

Correction of a vertex encephalocele related to amniotic band syndrome

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DESCRIPTION

An encephalocele is a neural tube defect characterised by the displacement of cranial contents outside of the skull. Often, these congenital malformations are due to a primary axial mesodermal defect and occur in 0.8–3.0 of 10 000 live births.^{1 2} In rare cases, encephaloceles are manifestations of amniotic band syndrome (ABS), best defined as multiple and often-diverse secondary deformities caused by the formation of thin membranous bands in utero.³ While encephaloceles related to primary neural tube defects are often found at a midline occipital or frontal location, ABS-related encephaloceles may involve multiple defects with variable locations.^{1 2}

The craniofacial defects associated with ABS usually involve nasal deformity and asymmetry. In contrast to typical encephaloceles, those related to ABS may only be covered with rudimentary meninges rather than true skin.⁴ Correction of the lesion is necessary to allow for more normal brain growth and development, as the presence of the excess tissue may differentially drive cranial skeleton formation.^{1 2} Empiric antibiotic therapy and surgical correction must be undertaken promptly to reduce the risk of infection.² Operative objectives include removal of the herniated cranial contents, closure of the dural defect and closure of the cranial defect, when feasible. Large encephaloceles can present a challenge to the neurosurgeon, especially in terms of positioning and obtaining a watertight dural closure. We present a case of a large atypical vertex encephalocele in a premature neonate with associated complex craniofacial malformations believed to be related to ABS.

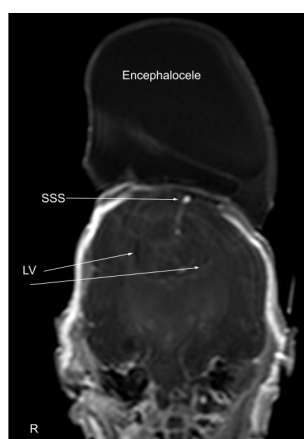


Figure 1 Coronal slice of T1 postcontrast MRI. LV, lateral ventricle; R, right side; SSS, superior sagittal sinus.



Figure 2 (A) Preoperative image demonstrating vertex encephalocele; (B) associated obliquely oriented cleft palate; (C) close-up view of the preoperative cranial vertex with visible brain covered by arachnoid [F] and numerous abrasions (small arrow) presumably related to amniotic bands; (D) preoperative lateral view demonstrating relative encephalocele size and positioning with a combination of gel rolls to elevate the vertex. A, anterior; E, encephalocele; L, left; P, posterior; R, right.

A 1.9kg neonate born at 33 weeks gestation presented for encephalocele repair at 72 hours of life. Preoperative MRI suggested that the superior sagittal sinus remained intracranial and no hydrocephalus was present (figure 1). On examination, an atypical obliquely oriented bilateral cleft palate

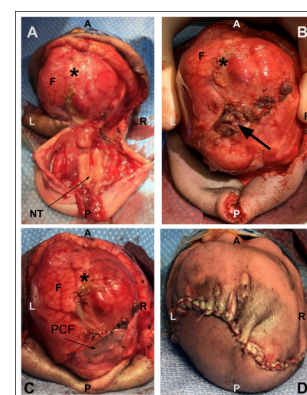


Figure 3 (A) Opened encephalocele with neural tissue; (B) neural tissue truncated, remnants of pedicle visible off midline (thick arrow); (C) pericranial flap, rotated off the occipital bone, closing pedicle defect; (D) skin closure. A, anterior; F, right frontal lobe covered only with arachnoid; L, left; NT, neural tissue; P, posterior; PCF, pericranial flap; R, right. The asterisk (*) denotes midline.



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Images in...

and a large vertex encephalocele were noted measuring about 10 cm by 6 cm (figure 2A,B). The lesion was covered with rudimentary skin, although only meninges were present in some areas. The bony defect was notably far larger than the pedicle of the encephalocele (figure 2). Head circumference was 24 cm (<3rd percentile).

In the operating room, she was positioned supine with head on a small gel donut (figure 2D). After routine induction of anaesthesia and intubation, the patient was positioned and prepped with betadine. Initial dissection was done in the plane between the visible rudimentary meninges and scalp. Once developed, a curvilinear bicoronal incision was made to expose the limited pericranium and cranium anteriorly and laterally. The encephalocele was opened with the fluid contents drained (figure 3A), and the non-functional nervous tissue was truncated with bipolar cautery and scissors at the base of the pedicle (figure 3B). A pericranial flap was rotated from over the superior occipital bone to close the dural defect (figure 3C). No drainage of cerebrospinal fluid was noted. The rudimentary skin and meninges were removed from the encephalocele sac, and the viable appearing skin tissue in the base was rotated for closing the skin (figure 3D). The patient returned to the neonatal intensive

care unit, was extubated on postoperative day 1 and weaned to room air. Postoperative MRI obtained on postoperative day 3 did not demonstrate any interval development of hydrocephalus. Empiric broad-spectrum antibiotics were continued through postoperative day 3.

Two months after surgery, her incision had completely healed and she was discharged home, still without evidence of hydrocephalus. However, at her outpatient clinic visit with the neurosurgeon 2 weeks later, it was noted that her encephalocele defect appeared to be larger. Fast-sequence MRI revealed significant dilation of the lateral ventricles, consistent with hydrocephalus. This patient is now scheduled to have a ventriculoperitoneal shunt to treat her hydrocephalus, a common result in patients with encephaloceles.

Contributors AMY-K was involved in the clinical care of the patient, manuscript preparation and editing, obtaining images and approval of the final manuscript. ACP was involved in the clinical care of the patient, manuscript preparation and editing, obtaining images and consent and approval of the final manuscript. CMB was involved in the clinical care of the patient, manuscript preparation and editing, obtaining images and approval of the final manuscript. SKR was involved in the clinical care of the patient, manuscript preparation and editing, obtaining images and approval of the final manuscript.

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Learning points

- Encephaloceles caused by amniotic band syndrome are rare and more complex than typical encephaloceles, presenting a considerable challenge to the neurosurgical team.
- An MRI of the brain with and without contrast or a CT venogram is critical prior to operating on an encephalocele, especially one in the vertex location, to understand the patient's vascular anatomy.
- Meticulous attention and planning prior to incision are critical to formulating a closure plan for both the skin and dura to minimise the risk of cerebrospinal fluid leak.

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