

## Huge interparietal posterior fontanel meningoencephalocele

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Rosildo JFC, Santos MFD, Santa Barbara RC. Huge interparietal posterior fontanel meningoencephalocele. *Autopsy Case Rep* [Internet]. 2015; 5(1):43-48. <http://dx.doi.org/10.4322/acr.2014.049>

### ABSTRACT

Congenital encephalocele is a neural tube defect characterized by a sac-like protrusion of the brain, meninges, and other intracranial structures through the skull, which is caused by an embryonic development abnormality. The most common location is at the occipital bone, and its incidence varies according to different world regions. We report a case of an 1-month and 7-day-old male child with a huge interparietal-posterior fontanel meningoencephalocele, a rare occurrence. Physical examination and volumetric computed tomography were diagnostic. The encephalocele was surgically resected. Intradural and extradural approaches were performed; the bone defect was not primarily closed. Two days after surgery, the patient developed hydrocephaly requiring ventriculoperitoneal shunting. The surgical treatment of the meningoencephalocele of the interparietal-posterior fontanel may be accompanied by technical challenges and followed by complications due to the presence of large blood vessels under the overlying skin. In these cases, huge sacs herniate through large bone defects including meninges, brain, and blood vessels. The latter present communication with the superior sagittal sinus and ventricular system. A favorable surgical outcome generally follows an accurate strategy taking into account individual features of the lesion.

### Keywords

Encephalocele; Brain; Meninges; Physical Examination; Hydrocephalus

### CASE REPORT

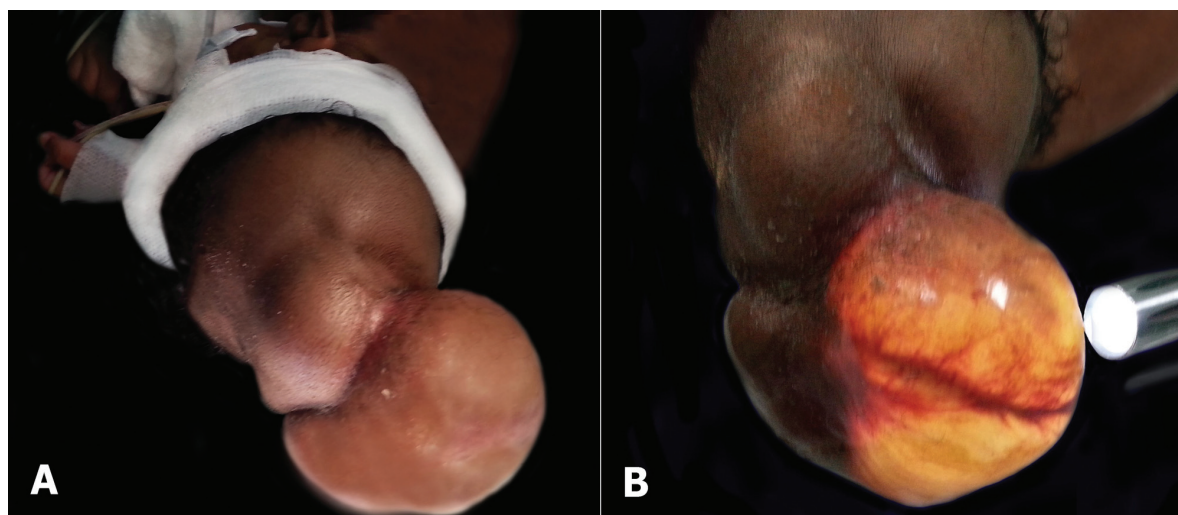
The first child of his parents, a 37 days old boy, born through a normal vaginal delivery at term, was admitted at the Department of Neurosurgery with the diagnosis of a firm interparietal-posterior fontanel tumor measuring 21 × 7.7 × 7 cm protruding through a wide circular skull defect, consistent with an encephalocele. The encephalocele had a sessile base and also scattered skin ulcerations. The malformation was cystic on its superior part where trans-illumination was positive (Figure 1).

The child's head circumference measured 38 cm; the hairline and ears were normally set. The fontanel had normal tension on both the supine and the vertical positions. The neck and all four limbs had normal configuration and there were no movement abnormalities. Volumetric computed tomography (VCT) of the head showed a malformation on the midline measuring 22.2 × 8.7 × 8.3 cm protruding through an annular skull defect at the interparietal posterior fontanel region measuring 6.38 × 6.67 cm. The protruded malformation showed two large

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**Figure 1.** Interparietal posterior fontanel meningoencephalocele (A and B); positive sac transillumination (B).

blood vessels in the midline under the overlying skin, which extended to the frontal region (Figure 2). The encephalocele sac was filled by cerebral tissue, meninges, and large blood vessels, which were in communication with the superior sagittal sinus. The sac had various partitions, which were in communication with the ventricular system (Figure 3).

Therefore, the diagnosis of meningoencephalocele was confirmed. Other imaging studies such as plain chest x-ray and abdominal ultrasonography were performed and ruled out additional defects. Ancillary blood tests were normal.

The patient was operated on in the supine position with his head tilted to the left. After antisepsis of the surgical field, a stitch was set at the midline of the interparietal scalp, 1 cm before the sac neck, in order to occlude the large scalp blood vessel revealed on the VCT.

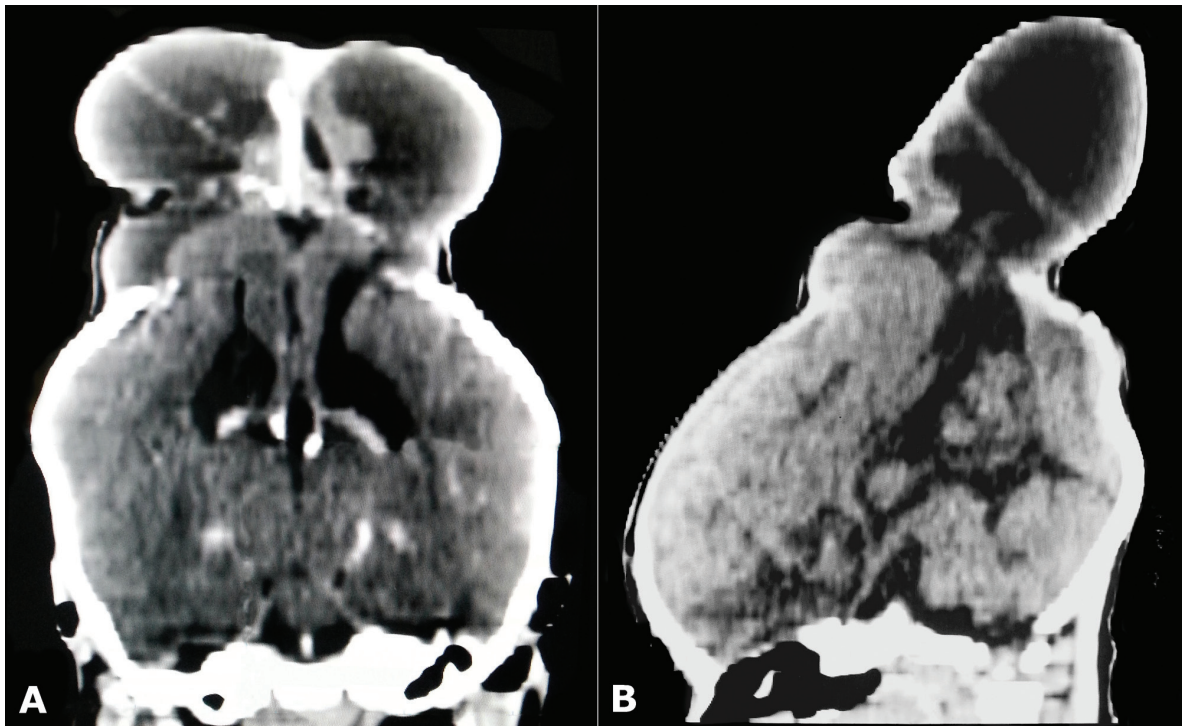
The amount of 255 mL of cerebrospinal fluid was drained from the encephalocele cystic portion. An incision beginning at the puncture site, 1.5 cm laterally shifted to the right side of midline, was extended, rounded in shape, until the contralateral paramedian region. The sac layers were all dissected until the limits of the bone defect (Figure 4).

The duramater was opened and all the non-viable and gliotic brain tissue was excised; meticulous hemostasis observed, preserving as many of the vast number of veins as possible. The surgical wound was sutured with absorbable monocryl 2-0 (Figure 5A).

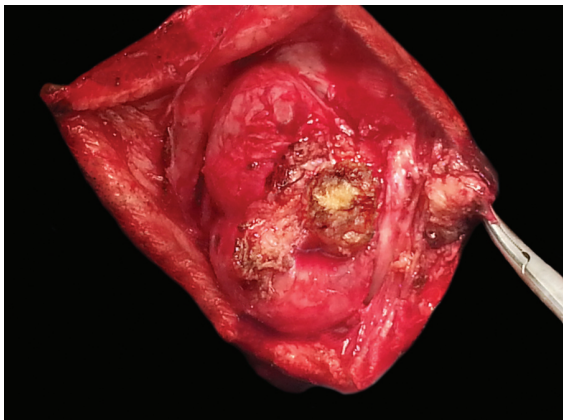


**Figure 2.** Brain volumetric CT showing a large epicranial blood vessel extending from the frontal region to the encephalocele.

The immediate postoperative was uneventful until the second day when the patient developed hydrocephalus, which required ventriculoperitoneal shunting. The patient was discharged on the fifth day after the second surgical procedure and was well on the regular follow-up at the neurosurgery outpatient service (Figure 5B). The patient's mother and the



**Figure 3.** Brain CT scan showing the encephalocele sac content (coronal (A) and sagittal (B) views).



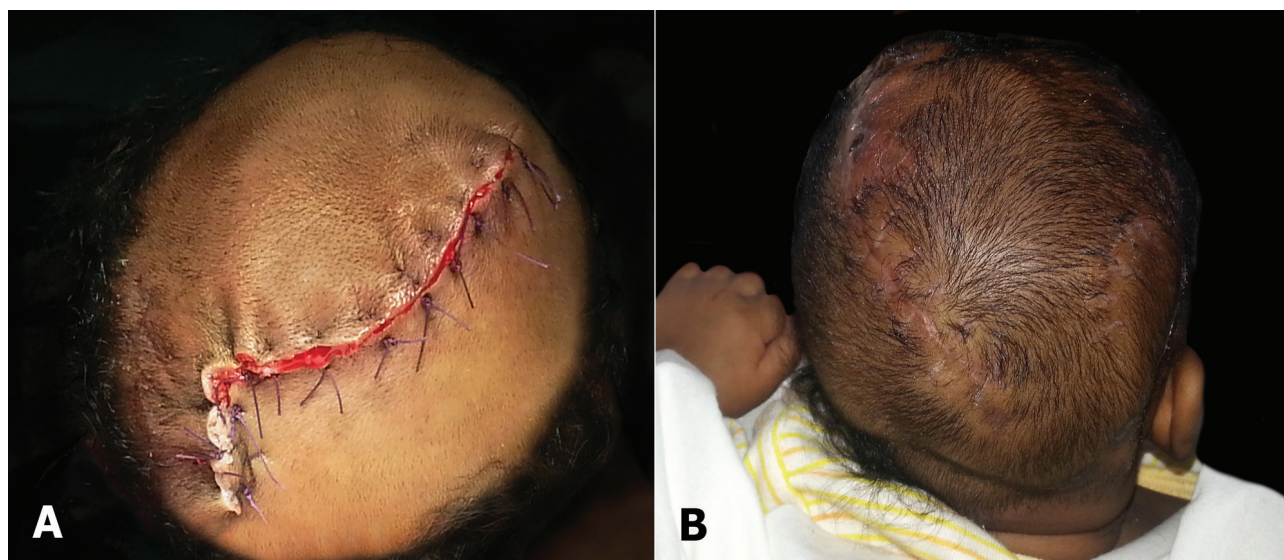
**Figure 4.** Operating view showing sac dissection.

clinic's ethic committee signed their consent for this publication.

## DISCUSSION

Congenital encephalocele is a neural tube defect characterized by a sac-like protrusion of the brain, its overlying membranes, and other intracranial structures through a skull defect. It is the neural tube defect of lowest incidence, variable according to different world regions. Occipital encephalocele is the most common type represented by 75–80% of all cases, while the

parietal type represents only 10%.<sup>1,2</sup> A large number of encephalocele classifications has been proposed according to the (1) origin (congenital or acquired); (2) sac contents (meninges: meningocele, brain: encephalocele; ventricular system, meninges and brain: hydromeningoencephalocele); (3) location (occipital, sincipital, basal, anterior, posterior, supratocrular, infratorcular); (4) intensity of protrusion (grades I, II, and III); and (5) morphological, aesthetical, and functional (internal, external, sincipital, craniofacial malformation). Anterior encephalocele refers to the occurrence anteriorly to the coronal suture; parietal when it is located between the lambdoid and the coronal sutures; and occipital or posterior when it occurs at the lambdoid suture.<sup>3,4</sup> Occipital encephaloceles are the most common type, while the interparietal is uncommon. The interparietal encephalocele associated with the posterior fontanel location is rare; our research revealed no publications involving these two locations simultaneously. Other remarkable features of this case include the great sac volume, the large bone defect, and the presence of large blood vessels in the midline under overlying skin and into the sac in communication with the superior sagittal sinus.<sup>5</sup>



**Figure 5.** Sutured surgical wound (A) and scar incisions 15 days after surgery (B).

The exact etiology of encephalocele is not entirely defined. There are some hypotheses based on experimental studies. The occurrence among brothers and twins points towards a genetic origin of some types. Other authors highlighted environmental factors in the genesis of encephalocele, such as drugs (high doses of vitamin A, valproic acid, cytostatic agents, and folic acid deficiency); hyperthermia; maternal chronic diseases; and radiation effects.

Congenital encephalocele originates from the lack of separation of the neuroectoderm and the ectodermic surface during neural tube formation, blocking the interposition of the mesoderm between the two germinative layers.

Another hypothesis concerning the secondary protrusion of intracranial structures relates to a primary bone defect due to a failure of cartilage formation during skull ossification. The intracranial hypertension during intrauterine life could be of paramount importance in causing intracranial structure herniation through the bone defect.<sup>6,7</sup> The theoretical relation of pressure and force of liquids and solids against surfaces ( $P = F/S$ ), generates an outwards vectorial force ("protrusion force") perpendicular to the deficient bone surface that could enlarge some parts of the brain and other intracranial structures, in case of intracranial pressure elevation during intrauterine life.

Generally, the diagnosis of encephalocele is easy. In these cases, noticeable deformities are present and diagnosis may be made immediately

after birth. Sometimes, the diagnosis is challenging and incidentally discovered during a routine physical examination in asymptomatic patients. Clinical features vary depending on multiple factors, such as the location and volume of the malformation, the grade, sac contents and associated complications. Symptoms include mental retardation, nasal congestion, ocular symptoms, and craniofacial bulging with or without cerebrospinal fluid leakage.<sup>8</sup>

Intrauterine diagnosis of encephalocele became feasible by alpha-fetoprotein level determination and by ultrasonography. After birth, the diagnosis is based on physical examination and imaging studies, which evaluate the encephalocele and other possibly associated malformations.

Sac transillumination must be performed whenever possible to detect solid contents within the sac. Cervical spinal x-ray is required to study the anatomy of vertebrae, as well as head CT and magnetic resonance imaging (MRI). The latter is the imaging examination of choice because of its higher specificity and sensibility to define the sac contents, complemented with angio MRI to better study the malformation's vascular pattern.<sup>9</sup> In this case, MRI was not performed in accordance to the patient's family decision. The diagnosis was confirmed by VCT, which showed the presence of blood vessels under the overlying skin from the frontal region of the skull to the malformation. Other encephalocele sac blood vessels were in communication with the sagittal sinus. VCT was also useful to study the skull defect.

These details were important for surgical planning, preventing complications, and technical challenges.

Surgical treatment of encephaloceles consists of resecting the redundant layers (skin and meninges), repositioning the protruding elements back into the skull, and correcting the deformities. Aspiration of the cerebrospinal fluid before the incision—in patients with a large encephalocele as in our case—helps the dissection of the sac. The surgical procedure may be performed by endoscopy or by open surgery, as in this case.

Open surgery techniques may access the lesion either intradurally or extradurally, or both. Some surgeons suggest that small bone defects do not necessarily need surgical correction.<sup>10,11</sup> In this case report, intradural and extradural approaches were performed. The duramater was opened to permit the evaluation of the inner defect. Brain tissue with gliosis was then excised. The duramater was sutured in watertight fashion followed by multiple-layer suture of the skin. During the surgical procedure, the sac was dissected to its limit with the skull defect, which was not primarily closed until intracranial pressure control and all tissues repair was completed. The surgical wound suture was held with an adequate tension. Every layer was sutured separately to create a strong surface, generating a “contention force” to avoid further protrusion and to permit the reduction of the bone defect as the child’s skull grows with age.

A large number of factors influence the outcome of encephalocele surgical treatment; namely, the location, the size, the amount of the herniated brain, the presence of blood vessels into the sac, the presence of hydrocephalus, and additional birth defects.<sup>12</sup> Surgeons’ expertise on this type of malformation is also considered a risk factor for surgical results.

Despite the unusual location, the presence of large blood vessels, the size of the sac and skull defect, and the venous drainage to the sagittal sinus, no surgical complications was observed in this case.

## CONCLUSIONS

Even in the presence of non-negligible number of complicating factors that may hamper the surgical treatment, a thorough imaging study followed by meticulous surgical planning by an experienced

neurosurgeon allows good outcomes in operative treatment of huge encephalocele.

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**Conflict of interest:** None

**Submitted on:** October 12, 2014

**Accepted on:** November 10, 2014

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