A 4-Week-Old Infant with Scalp Swelling in the Parietal Region

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A 4-week-old boy presented with a small parietal lump that had been present since birth. The child was a term baby who had been delivered vaginally. There was no history of birth trauma. The lump was soft on clinical examination, and magnetic resonance imaging (MRI) of the brain was performed.

MRI showed a well-defined, small, midline, spherical subcutaneous lesion in the parietal region. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images. The postcontrast study showed mild peripheral enhancement (Figure 1).

There was also a linear T2-weighted hyperintense tract along the interhemispheric fissure extending to the scalp lesion through a small bony defect. The study also showed mild enhancement of the tract (Figure 2), and there was associated tenting of the tentorium and prominent supra- cerebellar cistern (Figure 3). The dural venous sinuses were otherwise patent.

Figure 1. (A) Axial T2-weighted and (B) T1-weighted images show a heterogeneous subcutaneous T2-weighted hyperintense and T1-weighted hypointense lesion in the parietal region. (C) Postcontrast axial T1-weighted image shows minimal heterogeneous contrast enhancement.

Figure 2. (A) Sagittal T2 and (B) postcontrast T1-weighted images demonstrate an enhancing tract (small white arrows) coursing through a small skull defect connecting the dura mater and the scalp swelling. Also note the persistent falcine sinus (long white arrow in A).

For diagnosis, see page 313

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Diagnosis:

Atretic Parietal Encephalocele with Persistent Falcine Sinus

The MRI showed an atretic parietal encephalocele as well as an anomalous persistent falcine sinus. The intraoperative findings showed a 2 cm × 2 cm atretic encephalocele at the parietal region with a fairly thick stalk containing multiple vessels. The patient underwent surgical excision of atretic encephalocele.

DISCUSSION

Scalp lumps in children are a common entity and pose a diagnostic challenge to clinicians. Atretic encephalocele is a great mimicker of other benign soft tissue lumps of the scalp. It is also commonly associated with vascular and intracranial abnormalities.1,2

The term “encephalocele” refers to extracranial herniation of intracranial contents through a skull defect. When there is herniation of meninges and cerebrospinal fluid it is referred to as meningocele. When it is associated with herniation of brain parenchyma it is referred to as meningoencephalocele. They may be diagnosed antenatally with either ultrasound or MRI.3,4

Atretic encephalocele was initially described by James and Lassmann in 1972 as an encephalocele in degenerative form. The term “atretic encephalocele” is used when the herniation of meninges is associated with remnants of glial or neural tissue and associated fibrosis.6 They occur typically in the parietal region but may also be encountered in occipital regions.1

The common theories that have been proposed to explain the pathogenesis of this condition include incomplete involution of a meningoencephalocele that was formed early in fetal life and persistent nuchal bleb.7

They are usually a small, flat, midline solid or cystic lesion in the vertex due to variable amount of proliferation of fibrous tissue, blood vessels, and neural tissue.8,9 Presence of a linear fibrous stalk that connects to the dura mater through a small skull defect is an important clue in the diagnosis of atretic parietal encephalocele.1,10

This entity is commonly associated with intracranial venous abnormality similar to enlarged bilateral foramina, such as absence or hypoplasia or vertically oriented embryonic straight sinus with an alternative venous drainage via persistent falcine sinus.7,11 Other associated abnormalities include abnormally high position of the falx and tentorial junction with a prominent superior cerebellar cistern, fenestration of the superior sagittal sinus, midline defects such as porencephaly, and corpus callosal agenesis.7,10

The common differential diagnosis of atretic encephalocele in infants includes encephalocele, lipoma, vascular malformation, sinus pericranii, and dermoid and epidermoid cysts, which can be readily differentiated with MRI. Sinus pericranii usually contains venous blood, and lipoma can be recognized by its fat content. Dermoid cysts usually demonstrate a varying degree of fat, and epidermoid cysts appear bright on diffusion-weighted images.1,2

In general, atretic encephalocele has a good prognosis.2 Patients may experience pain because of stretching of dura mater and may also have overlying skin ulcerations. Hence, surgery is necessary in atretic encephalocele for more than just cosmetic reasons.8

CONCLUSION

This case illustrates the presentation, diagnosis, and management of atretic parietal encephalocele in an infant. Atretic parietal encephalocele is a distinct degenerative form of encephalocele having a very good prognosis. MRI is the imaging modality of choice to diagnose the condition and to look for associated vascular and intracranial abnormalities. Preoperative diagnosis can be confidently made with MRI and readily differentiated from a wide variety of conditions presenting with scalp swelling, ranging from dermoid to encephalocele.

REFERENCES

5. James CC, Lassman LP. Spinal Dysra-