MR Images of Spontaneously Involuted Atretic Cephalocele Concomitant with Persistent Falcine Sinus in an Adult

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Atretic cephalocelces are defined as skin-covered midline subscalp lesions that contain meninges and rest of glial and/or central nervous system tissue. When the straight sinus is absent or rudimentary, the falcine sinus can be recanalized to enable venous drainage. Although the atretic cephalocele or persistent falcine sinus has largely been described in the pediatric population, it is a rarely observed in the adult population. We report a unique case of spontaneously involuted atretic cephalocele coexistent with persistent falcine sinus in an adult. MR images and MR venography were useful for diagnosis and accurate anatomical depiction.

Index words : Brain, MR images, MR venography
Persistent falcine sinus
Atretic cephalocele

Introduction

Atretic cephaloceles are midline anomalies, usually located in the interparietal or occipital region and covered by abnormal skin, and associated with venous anomalies including absence of the straight sinus or duplication of the longitudinal sinus (1). Cephalocele is known to be a remnant of a neural tube defect as well as a form of occult cranium bifidum with rudimentary cephalic hernia (2). In spite of embryologic attention paid to theories of these lesions, it remains under the controversial debate. The falcine sinus is a normal structure located in the falx cerebri, normally involuting after birth (3). When the straight sinus is absent or rudimentary, the falcine sinus can be recanalized to enable venous drainage in response to the arteric straight sinus (3, 4). The atretic cephalocele or persistent falcine sinus has largely been described in the pediatric population, and rarely observed in the adult population (4, 6). To the best of our knowledge, MR image findings of spontaneously involutes atretic cephalocele coexistent with persistent falcine sinus has not been documented before in literature. We describe a unique case of spontaneously involuted atretic cephalocele coexistent with persistent falcine sinus in an adult.

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Case Report

A 45-year-old man presented to our out-patient department with presented chronic intermittent headache and palpable scalp mass in the middle of the high parietal area. The patient had no history of blunt head trauma or cranial surgery. Personal medical history included hypertension and non-insulin dependent diabetes mellitus. Since his birth, a soft tissue mass was observed in the middle of the high parietal area. Physical examination results were normal except for an elevated and mild tender soft tissue mass located at the posterior midline of the vertex. The scalp appeared moderate alopecia. No change in shape or size was noted when he made strong expiration with Valsalva maneuver. On neurological examination, he showed no pathognomonic neurologic response but healthy. Laboratory examination showed a normal range.

Plain radiograph of the skull revealed a coin sized well-defined bony defect at interparietal region (Fig. 1a). MR images revealed 2 cm-sized calvarial defect as well as an U-shaped hypointense subscalp lesion extending to congenital bony defect, and meninges covered the residual cephalocele with its continuity (Fig. 1b, c). Contrast enhanced T1WI showed non-enhancing hypointense subscalp lesion and anomalous falcine sinus (Fig. 1d). Anterior cerebellar vermis atrophy appeared on the MR features. Gadolinium-enhanced three-dimensional gradient-echo MR venography (3D MRV) clearly visualized the fetal falcine sinus with tapered stenotic ending (Fig. 1e). According to the 3D MRV, there was no visible normal

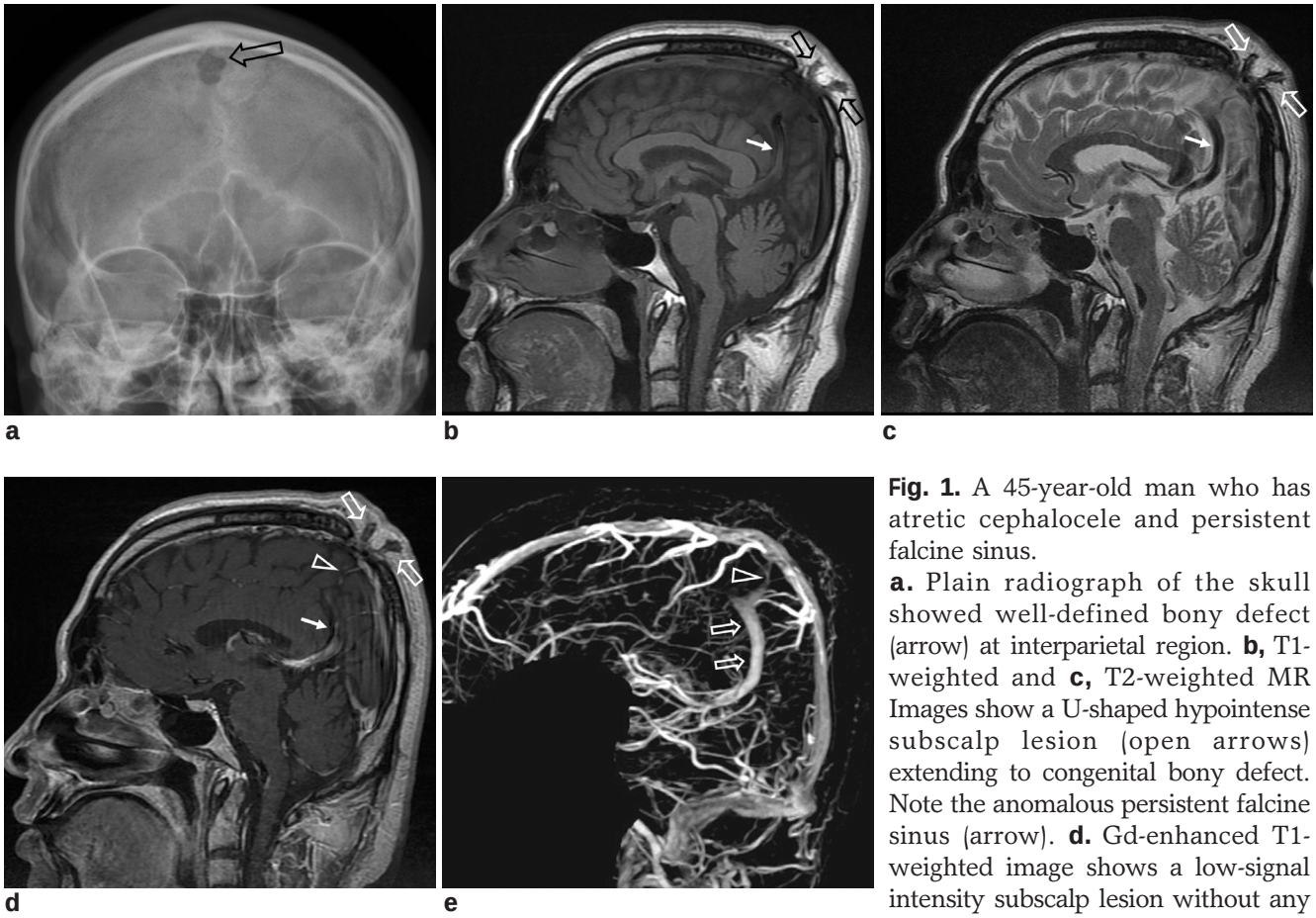

Fig. 1. A 45-year-old man who has atretic cephalocele and persistent falcine sinus.
a. Plain radiograph of the skull showed well-defined bony defect (arrow) at interparietal region. **b,** T1-weighted and **c,** T2-weighted MR Images show a U-shaped hypointense subscalp lesion (open arrows) extending to congenital bony defect. Note the anomalous persistent falcine sinus (arrow). **d.** Gd-enhanced T1-weighted image shows a low-signal intensity subscalp lesion without any significant enhancement (open arrows), and the anomalous persistent falcine sinus (arrow) and stenotic drain vein (arrowhead). The distal portion of the persistent falcine sinus, however, was not visualized clearly. **e.** Contrast-enhanced 3D MR venography clearly demonstrates marked stenosis of outflow (arrowhead) of the persistent falcine sinus (open arrows) at the junction of falcine and superior sagittal sinus.

straight sinus, but tight and small venous opening of the prominent persistent falcine sinus was definitely drained to the superior sagittal sinus through the stenotic vein. The end of the tapered venous defect was not cut off, but was narrowed from proximal vein to distal. As a result of MR image findings, surgery was not performed and planed follow up.

Discussion

Cephaloceles are congenital herniations of intracranial structures through a skull defect. The expression 'atretic', 'abortive', 'occult' and 'rudimentary' cephalocele is meaning spontaneous arrest in the development of malformation, and referring to a skin-covered subscalp lesion that consists of meninges and rests of glial and/or central nerve tissues (6). Although the embryopathogenesis of atretic cephaloceles is still obscure, many different explanations have been described the developmental origin of atretic cephalocele, those assuming a failure or a remnant of neural tube closure, excessive doses of vitamin A, X-ray exposure, trypan blue and other teratogens can lead to cephaloceles (7). Cephaloceles are related to the midline closure of the neural tube and are sometimes associated with congenital anomaly such as porencephaly or Dandy-Walker malformation, Chiari malformation, and agenesis of the corpus callosum (7, 8). Parietal cephaloceles carried a much less favorable prognosis than those in the occipital region, regardless of the type of cephalocele; they were associated with cerebral malformations more frequently and were more severe than occipital cephaloceles (8).

On MR images, 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 (9). The differential diagnosis of cystic cephalocele includes dermoid cyst and sinus pericranii (5, 6). On a plain skull radiograph, dermoid cyst shows a rounded defect with partially sclerosed margins as opposed to atretic cephalocele where the defect is oval or elongated. The edges of the dermoid cyst on CT narrow from outside inwards, the opposite being seen in atretic cephalocele, in which case the margins narrow from inside outwards. Dermoid cysts do not enhance with contrast where as atretic cephaloceles enhance vividly. Sinus pericranii are soft bulging masses in the scalp situated close to the midline in the parietal area. They represent dilated veins that communicate with the dural sinuses by way of an emissary vein. In the present case, we did not observe the findings of subscalp cystic or enhancing lesion, and there was U-shaped contracted sac suggesting spontaneous involution of atretic cephalocele. These are unique findings of spontaneous involution of atretic cephalocele, and differ from the subscalp cystic lesions or pediatric atretic cephalocele. In our case, it is interesting to note that the patient had a normal mental development during his childhood and the findings were totally incidental. The presence of atretic cephalocele does not necessarily add to the bad prognostic value as seen in our case.

The falcine sinus is defined as an ascending, rather than descending midline single vein bridges the great vein of Galen and sagittal sinus above the torcular. In our case of atretic cephalocele, the persistent falcine sinus ascended through the interhemisphere in the parietal lobe and joined the superior sagittal sinus at interparietal position. Venous anomalies are known to be associated with atretic and other cephaloceles, and they include absence of the straight sinus and duplication of the longitudinal sinus (1). Absence of the straight sinus and the persistence of the fetal falcine sinus suggest the presence of a normal accessory sinus that can be observed in fetus. In patient with posterior cephaloceles, the presence of an adhesion between the skin and the brain structures may have prevented the normal straight sinus from descending (1). Sener et al. described that persistent falcine sinus was associated with arteriovenous malformations, total absence of the corpus callosum, acrocephalosyndactyly (Apert's syndrome), osteogenesis imperfecta, Chiari II malformation (3).

When the straight sinus is absent or rudimentary, the falcine sinus can be recanalized to enable venous drainage in response to the atretic straight sinus. However, the falcine sinus can be detected in patients with absent, rudimentary or entirely normal straight sinuses, and this suggested that a mesenchymal disorder can be the primary cause for an open falcine sinus either in isolation or in association with variable changes in the straight sinus (3). It was not clear whether the falcine sinus became recanalized due to

the obstruction of the sigmoid sinus and potential increased venous pressure or had always been present as a congenital variation. Which one of the reasonable mechanisms causes the venous flow patency in falcine sinus is still obscure.

In our case, as well as the persistent falcine sinus with stenotic ending continuously drained to superior sagittal sinus, spontaneously involuted atretic cephalocele was identified on MR images. 3D MRV clearly depicted marked stenosis of outflow of the persistent falcine sinus at the junction of falcine and superior sagittal sinus. Gadolinium chelate reduces the spin saturation and is best administered as a bolus to avoid enhancement of chronically thrombosed venous structures, and 3D MRV is often superior to two-dimensional time-of-flight (2D TOF) in the delineation of major cerebral venous structure (10).

We have experienced a unique case of spontaneously involuted atretic cephalocele coexistent with persistent falcine sinus in an adult, and MR images have not been presented in literature. This particular abnormality can be considered as a primary or metastatic bony lesion or benign subscalp lesion. Radiologist should be aware of this relatively rare condition and the imaging findings, and is therefore helpful in the diagnosis and management of the patient.

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성인에서 지속적인 낫형동을 동반한 자연퇴축된 위축뇌탈출증의 MR영상소견

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위축뇌탈출증(atrectic cephalocele)은 중앙의 두피하부에 있는 피부로 덮힌 병변으로서 수막과 아교세포나 중추신경계 조직의 잔유물들을 포함한다. 수직동이 없거나 잔유물로 남아있는 경우 낫형동(falcin sinus)이 다시 관류하게 되어 정맥의 배출이 가능하게 된다. 위축된 뇌탈출증 또는 지속적인 낫형동은 소아에서 주로 기술되어 왔으며, 성인에서는 매우 드문 병변이다. 저자들은 성인에서 자연퇴화된 위축뇌탈출증이 지속적인 낫형동을 동반한 매우 드문 증례를 보고하고자 하며 MR영상과 MR정맥조영술은 정확한 해부학적 묘사와 진단에 유용하였다.

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