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Cervical Pedicle Screw Fixation in a Patient With Larsen Syndrome: A Case Report

Larsen Sendromlu bir Olguda Servikal Pediküler Vida Fiksasyonu: Olgu Sunumu

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ABSTRACT

Larsen syndrome is characterized by flat face with hypertelorism, multiple and bilateral congenital dislocations and juxta-calcaneal accessory bone or bifid calcaneus due to a generalized mesenchymal defect involving connective tissue. This rare syndrome sometimes causes severe cervical spinal kyphosis in childhood. In this report a six-year-old female patient with severe cervicothoracic kyphosis due to Larsen syndrome is presented. The patient was operated on through the posterior approach and cervical and upper thoracic pedicle screws were used for correction of the deformity. Larsen syndrome and surgical correction techniques for the severe cervical kyphotic deformities are discussed with the relevant literature.

KEY WORDS: Cervical kyphosis, Larsen syndrome, Spinal deformity correction

ÖZ

Larsen sendromu konnektif dokuyu ilgilendiren generalize mezenkimal defekt sonucu ortaya çıkmış olan hipertelorizmin eşlik ettiği yassı yüz, multiple ve bilateral konjenital dislokasyonlar ve juxta-calcaneal aksesuar kemik veya bifid kalkaneus ile karakterizedir. Bu ender görülen sendrom bazen çocukluk çağında ağır servikal spinal kifoza neden olur. Bu raporda Larsen sendromu nedeniyle ağır servikotorasik kifozu olan altı yaşında bir kadın hasta sunulmaktadır. Hasta posterior yaklaşımla opere edilerek, deformitenin düzeltilmesi için servikal ve üst torakal pedikül vidaları kullanıldı. Larsen sendromu ve ağır servikal kifotik deformiteleri düzeltme teknikleri ilişkili literatür ile tartışıldı.

ANAHTAR SÖZCÜKLER: Larsen sendromu, Servikal kifoz, Spinal deformite düzeltilmesi

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INTRODUCTION

Larsen and associates first described this rare syndrome in 1950 (21). It is characterized by a flat face with depressed nasal bridge, bulging forehead and widely spaced eyes (hypertelorism), multiple and bilateral congenital dislocations, usually of the hips, knees, and radial heads, and a juxta-calcaneal accessory bone or bifid calcaneus, which, if present, is diagnostic for Larsen syndrome. Inheritance of Larsen's syndrome is by dominant and recessive autosomal transmission (7, 21). The cause of the extensive abnormalities is a generalized mesenchymal defect involving connective tissue (3, 25). Cardiovascular lesions are not uncommon. These consist of congenital anomalies, e.g., ventricular or atrial septal defect, aortic dilation and insufficiency, mitral valve prolapse and insufficiency, and ductus arteriosus aneurysm (7). Maldeveloped cartilage of the larynx and the tracheal rings ('flabby cartilage') may cause respiratory difficulty (3, 7, 25). Affected children usually have normal intelligence and have minimal disability after correction of the orthopedic deformities (7, 25, 27). Larsen syndrome patients are usually treated by neurosurgeons because of the various associated anomalies and particularly those involving the spinal column. The main anomaly is abnormal segmentation of the cervical spine, occurring in about 20 percent of the reported cases and resulting in midcervical kyphosis, cervicothoracic lordosis or kyphosis, and progressive spinal instability (3, 7, 9, 17, 18, 25, 27). In this report a six-year-old girl with severe cervical kyphosis due to Larsen syndrome is presented and surgical correction techniques of the severe cervical kyphotic deformities are discussed with the relevant literature.

CASE REPORT

A six-year-old female patient presented with progressive paraparesis, urinary incontinence and difficulty in swallowing. Her neurological examination revealed paraparesis of the lower extremities (2/5), bilateral positive Babinsky sign, hyperactivity of deep tendon reflexes and generalized muscle atrophy. There was significant retardation of physical development due to dysphagia causing malnutrition. She had the typical face of Larsen syndrome. She was not able to hold his head straight, and the deformity in the cervical and upper thoracic region was obvious. She had been operated on nine times because of the multiple deformities of her extremities by plastic and orthopedic surgeons after she was diagnosed as Larsen syndrome. Her mental development was normal. Her pre-operative antero-posterior and lateral cervical x-ray films revealed both kyphosis and scoliosis of the cervical and thoracic spine. The cervical magnetic resonance imaging (MRI) showed 105 degrees of kyphosis between C5 and T4, and signal intensity changes of myelomalasia caused by spinal canal narrowing due to severe kyphosis at C7-T1 (Figure 1). Since it is a well-known entity that general anesthetics may induce malignant hyperthermia in patients with Larsen's syndrome (2, 9) the necessary precautions were taken including preparing Dandrole and operating under elective conditions.

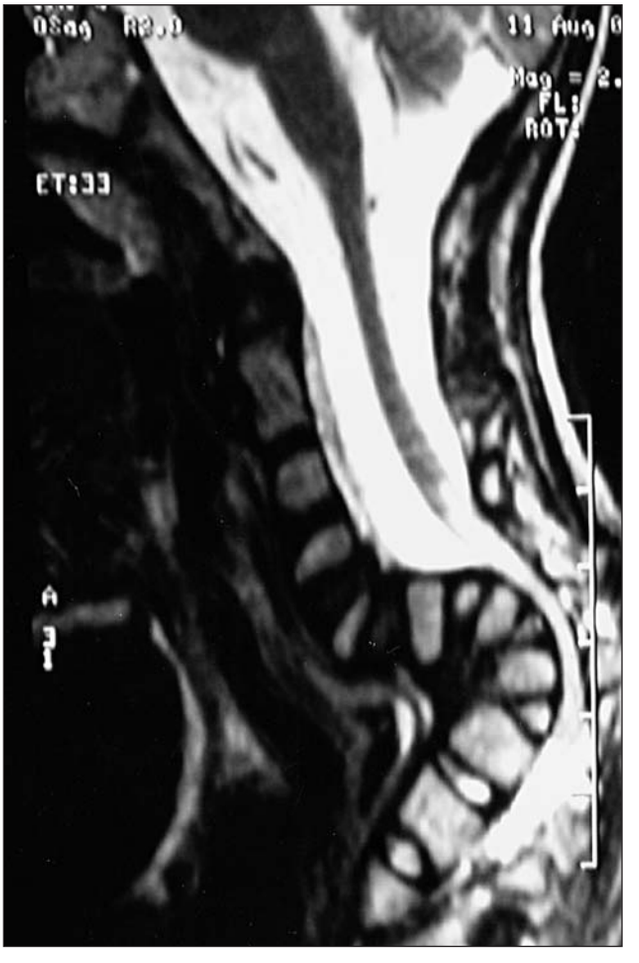

Figure 1: Preoperative sagittal T2-weighted MRI of the patient.

SURGERY

It was possible to classify the neurosurgical problems that had to be corrected into 3 groups. a) Severe spinal kyphotic deformity resulting in abnormal posture of the head causing dysphagia and malnutrition, b) Spinal canal stenosis causing progressive paraparesis and incontinence, c) Instability.

These three main problems became our guide for the technical planning of the operation. Total laminectomy for the decompression of the spinal canal and for the correction of the kyphotic deformity and providing stability together with traction and fixation with cervical and upper thoracic pedicle screws after the release of posterior elements was planned and the patient was operated on under elective conditions. Using the posterior approach, a skin incision from just above the foramen magnum to the 5th thoracic vertebra was made. After blunt dissection of paravertebral muscles, lateral masses of the cervical vertebrae and transverse processes of the thoracic vertebrae were exposed. Pedicle screws of 14 mm length and 3 mm width were placed into the C2, C4, C6, T1, and T3 vertebrae. All facet joints between axis and T4, and all costo-transverse junctions were cut with high-speed drill for the release of posterior elements. Laminas were then cut bilaterally from lateral masses and total laminectomy from C5 to T1 were done. Pedicle screws were fixed at maximum traction on the sagittal plane. Her swallowing returned to normal immediately after the operation and she began to take food orally. There were no complications in the post-operative period and she was discharged from the hospital 10 days after the operation.

Follow-Up:

In the control examination three months after a special rehabilitation program, she was able to walk without help; she had full control of urination and hac put on weight. Since she refused to stay in the MR device, her control images were taken by direct X-ray films and three-dimensional computed tomography (Figures 2 A, B, Figure 3). These showed that the kyphosis had decreased to 35 degrees and there was no problem with the cervical fixation device.

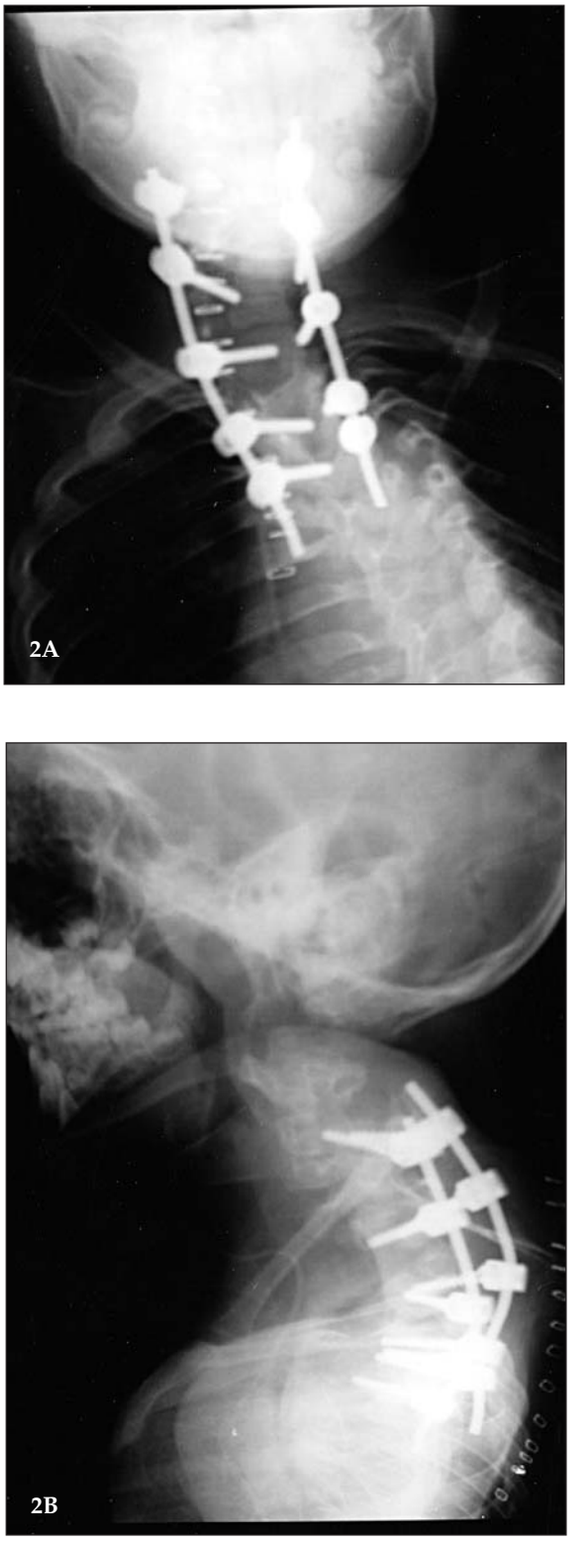

Figure 2: Postoperative direct anteroposterior (**A**) and lateral (**B**) roentgenograms of the patient.

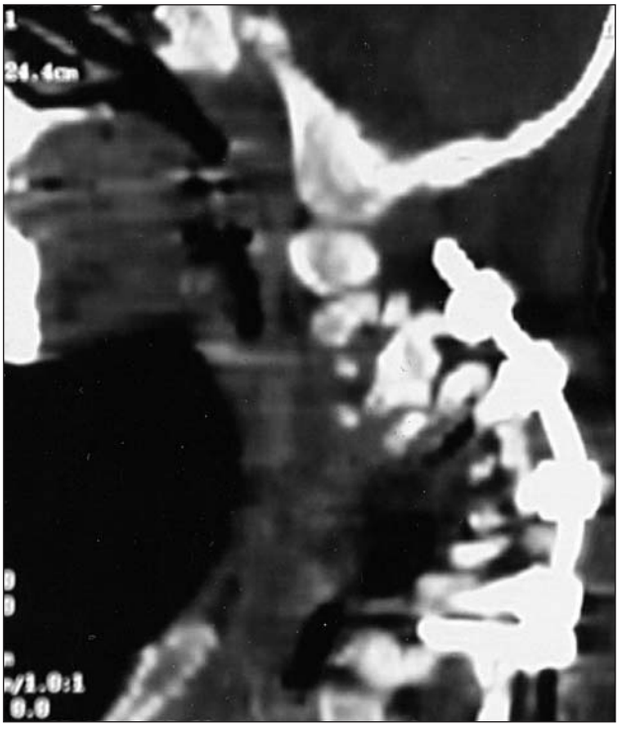

Figure 3: Postoperative three-dimensional sagittal reconstruction CT of the patient.

DISCUSSION

The clinical manifestations of a patient with Larsen syndrome are easily recognized because of the multiple dislocations of the joints (hips, knees, and elbows), the deformities of the foot (equinovarus or equinovalgus), and the dysmorphic facies, which were all originally described in 1950 (21). Although these orthopaedic manifestations have been previously identified, there are only few published cases of cervical kyphosis associated with Larsen syndrome (14, 15, 18, 24). Laville et al. (22) reviewed thirty-eight patients with Larsen syndrome who came from an isolated geographical area and did not find any who had cervical kyphosis. In that series, however, the absence of the accessory calcaneal apophysis, which is a characteristic radiographic finding in classic Larsen syndrome, casts doubt on the diagnosis and, hence, on the validity of the failure to observe cervical deformity. It is therefore assumed that cervical deformity has been underdiagnosed in patients with this syndrome (7). The potential morbidity and mortality due to the cervical deformity are obvious. Many patients who have Larsen syndrome are described as being hypotonic, a condition that contributes to a delay in the achievement of motor skills, such as the ability to walk. Johnston et al. (1) assert that the hypotonia that is characteristic of Larsen syndrome may be a result of early chronic compression of the spinal cord that remains undetected because of the absence of obvious long-tract signs, its low-grade chronicity, and the attention drawn by the often dramatic dislocations of the joints and deformities of the feet. In our patient, the severe cervical deformity was diagnosed with the appearance of incontinence and lower extremity weakness after several orthopaedic interventions.

The goals of surgery for the treatment of cervical kyphosis are correction and stabilization of the deformity and decompression of the neural elements. Correction of the severe kyphosis in the cervical spine has been one of the challenging problems in the field of spinal surgery. There have been several reports regarding surgical correction of kyphosis in the cervical spine by anterior release and fusion (4, 13), posterior release and fusion (10), and combined anterior-posterior release and fusion (11, 12, 19). In principle, an anterior decompression followed by correction and fixation by a posterior construct is advised for effective correction of the cervical kyphosis (5). Anterior decompression is also necessary to obtain maximum neurological recovery for patients who have myelopathy due to cervical kyphosis. However, an anterior decompression and arthrodesis is not a proper procedure for very young children who have Larsen syndrome because anterior growth and, hence, the potential for correction are eliminated. In our six-year-old patient, we did not perform an anterior decompression and, hence, a limited correction of kyphosis could be achieved. From a biomechanical point of view, posterior fixation devices have an advantage over anterior devices. Based on results obtained from the comparative biomechanical studies of cervical fixation procedures, there is relatively little difference in stability between lateral mass screw-plate fixation and conventional nonscrew posterior fixation procedures (20, 26). On the other hand, pedicle screw fixation systems have been widely used for reconstruction of the thoracic and lumbar spine because of their biomechanical superiority. The pedicle of the cervical spine is also a strong structural element of the vertebrae, as it is in the thoracic and lumbar spine. Recent biomechanical studies by Kotani et al. (26), Jones et al. (6), and Bozkus et al. (16) have shown that the stability provided by

pedicle screw fixation in the cervical spine is greater than that achieved with other cervical internal fixation procedures, including lateral mass plate-screw fixation and sublaminar wiring. However, the procedure in the cervical spine has been criticized for the risk to neurovascular structures, except at the C2 level (8, 23). We placed a total of ten pedicle screws from the C2 to T3 vertebra in our patient without any complication.

CONCLUSION

The prevalence of cervical kyphosis in Larsen syndrome is probably underestimated. Diagnosis of this deformity should be a priority in the initial evaluation of patients who have this syndrome. Early diagnosis followed by operative stabilization avoids neurological deficits in these patients. Posterior arthrodesis for cervical kyphosis should take part in the staging of the multiple orthopedic procedures that are necessary for a patient who has Larsen syndrome, before the development of a severe rigid kyphosis or myelopathy.

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