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CASE-BASED UPDATE

# Lateral cephaloceles: case-based update

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**Abstract**
*Background* Masses of the lateral skull are not uncommon and include lipomas and epidermoids. However, meningoceles of the lateral skull are very rare and are often misdiagnosed. Even rarer are atretic encephaloceles of this region.
*Illustrative case* We report a newborn with a right atretic encephalocele of the asterion. Lesions of the anterolateral and posterolateral fontanelles should include lateral cephaloceles/encephaloceles in the differential diagnosis. The literature and embryology of these rare entities are discussed.

**Keywords** Meningocele · Lateral skull defect · Asterion · Skull · Lateral · Neurosurgery · Pediatrics

## Background

Cranium bifidum is a congenital defect of the skull that is commonly located in the midline that may allow protrusion of intracranial contents. A herniation containing meninges and cerebrospinal fluid is defined as a meningocele and in addition, a cephalocele including brain tissues is termed meningoencephalocele [1]. Cephaloceles are further subdivided according to their location (Table 1) [2]. Most cases have been reported in or near the midline of the nasal, nasopharyngeal, buccal, naso-orbital, metopic, interparietal, occipital, and suboccipital areas. However, the occipital region is the most common site [3, 4].

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## Etiology

The causes of cephaloceles are based on the pathology that leads to a bony defect. Some acquired pathologies include trauma, surgery, and neoplasm [5, 6]. Congenital cephaloceles are postulated to be the result of a neural tube defect [7] with various etiologies listed in Table 2.

## Embryology

Classically, cephaloceles have been hypothesized to be caused by a neural tube defect [7]. However, the exact mechanism of neural tube closure is debatable. Conventionally, neural tube closure in humans has been portrayed as a "zipper model." This model represents one initiation site of closure in the lower cervical region that proceeds simultaneously to the rostral and caudal ends of the neural tube [8–13]. In contrast, Van Allen et al. [7] postulated that there are multiple sites of initiation of neural tube closure. They hypothesize that closure of the human neural tube follows the same pattern as demonstrated in animal models. These authors argued that this model provides a more comprehensive explanation of the diverse presentations of cephaloceles. However, the multi-site initiation theory focused on cephaloceles located in or near the midline. Therefore, the failure of neural tube closure does not completely explain the development of lateral cephaloceles. To the best of our knowledge, there are two differing

**Table 1** Types of meningoceles/encephaloceles (from Lumenta et al. [12])

| Region of skull | Localization |
|---|---|
| Occipital | Supratentorial |
| | Infratentorial |
| Occipitalcervical | Occipitocervical |
| Parietal | Interfrontal |
| | Interparietal |
| | Anterior frontal |
| | Posterior frontal |
| Temporal | Anterolateral fontanelle (pterion) |
| | Posterolateral fontanelle (asterion) |
| Sincipital (Frontal) | Frontoethmoidal |
| | Nasofrontal |
| | Nasoethmoidal |
| | Naso-orbital |
| | Craniofacial cleft |
| Basal | Sphenopharyngeal |
| | Spheno-orbtital |
| | Sphenomaxillary |
| | Sphenoethmoidal |
| | Transethmoidal |
| | Transsphenoidal |
| | Basioccipital |

theories of the origin of lateral cephaloceles in the literature. Van Allen et al. [7] postulated these as neural tube defects. They proposed that lateral cephaloceles are most likely due to a secondary reopening of the neural tube. In contrast, Martinez-Lage et al. [3] and Nagulich et al. [4] suggested that ectopic deposits of meninges or choroid plexus form a hernia filled with CSF. The herniation then

**Table 2** From Burgener et al. [2] and Papanikolaou et al. [15]

| Method of development of cephaloceles | Cause |
|---|---|
| Spontaneous | Congenital |
| | Idopathic |
| Non traumatic | Chronic otitis media with or without cholestereartoma |
| | Neoplastic |
| | Inflammation/infection |
| | Dermal sinus and vascular abnormalities |
| | Miscellaneous |
| Iatrogenic | Surgery |
| | Irradiation |
| Traumatic | Blunt force trauma |
| | Penetrating trauma |

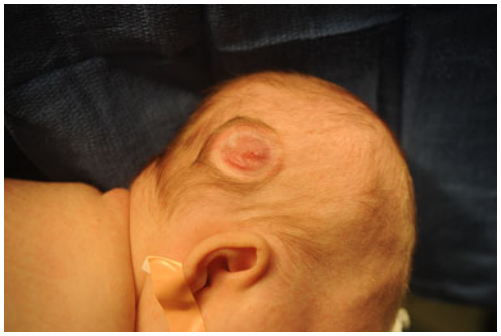

**Fig. 1** Atretic meningocele of the asterion in the patient presented herein

protrudes through a fontanelle that was in our case the posterolateral fontanelle. Such protrusions may also represent non neural tube defects with simply herniation of intracranial contents through a weakened area of the lateral meninges and skull. Lastly, these processes may represent arrest of the formation of a true meningocele.

Apparently, cephaloceles of the lateral skull are not associated with intracranial brain defects that are common in for example, parietal cephaloceles. These defects include commissure dysgenesis, venous derailment, and hindbrain anomalies [14]. For cephaloceles in general, Yokota et al. [14] posited that these represent meningoceles that are "restored" in utero and that the atretic process is largely influenced by internal hydrostatic pressures.

## Incidence

Congenital cephaloceles are a rare anomaly. In the USA, these occur approximately one out of every 10,000 births [15]. The occurrence of lateral cephaloceles is even less

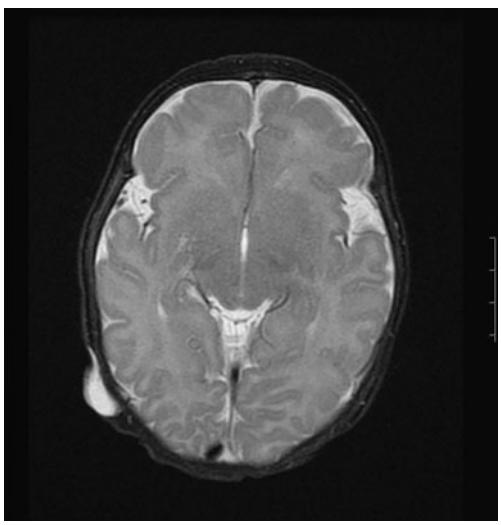

**Fig. 2** T2-weighted MRI noting the right-sided extracranial CSF collection overlying the region of the asterion

common. To the best of our knowledge, only 17 cases have been reported in the literature since 1967 [3, 4, 16–19]. The scarce amount of lateral cephaloceles throughout the years lends credence to the hypothesis proposed by Martinez-Lage et al. [3] and Nagulich et al. [4].

Congenital meningoceles and meningoencephaloceles of the lateral region are very rare. To the best of our knowledge only 17 cases have been reported in the literature. Sixteen of those cases reported a herniation at the pterion [4, 16–19]. Only one case reported by Martinez-Lage et al. [3] in 1982 was of a meningocele of the asterion. As an illustrative case, we report an atretic encephalocele of the asterion.

## Management

Meningoceles of the lateral skull are treated similarly to meningoceles of the midline skull. If ischemic herniated tissues are present within the meningocele, these are often transected flush with the skull defect. Additional skin and dura are trimmed and the wound closed in standard layers. The bony defect may require cranioplasty if large. Although meningoceles of the lateral skull are rare and thus their natural history is uncertain, these patients are also monitored for the development of hydrocephalus and treated accordingly.

## Illustrative case

We report a 1-month-old, white female with a right-sided scalp mass present over the asterion since birth (Figs. 1 and 2). No additional medical or structural issues were found in this patient. CT imaging revealed normal intracranial contents with no obvious connections to the extracranial mass. The child had prospered with no signs of infection or neurological compromise. At operation, the skin lesion was removed. A very small amount of what was believed to be CSF was present. The defect was excised and a remnant of connection to the intracranial cavity was observed but found to be insignificant and no longer communicating with the extracranial lesion. No gross meningeal or brain tissues were found in the mass. Pathological findings were that of dermal fibrosis and proliferation of meningothelial cells with associated prominent vascular proliferation consistent with an atretic meningocele. At 2 years follow up, the child is well and is making appropriate developmental milestones. The patient did not develop hydrocephalus.

## Conclusion

Although apparently very rare, an atretic meningocele of the asterion should be placed in the differential diagnosis of lateral skull masses. Midline brain abnormalities that are commonly found in association with for example, parietal cephaloceles [14], appear to be uncommon with lateral cephaloceles.

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