Atretic parietal cephalocele: Prognosis and embryological perspective with key radiological characteristics

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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Contrast agent-other
Procedure: Diagnostic procedure
Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 5 weeks, male

Clinical History:

A five-week-old male patient presented for evaluation of posterior scalp swelling. Physical examination revealed a soft palpable interparietal subgaleal mass. A normal neurologic exam was otherwise evident.

Imaging Findings:

A magnetic resonance (MR) image with and without contrast demonstrated a heterogeneous subcutaneous scalp lesion with intracranial extension, measuring 1.2 x 1.6 cm. This cystic parietal scalp swelling communicated intracranially with a posterior interhemispheric cyst that traversed along a persistent falcine sinus. Furthermore, characteristic radiological findings such as identification of a cigar-shaped cerebrospinal fluid tract within the interhemispheric fissure, prominent superior cerebellar cistern, superior peaking of the tentorium and a spinning-top configuration of the tentorial incisura were visualized on MR sequences. The accompanying MR venogram demonstrated a persistent falcine venous sinus and fenestration of the superior sagittal sinus. Additionally, the MR venogram showed a non-developed straight sinus with the internal cerebral veins and the vein of Galen draining into the falcine sinus.

A neonatal intracranial ultrasound was performed that showed a hypoechoic, parietal subgaleal fluid collection with internal echogenicity representing fibrous tissue. A hypoechoic fibrous tract was also visualized.

Discussion:

Background:
Cephaloceles are congenital herniations of intracranial structures [1]. While cephalocele describes any diverticulated central nervous system tissue, an atretic cephalocele defines a skin-covered herniation of dura, fibrous tissue and dysplastic brain parenchyma. Atretic parietal cephaloceles are rare, encompassing 1% of all cerebrospinal congenital malformations [2].

Pathogenesis remains elusive. While some argue that they are a result of failed neural tube closure, others maintain
that the tube closes normally and then reopens abnormally. Specific hypotheses include persistence of the nuchal bleb, persistence of neural crest remnants, and the near resolution of involuted meningoencephaloceles formed in utero [3, 4, 5]. Patterson et al. have described the concept of an embryonic straight sinus; however, this is controversial and can be equivalent to a persistent falcine sinus on MR sequences [6].

Clinical Perspective:
Lesions present as a posterior scalp mass. Imaging is necessary for diagnosis. While general prognosis is good, it remains variable and depends on several factors, including tissue contents and the presence of associated congenital abnormalities, such as Walker-Warburg syndrome [6].

Imaging Perspective:
Recognition of a subgaleal cystic lesion with intracranial extension through a bony defect is identified on MR imaging [6]. MR venogram sequences can demonstrate focal fenestration of the superior sagittal sinus at the level of the lesion and a persistent falcine sinus directed towards the lesion [6]. Additional key MR features include a “cigar-shaped” CSF tract within the interhemispheric fissure and a “spinning-top” configuration of the tentorial incisura that is formed due to a prominent superior cerebellar cistern and superiorly peaked posterior tentorium [6]. A fibrous stalk is a subtle finding that can be seen on T1-weighted post-contrast sequences by delineating the adjacent enhancing veins.

Patterson et al. have described the vertical embryonic positioning of the straight sinus as a key radiologic feature [6]. However, this is controversial and has been thought of as equivalent to a persistent falcine sinus. Needless to say, this has no significance on prognosis [8].

Outcome:
Management is via surgery. Although curative, prognosis depends not on the presence of the cephalocele, but rather on the coexistence of other neurologic malformations, such as ventricular dilatation, quadrigeminal cistern lipoma and Walker-Warburg syndrome [5, 6, 7]. Thus, imaging is critical to evaluate for ancillary lesions.

Take Home Message:
Atretic cephalocele should be included in the differential diagnosis of a posterior scalp lesion. Key radiological findings on MRI are crucial in identification. Prognosis depends on associated intracranial anomalies.

**Differential Diagnosis List:**
- Atretic parietal cephalocele.
- Dermoid or epidermoid cyst.
- Infantile haemangioma.
- Sinus pericranii.
- Cephalohematoma.
- Sebaceous cyst.

**Final Diagnosis:**
Atretic parietal cephalocele.

**References:**
**Description:** Note the parietal cephalocele (red arrow) communicating with the posterior interhemispheric cyst (blue arrow). A prominence of the superior cerebellar cistern (green arrow) and a persistent falcine sinus (orange arrow) are also noted. **Origin:** Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse, NY.
Figure 2

Description: Note the cigar-shaped CSF tract (green arrow) communicating with the cephalocele (red arrow), with the tract inferior to the falcine sinus (blue arrow). Prominence of the superior cerebellar cistern (orange arrow) is redemonstrated. Origin: Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse NY.
Description: Note the spinning-top configuration (red arrow) of the tentorial incisura resulting from the prominent superior cerebellar cistern and the high positioning tentorium. Origin: Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse NY.
Figure 4

Description: Note the superior peaking (high positioning) of the tentorium. Origin: Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse NY.
Description: Note the persistent falcine sinus (red arrow) with the internal cerebral veins and the vein of Galen draining into the falcine sinus. Origin: Department of Radiology, SUNY Upstate University Hospital, Syracuse NY.
Description: Note the focal fenestration of the superior sagittal sinus (red arrow). Origin: Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse NY.
Description: Note the hypoechoic, parietal subgaleal fluid collection (red arrow) with heterogeneous internal echogenicity (blue arrow) representing fibrous tissue. A hypoechoic fibrous tract (orange arrow) is identified coursing toward the cranial defect. Origin: Department of Radiology, SUNY Upstate Medical University Hospital, Syracuse NY