

Split Cord Malformation Type I and Lipomyelomeningocele: A Case Report

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- ✓ Spina bifida is a term commonly used to refer to myelomeningocele, lumbosacral lipomas, split cord malformation (diastematomyelia), meningoceles and some other neural tube defects. Split cord malformations with a fibrous, cartilaginous or bony band or spicule separating the spinal cord into hemicords, can occur as an isolated defect or either with a myelomeningocele or a lipomyelomeningocele. The occurrence of split cord malformations with lipomyelomeningocele is relatively rare in the pediatric neurosurgical practice. In this report, we present a one month old baby who had right leg monoparesis, with lipomyelomeningocele and split cord malformations with lipomyelomeningocele is relatively rare in the pediatric neurosurgical practice.

Key words: Spina bifida, split cord malformations, lipomyelomeningocele

- ✓ **Bir Split Kord Malformasyonu Tip I ve Lipomyelomeningosel Olgusu: Olgu Bildirimi**
Spina bifida terimi genellikle myelomeningosel, lumbosakral lipom, split kord malformasyonu (diastematomyeli), meningosel ve diğer bazı nöral tüp defektlerini tanımlamak için kullanılmaktadır. Split kord malformasyonu spinal kordun fibröz, kıkırdak, kemik bant veya bir kemik spikül tarafından iki hemikorda ayrılmasıdır, bu malformasyon tek başına veya myelomeningosel veya lipomyelomeningosel gibi anomalilerle birlikte görülebilir. Split kord malformasyonları ve lipomyelomeningoselin birlikte görülmesi pediatrik nöroşirürji pratiğinde sık değildir.

Bu yazıda, sağ bacak monoparezisi bulunan, lipomyelomeningosel ve split kord malformasyonu tip I'i olan bir aylık bir bebek olgu sunulmaktadır.

Anahtar kelimeler: Spina bifida, split kord malformasyonu, lipomyelomeningosel

INTRODUCTION

Split cord malformations (SCMs) type I is a term used to describe two hemicords each lying within its own dural sac and separated from its counterpart by the midline cuff of dura that surrounds a bony or fibrocartilaginous spur⁽¹⁾. A lipomyelomeningocele is a congenital lesion associated with spina bifida⁽²⁾. These lesions are accompanied by other forms of occult dysraphism such as tethered spinal cord, dermal sinus tract, and diastematomyelia (SCMs, type I and II)⁽²⁾.

In this article, we report a case with lipomyelomeningocele and SCM type I.

CASE REPORT

A 1 month old female fullterm baby was admitted to department of neurosurgery with a congenital 12x24 cm diameter thoracolumbar lesion covered with a skin. Neurological examination revealed only right leg monoparesis.

Radiography showed a large cleft within the bodies and neural arches of the vertebrae

extending from eighth thoracic to third sacral level. Ultrasound examination defined two anechoic sacs and bilateral heterogeneous solid masses which were evaluated as lipomatous tissue. Fusion anomaly was also depicted. CT images of 6 mm thick contiguous slices showed a large cleft involving bodies and neural arches of vertebrae from the low thoracic to the sacral level. There were two separate CSF containing sacs, of which each was associated with a large lipoma that extends from the subcutaneous fat of the back. CT was able to define duplication of the spinal cord and formation of two hemicords which were splitted by a large bony spur (Figure).

Surgery was consistent with the radiological findings. Total and subtotal resection was applied to diastematomyelic spur and lipomas, respectively, and followed

by repair of the sacs. The right monoparesis improved slightly after surgery.

DISCUSSION

SCMs are anomalies in which the cord is split over a portion of its length to form a double neural tube⁽¹⁾. Diastematomyelia is a congenital splitting of any part of the spinal cord. Hertwig⁽³⁾ in 1892 first used this term to describe SCMs. Herren and Edwards⁽⁴⁾ in 1940 used the term diplomyelia to describe a complete segmental duplication of the spinal cord. A type I SCM contains two hemicords each lying within its own dural sac and separated from its counterpart by the midline cuff of dura that surrounds a bony or fibrocartilaginous spur⁽¹⁾. A type II SCM contains both hemicords lying within a single dural sheath and separated by a finer, more delicate intradural fibrous midline septum⁽¹⁾.

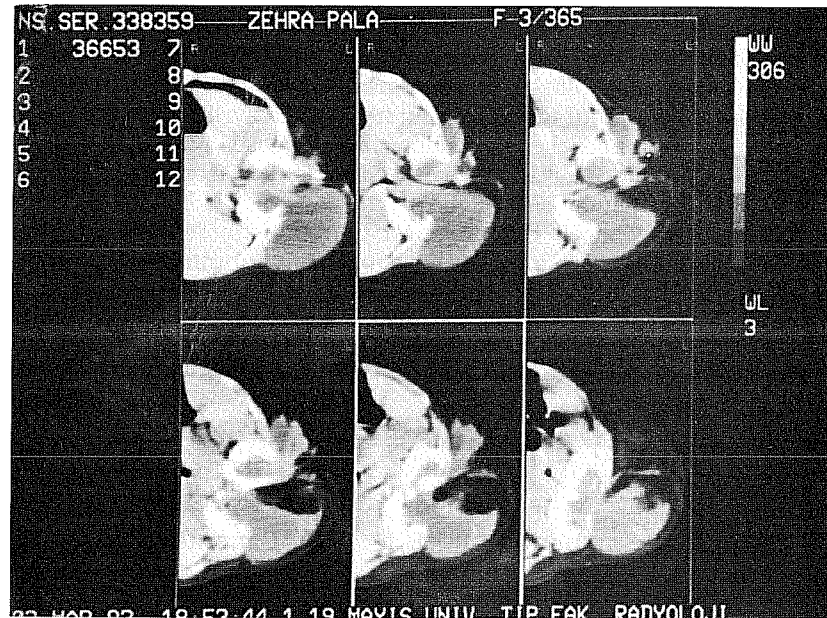


Figure : Contiguous CT slices at the thoracolumbar level show two separate CSF containing sacs, lipomas, bony spur splitting the spinal cord, and large cleft within the vertebrae. Each hemicord diverges extending to its own adjacent sac and lipomatous tissue.

The etiology of spina bifida is unknown. These malformations can be caused by teratogens such as excessive alcohol intake, carbamazepine, valproic acid, hiperthermia and obesity⁽⁵⁾. Folic acid and other vitamin and mineral deficiencies have been associated with an increased incidence of spina bifida⁽⁵⁾.

Lipomyelomeningoceles are common congenital central nervous system malformations in pediatric neurosurgical practice. Schut and et al⁽⁶⁾, reported that their incidence was 25% that of myelomeningoceles, far out numbering other dysraphic forms such as diastamatomyelia, dermal sinuses, and thickened filum terminales.

The symptoms of lipomyelomeningoceles may be produced by several possible mechanism such as abnormal formation of the spinal cord, local mass effect from the fatty mass growing within the spinal canal, and traction on the spinal cord⁽²⁾.

The radiological evaluation for the child suspected of harboring a SCM and lipomyelomeningocele includes direct radiography, CT, magnetic resonance imaging (MRI), myelography, postmyelogram CT and ultrasonography⁽¹⁾.

Direct radiography and CT demonstrates the posterior fusion defect and associated bony malformations of the vertebrae. CT is found to be effective for preoperative evaluation in this case, since each structure has quite different attenuation values. MRI has been very effective in the screening and diagnosis of patients with occult spinal disraphism⁽⁷⁾. The presence of two hemicords and median nerve roots, the low lying position of the conus medullaris, details of the relationships between the two hemicords, myelomeningocele manqué, and other associated malformations such as lipomyelomeningocele are frequently obscured

on MRI⁽⁸⁾. With a MRI performed in both sagittal and axial planes, lipomyelomeningoceles are readily visualized on T1-weighted images as areas of increased signal intensity because of their short relaxation times⁽²⁾. The details of the malformation are seen to best advantage with myelography and CT myelography.

Ultrasonography can provide images of the intradural contents. The finding of an echogenic mass in the lumbosacral area may suggest the fatty mass, but it may be impossible to exclude myelomeningocele.

The surgical management of both type I and II SCM involves exploration of the entire malformation and excision of all tethering elements⁽¹⁾. After the lipoma has been largely removed from the spinal cord, it is sometimes possible to reapproximate the pial edges of the cord to reconstitute the normal tubular configuration. When the cord is free of adhesions (bony spur, fibrous band, and others), the dura is closed⁽¹⁾. In our case, the improving of the right leg paresis was thought to be due to the result of the excision of tethering elements (bony spur and lipoma).

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