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Atretic Parietal Cephaloceles Revisited: An Enlarging Clinical and Imaging Spectrum?

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PURPOSE: **We describe imaging features that are clues to the diagnosis of atretic cephaloceles and discuss clinical findings and a possible mechanism by which these lesions develop.**

METHODS: **Eight children (five girls and three boys) ranging in age from 1 day to 3 years 4 months with midline subscalp lesions underwent radiologic examination with CT or MR imaging. In all cases, the lesions were surgically excised and subjected to pathologic examination. Imaging studies and medical records were reviewed retrospectively.**

RESULTS: **Six of eight children had vertical embryonic positioning of the straight sinus with a prominent superior cerebellar cistern. A "spinning-top" configuration of the tentorial incisura, a "cigar-shaped" CSF tract within the interhemispheric fissure, fenestration of the superior sagittal sinus, and "peaking" of the tentorium were associated findings helpful in making this diagnosis. Two of the eight children had findings indistinguishable from focal dermoid, six were developmentally normal, one had mild motor delay, and one died at the age of 3 years. Pathologic examination revealed glial, meningeal (arachnoid), fibrous, and dermal elements.**

CONCLUSION: **Characteristic findings on MR images and CT scans provide clues to the diagnosis of atretic cephalocele. However, even in the presence of abnormal imaging findings, these children may be developmentally normal.**

In 1964, McLaurin (1) reported seven children with a diagnosis of parietal meningocele among a series of 13 children with parietal cephaloceles. Since that time, a number of investigators have reported small, skin-covered midline subscalp masses or cysts variably described as atretic cephaloceles, atypical meningoceles, rudimentary meningoceles, meningeal heterotopias, and meningoceles manqué. These rare lesions generally occur within a few centimeters of the lambda and contain meninges and neural rests, with approximately half the reported cases parietal in location (2–5).

We report eight children with parietal atretic cephaloceles, the majority of which have had a more benign clinical course, in contrast to those reported in recent series (6, 7). We present the imaging features that provide clues to this unusual diagnosis, expand upon the radiologic and clinical spectra reported to date, and discuss a possible mechanism by which atretic cephaloceles develop.

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Methods

Our patient population consisted of eight children (five girls and three boys) who were seen in the neurosurgery department between 1985 and 1994 for evaluation of a midline parietal subscalp nodule or cyst. The children ranged in age from 1 day to 3 years 4 months at the time of presentation. All children had at least one cross-sectional imaging study (six of eight had CT, five of eight had MR imaging).

All patients were treated with surgical excision of the mass or cyst. In cases in which a fibrous tract extended intracranially, the tract was divided at the level of the dura and the dura was oversewn. Clinical follow-up ranged from 6 months to 9 years. Pathologic correlation was available in all patients. Neuroimaging studies and medical records were reviewed retrospectively.

Results

Clinical and imaging findings are summarized in the Table. On the basis of neuroimaging studies, two subsets of children emerged: those with and those without vertical embryonic positioning of the straight sinus.

Embryonic Straight Sinus Positioning

Six of the eight children (cases 1 through 6) in our study group had evidence of vertical embryonic positioning of the straight sinus. In two cases, the abnor-

Clinical and imaging correlation

Case	Age/Sex	Embryonic Straight Sinus	Additional Findings	History/Surgical Findings	Clinical Outcome
1	40 mo/F	+	. . .	Midline posterior parietal scalp mass/ cephalocele with stalk attached to dura	Normal
2	6 d/F	+	Moderate ventricular dilatation, gray matter heterotopia	Palpable vertex mass/cephalocele stalk extending through skull	Normal
3	3 mo/M	+	Mild ventricular enlargement, quadrigeminal cistern lipoma	Soft-tissue prominence/cephalocele with stalk	Normal
4	1 wk/F	+	. . .	Palpable mass near posterior fontanelle/cephalocele with stalk extending through dura	Mild motor delay
5	1 d/M	+	Moderate ventricular enlargement, lissencephaly, metencephalic hypoplasia	Posterior midline mass/cephalocele with stalk extending through skull	Died at age 3 y
6	3 mo/M	+	. . .	Posterior midline parietal mass/cephalocele with stalk penetrating dura	Normal
7	5 mo/F	−	. . .	"Swelling" over posterior fontanelle/cephalocele inserting into fascia	Normal
8	3 d/F	−	. . .	Mass over posterior fontanelle/encephalocele with stalk attached to dura	Normal

mally positioned straight sinus could be seen directly on sagittal MR images, identifiable as a linear flow void extending posteriorly and superiorly within the posterior interhemispheric fissure to the base of the subscalp lesion. Prominence of the superior cerebellar cistern and suprapineal recess of the third ventricle was also present (cases 2 and 4) (Figs 1 and 2).

Axial images from both CT and MR examinations were remarkably consistent in appearance. In all cases, the tentorial incisura demonstrated a "spinning-top" configuration, believed to represent a combination of a high falx/tentorial junction associated with prominence of the subjacent superior cerebellar cistern. In addition, an ovoid, "cigar-shaped" CSF tract could be sequentially followed superiorly within the posterior interhemispheric fissure extending to the base of the subscalp cyst or nodule. In all cases, this tract maintained a position posterior and inferior to the straight sinus. Coronal images from two MR examinations were remarkable for superior "peaking" of the posterior tentorium, with the elevated straight sinus identifiable as a linear flow void at the apex of the tentorium (case 6) (Fig 3). On contrast-enhanced CT scans, fenestration of the superior sagittal sinus by the CSF tract could be identified in four patients (case 1) (Fig 4). In cases in which bone windows were obtained, a narrow tract could be further traced through a tiny midline calvarial defect beneath the subscalp lesion (case 3) (Fig 5).

Of the six children with embryonic straight sinus position, three had additional intracranial anomalies associated with at least mild lateral ventricular dilatation. The most severely affected child had Walker-Warburg syndrome (case 5) with characteristic lissencephaly, hypoplasia of the metencephalon, and moderate to marked ventricular enlargement. This patient had profound psychomotor delay and died at 3 years of age. A second patient (case 2) had nodular subependymal gray matter heterotopia along the lateral margin of the right lateral ventricle in addition to moderate asymmetric ventricular enlargement. To date, this child is developmentally normal without seizure activity. The third child (case 3) had mild lateral ventricular enlargement and a very small lipoma in the quadrigeminal plate cistern. This child had relatively mild motor delay as an infant, but is otherwise developmentally normal.

The surgical approach to all lesions was the same, with excision of an ellipse of scalp surrounding the nodule or cyst. In the cases in which a fibrous tract was identified at the base of the lesion, dissection was carried out only to the level of the dura with amputation of the tract at that level. The dura was then closed. No bone grafts were used. By pathologic examination, the lesions were composed of dermal, meningeal, and glial elements as well as fibrous tissue. Dural elements were not identified in any of the lesions.

Without Embryonic Straight Sinus Position

The remaining two children (cases 7 and 8) did not show evidence of embryonic straight sinus positioning. In both cases, findings on neuroimaging studies were nonspecific and indistinguishable from a focal dermoid. At surgery, one child (case 8) had a small fibrous tract extending intracranially from the subcutaneous nodule and terminating in the underlying dura. The other patient (case 7) had focal thinning of the calvaria beneath the subscalp lesion with no evidence of a true bony ostium. Dermal, meningeal, and glial elements were present on pathologic examination. Developmental history in both children was unremarkable.

Discussion

The development of atretic cephaloceles and their classification remain a source of controversy. A vari-

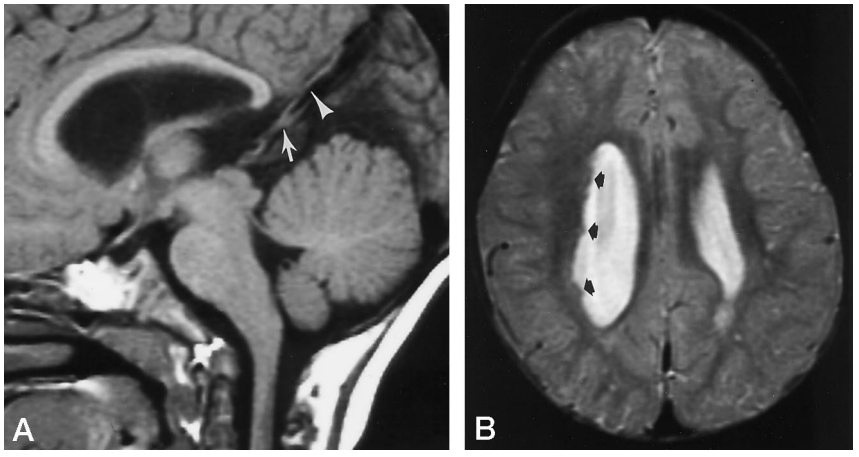


FIG 1. Case 2.

A, Sagittal T1-weighted (500/10) noncontrast MR image shows a flow void extending posteriorly and superiorly from the area of the tectum. This represents the embryonic positioning of the straight sinus (*arrowhead*). Irregular linear soft-tissue signal (fibrous tract) accompanies the flow void of the straight sinus (*arrow*).

B, Axial T2-weighted (2500/100) MR image shows subtle areas of heterotopic gray matter along the ependymal surface of the enlarged right lateral ventricle (*arrows*).

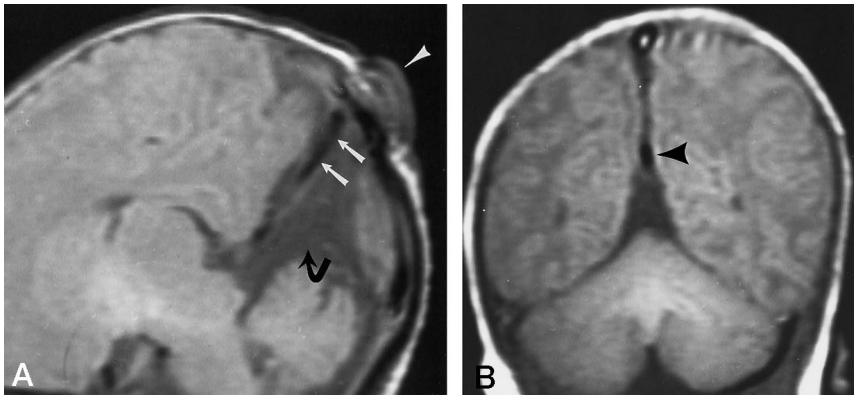


FIG 2. Case 4.

A, Sagittal T1-weighted (500/10) noncontrast MR image shows flow void (*straight arrows*) of the abnormally positioned straight sinus extending to the area of the soft-tissue subscalp nodule (*arrowhead*). Also note prominence of the superior cerebellar cistern (*curved arrow*).

B, Coronal T1-weighted (500/10) MR image shows abnormal superior position of the straight sinus (*arrowhead*) with prominence of the superior cerebellar cistern.

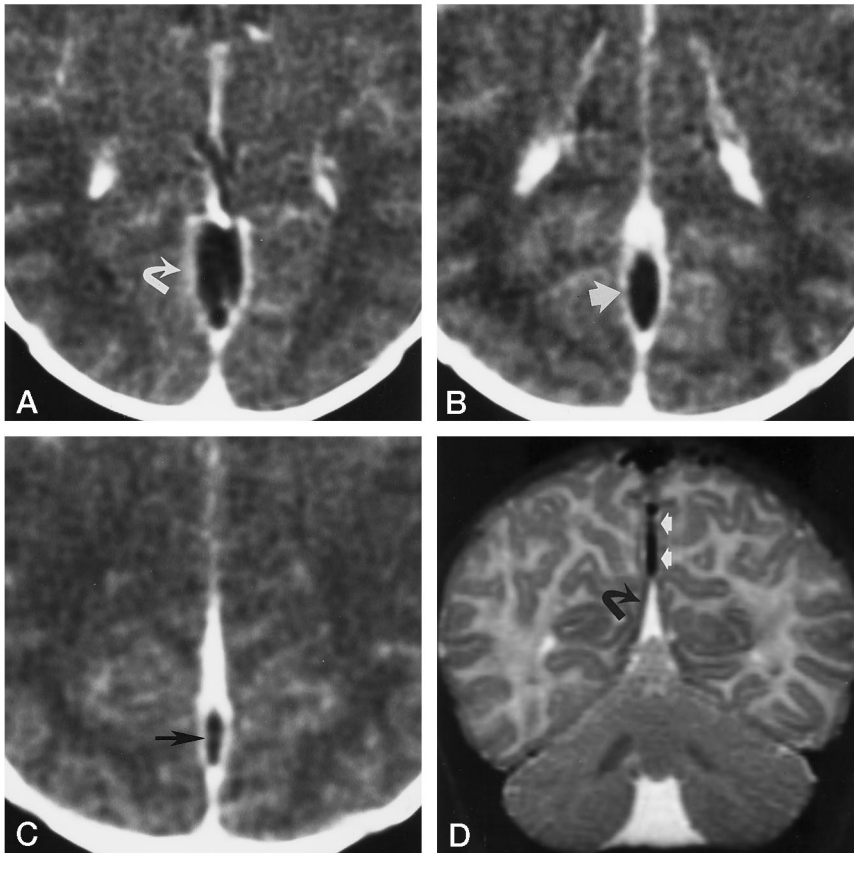


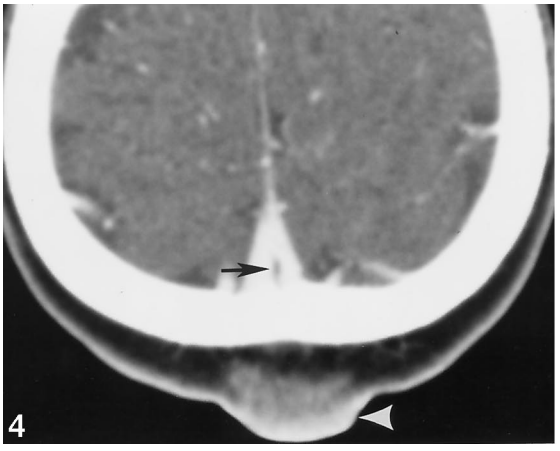
FIG 3. Case 6.

A, Axial contrast-enhanced CT scan shows the typical "spinning-top" configuration of the tentorial incisura (*arrow*) resulting from the prominent superior cerebellar cistern associated with the high position of the tentorium.

B, Axial contrast-enhanced CT scan reveals a "cigar-shaped" CSF tract (*arrow*) in the area of the abnormally positioned enhancing straight sinus.

C, Slightly higher image in same patient follows the course of the CSF tract (*arrow*).

D, Coronal fast spin-echo (5400/125eff) MR image shows high position of the tentorium (superior "peaking") (*curved arrow*) associated with linear flow void in a more superior midline position (*straight arrows*). Superior extension of the superior cerebellar cistern is also present.



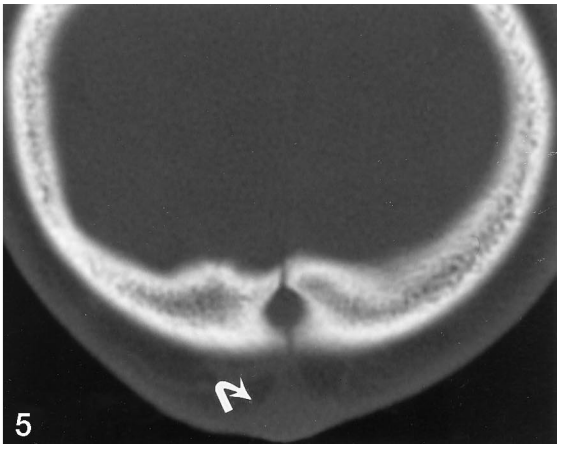


FIG 4. Case 1: Axial contrast-enhanced CT scan shows small, linear area of (CSF) soft-tissue attenuation (*arrow*) within the superior sagittal sinus, representing fenestration of this structure by the CSF tract (*arrowhead*).

FIG 5. Case 3: Bone windows from axial contrast-enhanced CT study. Bony defects of both the inner and outer tables of the parietal bone are present along with a more focal defect in the area of the diploe and a faintly visualized subcutaneous soft-tissue mass (*arrow*).

ety of pathogenetic explanations have been described, including nearly complete resolution of a larger meningoencephalocele formed in early fetal life, persistence of a fetal nuchal bleb caused by early embryonic cerebral "blow-out," and persistence of neural crest remnants (2, 4, 8).

Regardless of the mechanism of atretic cephalocele formation, embryonic positioning of the straight sinus has frequently been identified in these lesions and deserves mention both as a marker of the timing of the embryologic insult and as a clue to radiologic diagnosis (2, 4, 6, 8). During stage 7a of cranial venous development, corresponding to a crown-rump length of approximately 80 mm, the straight sinus is nearly vertical in a ventral-dorsal plane (9). At this time, the superior sagittal sinus is in the process of formation, as the result of posterior migration of the primitive transverse sinuses (marginal sinuses) and their subsequent coalescence in the midline. With expansion of the cerebral hemispheres, the straight sinus and tentorium acquire a more horizontal course, reaching the typical adult configuration after approximately the third month of gestation (9). If a midline fibrous strand were present connecting the mesencephalic tectum to the overlying membranous cranium, it is logical to assume that there could be an interruption of the normal progression of movement of the overlying straight sinus and midline tentorium to the normal adult positioning. Similarly, such a strand would need to be accommodated by the developing superior sagittal sinus, coalescing from the paired lateral primitive transverse sinuses. The frequent fenestration or high bifurcation of the superior sagittal sinus is therefore understandable. If the fibrous strand becomes discontinuous during this period of venous development, the straight sinus may achieve normal adult positioning. However, even in this case, a short fibrous stalk may be intimately related to the superior sagittal sinus, a possibility that must be borne in mind at the time of cyst or nodule excision.

The precise timing of formation of the fibrous strand cannot be determined on the basis of venous development other than to say that the stalk predates the appearance of the superior sagittal sinus (at approximately 10 weeks' gestation) (9). Therefore, a number of plausible explanations remain regarding the origin and timing of the embryologic event that culminates in formation of the subscalp cyst/nodule and associated stalk. Perhaps the most compelling is the concept of the "remnant nuchal bleb" (8). Inoue and colleagues (8) have drawn our attention to the work of Ingalls (10), who noted the relative frequency of small midline scalp cysts in early embryos. The position of these cysts over the developing anterior rhombencephalon is noteworthy. These cysts could reflect transient overdistention of the rhombencephalic vesicle (11).

The additional findings in cases 2, 3, and 5 in the current series are difficult to explain in any unifying theory of atretic cephalocele development. Case 5 had characteristic stigmata of Walker-Warburg syndrome, an autosomal recessive disorder with atretic cephaloceles in 25% to 50% of affected children (12). To our knowledge, quadrigeminal cistern lipoma (case 3) and subependymal gray matter heterotopia (case 2) have not been described in association with atretic cephalocele.

Yokota et al (6) have stressed the importance of location of the atretic cephalocele as related to the prevalence of additional intracranial anomalies as well as clinical outcome. In their series, five patients with parietal atretic cephaloceles had marked cerebral dysplasia with dorsal cyst malformation. All these patients were severely retarded and died. The five children with occipital atretic cephaloceles developed normally (6). In contrast to the children in their series, seven of eight of our patients did not have marked cerebral dysplasia, and the majority of the patients in our series (six of eight) exhibited normal development, so that our findings are in agreement with the most recent work of Martinez-Lage (2) and

dispute the importance of location of the cephalocele as a determinant of clinical outcome and predictor of intracranial findings on imaging. While the conclusions reached in both series differ, the total number of patients is quite small and larger series will be needed before more definitive comments can be made regarding this disorder.

Conclusion

Parietal atretic cephaloceles represent rare developmental anomalies that may have broader imaging and clinical spectra than once thought. Characteristic imaging findings are helpful in diagnosing this disorder. In our experience, the clinical outcome of these patients as related to imaging findings is more variable than previously reported. Specifically, some of these patients are relatively normal clinically.

Acknowledgment

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