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Delayed Presentation of Lumbosacral Myelomeningocele with Tethered Cord Syndrome: Endogenous Tissue Expansion Enabling Primary Closure Without Flap Reconstruction

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A B S T R A C T

Myelomeningocele (MMC) represents the most common open neural tube defect, typically presenting at birth or within the neonatal period. Delayed presentation beyond infancy in resource-limited settings remains exceptionally rare and poses unique surgical challenges, particularly regarding soft-tissue reconstruction and prevention of cerebrospinal fluid (CSF) leakage. This case highlights an unusual 4-year delayed presentation with progressive neurological deterioration including tethered cord syndrome and severe functional impairment requiring comprehensive multidisciplinary intervention. A 4-year-old male from rural Banda Aceh presented with a massive lumbosacral myelomeningocele, significant developmental delay, progressive lower extremity paresis (strength 2/5 bilaterally), and documented tethered spinal cord at L4. The patient demonstrated profound motor and sensory deficits with absent deep tendon reflexes and saddle distribution hypoesthesia. Neurosurgical intervention involved careful meningeal sac dissection, separation of functional from nonfunctional neural tissue, adhesiolysis, filum terminale sectioning, neural placode tubularization, and watertight dural closure. Remarkably, the chronic CSF accumulation had functioned as an endogenous tissue expander, providing sufficient skin laxity to enable primary midline closure without requiring flap reconstruction. Early postoperative assessment demonstrated neurological stabilization with subtle functional improvement and complete wound healing without complications. In conclusion, this case demonstrates the importance of multidisciplinary management in delayed neural tube defect presentation and introduces the concept of endogenous tissue expansion via chronic CSF accumulation as a novel biomechanical phenomenon enabling simplified soft-tissue reconstruction. Primary closure without flap reconstruction successfully prevented CSF leakage while preserving neurological stability in the postoperative period. Recognition of such mechanisms may inform surgical strategy in resource-limited settings where complex reconstructive options remain unavailable. The case highlights that even delayed presentations warrant aggressive surgical intervention when specialized expertise becomes available.

1. Introduction

Neural tube defects (NTDs) represent one of the most common congenital malformations affecting the central nervous system, with an estimated global prevalence ranging from 1 to 10 cases per 1,000 live births, demonstrating substantial geographic and population-based variation.[1] Myelomeningocele (MMC), classified as an open NTD, accounts for approximately 80% of all documented neural tube defects and remains the most common congenital

malformation requiring neurosurgical intervention.[1,2] The pathogenesis of MMC involves incomplete neural tube closure during embryonic weeks 3 to 4, resulting in herniation of neural tissue, meninges, and cerebrospinal fluid through the defect in the vertebral column and overlying soft tissues.[2] In developed nations with prenatal screening programs and folic acid supplementation initiatives, the incidence has declined substantially; however, in resource-limited settings with limited access to micronutrient supplementation and prenatal diagnostic services, NTDs continue to represent a significant public health burden, particularly in regions with inadequate obstetrical care infrastructure.[3]

The traditional clinical presentation of MMC occurs during the immediate postnatal period or within the first weeks of life, when the characteristic skin-covered or exposed cystic mass over the lumbosacral region becomes apparent to caregivers and healthcare providers. Early recognition and prompt neurosurgical closure within 24 to 72 hours of birth represent the accepted standard of care, aiming to prevent infection, meningitis, and further neurological deterioration from traction injury to the neural elements.[2,4] However, in regions with limited prenatal care infrastructure and delayed access to specialized neurosurgical services, presentation may be significantly delayed. Such delayed presentations pose formidable diagnostic and therapeutic challenges, as the pathophysiology evolves substantially over time, chronic CSF leakage may lead to persistent infection, and the associated neurological deficits may become partially irreversible.[4] The longer the interval between defect formation and surgical closure, the greater the cumulative neurological injury and the more challenging the soft-tissue reconstruction becomes.

Tethered cord syndrome (TCS) represents a progressive neurological condition characterized by traction-induced ischemia of the spinal cord due to abnormal fixation of the spinal cord to surrounding structures, preventing normal rostrocaudal excursion during spinal movement.[5,6] In patients with MMC, the combination of primary dysplasia and secondary tethering from scar tissue formation, fibrosis, and adhesions substantially increases the risk of progressive neurological deterioration beyond the immediate postoperative period.[6] The pathophysiology involves mechanical compression, vascular compromise, and metabolic dysfunction within the spinal cord, triggering an ischemic cascade that results in progressive motor weakness, sensory loss, bladder and bowel dysfunction, and orthopedic deformities.[5,6] The severity of tethering and the resulting neurological dysfunction depend upon the magnitude of traction forces, the duration of traction, and the spinal cord's capacity for ischemic tolerance.

Soft-tissue reconstruction following neurosurgical closure of MMC defects presents considerable technical challenges, particularly in cases with massive defects exceeding 10 centimeters in diameter or located in anatomically challenging regions such as the sacral area.[7,8] Traditional approaches have relied upon various flap-based techniques, including myofascial advancement flaps, perforator based flaps, and free flap transfer, to achieve watertight dural closure and prevent cerebrospinal fluid leakage while providing adequate soft-tissue coverage.[7-9] However, in resource-limited settings with limited access to complex reconstructive surgical expertise and imaging modalities, such flap-based approaches may not be feasible.[9] Furthermore, the high cost of specialized instrumentation and the extended operative time required for flap surgery may be prohibitive in settings with limited surgical resources.[9] Alternative simplified approaches emphasizing primary closure techniques when anatomically feasible may offer pragmatic solutions while maintaining adequate outcomes and patient safety.[8,10]

This case report presents an exceptionally rare instance of delayed lumbosacral myelomeningocele presenting at 4 years of age with documented tethered cord syndrome, complicated by severe neurological deficits, including progressive lower extremity paresis and profound sensory loss. The clinical course demonstrates the novel phenomenon of chronic

cerebrospinal fluid accumulation functioning as an endogenous tissue expander, providing sufficient skin laxity to enable primary midline soft-tissue closure without requiring complex flap reconstruction. This case underscores the importance of multidisciplinary management, the biomechanical principles underlying soft-tissue reconstruction in neural tube defects, and the relevance of adaptive tissue expansion mechanisms in resource-limited surgical settings. The aim of this report is to document this unusual presentation, elaborate on the surgical technique employed, discuss the postoperative clinical course, and contextualize the findings within the existing literature on neural tube defect management, with emphasis on practical approaches suitable for resource-limited environments.

2. Case Presentation

Patient background and clinical history

A 4-year-old male child was referred to the Neurosurgery Division at Zainoel Abidin Regional General Hospital, Banda Aceh, Indonesia, with a chief complaint of a progressively enlarging mass over the lumbosacral region associated with progressive difficulty in weight-bearing and progressive weakness in both lower extremities. The child was born in a rural area with limited prenatal care access and was noted at birth to have a known open spinal defect in the lumbosacral region, subsequently covered with skin by local healthcare providers utilizing basic sterile technique. The family reported that the mass had been gradually enlarging over the 4-year period, with intermittent episodes of fluid drainage and occasional malodorous discharge suggestive of intermittent infection (Table 1). Developmental history revealed a significant delay in the attainment of major milestones; the child did not begin to walk independently until age 3 years, with progressive gait disturbance and increasing inability to bear weight on the lower extremities, particularly the left side. The family also reported recurrent episodes of rectal prolapse beginning approximately 2 years prior to presentation, suggesting chronic pelvic floor dysfunction and progressive cauda equina involvement. These episodes of prolapse occurred during defecation and were typically self-reducing but caused significant parental distress and concern. The past medical history was significant for the documented spina bifida at birth, identified by local healthcare workers who performed basic wound management without formal surgical evaluation or intervention. The patient had never undergone formal neurosurgical repair, and there was no documented history of meningitis or ventriculoperitoneal shunt placement, though such documentation may have been unavailable in the rural setting of birth. The patient had not received formal rehabilitation or orthopedic intervention, though family members described attempts at basic exercises and stretching. The family history was unremarkable for neural tube defects or other congenital malformations, with no consanguinity reported. The patient lived in a rural agricultural community approximately 250 kilometers from the nearest specialized neurosurgical center, with limited access to specialized medical care, imaging services, and rehabilitation resources. On admission to our institution, the parents expressed significant anxiety regarding the progressive functional decline and the visible mass, which had become increasingly difficult to manage as the child approached school age. The family sought evaluation after reading about pediatric neurosurgical services available at our tertiary care institution.

Physical and neurological examination

General examination revealed a developmentally delayed 4-year-old male child with height and weight below the 5th percentile for his age, consistent with the effects of malnutrition and chronic neurological impairment. The patient demonstrated significant neuromotor impairment with the inability to ambulate independently without support, requiring constant parental assistance for all mobility.

Table 1. Demographic and clinical characteristics.

Parameter	Finding
Age	4 years
Gender	Male
Location	Rural Banda Aceh, Indonesia
Chief complaint	Lumbosacral mass, lower extremity weakness
Duration	Since birth (4-year delayed presentation)
Developmental milestones	Significantly delayed
Weight bearing	Progressive deterioration
Spina bifida history	Known since birth
Rectal prolapse	Documented episodes
Physical exam	Massive dome-shaped cystic mass
Defect extent	L1 to sacrum
Conus position	L4 (low-lying, tethered)

Vital signs were within normal limits, with heart rate 98 beats per minute, blood pressure 102/64 millimeters of mercury, respiratory rate 22 breaths per minute, and temperature 37.2 degrees Celsius. The skin overlying the lumbosacral region showed evidence of previous drainage and occasional weeping, with a well-epithelialized surface but with visible areas of prior erosion and maceration. The most striking finding on physical examination was a massive, smooth, dome-shaped cystic mass measuring approximately 12 centimeters in maximum diameter over the lumbosacral region, extending from approximately the L2-L3 level to the sacrococcygeal junction. The mass was soft, non-tender to gentle palpation, fluctuant with transillumination, and non-pulsatile, consistent with a fluid-filled lesion. The overlying skin appeared intact but markedly taut with visible superficial venous distension and increased blanching with applied pressure, suggesting chronic mass effect and vascular compromise. Neurological examination demonstrated profound motor deficits affecting the lower extremities bilaterally with clear lower motor neuron characteristics. Bilateral lower extremity strength was graded at 2/5 throughout the L4-S1 myotomes using the Medical Research Council scale, indicating the ability to move against gravity but without resistance. Muscle bulk was diminished bilaterally, with evident atrophy of the quadriceps, hamstrings, and calf musculature. Deep tendon reflexes were abolished; both patellar and Achilles reflexes were absent bilaterally, consistent with lower motor neuron involvement at multiple segmental levels. Sensory examination revealed profound hypoesthesia in a saddle distribution affecting the perineum, perirectal region, medial thighs, and distal lower extremities; vibration sense was markedly diminished distally and absent at the toes. No response to pinprick was elicited below the knee bilaterally, with some preservation of light touch at the proximal thighs. Proprioception was mildly impaired in the ankles. Bowel and bladder examination revealed evidence of loss of tone in the anal sphincter, with inability to voluntarily contract the external anal sphincter and obvious laxity on external palpation. The patient was unable to achieve urinary continence and required continuous catheterization throughout the day and night. Lower extremity tone was decreased throughout, with minimal resistance to passive movement in all planes. Examination of the feet revealed equinovarus deformity bilaterally, with

fixed contracture of the ankle plantarflexors and inversion, suggestive of chronic neurological involvement and prolonged unopposed muscle action. A summary of neurological examination findings is presented in Table 2, which documents all major neurological parameters and subsequent postoperative changes.

Table 2. Neurological examination findings.

Parameter	Pre-operative	Post-operative	Interpretation
Motor Strength (L4-S1)	2/5 bilateral	3/5 bilateral	Improved
Patellar DTR	No	No	No change
Achilles DTR	No	No	No change
Saddle Sensation	Profound hypoesthesia	Partial recovery	Improved
Distal LE Sensation	Profound hypoesthesia	Stable	Stabilized
Bladder Function	Incontinent	Spontaneous voiding	Significant improvement
Rectal Tone	Reduced	Improved	Improved

Radiological investigation and imaging findings

Multislice computed tomography (MSCT) of the lumbosacral spine was performed to define the anatomical extent of the defect and assess spinal canal contents. The study demonstrated a large posterior element defect involving the laminae and spinous processes from L1 through the sacrum, with a prominent midline bony gap consistent with spina bifida. The spinal canal was markedly enlarged at the level of the defect, accommodating a large fluid collection measuring approximately 12 centimeters in its greatest dimension. Axial images demonstrated low-lying termination of the spinal cord at the L4 vertebral level, with evidence of a thickened filum terminale measuring approximately 4 millimeters in diameter. The conus medullaris appeared tethered, with the filum terminale appearing as a thick, low-intensity structure in stark contrast to normal filum terminale morphology. The neural placode was visualized adherent to the posterior aspect of the meningeal sac, demonstrating a flattened rather than tubular morphology. Sagittal reconstructions demonstrated the full extent of the bony defect and the relationship of the neural elements to surrounding structures. The posterior elements of L1 through L4 and the sacrum showed characteristic dysplasia with incomplete ossification of the laminae and absence of normal spinous process development. Magnetic resonance imaging (MRI) of the lumbosacral spine was performed to further delineate the neural elements and assess for associated abnormalities such as syrinx formation or hydrocephalus. The study confirmed the presence of a large fluid collection within the lumbosacral meningeal sac, measuring approximately 12 centimeters in maximum diameter with uniform cerebrospinal fluid signal intensity on T2-weighted

imaging. The spinal cord terminated at the L4 level with a prominent, thickened filum terminale that appeared as a low signal structure continuous with the conus. Sagittal T2-weighted images demonstrated the neural placode within the sac, with evident adhesions to the dural lining along the posterior aspect. No syrinx was identified within the spinal cord proper, though the conus appeared somewhat expanded relative to normal anatomy. Axial T2 imaging confirmed the presence of neural tissue within the posterior portion of the sac, with surrounding cerebrospinal fluid creating the characteristic 'CSF layer' around the neural elements. No hydrocephalus was noted on intracranial images, with normal ventricular size and no evidence of Chiari malformation. No tethering point other than the filum terminale was identified on detailed analysis. The overall imaging studies confirmed the diagnosis of lumbosacral myelomeningocele with tethered cord syndrome due to thickened filum terminale and low-lying conus medullaris. The imaging information was critical for surgical planning, as it identified the primary site of tethering (filum terminale) and assessed the extent of neural tissue dysplasia.

Surgical intervention and technical approach

The multidisciplinary team, comprising neurosurgery and plastic reconstructive surgery, met to formulate a comprehensive surgical plan. The goals of intervention were defined as: (1) prevention of infection and CSF leakage, (2) careful separation of functional neural tissue from nonfunctional elements, (3) relief of tethering through filum terminale section, (4) restoration of dural continuity with watertight closure, and (5) achievement of adequate soft-tissue coverage. Given the massive size of the defect and the patient's resource-limited setting, complex flap reconstruction was deemed unfeasible both technically and economically. The team recognized that the chronic CSF accumulation had resulted in significant skin laxity over the lumbosacral region, potentially allowing primary midline closure if dural closure could be achieved without excessive tension. This recognition of the endogenous tissue expansion opportunity became central to surgical planning. The patient was placed in the prone position under general anesthesia with endotracheal intubation and controlled mechanical ventilation. Standard neurosurgical preparatory protocols were followed, including positioning on a radiolucent operating table, securing the head with a horseshoe headrest, and ensuring adequate padding at all pressure points including the chest, pelvis, and lower extremities. Intraoperative neuromonitoring with somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) was established using standard methodology, with baseline responses recorded prior to incision. The operative field was prepped in a sterile manner with povidone-iodine scrub solution and sterile draping, maintaining a wide sterile field to permit potential expansion of the surgical approach if needed. An incision was fashioned encircling the meningeal sac, designed to allow complete exposure of the lesion while preserving adequate skin margins for closure. The incision was initiated with a 15-blade scalpel at the junction of the epithelium-covered sac and the surrounding normal skin, circumscribing the lesion at approximately 5 millimeters from the visible margin of the mass to allow appropriate tissue handling. Upon opening the meningeal sac, clear cerebrospinal fluid was encountered in substantial quantity, confirming the fluid-filled nature of the lesion and suggesting the sac had maintained adequate integrity despite 4 years of external pressure and intermittent drainage. Careful dissection of the meningeal sac from the overlying skin was performed using blunt and sharp dissection techniques, with particular attention paid to preserving sufficient skin for eventual closure. The neural placode was identified within the posterior aspect of the sac, demonstrating a flattened, ribbon-like appearance consistent with severe dysplasia. The neural placode was found to be firmly adherent to the posterior dura along its midline, with nerve roots radiating laterally from the placode and adhering to the lateral aspects of the dural sac. Careful sharp dissection using microsurgical technique and a surgical operating microscope separated the neural

placode from the dura. The nerve roots were identified and carefully dissected from their adhesions to the dura and meningeal sac, with meticulous care taken to preserve functional root continuity using electrophysiologic guidance. The filum terminale was identified as a thickened structure extending inferiorly from the neural placode, measuring approximately 4 millimeters in diameter. Gentle traction on the spinal cord confirmed restricted rostrocaudal mobility, confirming the tethering. The filum terminale was carefully dissected free from surrounding structures using microsurgical technique. A distinction was made between functional neural tissue, representing elements with preserved motor function as assessed by intraoperative electrical stimulation, and nonfunctional neural tissue, identified as dysplastic elements without apparent motor contribution. Electrophysiologic stimulation using bipolar electrodes was performed to map motor responses and identify functional elements, with careful documentation of response characteristics. Based on intraoperative monitoring findings and anatomical assessment, nonfunctional neural tissue was carefully separated using microsurgical dissection and monopolar electrocautery set at conservative energy settings to minimize thermal injury to adjacent neural structures. The thickened filum terminale, confirmed as the primary tethering structure through anatomical and electrophysiologic means, was sharply divided using microsurgical scissors, and the cut ends were cauterized with bipolar electrocautery at low power to achieve hemostasis and prevent regeneration-induced retethering. Following adequate mobilization and tethering release, the remaining functional neural placode was gently positioned with appropriate tension relief, allowing the spinal cord to assume a more relaxed configuration. The neural placode was then carefully tubularized by approximating the lateral edges using 8-0 nylon sutures in an interrupted fashion, reconstructing a three-dimensional neural structure from the flattened dysplastic placode. Meticulous attention was paid to prevent excessive tension during tubularization, which could compromise vascular perfusion and worsen postoperative neurological outcomes. Following neural placode tubularization, dural closure was achieved. The dura was closed in a watertight fashion using 5-0 absorbable polyglactin (Vicryl) suture on a FS-2 needle, employing interrupted sutures in a linear fashion along the midline. Multiple passes were employed to ensure complete closure and hemostasis. The suture line was inspected meticulously to confirm watertight integrity, with no visualization of CSF leakage through the suture line. For soft-tissue closure, the plastic surgery team assessed the available skin laxity by gentle retraction of the skin edges toward the midline. Remarkably, the chronic cerebrospinal fluid accumulation had resulted in gradual expansion of the overlying skin and soft tissues over the 4-year period, providing substantial laxity sufficient for tension-free primary closure. Primary closure of the skin defect in the midline was achieved using 4-0 nylon suture in interrupted fashion, without requiring any flap-based reconstruction or tissue mobilization flaps. A subcutaneous drain was not placed, given that the dural closure was confirmed to be watertight and the risk of CSF leakage was deemed minimal. The skin was closed with meticulous attention to tension-free closure, ensuring that no undue stress was placed on the incision with careful matching of skin edges. A pressure dressing was applied following closure to minimize postoperative edema. Operative time was 180 minutes, with the majority of time devoted to careful neural dissection and dural closure rather than soft-tissue work. Estimated blood loss was minimal, approximately 100 milliliters, attributable to the avascular nature of the operative field and meticulous hemostasis throughout the procedure (Table 3).

Table 3. Surgical procedure details.

Step	Procedure	Finding/Technique	Outcome
1	Skin incision	Encircling MM sac	Access achieved
2	Sac dissection	Clear CSF	Intact drainage
3	Neural exploration	Placode adherent to dura	Functional tissue identified
4	Tissue separation	Sharp dissection	Safe separation
5	Detethering	Filum terminale sectioned (bipolar)	Complete release
6	Tubularization	Neural placode tubularized	Anatomic restoration
7	Dural closure	5-0 absorbable, watertight	No CSF leak
8	Soft-tissue	Primary midline closure	Tension-free, healed

Postoperative course and clinical outcomes

The postoperative period was characterized by uncomplicated wound healing and clinical stability. The patient was maintained on perioperative antibiotic prophylaxis with third-generation cephalosporin (ceftriaxone 1 gram intravenously every 12 hours) for 48 hours postoperatively, following institutional protocols for neurological surgery. Pain management was achieved with acetaminophen (15 milligrams per kilogram every 6 hours) and non-steroidal anti-inflammatory agents (ibuprofen 10 milligrams per kilogram every 8 hours), with judicious use of parenteral opioids reserved for breakthrough pain management. The wound was inspected daily, with dressing changes performed using sterile technique and antiseptic solution. By postoperative day 3, the incision demonstrated normal primary intention healing with no erythema, warmth, fluctuance, or drainage suggestive of infection or CSF leakage. No signs of CSF leakage, pseudomeningocele formation, or meningeal irritation were observed. The patient demonstrated stable vital signs throughout the postoperative hospitalization, with no fever or hemodynamic instability. Neurologically, the patient showed stabilization of motor function, with no further deterioration of baseline strength or emergence of new neurological deficits. By postoperative day 5, a subtle improvement in bilateral lower extremity strength was noted, with strength grading improving from 2/5 to 2+/5 throughout the L4-S1 myotomes. The patient demonstrated improved ability to maintain head and neck control and showed improved engagement and alertness compared to the preoperative baseline. Sensory examination demonstrated slight improvement in distal light touch sensation and pinprick response in the lower extremities compared to the preoperative baseline.

Urological status improved considerably in the immediate postoperative period, suggesting that decompression of the cauda equina resulted in functional improvement of autonomic function. The continuous urinary catheter was maintained for 72

hours postoperatively and was then removed, with the patient demonstrating spontaneous voiding without retention or incontinence. Postoperative postvoid residual volume measured by ultrasound was 45 milliliters, well below the abnormal threshold of 100 milliliters, suggesting significant improvement in bladder function and voiding efficiency. Anal sphincter tone improved slightly on digital examination, though the patient continued to require assistance with bowel management given the prolonged duration of neurological impairment. The equinovarus deformity of the feet remained unchanged, consistent with the chronic nature of the deformity and the expected time course for orthopedic correction through physical rehabilitation. The patient was discharged home on postoperative day 7 with an intact incision and absence of complications, with scar appearance appearing excellent. Discharge medications included oral antibiotics (amoxicillin-clavulanate 22.5 milligrams per kilogram per day divided in two doses) for 7 additional days and analgesia as needed. The family was counseled extensively on incision care, signs of infection, neurological warning signs requiring emergency evaluation, and the importance of physical rehabilitation. Follow-up evaluation at 4 weeks postoperatively demonstrated complete wound healing with no scar complications, hypertrophy, or contracture. The patient exhibited continued stabilization of neurological function, with subtle ongoing improvement in motor strength and preserved sensory examination findings. The patient had begun physical rehabilitation with a focus on assisted ambulation and motor strengthening, with sessions conducted 3 times weekly. The family reported improved child engagement and alertness compared to baseline, with improved responsiveness to family members and increased participation in family activities. At 12-week follow-up, the patient demonstrated further gradual improvement in motor function, with strength improving to 3-/5 in bilateral hip flexion and knee extension. Sensory function remained relatively stable with mild continued improvement in distal perception. The incision healed without complications, including no scar hypertrophy, contracture, or keloid formation. There was no evidence of CSF leakage or pseudomeningocele formation on clinical examination. The family remained satisfied with the surgical outcome and the prevention of further neurological decline. The case demonstrates the potential for late surgical intervention to stabilize and partially reverse neurological deficits even after a prolonged duration of tethering.

3. Discussion

Neural tube defects result from failure of the neural tube to close during embryonic weeks 3 to 4, with critical regulatory mechanisms involving multiple genes, environmental factors, and epigenetic influences.[1,20] The neural tube normally closes in a bidirectional fashion, beginning at the cervical region (closure site 1) and extending both cranially and caudally; failure of caudal closure results in lumbosacral defects characteristic of MMC.[1,20] The primary defect involves incomplete fusion of the neural folds, with subsequent herniation of dysplastic neural tissue (neural placode), meninges, and cerebrospinal fluid through the defect in the vertebral column, dura, and overlying skin. The neural tissue in MMC is markedly dysplastic, lacking normal cytoarchitecture and organization; rather than a normal spinal cord with discrete dorsal and ventral horns and a central canal, the tissue forms a flattened neural placode tethered to the dura mater.[2] In utero, the developing neural placode is exposed to a hostile intrauterine environment within the myelomeningocele, including exposure to amniotic fluid, urine, and mechanical trauma during fetal movement.[2] This in utero exposure, termed the 'two-hit' hypothesis, results in progressive neurological damage beyond that caused by the primary neural tube defect itself.[2] Additionally, traction-induced ischemia from progressive fibrosis and adhesion formation contributes to progressive deterioration in both the prenatal and postnatal periods.[2] The structural dysplasia and functional impairment in MMC result from both the primary

failure of neural tube closure and the secondary ischemic and chemical injury sustained during gestation.

Tethered cord syndrome represents a progressive neurological condition characterized by abnormal fixation of the spinal cord, preventing normal rostrocaudal excursion that would otherwise occur during spinal flexion and extension.[5,6] In patients with MMC, tethering occurs due to multiple mechanisms, including the primary dysplasia of the neural placode and its adherence to the dura, fibrosis and scar tissue formation during wound healing following repair, and hypertrophied or thickened filum terminale.[5,6] The pathophysiological consequence of cord tethering is mechanical traction-induced ischemia, which initiates a cascade of metabolic dysfunction, inflammatory injury, and progressive neuronal loss.[5,6] The ischemic cascade in tethered cord syndrome begins with mechanical compression and traction, which impairs microvascular blood flow to the spinal cord. This vascular compromise triggers anaerobic metabolism, lactate accumulation, and intracellular acidosis. The resulting metabolic dysfunction activates excitotoxic pathways through excessive glutamate release, leading to uncontrolled calcium influx through NMDA and AMPA receptors. Calcium overload triggers enzymatic cascades, including activation of calpains, phospholipases, and endonucleases, resulting in cytoskeletal breakdown, proteolysis, and DNA fragmentation.[6] Concurrent activation of apoptotic pathways through caspase-mediated mechanisms contributes to delayed neuronal death extending hours to days beyond the initial ischemic insult.[6] The cumulative effect is progressive degeneration of motor neurons and dorsal root ganglion cells, manifesting clinically as progressive weakness, sensory loss, and dysfunction of autonomic systems controlling bladder and bowel function.[6] In this patient, the prolonged duration of tethering over 4 years had allowed the ischemic cascade to substantially progress, explaining the profound motor and sensory deficits at presentation.

The presentation of myelomeningocele at 4 years of age represents an exceptionally rare occurrence in the medical literature.[13] The vast majority of MMC cases present either as prenatally diagnosed lesions (in settings with prenatal ultrasound screening) or within the immediate neonatal period (typically within 24 to 72 hours of birth).[2,4] Delayed presentation beyond infancy is exceptionally uncommon and has been documented in only isolated case reports from resource-limited settings.[13] In resource-limited regions, delayed presentation occurs due to multiple intersecting factors: absence of prenatal diagnostic services, including ultrasound screening and prenatal counseling; limited access to specialized neurosurgical services in geographically remote areas; cultural and socioeconomic factors affecting healthcare-seeking behavior; and inadequate infrastructure for neonatal surgical intervention.[13] In this patient's case, delivery occurred in a rural setting with minimal prenatal care, and the family initially managed the defect with local coverage without seeking specialist evaluation. Progressive neurological decline occurred gradually over 4 years, likely with parental attribution of developmental delay to malnutrition or other causes, delaying specialist referral. The chronic nature of the presentation allowed for secondary adaptive changes, including the remarkable phenomenon of endogenous tissue expansion through chronic CSF accumulation, which became therapeutically advantageous in surgical planning.

The soft-tissue reconstruction of large neural tube defects represents a significant challenge in plastic and reconstructive surgery, with substantial variation in approach based on defect size, location, patient age, and available surgical resources.[7-9] The fundamental objectives of soft-tissue reconstruction include: (1) prevention of cerebrospinal fluid leakage and associated meningitis; (2) provision of vascularized tissue coverage to prevent wound breakdown and infection; (3) achievement of tension-free closure to minimize wound complications and optimize aesthetic outcomes; and (4) provision of adequate padding over the repaired neural elements to

protect against external trauma.[7,8] For large defects exceeding 10 centimeters in diameter, traditional approaches have emphasized the use of myofascial or fasciocutaneous flaps to achieve adequate coverage.[7-9] The latissimus dorsi myocutaneous flap, which is based on the thoracodorsal neurovascular pedicle and provides robust vascularized tissue coverage, has been widely employed for lumbosacral defects.[9] The lumbar artery perforator (LAP) flap, based on perforators arising from the lumbar arteries, has more recently gained popularity for lower lumbosacral defects, providing adequate tissue without sacrificing major donor-site musculature.[9] Free flap transfer, including the rectus femoris free flap or latissimus dorsi free flap, may be employed when local flaps are unavailable or inadequate.[9] However, all flap-based approaches require sophisticated microvascular surgical expertise, specialized instrumentation, extended operative time, and vascular imaging preoperatively, limiting applicability in resource-limited settings.[9] In settings with limited access to flap-based reconstructive surgery, primary closure techniques emphasizing tissue mobilization and tension-free approximation offer a pragmatic alternative.[8,10] Primary closure depends upon the availability of adequate soft-tissue laxity to permit midline approximation without excessive tension. Tissue laxity may be achieved through: (1) pre-operative tissue expansion using external or implantable expanders; (2) progressive intraoperative mobilization of adjacent tissues through wide undermining and myofascial release; or (3) fortunate anatomical circumstances providing natural tissue redundancy, as occurred in this case through chronic fluid accumulation.[8,10] When primary closure can be safely achieved without tension, advantages include simplified operative technique, shorter operative time, avoidance of complex microvascular anastomosis, elimination of donor-site morbidity, and reduced cost, making it particularly suitable for resource-limited settings.

The phenomenon of tissue expansion has been well-characterized in plastic and reconstructive surgery, with applications ranging from burn reconstruction to large defect coverage to aesthetic body contouring.[11] Tissue expansion occurs through progressive mechanical stretch applied by an expandable implant gradually filled with saline over weeks to months, triggering adaptive vascular proliferation, dermal thickening, increased elastin production, and increased fibroblast activity, all of which increase the biomechanical properties of the expanded tissue.[11] The expanded tissue becomes increasingly lax and capable of accommodating larger defects without tension. The case presented herein introduces the concept of endogenous tissue expansion occurring spontaneously through chronic cerebrospinal fluid accumulation within the myelomeningocele sac. Rather than using an artificial expandable implant progressively filled with saline, the chronic accumulation of cerebrospinal fluid within the myelomeningocele over 4 years provided a sustained, gradually enlarging mass effect that progressively stretched the overlying skin and soft tissues. This chronic mechanical stretch likely triggered similar adaptive tissue responses to those seen with artificial tissue expansion, including vascular proliferation, increased elasticity, and reduced tissue tension. Unlike artificial tissue expansion, which requires monthly clinic visits for progressive expansion and extends over 2 to 4 months, this endogenous expansion occurred passively without clinical intervention. Upon preoperative assessment, the massive laxity of the overlying skin and subcutaneous tissues allowed primary midline closure without requiring flap-based reconstruction. This observation suggests that the delayed presentation, while associated with progressive neurological deterioration from tethering, paradoxically conferred a surgical advantage through the development of natural tissue redundancy that simplified reconstruction. The chronic fluid accumulation appears to have functioned as an endogenous tissue expander, creating conditions that permitted primary closure without the requirement for complex flap-based reconstruction. This mechanism may offer insights for the management of delayed presentations

in resource-limited settings where complex reconstructive options are unavailable or unaffordable. Recognition of this phenomenon may inform preoperative planning and surgical strategy in analogous cases.[12-15]

The decision to employ primary closure versus flap-based reconstruction in neural tube defect repair involves consideration of multiple factors, including defect size, defect location, availability of surrounding tissue laxity, local tissue quality and perfusion, patient age and healing capacity, surgeon expertise, available institutional resources, and cost considerations.[8,10] Large defects exceeding 10 to 12 centimeters have traditionally been considered candidates for flap-based reconstruction due to concerns regarding tension-free closure, whereas smaller defects may be amenable to primary closure.[8] However, this conventional approach does not account for individual variations in tissue laxity, which may permit primary closure of larger defects when adequate laxity is present. In the case presented herein, the defect measured approximately 12 centimeters in maximum diameter, which by conventional criteria would be considered large and potentially requiring flap reconstruction. However, the chronic accumulation of cerebrospinal fluid had created substantial tissue laxity, permitting primary closure without tension. This decision was made collaboratively by the plastic and neurosurgical teams after careful preoperative assessment and intraoperative reassessment following dural closure. The successful outcome, with no CSF leakage or wound complications, validates the decision to employ primary closure in this specific case. The cost savings and simplified operative approach would not have been possible with flap-based reconstruction, making this an important lesson for resource-limited settings.[8,10]

Table 4 presents a comparison of this case with similar cases documented in the medical literature, including age at presentation, defect level, closure technique employed, complications encountered, and clinical outcomes. The delayed presentation at 4 years of age remains exceptionally rare, with most comparable cases presenting within the first year of life. Similarly, the use of primary closure for a defect of this magnitude (12 centimeters) without flap-based reconstruction represents an unusual approach, as most large defects in the literature are managed with flap-based reconstruction. The absence of CSF leakage complications and the successful achievement of wound healing without flap placement distinguish this case in the literature context. The postoperative improvement in neurological function, despite the prolonged duration of tethering, indicates that neurolysis and filum section may result in clinical benefit even in cases with delayed presentation and established neurological deficits.[16-20]

The successful management of complex neural tube defects requires a collaborative, multidisciplinary approach involving neurosurgery, plastic and reconstructive surgery, orthopedic surgery, pediatrics, anesthesiology, and specialized nursing and rehabilitation services.[21-23] In this case, close collaboration between neurosurgery and plastic reconstructive surgery was essential in achieving optimal outcomes. The neurosurgical team's expertise in microsurgical technique, careful neural tissue dissection, spinal cord monitoring, and dural closure ensured protection of neural function and prevention of CSF leakage. The plastic surgical team's expertise in soft-tissue assessment, flap design, wound closure, and complication prevention ensured adequate coverage and successful wound healing. Shared preoperative planning and communication permitted the formulation of a coherent surgical strategy that avoided unnecessarily complex reconstruction while maintaining safety.

Table 4. Comparison with literature.

Feature	Present case	Diab 2021	Ntimbani 2020	Lim 2024	Chalhoub 2025
Age at surgery	4 years	Variable	Neonatal-infant	Neonatal-10y	Pediatric
Defect level	L1-sacrum	Various	Various	Various	Various
Tethered cord	Yes (L4)	Common	Common	Possible	Secondary
Closure type	Primary	Flaps	Variable	Variable	Staged
Flap required	No	Often yes	Defect-dependent	Often yes	Yes
CSF leak	None	3-12%	3-12%	5.6%	Recurrent
Wound infection	None	2-15%	3-29%	8.3%	Secondary
Key novelty	Endogenous expansion	Late data	Review	Outcomes	Salvage

Beyond the acute surgical phase, comprehensive management should include urological assessment and management of neurogenic bladder, bowel management programs, orthopedic surveillance for scoliosis and lower extremity deformities, and intensive rehabilitation focusing on motor strengthening and functional recovery.[21] In this case, immediate postoperative improvement in bladder function permitted discontinuation of catheterization within 72 hours, suggesting that decompression of the neural elements resulted in a significant functional benefit. Ongoing physical rehabilitation with a focus on motor strengthening and gait training will be essential for optimizing long-term functional outcomes. Follow-up neuroimaging may be considered at regular intervals to assess for retethering or other postoperative complications, though the patient's clinical stability does not currently mandate urgent follow-up imaging. The case presented demonstrates important lessons for the surgical management of neural tube defects in resource-limited settings where access to complex reconstructive surgical expertise, specialized imaging modalities, and expensive implantable devices may be unavailable. First, simplified surgical approaches emphasizing meticulous neurosurgical technique and primary soft-tissue closure, when anatomically feasible, may achieve comparable outcomes to flap-based reconstruction while substantially reducing operative time, cost, and complexity. Second, recognition of anatomical variations and adaptive tissue changes, such as the endogenous tissue expansion observed in this case, may permit decision-making that optimally utilizes available local tissue resources. Third, multidisciplinary collaboration between available surgical specialists, even in resource-limited settings, can significantly enhance surgical safety and outcomes. Fourth, even in settings with delayed presentation and prolonged neurological compromise, neurosurgical intervention may result in functional improvement and prevention of further deterioration, justifying operative intervention despite the inherent risks. The case serves as a paradigm for pragmatic surgical management adapted to available resources while maintaining patient safety and optimizing outcomes.

4. Conclusion

This case report presents an exceptionally rare instance of delayed lumbosacral myelomeningocele

presenting at 4 years of age with concurrent tethered cord syndrome, documented progressive neurological decline, and profound motor and sensory deficits. The clinical presentation illustrates the importance of prompt recognition and specialized surgical intervention, even in cases presenting substantially delayed from birth. The surgical approach employed, incorporating meticulous neurosurgical technique for neural tissue preservation and tethering release, combined with simplified primary soft-tissue closure enabled by endogenous tissue expansion, resulted in uncomplicated wound healing, prevention of cerebrospinal fluid leakage, and stabilization with modest improvement in neurological function during early follow-up. The case introduces the concept of endogenous tissue expansion via chronic cerebrospinal fluid accumulation as a novel biomechanical phenomenon that may facilitate simplified soft-tissue reconstruction in neural tube defect closure. Recognition of this mechanism may inform surgical strategy and decision-making in resource-limited settings where access to complex flap-based reconstructive surgery is unavailable or economically unfeasible. The successful outcome achieved through multidisciplinary collaboration between neurosurgery and plastic reconstructive surgery, utilizing available local resources and simplified operative techniques, demonstrates the feasibility of achieving excellent outcomes even in resource-limited environments. Future management of this patient should include continued clinical surveillance with assessment for retethering or other postoperative complications, intensive physical rehabilitation to optimize motor recovery, urological surveillance and management of neurogenic bladder to prevent upper urinary tract complications, orthopedic assessment and management of lower extremity deformities, and comprehensive neurodevelopmental assessment to document long-term functional outcomes. While the immediate postoperative period demonstrated clinical stability and early improvement, the long-term neurological trajectory remains uncertain given the prolonged duration of preoperative tethering. Continued follow-up and detailed documentation of outcomes will contribute to the medical literature on delayed neural tube defect presentation and long-term outcomes of operative intervention in such cases. This case underscores several important principles: (1) neural tube defects may present with substantial delay in resource-limited settings, and delayed presentation does not preclude operative intervention when specialized expertise becomes available; (2) meticulous microsurgical technique in dissection of dysplastic neural tissue may permit functional preservation and modest functional improvement even in cases with prolonged preoperative neurological compromise; (3) simplified primary closure techniques, when supported by adequate tissue laxity, may achieve comparable outcomes to complex flap-based reconstruction while substantially reducing operative complexity and cost; (4) recognition of adaptive tissue changes and anatomical variations may inform optimal surgical decision-making in individual cases; and (5) multidisciplinary collaboration remains essential in optimizing outcomes, even in resource-limited settings with restricted access to specialized expertise and resources. The successful management of this complex case demonstrates that pragmatic, technically sound surgical approaches, grounded in fundamental principles of neurosurgery and soft-tissue reconstruction, can achieve excellent results regardless of resource limitations.

5. References

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