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Soft tissue coverage of delayed presentation of meningomyocele by modified keystone flap

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Abstract

Neural tube defects are congenital abnormalities of neural development with a wide spectrum of clinical manifestations. Meningomyelocele a type of spina bifida , in which the spinal cord and meninges protrude through an opening.

Early surgical interventions are required to reduce the mortality rate by preventing early cerebrospinal fluid leakage and related infections. Simple closures can repair small defects. Simple closure can be challenging in larger defects, leading to wound dehiscence and subsequent cerebrospinal fluid leakage. This case report discusses the successful use of modified keystone flap repair for large lumbosacral meningomyelocele defects.

We report the 18th day of life, term female baby with lumbosacral meningomyelocele, who underwent neural placode reduction with repair of dura followed by modified keystone flap repair to achieve adequate neural closure and prevent subsequent infection. The flap preserved vascularity and decreased the need for complex grafts and revision surgery with no sign of flap necrosis, fistula, or cerebrospinal fluid leakage.

This case highlights that modified keystone flap repair is a safe, simple, successful, and versatile technique that can be used to cover large meningomyelocele defects.

Keywords: keystone flap; meningomyelocele; neural tube defect

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Introduction

Neural tube defects (NTDs) are the most common severe central nervous system anomalies, with a prevalence that varies according to ethnicity, geography, gender, and country. The incidence rate is 1 to 2 cases per 1000 births [1] NTDs involve aberrant development of the vertebrae, paravertebral muscles, skin, and central nervous system, with the possibility of other anatomical anomalies coexisting. The most common NTDs are spina bifida and anencephaly.[2]

Spina bifida is the embryological failure of fusion of one or more vertebral arches (laminae), and its subtypes are classified based on the degree of neuroectoderm involvement. Spina bifida occulta has a hidden external boundary, with no neural tissue involvement.[2] Meningocele is a closed neural tube defect where only the meninges herniate through the defect, with no loss of CSF during development. In contrast, myelomeningocele is an open neural tube defect where both the meninges and spinal cord protrude through the defect, with intermittent or continuous CSF leakage.[3] Meningocele affects only the spine, while myelomeningocele, the most common form of open NTD, involves the entire CNS.[3] Neurological deficits vary depending on the level of the defect.[4]

Being one of the most common congenital malformations, the global prevalence of myelomeningocele has been reported to be 0.8–1 per 1000 live births.[5] Although compatible with life, myelomeningocele is associated with lifelong morbidity, including paralysis, bowel and bladder dysfunction, hydrocephalus, and severe mental retardation in up to 15% of cases.[6]

Case Report

A term female infant weighing 4 kg, delivered via emergency LSCS for hydrocephalus, presented to our OPD on the 18th day of life with swelling over the back, serous discharge, and inability to move both lower limbs. The infant had no history of fever, or inappropriate crying, and was feeding well. The mother reported taking folic acid during her pregnancy but no other medications.

On general examination, the infant's vitals were stable. Head circumference was 37.8 cm, with a wide and open anterior fontanelle, and a closed posterior fontanelle. There were no other gross anomalies present. Neurologically, sucking, rooting, and Moro reflexes were present. Power was 0/5 in both lower limbs, and reflexes were absent bilaterally in lower limbs.

Local examination of the back revealed bluish cystic swelling in the lumbar region, measuring 5x7 cm, elliptical with a smooth surface, fluctuating in consistency, and with serous discharge. MRI confirmed spina bifida aperta with meningomyelocele, tethered cord, tonsillar herniation with kinking at the cervical-medullary junction, and syrinx at the C7-D12 level, consistent with Chiari II malformation.

The patient underwent repair of the meningomyelocele using a modified keystone perforator flap on the 21st day of life. There was a 7x7 cm defect after the neurosurgery team repaired the meninges, Figure 1.

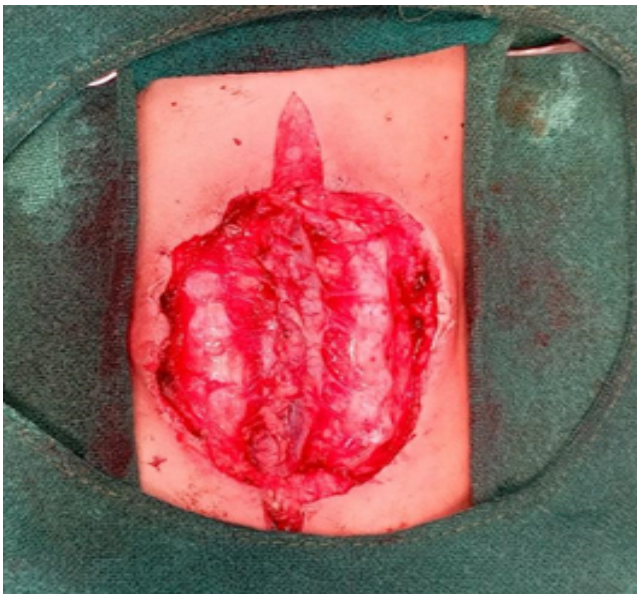

Figure 1. Soft tissue defect measuring 7 x 6 cm after repair of meningeal layer

The perforator on either side was marked along the lateral surface of the defect with Doppler. A standard keystone flap was designed and raised in a subcutaneous plane up to the marked perforator bilaterally from either side of the soft tissue defect. Medial advancement of the bilateral flap was done but the flap was not enough to cover the defect on the midline. So, the superior and inferior triangle of the keystone flap was raised on either side, for the superior triangle mobility dissection was done from the superior to the inferior direction in the subfascial plane on the bilateral side. Similarly, for inferior triangle mobility dissection was done from the inferior to the superior direction in the subfascial plane on the bilateral side. Perforator dissection into the muscles was done to provide extra mobility. The raised keystone flap and its four triangles on cranial and caudal end of the keystone flap increased the arc of movement and enhanced the mobility toward the midline to close the defect by omega variation, Figure 2.

Postoperatively, the infant developed bulging of the anterior fontanelle, and a CT scan revealed obstructive

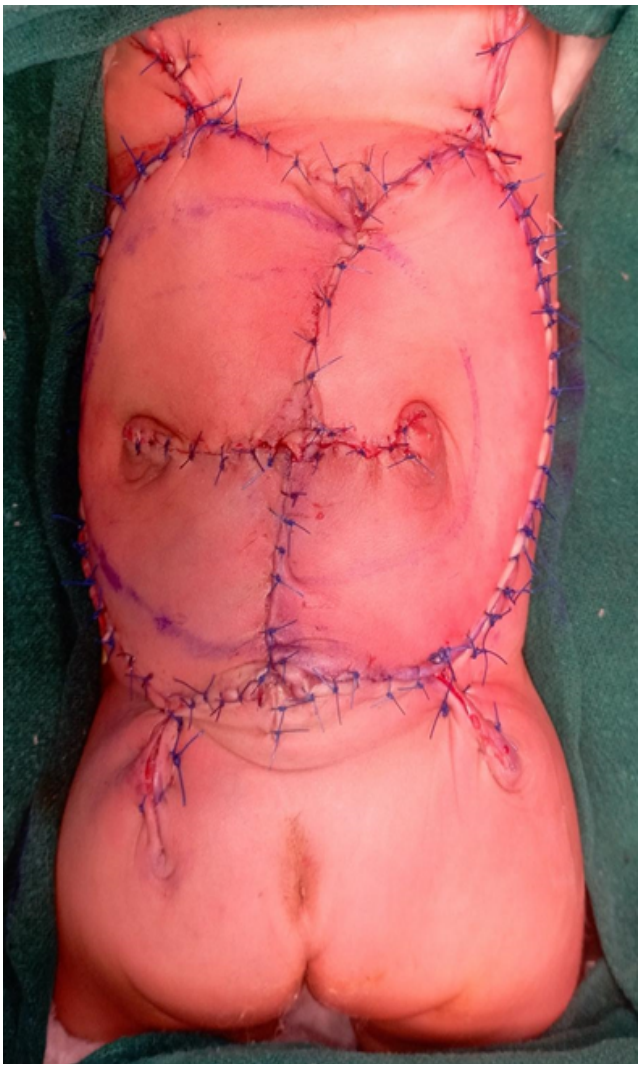

Figure 2. Immediate result after modified keystone flap to cover the defect

hydrocephalus, necessitating emergency right-sided ventriculoperitoneal shunt placement on the 8th postoperative day. The patient also developed a surgical site infection at the flap site, which was managed with gentamicin dressings and treated non-operatively. The wound healed by the 21st postoperative day, Figure 3.

Discussion

The etiology of NTDs is multifactorial, with genetic, geographical, socioeconomic, and nutritional factors such as folic acid deficiency playing key roles. The lumbosacral region is the most common site of these defects, accounting for approximately 75% of cases.[7] Although fetal surgery for myelomeningocele is now performed in some centers, many institutions continue postnatal repair due to parental concerns and financial constraints.[5] Early surgical intervention within 72 hours of birth significantly reduces hospitalization duration, antibiotic usage and complication rates.[8]

Our patient presented late, and surgery was performed based on evidence of successful repair

Figure 3. Healed wound on the 21st postoperative day

as late as 52 days postnatally.[4] Factors influencing surgical treatment of myelomeningocele include defect size, location, presence of kyphosis, and the quality of surrounding tissue.[4] The goals of surgery are to preserve neural tissue function and prevent secondary infection.[5] In ideal cases, skin closure must be tension-free, durable, sensitive, and have adequate blood flow.[9]

For small defects (<5 cm), direct closure is possible without complications [4,7]. For larger defects, flaps such as the advancement flap, pedicled flap, double Z-plasty, and Limberg flap are used to avoid tension on the suture line.[7] When comparing standard keystone flaps with another advancement flap, the greatest advantage comes due to the simplicity of the flap which can be performed on a single stage without donor site morbidity. Some limitations and disadvantages of traditional keystone flap repair include longer incision, limited maneuverability, high tension along flap edges, and vascular compromise. This can be obviated by a modified keystone perforator flap. Therefore, in this case, we employed the modified keystone perforator flap which has proven effective in repairing traumatic, oncologic, and congenital soft tissue defects.[10] The flap can be raised in the conventional method on either side of the defect over the back and move the flap towards the midline, however, in this case, where the midline could not be closed without tension, the same flap was modified in omega fashion to close the defect. This provided additional maneuverability,

redistribution of tissue laxity, and tension, where the incision was already given. This flap is nourished by multiple perforators and provides reliable blood flow, making it a versatile option.[9]

Our patient required ventriculoperitoneal shunt placement for obstructive hydrocephalus on the 8th postoperative day. A study with 14 years follow in 5 patients showed, CSF shunt insertion is often necessary in patients with myelomeningocele .[11] The keystone flap's use has been associated with reduced morbidity, hospital stay, and complication rates.[11]

Conclusion

Neural Tube Defects (NTDs) remain a significant health burden in the developing world, often resulting in severe outcomes for the child and family. Selecting a reliable flap coverage technique when correcting large myelomeningocele defects is crucial to minimizing complications and improving the child's quality of life. In our experience, the modified keystone flap proved to be an excellent option for large myelomeningocele repair due to its extensive coverage, tension-free closure, and durability.

Informed Consent

The legal guardian provided informed consent for both the surgical procedure and the publication of the case report, and the patient provided consent for the use of clinical images.

Author's Contribution

Concept, design, planning: PG, PP, SP, SK, KK, SR, ARTR, AG; Literature review: PG, SK, KK, SR, ARTR; Data collection: PP, SP, ARTR, AG; Data analysis: PG, PP, SP, SK, KK; Draft manuscript: PG, PP, SP, SK, KK, SR, ARTR, AG; Revision of draft: PG, PP, SP, SK, KK, SR, ARTR, AG; Final manuscript: PG, PP, SP, SK, KK, SR, ARTR, AG; Accountability of the work: PG, PP, SP, SK, KK, SR, ARTR, AG.

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