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Atretic Parietal Cephalocele with Venous Contents, Hair-Collar-and-Tuft-Sign and Associated Venous Anomaly

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Abstract **Case Report**

Atreticcephaloceles (AC) are midline anomalies covered by abnormal skin and are usually located in the parietal or occipital region [1-3]. The coexistence of venous anomalies has been frequently reported in patients with AC [1-7].We report a case of atreticcephalocele (AC) characterized by the presence of abnormalities of the superficial venous system and a Hair-Collar-and-Tuft-Sign.

Keywords: Atretic Parietal Cephalocele Hair-Collar-and-Tuft-Sign.

CASE REPORT

A 3-months-old boy, born at term was admitted to our hospital because of apainless midline scalp swelling over the parietal region.

At the physical examination, the scalp mass was soft, roundish, non-mobile, located in the parietal midline, cranially to the lambda, and slightly hypertrichotic (Figure 1), about 1 cm in diameter. The lesion did not show pulsation and did not change in size, shape or tension even when the child was crying.

The US study, showed a subcutaneous irregularly hypoechoic mass, with a diameter of 20 x 12mm (Figure 2) and with a solid fibrous tract connecting the extracranial lesion, through a small bony cranium defect, to the intracranial space and further to the superior sagittal sinus (fenestrated at this location). This mass lights up with colour Doppler sonography and a blood vessel with a low flow signal was detected in the fibrous tract.

The CT study was performed with the spiral technique, followed by multiplanar and volumetric reconstructions with and without contrast, using a 16-slice multidetector CT scanner, for a more precise evaluation of the skull defect and to exclude other bony anomalies.

It showed (Figure 3) a heterogeneous subcutaneous scalp lesion with intracranial extension, measuring 21 x 12 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 and the superior peaking of the tentorium.

Additionally, the CT also showed a fenestration of the superior sagittal sinus, a non-developed straight sinus and the internal cerebral veinsdraining into the falcine sinus.

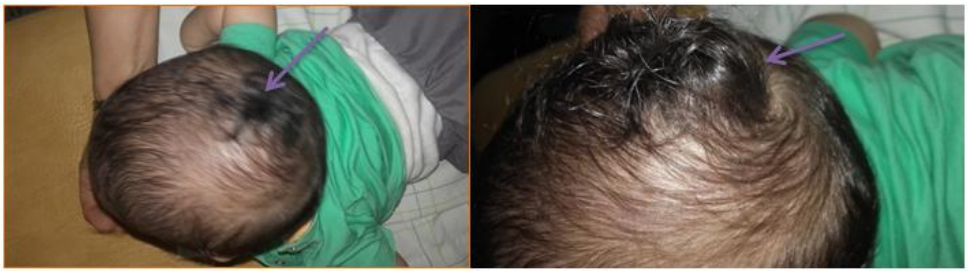

Fig-1: Clear hair-collar-and-tuft-sign

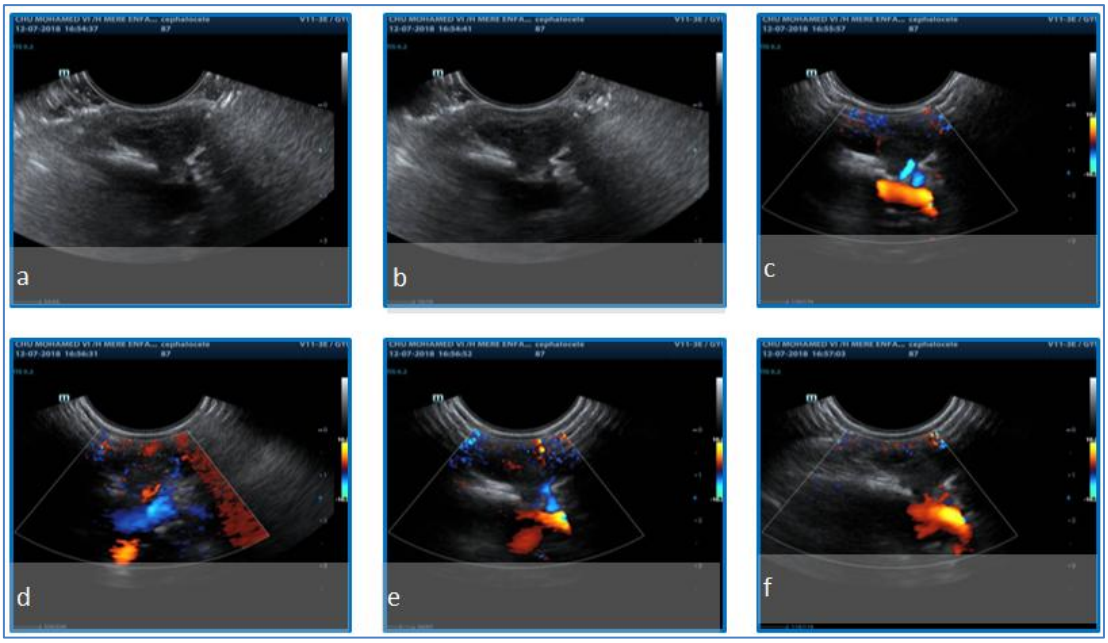

Fig-2: Sonography of the cephalocele, coronal and sagittal views: subcutaneous irregularly hypoechoic mass (blue arrow), with a diameter of 20 x 12mm (a-f) and with a solid fibrous tract connecting the extracranial lesion, through a small bony cranium defect (orange arrow), to the intracranial space and further to the superior sagittal sinus (green arrow). This mass lights up with colour Doppler sonography and a blood vessel with a low flux signal was detected in the fibrous tract (red arrow)

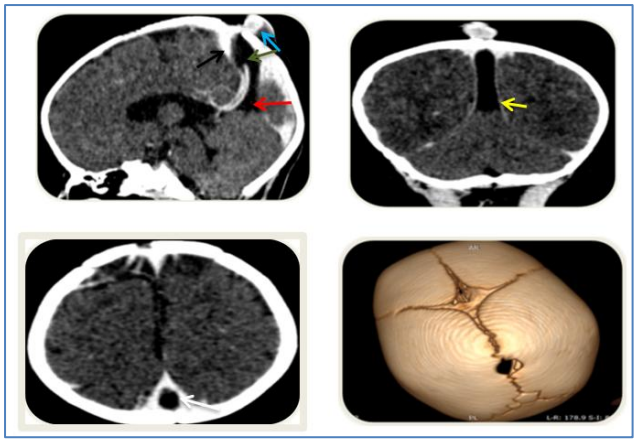

Fig-3: CT with contrast in the parenchymatous window and VRT:

Heterogeneous subcutaneous scalp lesion (blue arrow) with intracranial extension. This cystic parietal scalp swelling communicated intracranially with a posterior interhemisphericcyst(grenn arrow).

A cigar-shaped cerebrospinal fluid tract within the interhemispheric fissure, prominent superior cerebellar cistern (red arrow) and the superior peaking of the tentorium (orange arrow)

A persistent falcine venous sinus (black arrow) and fenestration of the superior sagittal sinus (white arrow), a non-developed straight sinus and the internal cerebral veins draining into the falcine sinus.

DISCUSSION

Atreticcephaloceles represent 37.5% of all types of cephaloceles[8].They are abortive rudimentary cephaloceles, benign malformative lesions consisting of meningealand vestigial tissues (arachnoid, glial or CNS rests).Therefore they have a more favourable prognosis than the true encephaloceles[2].

The development of an AC is usually explained by a partial failure of the neural tube to close. The cranial bone defect is secondary to the formation of the AC, and it is due to a failure of mesodermal interposition between the cutaneous ectoderm and neuroectoderm[8].

Patterson *et al*. divided children with AC into two subsets: those with and those without vertical embryonic positioning of the straight sinus. Embryonic positioning of the straight sinus in these lesions has been frequently found, and it is a marker of the timing of the embryologic insult that caused the formation of the AC [7].

Vertical embryonic positioning of the straight sinus is characterized by anomalous veins, including internal cerebral veins, the great vein of Galen, and avertically positioned straight sinus in the falcine sinus, which extends superiorly within the large cistern in the posterior interhemispheric fissure (Fig. 4a, b). While the vertical straight sinus drainsinto the superior sagittal sinus, the cerebrospinal fluid tract maintains a position posterior to the anomalousveins, runs through the superior sagittal sinus, and extends to the parietal ACthrough a skull defect. These venous anomalies are exclusively encountered when the AC is found above the torcular [4, 5,9, 10].

Formation of vertical embryonic positioning of the straight sinus has been explained by the fibrous strand, which is part of the AC, preventing movement of the straight sinus from the embryonic vertical position to the adult horizontal position [7].

Cephaloceles can be detected by prenatal US but as they are very rare, and sometimes very small, they are not expected by the examiner. There is a wide range of clinical presentations of patients with AC. A child may be normal with regard to neurodevelopmental milestones, or may have severe mental retardations if the AC is associated with other intracranial anomalies: malformations of cortical development,Walker–Warburg syndrome, Dandy–Walker syndrome or ventriculomegaly[11].

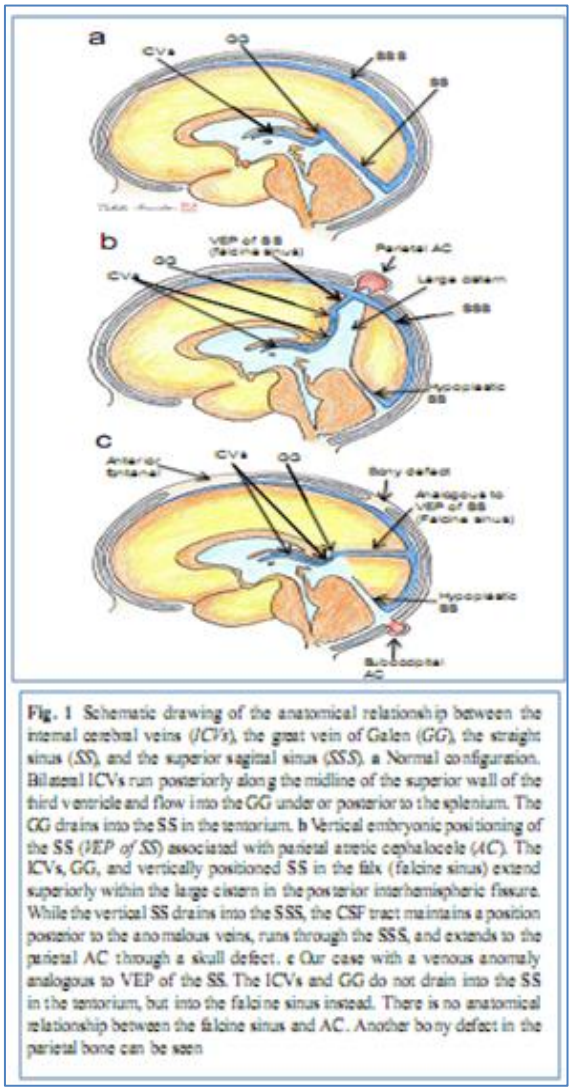

Fig. 1 Schematic drawing of the anatomical relationship between the internal cerebral veins (*ICVs*), the great vein of Galen (*GG*), the straight sinus (*SS*), and the superior sagittal sinus (*SSS*). **a** Normal configuration. Bilateral ICVs run posteriorly along the midline of the superior wall of the third ventricle and flow into the *GG* under or posterior to the splenium. The *GG* drains into the SS in the tentorium. **b** Vertical embryonic positioning of the SS (*VEP of SS*) associated with parietal atretic cephalocele (*AC*). The ICVs, *GG*, and vertically positioned SS in the falx (falcine sinus) extend superiorly within the large cistern in the posterior interhemispheric fissure. While the vertical SS drains into the SSS, the CSF tract maintains a position posterior to the anomalous veins, runs through the SSS, and extends to the parietal AC through a skull defect. **c** Our case with a venous anomaly analogous to VEP of the SS. The ICVs and *GG* do not drain into the SS in the tentorium, but into the falcine sinus instead. There is no anatomical relationship between the falcine sinus and AC. Another bony defect in the parietal bone can be seen

Fig-4: Nobuya Murakami, Takato Morioka, Nobuko Kawamura, Satoshi O. Suzuki, Ryutaro Kira (2016) Venous anomaly analogous to vertical embryonic positioning of the straight sinus associated with atreticcephalocele at the suboccipital region Childs NervSyst DOI 10.1007/s00381-016-3134-y.

CONCLUSION

Atreticcephalocele should be included in the differential diagnosis of a posterior scalp lesion. Key radiological findings on MRI or at fault the scanner are crucial in identification. Prognosis depends on associated intracranial anomalies.

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