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PERSISTENT EMBRYONIC FALCINE SINUS PRESENTING AS ATRETIC PARIETAL CEPHALOCELE: CASE REPORT

Radiodiagnosis

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ABSTRACT

Cephaloceles are congenital neural tube defect causing herniations of intracranial structures (dura, fibrous tissue and dysplastic brain tissue) through a skull defect. Frequently associated with other intracranial anomalies like grey matter heterotopia, Vein of Galen malformations, Ventriculomegaly, Walker–Warburg syndrome, lobar holoprosencephaly, Dandy–Walker syndrome, hypogenesis of the corpus callosum, interhemispheric cysts, microphthalmia, and retro-ocular cysts.We are presenting a rare case of persistent embryonic sinus presenting with parietal cephalocele.

KEYWORDS

Atretic parietal cephalocele, Persistent embryonic falcine sinus, Magnetic resonance imaging

CASE HISTORY

A 4 days old male baby presented with a painless midline high parietal swelling over the scalp since birth. On examination, the swelling was soft palpable fluctuant extracalvarial mass. His neurological examination was otherwise normal. No focal neurological deficit. No significant obstetric history. No previous antenatal ultrasounds done. Birth history is normal.

IMAGING FINDINGS

CT Brain plain, sagittal and coronal sections demonstrated a linear fibrous stalk like structure extending from cerebral parenchyma into the cystic scalp mass with a midline osseous defect in parietal region (Fig 1).

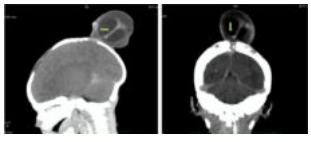

Fig 1: CT sagittal and coronal sections showing a fluid attenuation cystic lesion with a calvarial defect and linear hyperechoic stalk like structure extending intracranially.

Magnetic resonance (MR) image axial, coronal and sagittal sections showed a T1 hypointense, T2 hyperintense circumscribed cystic lesion in the soft tissues of the central parietal scalp with a connecting with a narrow fibrous stalk passed intracranially through a small osseous defect in the sagittal suture suggesting persistent falcine vein. A cerebrospinal fluid tract is seen leading towards the defect in the calvaria. Elevated straight sinus and prominent superior cerebellar cistern are seen with superior displacement of tentorium giving a spinning top appearance.

MR Venogram showed a falcine sinus with small fenestration in superior sagittal sinus. Findings were consistent with persistent embryonic falcine sinus with associated atretic parietal cephalocele (Fig 2). Operative findings showed a white fibrous stalk like structure within the cephalocele suggesting persistent falcine sinus (Fig 3).

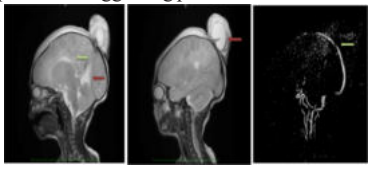

Fig 2: MRI brain sagittal sections showing a CSF intensity lesion with thin fibrous stalk like structure. Prominence of the superior cerebellar cistern and suprapineal recess noted. Superior peaking of the posterior tentorium with spinning top configuration of the tentorial incisura is seen.MR venogram showing focal fenestration in superior sagittal sinus posteriorly with a falcine sinus.

Fig 3: Intraoperative findings showing a cystic scalp swelling with a white fibrous stalk like structure depicting embryonic falcine sinus.

DISCUSSION

Atretic parietal cephalocele presents as a subgaleal soft tissue mass with an intracranial extension via a sharply demarcated calvarial defect (cranium bifidum), CSF tract and a vertically oriented falcine vein pointing to the subcutaneous scalp mass. They occur with an incidence of 1 in 3500 - 5000 live births [1, 2]. Atretic cephaloceles are considered as rare degenerative form of encephaloceles with an incidence of 4 – 17 % of all encephaloceles and most commonly located in parietal region. [3]. Main etio-pathogenesis include the persistence of neural bleb and neural crest remnants with resolution of involuted meningoencephaloceles in-utero [6,12]. Persistent falcine sinus is considered as a persistent vascular channel of sagittal plexus demonstrated as caudal anastomotic loops seen along the falx cerebri and showing communication with superior and inferior sagittal sinuses [8]. Patients most commonly present with a posterior scalp mass. Usually it is associated with other congenital abnormalities like walker-warburg syndrome, grey matter heterotopia, Vein of Galen malformations, Ventriculomegaly, lobar holoprosencephaly, Dandy–Walker syndrome, hypogenesis of the corpus callosum, interhemispheric cysts, microphthalmia, chiari II malformations, Apert's syndrome and retro-ocular cysts [9,10].

Falcine sinus can be classified basing on the communication with superior sagittal sinus. Type I falcine sinus shows no communication with the superior sagittal sinus, Type II falcine sinus shows faint communication with the superior sagittal sinus, and Type III falcine sinus shows significant communication with the superior sagittal sinus [11].

Hence imaging plays an important role in diagnosis and to detect associated abnormalities. MR imaging with venogram helps to demonstrates embryonic falcine sinus as a fibrous stalk connecting the cephalocele with focal fenestration of superior sagittal sinus at the level of atretic parietal cephalocele connecting to the lesion. Prominence of the superior cerebellar cistern and suprapineal recess. Superior peaking of the posterior tentorium with spinning top configuration of the tentorial incisura [5]. However, prognosis depends on the associated intracranial anomalies. With no other CNS abnormalities, majority of these patients have a good prognosis [4].

Differentials to consider include Sinus pericranii, Dermoid or Epidermoid cyst, Cephalhaematoma, Sebaceous cyst, cystic hygroma and Vascular lesions like Haemangioma [7].

CONCLUSION

Atretic parietal cephaloceles are considered as a rare congwnital anomaly. Hence clinician should be aware of atretic parietal cephalocele with underlying persistent embryonic falcine sinus and should be considered as one of the differentials in a paediatric patient with parietal scalp swelling. Imaging plays a crucial role in diagnosis of such conditions and helps the clinician in surgical management. MRI is considered in identifying and demonstration of the diagnosis.

SOURCE OF SUPPORT: Nil

CONFLICT OF INTEREST: None

INFORMED CONSENT FROM THE PATIENT: Yes

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