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CASE REPORT
Craniofacial/Pediatric

Repair of an Occipital Meningocele and Scalp Soft-tissue Reconstruction in a Newborn Patient

Jonathan D. Freedman, MD, PhD*
Michael B. Gehring, MD*
Brent R. O'Neill, MD†
Brooke French, MD*
David Khechoyan, MD*

Summary: The differential diagnosis of large congenital scalp defects includes aplasia cutis and encephalocele, among others. Treatment includes conservative management with dressings or operative management with dermal substitutes, skin grafting, local flaps, and free flaps. This case report discusses the technical considerations and reconstructive strategies for repair of a meningocele in a newborn with a large 5.5-cm scalp defect. The key strategies include preemptive cerebrospinal fluid (CSF) diversion with external ventricular drain to reduce the risk of CSF leak and mitigate wound-healing complications; careful identification and avoidance of key anatomic structures, such as the superior sagittal sinus, as anatomy may be significantly distorted due to the presence of a meningocele and after CSF diversion; and careful, thoughtful design of the local scalp flaps to maximize blood supply and to avoid tension on the final reconstruction. *(Plast Reconstr Surg Glob Open 2024; 12:e5663; doi: 10.1097/GOX.0000000000005663; Published online 8 March 2024.)*

The etiology of large congenital scalp defects includes aplasia cutis congenita (ACC) and encephaloceles. ACC presents with full-thickness skin defects with possible absence of bone and dura due to impaired fetal skin development.[1] Encephaloceles, a type of neural tube defect, can manifest as either meningoceles or meningoencephaloceles.[2] Scalp defects with thin membranes result when the surface ectoderm fails to separate from the underlying neuroectoderm as the neural tube closes in the third to fourth week of gestation.[3] Both ACC and encephalocele occur with an incidence of one in 10,000, and the most common location is the scalp vertex.

Untreated congenital scalp defects can lead to severe complications such as local infection, meningitis, sagittal sinus thrombosis or rupture, cerebrospinal fluid leak, and hyponatremia. Conservative treatment allows for the gradual re-epithelization of the defects over time.[4] Dressings include gauze with saline drips, various adherent and nonadherent coverings, antibiotic ointment, vaseline, and silver impregnated products.[5] Operative management allows for timely, definitive coverage, and reconstructive strategies may include application of dermal substitutes, skin grafting, local scalp flaps, and free tissue transfer. Surgical repair minimizes potential infection but risks injury to the major dural sinuses, dura, and brain parenchyma.

Treatment should be determined by the size and location of the defect and the patient's clinical condition and prognosis. When complications develop related to the defect, surgery becomes the preferred choice of treatment.[6] However, consideration should be given to preemptive surgical intervention to avoid the risk of major complications.

In this case report, a large congenital meningocele is presented, along with a discussion of technical considerations for reconstruction.

CASE

The patient was diagnosed prenatally with a large occipital meningocele on routine screening ultrasound and subsequent confirmatory fetal magnetic resonance (MR) imaging at 24 weeks gestational age. (**See figure, Supplemental Digital Content 1**, which shows T2 fetal MR sagittal image at 24 weeks showing abnormal, asymmetric cerebral folding and a posterior parietal skull meningocele, **http://links.lww.com/PRSGO/D100**.) Delivery was planned by C-section to avoid rupturing the meningocele. It was performed 1 week early at 39 weeks gestational age, following rupture of membranes.

*From the *Division of Plastic Surgery, Department of Surgery, University of Colorado, Aurora, Colo.; and †Department of Neurosurgery, Penn State Health, Hershey, Penn.*

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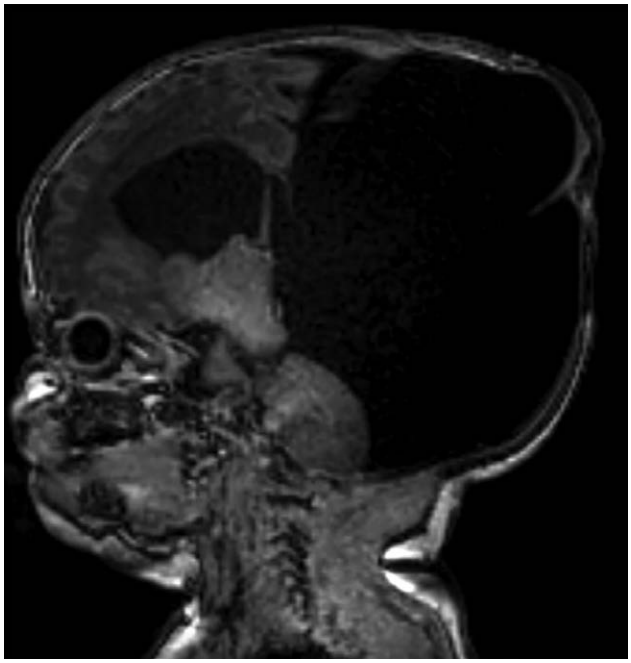

Fig. 1. T1 MR sagittal image at day 1 of life showing multiple brain abnormalities including polymicrogyria, gray matter heterotopia, and schizencephaly. The posterior cranial vault is occupied by a large meningocele that bulges through the occiput.

Postnatal MR brain imaging (Fig. 1) showed a large cystic structure in the posterior cranial vault with meninges bulging through diffuse splaying of the cranial sutures. (**See figure, Supplemental Digital Content 2**, which shows three-dimensional x-ray CT imaging at day 1 of life showing diffuse splaying of the cranial suture with widening of the posterior sagittal suture where the meningocele bulges, (**http://links.lww.com/PRSGO/D101**.) She was taken to the operating room on day of life 2 for the repair of the meningocele (Fig. 2A), placement of an external ventricular drain (EVD), and soft-tissue reconstruction with local scalp rotation-advancement flaps. Once the EVD was placed and some cerebrospinal fluid (CSF) was diverted, the major cranial sutures in the region, including bilateral lambdoid and sagittal sutures, collapsed and assumed a more normal width and configuration, which, in turn, facilitated the soft-tissue reconstruction by removing the tension on the soft-tissue repair. This maneuver led to some overlap of the occipital and parietal bones. The EVD was placed in the designed rotation flap (see Fig. 2A for markings) and subsequently tunneled away from the vascular base of the flap in the subgaleal plane and exited at a more remote location percutaneously (Fig. 2B). The rotation-advancement flaps were carefully inset with 4-0 Vicryl sutures for the galea, followed by loosely placed, simple interrupted 5-0 Monocryl sutures on the skin. The EVD was internalized to a shunt on day 12 for permanent CSF diversion. The patient was discharged on day 14 after treatment for seizures in the intensive care unit.

DISCUSSION

Congenital meningoceles were classified by Talamonti et al[7] as small (<5 cm), large (5–9 cm), and giant (>9 cm). The meningocele in this case was 5.5 cm and classified as large. Although there are many reports of nenonatal, congenital meningoceles with associated cranial cutaneous defects in the literature, there are few reports focused on the technical considerations for excision of the thin membrane and local scalp flap reconstruction. The membrane overlying the meningocele can be incredibly thin and fragile. Rupture is a surgical emergency due to the risk of meningitis and requires prompt EVD placement and soft-tissue closure. Otherwise, if there is no disruption of the membrane, the meningocele may be repaired electively in

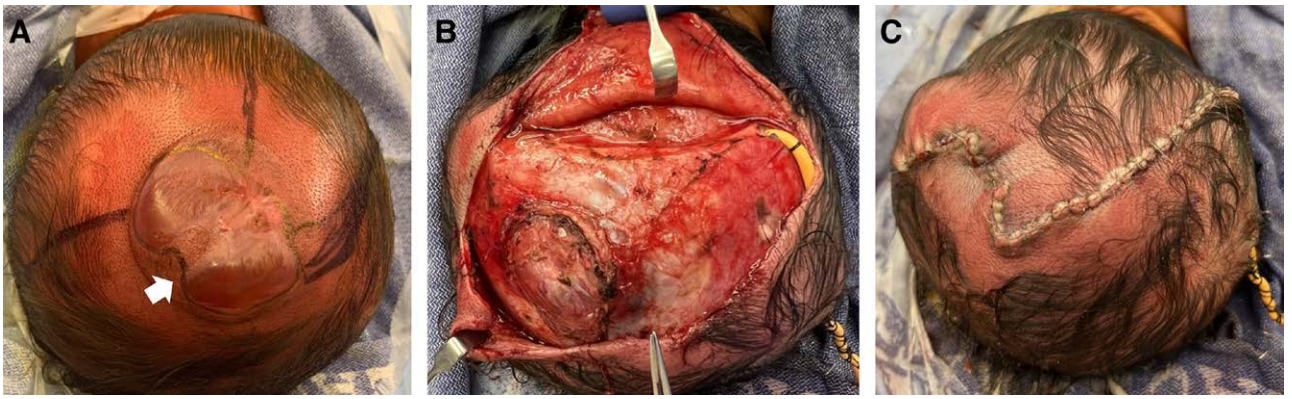

Fig. 2. Intraoperative photographs of meningocele resection and reconstruction. A, Prone view of the scalp with a thin membrane covering the meningocele approximately 5.5 cm in diameter. The surrounding skin is hyperemic, likely due to numerous emissary blood vessels. Shaving of the surrounding hair allowed definitive identification of the membrane-skin border and outlined. Note that a small skin bridge (arrow) was initially saved, but excised due to poor blood supply. The preoperative markings for local rotation flaps are drawn (left and right). The approximate course of the sagittal sinus is marked (posterior to anterior). The EVD placement was placed along the left-sided markings to avoid tethering of the flaps. B, Prone view of the scalp after meningocele excision and elevation of rotation flaps. The endoventricular drain was placed in the designed rotation flaps before tunneling. After the cerebrospinal fluid was drained, the parietal bones overlapped the occipital bones causing invagination of the dura and periosteum. The sagittal sinus was displaced from midline (forceps). C, Closure of the rotation advance flaps.

the first week of life. It is important to shave the newborn's hair to precisely identify the transition zone between normal skin and meningocele, thus minimizing the necessary excision (Fig. 2A). Before EVD placement and excision of the meningocele capsule, the potential design of the rotation flaps should be marked. This allows for the EVD to be placed in line with potential incisions and avoid tethering of the rotation-advancement flaps.

The skin surrounding the meningocele defect was hyperemic with some degree of venous congestion (Fig. 2A), which was likely caused by numerous, sizeable 2–3 mm emissary veins draining the scalp into the dura, sagittal sinus, or cortical veins. Hyperemia should alert the treating surgeon that large veins will be encountered in the dissection. The excision of the membrane should proceed from known to unknown, and the tissue plane may be indistinct near the membrane. Westcott scissors are useful for identifying the plane between the skin and the membrane overlying the meningocele. In this case, once the CSF was drained, the parietal and occipital bones overlapped (Fig. 2B), causing invagination of the dura and periosteum. This added to the difficulty of the dissection due to an uneven plane of the dissection between the bone and pericranium and galea. Additionally, after drainage of CSF, the superior sagittal sinus (Fig. 2B; forceps) may be displaced from midline and differ from location identified on preoperative imaging. Wide, broad-based rotation-advancement flaps should be designed based on the scalp vascular anatomy, with the 6:1 ratio of a rotation arc. Interrupted surface sutures should be utilized to minimize ischemia to the skin edges.

CONCLUSIONS

Large meningoceles require scalp closure. If the membrane remains intact, the defect can be closed electively in the first week of life. Rupture should be treated with urgency to avoid infection. Surgical considerations for excision and local flap closure include appropriate CSF diversion and EVD placement in the designed flaps for avoiding tethering and flap compromise, identification of the major venous dural sinsues, and careful flap design to yield tension-free closure.

Jonathan D. Freedman, MD, PhD
University of Colorado, Anschutz Medical Center
Division of Plastic and Reconstructive Surgery
12631 East 17th Ave, Aurora, CO 80045
E-mail: JonathanDFreedman@gmail.com

DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

Parents or guardians provided written consent for the use of the patient's image.

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