Sinir Sistemi Cerrahisi Derg 5(3-4):92-96, 2015
doi:10.5222/sscd.2015.092

Olgu Sunumu

Congenital Malformation Meningomyelocele at L5-S1 Level and Defective Thecal Sac Which is Terminated Subcutaneously at L3-4 Level: Presentation of a Rare Case and Surgical Technique

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Nutritional, environmental factors and genetic disorders are the main factors that play role in the development of neural tube defects (NTDs). In the spinal surgery, the most common congenital lesions presenting to medical attention are the diverse forms of the spinal dysraphism and caudal spinal anomalies. This report describes a rare case of congenital defective thecal sac that terminated in the skin at L3-4 level of the child who underwent surgery to repair meningomyelocele in our institute when he was 11- day old. After the urodynamic study was performed the surgery to repair swelling at L3-4 level had done. The child followed-up for four years from his birth. The child is doing well, without growth or mental retardation. Herein, the authors describe the management of multiple congenital malformations in children.

Keywords: Defective congenital thecal sac, laminoplasty, spina bifida occulta, intraoperative neurophysiological monitoring, urodynamic study

J Nervous Sys Surgery 2015; 5(3-4):92-96

L5-S1 Seviyesinde Meningomyelosel ve L3-4 Seviyesinde Cilt Altında Sonlanan Durası Konjenital Malformasyon: Nadir Olgu Sunumu ve Ameliyat Tekniği

Beslenme, çevre faktörleri ve genetic hastalıkları, nöral tüp defektleri gelişmesinde önemli oynayan faktörlerdir. Spinal cerrahisinde, disrafizm ve kaudal spinal anomaliler, konjenital lezyonlar arasında en sık bebeklere cerrahi bırakan lezyonlardır. Bu makalede, ender olarak 11 günlükken meninjomyelosel nedeniyle merkezimizde cerrahi olarak tedavi edilen bebekte saptanıp L3-4 seviyesinde ciltaltında biten konjenital defektif dura sunulmaktadır. Ürodinamik test yapıldıktan sonra L3-4 seviye arasında bulunan kese müdahele edilip tamir edildi. Çocuk, postoperative olarak 4 yıl takip edildi. Durumu gayet iyidir, zihinsel ve/veya yapısal gelişim geriliği saptanmamıştır. Bu raporun önemi yazarların keseli olan ve nörolojik olarak sağlıklı çocuklarda deficit olmadan nasıl müdahale edilebileceğini anlatmalarıdır.

Anahtar kelimeler: Defektif konjenital dura kesesi, laminoplasti, spina bifida okülta, intraoperatif nörofizyolojik monitorizasyon, ürodinamik çalışma

J Nervous Sys Surgery 2015; 5(3-4):92-96

Alındığı tarih: 20.11.2016
Kabul tarihi: 14.09.2017
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INTRODUCTION

The incidence of neural tube defects (NTDs) is 0.17–12 per 1000 live births worldwide (1-3). Meningomyelocele is one of the common spinal dysraphic lesions whose incidence had been reduced in developed countries due to supplementation of folic acid during pregnancy and prenatal diagnosis of dysraphic malformations almost leading to termination of the pregnancy. Meningomyelocele is generally associated with several malformations such as tethered cord, split cord malformation, diastematomyelia and vertebral fusion anomalies such as spina bifida occulta, block vertebra or butterfly-shaped vertebrae.

Several factors may play role in developing NTDs such as nutritional, environmental, genetic syndromes and mutations in multiple genes (2). However, folic acid deficiency in pregnancy is still the proven etiology. NTDs can be divided into two main subgroups according to the involved sites; cranial dysraphism and spinal dysraphism. On the other hand, some authors classified NTDs as open or closed NTD.

The consensus among most neurosurgeons is that the child who was born with meningomyelocele should be closed promptly. Several centres in USA and Europe started to treat infants during their intrauterine life (1,4). Herein, the authors describe the management of children involved with multiple spinal malformations after presenting surgical intervention to treat a rare case of meningomyelocele, spina bifida occulta and spinal cutaneous swelling.

CASE REPORT

A 20-month-old male child was born with the meningomyelocele. At the 11th day of his life, he had been treated by repairing the thecal sac and closing the skin at the L5-S1 level. He had a swelling sac which was covered with hair patch at L3-4 level. To avoid the complication may be occurred, the swelling at L3-4 had not been included in the surgical procedure. It was thought that swelling contains fibers of the medullary conus, therefore, neurosurgical team decided to perform urodynamic test to explore the contents of this cystic swelling then plan a second surgery when the child would be 18 months of age. After the first surgery, the child called to monthly control visits.

On his 18-month control child was neurologically intact. Excluding the hypertrichosis (tuft of hair) and swelling on his back, his physical examination was unremarkable. The neurosurgical team decided to perform urodynamic study (because of the controversies about use of MRI in children the authors did not perform MRI). The urodynamic study demonstrated residue in the bladder after urination (70 cc). As is well known radiography ca not confirm the presence of a neural structure (if any) in the swelling at L3-4 .

Surgical Procedure (Technical Note)

Under general anesthesia and intraoperative neurophysiological monitoring the paramedian vertical midline (fish- mouth shaped) incision between L1 to L5 was performed. To avoid cutting or incising any neural structure contained in this swelling the spinous process of L2 was identified, bilateral paraspinal muscles were dissected and spinous process of L2 was split using a fine-tip blade. The spina bifida occulta of L3-L4, and diastematomyelia at L3 level were seen, while the thecal sac was terminated in the skin at L3-4 level (Figure 1). Duraplasty was performed (Figure 2). L2, 3 and 4 laminoplastİes were performed using non-absorbable 2/0 sutures. The fascia and skin were sutured appropriately to their anatomic layers. Postoperative course was

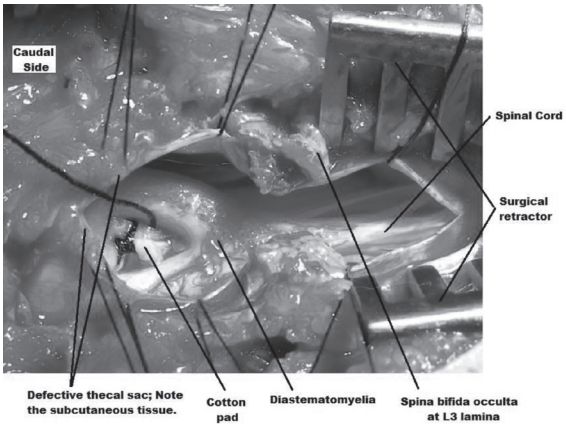

Figure 1. Photo had been taken through operation showing the spina bifida occulta of L3-L4, diastematomyelia at L3 level, and the thecal sac was terminated in the skin at L3-4 level. Note the subcutaneous tissue in the caudal part of the dura. There was no bony septum or fibrous band between both hemispinal cords.

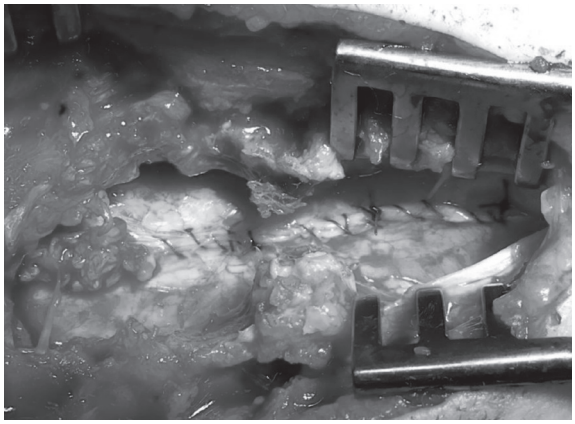

Figure 2. Duraplasty was performed after explore the spinal cord.

uneventful, no complication was detected, and the child was discharged after 3 days.

The child were followed-up for four years from his birth. He is developing well, without demonstrating growth or mental retardation. On his postoperative 30th month visit, the urodynamic study showed a slight decrease in residual volume (60 cc which is at age-matched upper limit of normal physiologic volume). Yearly control visits were recommended.

DISCUSSION

Meningomyelocele is the most common congenital anomaly of the central nervous system. It accounts for 98% of open spinal dysraphisms [5]. It is caused by defective neural tube. It is associated with other spinal cord anomalies. A hair tuft on our patient can be distinctive finding on physical examination. The malformations in our child were meningomyelocele, diastematomyelia, spina bifida occult, and spinal cutaneous swelling (cyst) between L3 and L4, however, his systematic and neurologic examinations were unremarkable.

Spina bifida is an incomplete fusion of halves of the ventral arches resulting in midline defect usually in lumbosacral region. Its manifestations vary, but generally the small bones (vertebrae) that make up the spine do not complete their developmental process, and gaps may be seen between them. The term spina bifida refers to the defective fusion of posterior spinal bony elements in open dysraphism. Myelomeningoceles and myeloceles are characterized by exposure of the placode through a midline defect in the back. In myelomeningoceles, expansion of the underlying subarachnoid space results in elevation of the placode above the cutaneous surface (open spina bifida with dorsal cyst), whereas in myeloceles, the placode is flush with the cutaneous surface [6].

Open spinal dysraphism originates from defective closure of the primary neural tube, which leads to the persistence of a segment of non-neurulated placode. Generally majority of cases are located at the lumbosacral level, with the placode being the conus [5,6]. Since neurulation does not occur, the cutaneous ectoderm does not detach from the neural ectoderm and remains in a lateral position. This results in a midline skin defect. Therefore, the external surface of the placode is directly visible on inspection [6].

In utero surgical repair has been applied in sev-

eral centers in the USA and Europe for many years (1,4) for the management of myelomeningocele. In utero surgical repair was evaluated in a controlled trial and short-term benefits for the newborn, including 50% reduction in the need for hydrocephalus shunting and significant improvement in spinal neurological function was demonstratred [(1)].

Our patient had been treated surgically by repair of his thecal sac at L5-S1 level. When we reoperated child we had seen that the laminae were intact without prior surgical interventişon. The neurosurgical team suggests that laminoplasty approach should be used in such pediatric patients. In pediatric patients we prefer to incise the cartilage of spinous processes by surgical blade, then use small retractors to expose the surgical field. After performing all surgical intervention we ought to close right and left sides of laminae using a strong non-absorbable sutures to perform laminoplasty. Laminoplasty and neurophysiological monitoring is essential in child who will undergo spinal surgery. Further investigations such as urodynamic studies have to be performed in children with meningomyelocele especially those with hair tufts.

In the management of children affected by myelomeningocele associated with other spinal malformations, first meningomyelocele should be closed promptly to protect child from meningitis that may be associated with open NTDs. The authors here suggest to wait 18 months to perform urodynamic test. The urologists comments of urodynamic test did not guide the neurosurgical team to decide if there wasa neural structure involved or not. Therefore, in second surgery surgeons did not incise the swelling directly but passed close the swelling using midline fish- mouth shaped incision to avoid injuring the neural fibers may pass through the cyst (swelling). The surgeons started dissection above L2 lamina (the intact lamina of swelling superior edge). We noticed that the thecal sac was terminated in subcutaneous layer (Figure 2). Dissection of the dural sac had been completed. Then the spinal cord and diastematomyelia were explored to ensure inexistence of split cord malformation . After duraplasty achieved using 5/0 absorbable (vicryl) sutures, intact L2 and defective L3 and 4 were repaired with laminoplasty using non-absorbable sutures.

CONCLUSION

LLaminoplasty and neurophysiological monitoring is essential in child who will undergo spinal surgery. The management for spinal cutaneous swelling (cyst) that has no risk of infection or meningitis in newborns first, urodynamic test should be performed at age of 18 months. If this study did not give details about the structures in the cyst surgeon has to avoid cutting or incising the swelling directly so as to save neural structures that may be contained in the cyst.

Competing Interest

The authors declare that they have no competing interests. All authors certify that they have NO affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Patient Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interest

There are no financial disclosures or conflicts of interest.

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