A success of lipomyelomeningoceles resection with intraoperative neurophysiological monitoring (IONM) guidance in a 10-year-old child: A case report

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ABSTRACT

Introduction: Lipomyelomeningoceles are one of the closed spinal dysraphisms, inherently associated with tethered cord. Its prevalence ranges between 0.3 and 0.6 per 10,000 live births. Deterioration of bladder and bowel function often manifests before the motor and sensory function. Intraoperative neurophysiological monitoring (IONM) is often used in lipomyelomeningocoele surgery to facilitate safe resection of lipomatous components and detethering of the spinal cord due to the absence of a clear boundary between the lipoma and neural placode. This case highlights the critical role of adequate IONM to reduce the complications of lipomyelomeningoceles resection surgery.

Case Presentation: Here we present a 10-year-old boy with a lump over the back since birth, progressively increased in size, and with a history of urinary and faecal incontinence since one year before admission. There was also a history of lower extremity weakness and pain. Repeat physical examination of the lumbosacral region revealed a single lump measured about 4.5 x 1.9 x 3.2 cm in size, soft consistency, and immobile. Magnetic resonance imaging of the lumbosacral region revealed a lipomyelomeningocoele with tethered cord protruding into the anatomical defect from sacral vertebrae S1-S4. Surgery was indicated because of neurological symptoms, and it was safely performed with the assistance of IONM. There were no postoperative complications after surgery, and there was an improvement in the patient's neurological symptoms.

Conclusion: IONM in spinal surgery has been proven useful to reduce the postoperative neurological complication by providing identification of neural structures topographically and functionally, therefore, giving a warning alarm during surgery, which can be immediately responded by the surgeons.

Keywords: lipomyelomeningoceles, intraoperative neurophysiological monitoring, tethered cord, spinal dysraphism, SSEP, MEP, EMG.

INTRODUCTION

Lipomyelomeningocele is categorized as one of spinal dysraphisms of which lipomatous tissue abnormally attaches to the spinal cord and its meninges, resulting in a closed neural tube defect. There are three classifications of it based on the anatomy of lipoma to the neural component: dorsal, transitional, and caudal.1,2 Its prevalence ranges between 0.3 and 0.6 per 10,000 live births and accounts for 14.4% of cases in spina bifida. The neural tube is formed during the 3rd week of embryonic development and the defect can occur as a result of environmental and genetic factors. Older age of pregnancy and Hispanic race increase the occurrence of lipomyelomeningoceles in infants.1,3,4

This disease is often associated with the tethered cord that later potentially show symptoms called tethered cord syndrome (TCS). These symptoms generally occur in the age over five years old including sensory and motor deficits of the lower extremities, back and leg pain, disturbances in urination and defecation, or musculoskeletal deformities due to the tethered cord disrupting cauda equina nerve root (L2 and below).1,2,4,5-11

However, lipomyelomeningocele can be examined with ultrasonography showing a subcutaneous mass in the lower sacral area at 36 weeks of gestation.1,2,5-9 The only treatment of choice is immediate surgery to release the spinal cord from the attached lipoma. In this case, the assisted surgery with intraoperative neurophysiological monitoring (IONM) that help to monitor spinal cord and spinal root function as well as to map the neural networks during surgery could result a good outcome.6-9 Studies found that combination of IONM such as spinal somatosensory evoked potential (SSEP), motor evoked potential (MEP), and electromyography (EMG) provided higher positive or negative predictive value and higher sensitivity than a single modality monitoring technique.6,8-10 In this case, we are presenting a successful case of lipomyelomeningoceles resection surgery assisted with SSEP and MEP monitoring in a child lipomyelomeningocele. This case highlights the critical role of adequate

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IONM to reduce the complications of lipomyelomeningocele resection surgery.

CASE PRESENTATION

A 10-year-old boy complained of being unable to defecate and urinate since one year before admission. Since then, the urination was drained by a catheter insertion for four months. The patient also complained the incontinence of urination and defecation four months later. There was no sensation of tingling, pain, or motoric weakness. History of trauma was denied. The patient is the fifth of five children. The delivery history was term, assisted by a traditional midwife, with a birth weight was 3600 grams. A lump in the lower back was increasing in size (Figure 1). The vaccination history was incomplete.

Physical examination revealed an immobile lump in the lumbosacral region measuring 4.5 x 1.9 x 3.2 cm, with a firm and spongy consistency (Figure 1). From the neurological examination, the patient was fully conscious with neurological deficits in the form of hypoesthesia at the level of L2 segment, followed by urinary and fecal incontinence. From the lumbosacral x-ray, a defect in the spinous processes of sacral vertebrae (VS) 1 and 2 which leaded to spina bifida. Lumbosacral magnetic resonance imaging (MRI) examination revealed a defect measuring 2.7 x 2.2 cm in the crest of VS 1-5 with meningeal prolapse, cerebrospinal fluid, cauda equina, and fat components leading to spina bifida aperta with lipomyelomeningocele and tethered cord (Figure 2).

The patient was diagnosed with lipomyelomeningocele in the lumbosacral region. The defect was in the crest of VS 1-5 with prolapse of the meninges, cerebrospinal fluid, cauda equina and fat component leading to spina bifida aperta with lipomyelomeningocele and tethered cord. The occurrence of neurological manifestations was the part of the tethered cord syndrome along with the upward growth of the spinal canal which caused the nerve roots in the cauda equina stretches. The lipomyelomeningocele resection was planned to the patient. However, in our patient there were unclear boundaries between the tethered cord, lipoma, neural tissue and connective tissue. Making this the resection was a high-risk procedure. Therefore, lipomyelomeningocele resection surgery was conducted assisted with IONM, including somatosensory evoked potential (SSEP) and motor...
evoked potential (MEP). This monitoring was used in order to identify the sacral roots to assist the resection of the lipoma component and release the spinal nerve entrapments. The IONM was used since it could identify the neural structures and reduce surgical errors by providing real-time notifications to surgical operators.

Since SSEP and MEP could be affected by anesthetic agents, our patient used intravenous propofol and fentanyl with short-acting neuromuscular blockade, rocuronium, to facilitate tracheal intubation and ventilation. The depth of neuromuscular blockade was measured by recording compound muscle action potential (CMAP) for up to 4 stimuli, called as train of four (TOF). TOF in our patient was 0% (Figure 3A) which means no residual neuromuscular agents that can affect the measurements. The SSEP monitored the neural network of the dorsal column pathway and the tibial nerve to evaluate the L4-S3 nerve roots (Figure 3B and C). In our patient’s SSEP there was no distress signal during surgery, since the amplitude never declined less than 60% or had latency increase by 10% or 2 milliseconds, defined as a distress signal. MEP measured the motoric function of tibialis anterior (L4-L5), abductor hallucis longus (L4-S3), gastrocnemius (S1-2) and anal sphincter were recorded. The overview of transcranial MEP durante operative and post-operative are presented in Figure 3D and E. Recording of the external anal sphincter was performed to preserve the function of both urethral and anal sphincters. The Tc MEP response originating from the CNS is considered as normal if the amplitude is above 100 V. It was used to assess the response of vital neural networks after detethering the spinal cord. Meanwhile, the direct nerve root stimulation was used to detect and differentiate neural and non-neural tissues the surgery. In our patient, there was no decline for more than 50% or rise above the threshold value of 100V of the amplitude, detected from baseline throughout the surgery.

IONM-assisted lipomyelomeningocele resection surgery was completed without any significant problem durante operative. The procedure did not injure the nerves in either the motor or sensory pathways.

Figure 3. Overview of intraoperative neurophysiological monitoring (IONM) of the patient. IONM included a “Train of four” test to determine the degree of muscle relaxation of the patient (A), somatosensory evoked potential (SSEP) of the patient (B and C), and transcranial motor evoked potential (MEP) durante (D) and post-operative (E) of the patient. TA R: right anterior tibial; AH R, L: right, left abductor hallucis; Gas R, L: right, left gastrocnemius; Sphinc R: right anal sphincter.

There was no postoperative motor deficit in the patient. Post-operative found that there were no worsening post-operative neurological deficits. Follow-up after three weeks post-operative, the patient’s urinary incontinence complaint improved and the sensation of urination can be felt, but still had fecal incontinence.

DISCUSSION

The patient was diagnosed with lipomyelomeningocele and MRI showing a defect in the crest of VS 1-5 with prolapse of the meninges, cerebrospinal fluid, cauda equina and fat component leading to spina bifida aperta with lipomyelomeningocele and tethered cord. The occurrence of neurological manifestations is the part of the tethered cord syndrome along with the upward growth of the spinal canal which causes the nerve roots in the cauda equina stretches. This is an indication for surgery and in this case was assisted with the IONM.11-15 A surgical procedure in cases of cauda equine syndrome must be carried out immediately in order to attain a better prognosis.8,14,15

One of the challenges of lipomyelomeningocele surgery is the unclear boundaries between the tethered cord, lipoma, neural tissue and connective tissue. In addition, reversible surgical complications such as leakage of cerebral spinal fluid (CSF), wound infection, and irreversible complications such as neurological and urological disturbances are common in lipomyelomeningocele cases. In our patient, considering the patient’s complaint of disturbing neurological disorders, lipomyelomeningocele surgery was a high-risk choice. Therefore, IONM with its mapping function was needed to identify the sacral roots within the lipoma during the resection of the lipoma component and releasing the spinal nerve entrapments through the filum terminale. Therefore IONM could reduce the complications during the procedure.7,12

The IONM could precisely identify the neural structures and reduce surgical errors by providing real-time notifications alarming the operators in order to be able to respond immediately when needed to minimize the unfavorable effects of
surgical procedures. In our case the monitoring was conducted using the modalities of SSEP and MEP. The SSEP monitored the neural network of the dorsal column pathway and the tibial nerve to evaluate the L4–S3 nerve roots. The MEP evaluates the function of some muscles such as quadriceps, gastrocnemius, tibialis anterior, anal sphincters or other muscles. The monitoring of external anal sphincters could assess the pudendal nerve, consisting of S2–4 roots.

However, various physiological and pharmacological factors could affect SSEP and MEP. For instances, physical parameters and anesthetic agents that affect the electrical conduction along the axon could change the generated potential waves. These also could be influenced by the physical parameters such as the length of the synaptic tract (the longer, the more sensitive) and the location of the limbs (the upper extremities are easier to receive signals with fewer pulses than the lower extremities). In addition, other physiological variables can affect neuromonitoring modalities, like systemic blood pressure, temperature, acid-base balance, hematocrit, and arterial blood gases. Inhaled or intravenous anesthetic agents and muscle relaxants (neuromuscular blockade) can affect the amplitude and latency of the SSEP and MEP. Our patient used intravenous propofol and fentanyl with short-acting neuromuscular blockade, namely rocuronium to facilitate tracheal intubation and ventilation. Propofol was chosen because it causes more minimal effect in suppressing the motoneuron excitability than the inhaled agents.

**CONCLUSION**

Lipomyelomeningocele resection with IONM guidance could reduce postoperative neurological complications by providing identification and functional preservation of the neural networks entrapment. The notifications of IONM contribute in suppressing the risk during the spinal surgery. Therefore, IONM-assisted lipomyelomeningocele resection is recommended in those medical centers that have the equipment and the expert.

**AUTHOR CONTRIBUTIONS**

Conceptualization, Y.I., T., D.A, and F.; validation, Y.I., T., D.A, and F.; formal analysis, Y.I., T., D.A, and F.; investigation, Y.I., T., D.A, and F.; resources, Y.I., T., D.A, and F.; data curation, D.A, and F.; writing—original draft preparation, Y.I., T.; writing—review and editing, Y.I., T., D.A, and F.; supervision, D.A, and F.; project administration, Y.I., T. All authors have read and agreed to the published version of the manuscript.

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**INFORMED CONSENT STATEMENT**

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**CONFLICTS OF INTEREST**

The authors declare no conflict of interest.

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