with an anatomic pathology similar to our patient, required an anterior procedure 15 years after a posterior decompression had been performed because of delayed progression of his neurologic deficit. In his review, Nagib et al suggested some management options available to these high risk patients. Three groups of patients were defined: 1) those with unstable fusion patterns, 2) those with associated craniocervical abnormalities, and 3) those with concomitant spinal canal stenosis. The proposed management strategy stated that those with unstable necks should be fused posteriorly, and that those with stenosis should undergo posterior decompression. Their recommendation for the group

with associated craniocervical abnormalities was for an attempt at reduction through traction, followed by posterior stabilization if the attempt at reduction was successful. If reduction is unsuccessful, either an anterior or posterior decompression should be performed depending upon the region of pathology. This followed a paradigm suggested earlier by Menezes et al (8).

Basilar invagination, if irreducible, can generally be treated with anterior decompression if there is no associated posterior pathology. A secondary posterior fusion is then required. However, if there are associated posterior craniocervical abnormalities, ventral and dorsal decompression may be required for the restoration and preservation of neurologic function.

Our patient initially underwent a posterior procedure because of his type I Chiari malformation and intraspinal tumor. Fusion was performed during the first procedure in anticipation of a second stage anterior procedure because of significant kinking of the cervicomedullary junction. Neurologic status of the 60-year-old patient with KFS improved significantly following posterior decompression and stabilization. The anterior decompression which has been planned previously was postponed due to the general status, age and improved neurologic status of the patient. The occiput C5-6 fusion performed with Stinmen pin and Songer cable in the first operation was not sufficient and this may be the reason for a second neurologic deterioration. Using an instrument which supports the occipital-cervical fusion with a lateral mass plaque could have resulted in a more stable biomechanic system. In addition, the sudden neurologic deterioration after the late postoperative period of the first procedure has manifested the

necessity of anterior as well as posterior intervention.

Despite such improvement, it might be better to perform two procedures at the same time for preservation of function in the early stage.

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