USE OF INTRAOPERATIVE NEUROPHYSIOLOGICAL MONITORING (IONM) IN A NEONATE WITH LIMITED DORSAL MYELOSCHISIS

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KEY CLINICAL MESSAGE:

Intraoperative neurophysiologic monitoring (IONM) is widely used in neurosurgery to recognize important neurological structures but can be challenging in the pediatric population due to incomplete neural development.

ABSTRACT:

INTRODUCTION: Limited dorsal myeloschisis (LDM) is a rare form of spinal dysraphism characterized by two constant features: a focal "closed" midline skin defect and a fibro neural tract connecting the skin lesion to the underlying spinal cord. Excision of the tract followed by reconstruction is the preferred management. Intraoperative neurophysiologic monitoring (IONM) is widely used in neurosurgery to recognize important neurological structures but can be challenging in the pediatric population due to incomplete neural development.

CASE REPORT: A 7-day-old male baby presented with a history of dorsal "cigarette burn" skin lesion noted at birth. Clinical assessment and imaging findings revealed a fibro neural tract extending from the skin surface into the lumbar spinal canal at the level of L5-S1. Excision of skin lesion/ tract and untethering of filum terminale with dural tube reconstruction under Intraoperative neurophysiological monitoring (INOM) was performed. The procedure went smoothly. Postoperatively, the child showed no neurological compromise or dermatological complications. In addition, the wound healed well, and the neonate was completely normal.

CONCLUSION: This case demonstrates the successful management of LDM through an IONM-guided complex reconstruction surgery in a neonate and the effective use of IONM in such challenging cases.

KEYWORDS: Limited Dorsal Myeloschisis, Spinal cord dysraphism, Extradural stalk, IONM, Untethering.

INTRODUCTION:

Limited Dorsal Myeloschisis (LDM) is a distinctive clinicopathological entity of spinal dysraphic malformations [1]. The criteria for LDM are a cutaneous abnormality and a fibro neural or fibro vascular-neural stalk that connects the base of the skin lesion to a focus on the dorsal midline of the spinal cord [1].

The pathophysiology of LDM is hypothesized to be a primary neurulation anomaly that results in incomplete disjunction between the cutaneous and neural ectoderms of the developing embryo [2]. Based on their cutaneous manifestations, LDMs are classified as saccular or non-saccular (flat) [1]. Congenital dermal sinus (CDS) is a condition of secondary neurulation, and the stalk of CDS comprises dermal elements rather than fibroglial tissue, making the distinction between CDS and LDM crucial. A CDS develops when the cutaneous ectoderm is "pulled" to the spinal cord, whereas the fibroglial connection happens when the neural ectoderm that makes up the spinal cord is "stretched" to the skin with an LDM.

For the diagnosis of LDM, magnetic resonance imaging (MRI) is the gold standard [3]. With visualization of the stalk connecting the skin or posterior mass to the underlying spinal cord, spinal imaging can show the characteristics of LDM [4].

CASE PRESENTATION:

A 7-day-old male baby was admitted with a history of an abnormal skin lesion on his lower back. The skin lesion was flat, with no swelling or discharge. Physical examination revealed a skin lesion which was flat with a dimple at the sacrum level resembling a cigarette burn (**Figure 1**). A spinal MRI revealed a fibro neural tract extending from the skin surface into the lumbar spinal canal at the level of L5-S1 with a syringohydromyelia and neurogenic bladder (**Figure 2**).

According to the Neurological grading system of LDMs [5], he had a grade 2 defect. Excision of skin lesion/tract and untethering of filum with dural tube reconstruction under INOM guidance was performed.

Figure 1: Cigarette burn skin lesion present on the sacral region of the child

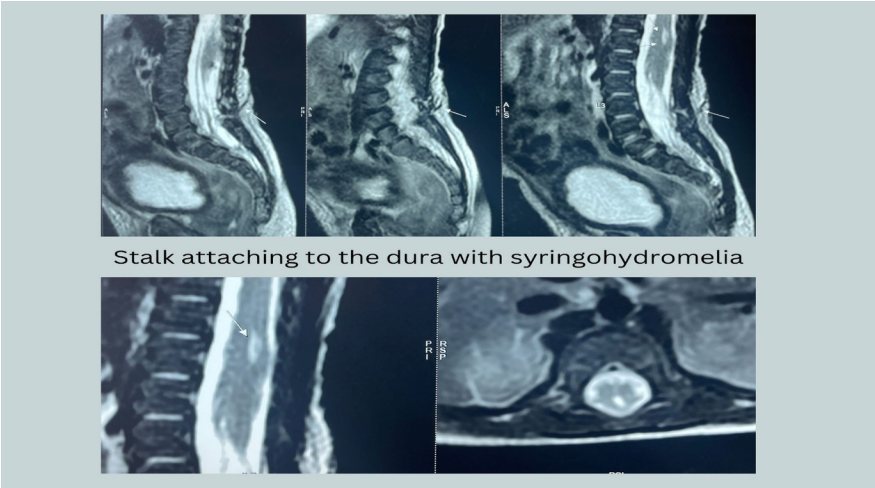

Figure 2: MRI showing a fibro neural tract extending from the skin surface into the lumbar spinal canal at the level of L5-S1

ANAESTHESIA:

Intravenous anesthesia induction was achieved by 2 mg/kg of Propofol with 0.5ml of preservative-free lidocaine, one mcg/kg fentanyl, and 0.5 mg/kg of Atracurium. Anaesthesia was maintained alongside bispectral index (BIS) monitoring with a manual infusion of propofol and 0.5mcg/kg/hr of fentanyl [6]. A manual

infusion regimen followed required a loading dose of 2mg/kg over 10 minutes followed by an infusion rate of 9 mg/kg/hr for the first 15 minutes, 6 mg/kg/hr from 15 to 30 minutes, 4 mg/kg/hr for maintenance for next 1 hour [7].

INTRA-OPERATIVE NEUROPHYSIOLOGICAL MONITORING (IONM):

INOMED IONM system was used in this patient (**Figure 3**). Modalities consisting of Free-run EMG and Triggered EMG for identifying and mapping vital nervous tissue were planned. The disposable needle electrodes (27 gauge) were positioned in the quadriceps (L2-4), anterior tibialis (L4-5), abductor hallucis (S1-2), and external anal sphincters (S2-4), groups of muscles that are innervated by the nerve roots and are at the highest risk of injury during the release of the tethered cord. Filters between 20 Hz (High Pass) and 2000 Hz (Low Pass), 50 Hz (Notch), gain 100 V/division, and time base five m/div were used to create the recording conditions for free-run and triggered EMG [8].

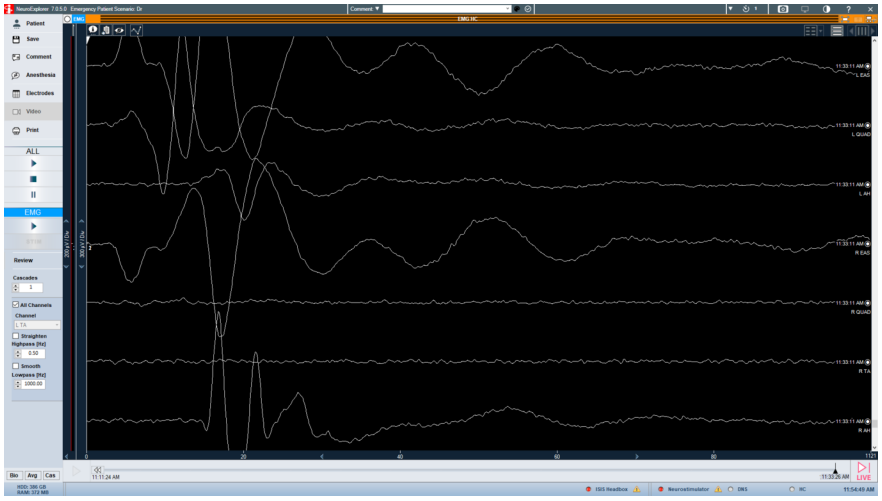

Figure 3: Intra-operative neurophysiological monitoring (IONM) used for the neonate

SURGICAL PROCEDURE:

The aim of surgery was to prevent the tethering effect on the cord by the fibro-neural stalk. A midline vertical skin incision encompassed the flat lesion. Then, the incision was deepened, and the tract was dissected off the fascial defect. There was the absence of L5-S1 posterior elements. L4 laminectomy was done. The dural tube, tract, and skin lesion were delineated separately.

The simulation threshold for the filum / non-functioning tract was at 100 times the root/rootlets. The dura was opened, and tract stimulation (10mA) was carried out to demonstrate the absence of neural elements. Then, the tract was disconnected with the dura fully opened. The filum was identified and, upon stimulation (10mA), showed no response (**Figure 4 A-E**). The quadriceps, anterior tibialis, Abductor Hallucis and External Anal Sphincter, were assessed before the closure by triggered EMGs at 1 mA current and the recordings are attached in the figure. The operation was concluded by performing dural reconstruction using 6-0 prolene. The lumbodorsal fascia was reconstructed. Lastly, the subcutaneous tissue and skin were closed in layers, and a sterile dressing was applied. The post-operative period was uneventful.

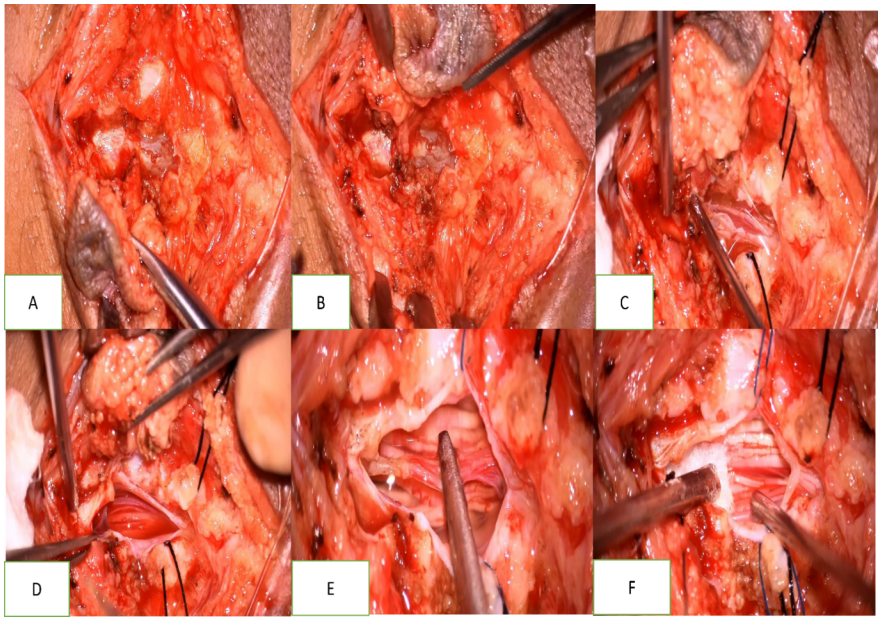

Fig 4: Limited dorsal myeloschisis (LDM) defect with lumbar crater-type flat (nonsaccular) resection. A, B: Ellipse of resected skin crater and subcutaneous tract passing through lumbodorsal fascia defect. C: An intradural exposure demonstrating cord-stalk union and stalk stimulation. D: dissection of stalk flush with cord surface. E: coagulating the filum, F: stimulating the cauda equina rootlets

DISCUSSION:

The recommended surgical management for LDM is full excision of the intradural stalk from its dural entry point to its merge point with the spinal cord [9]. IONM can aid in distinguishing the border between the two structures. This is important because incomplete elimination of the tethering elements can cause secondary deterioration and additional follow-up surgeries. Because anesthesia can affect synaptic connections and alter the evoked potential, anesthesia protocol, and management are crucial during intraoperative neurophysiological monitoring [8].

The majority of anesthetists favor the total intravenous anesthesia (TIVA) procedure due to the depressing effects of inhaling drugs on evoked potentials. In TIVA, propofol is typically preferred in addition to opioids or other analgesics. In severely ill newborns, patients without known or suspected mitochondrial illness, or short-duration procedures (3 hours), propofol infusion syndrome (PrIS) is typically not a cause for concern during anesthesia [10].

Children's context-sensitive half-lives are longer than those of adults; they are 10.4 vs. 6.7 min after a one-hour infusion and 19.6 vs. 9.5 min after four hours [7]. Although clinical significance is rarely present, infusion rates can be lowered as cases progress to prevent protracted recovery durations. In order to obtain the goal plasma concentration of 3 g/ml suggested by Morse et al., neonates need to receive a loading dose of 2 mg/kg followed by an infusion rate of 9 mg/kg/hr for the first 15 min, 7 mg/kg/hr from 15 to 30 min, 6 mg/kg/hr from 30 to 60 min, and 5 mg/kg/hr from 1 to 2 hours [7].

CONCLUSION:

This case explores the potential of conducting a successful early untethering and reconstruction surgery on a neonate under the guidance of INOM. To prevent further degeneration and untethering surgery, we advise employing INOM while removing the complete intradural LDM stalk during the initial treatment.

CONFLICTS OF INTEREST:

None declared.

ACKNOWLEDGEMENT:

None

AUTHOR CONTRIBUTION:

All the authors contributed equally in drafting, editing, revising, and finalizing the case report.

ETHICAL APPROVAL:

Ethical approval was not required for the case report as per the country's guidelines.

CONSENT:

Written informed consent was obtained from the patient to publish this report.

DATA AVAILABILITY STATEMENT:

The data supporting this article's findings are available from the corresponding author upon reasonable request.

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