ATRETIC PARIELT CEPHALOCELE

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Abstract

A posterior parietal midline subscalp swelling noticed in a 19 month boy, by his family. These nodule had been enlarging slowly since birth. Radiologic imaging revealed that, subscalp lesion is a cyst and there was a communication between cyst and subarachnoid space via a calvarial defect. The other finding was “Vertically positioned straight sinus”. With this imaging findings our diagnosis was “Atretic parietal cephalocele”. Calvarial subcutaneous swellings, may caused by simple lesions, like lipoma, sebaceous cyst or it can be a sign of intracranial pathology. Atretic parietal cephalocele and concomitant pathologies can be set forth in a detailed manner with radiological imaging techniques.

Keywords: atretic, cephalocele, CT, MR.

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Introduction

Cranial subcutaneous swellings included in a wide spectrum of differential diagnosis. Trying to understand the pathology only by physical examination may lead to wrong diagnosis. Sebaceous cyst, lipoma, inclusion cyst, cephalocele, sinus perikranii, hemangioma, benign and malignant tumors, may be considered as the causes of scalp swelling. In addition bone and soft tissue infections, post-traumatic lesions can create a similar appearance¹.

By using radiological imaging, we can demonstrate characteristic of these lesions, borders, relationships with intracranial structures and associated anomalies in a detailed manner.

We present “Atretic parietal cephalocele” as a rare case which should be considered in the differential diagnosis of subcutaneous swellings.

Case Report

A nineteen months-old boy, who has no abnormalities of physical and neurological development, was brought to pediatrics clinic by his family with complaints of a slowly growing swelling which persist for long time in scalp. In physical examination, a non-rigid mass about 1 cm in diameter was present in the posterior parietal region. He has a normal history of birth and medical-family history. Any disease associated with the central nervous system was unremarkable.

Patient referred to our radiology department for researching lesion characterization, origin and relationships with intracranial structures. A cranial MRI study performed with 1.5T MR (Achieva,Philips)scanner.T1W,T2W,T2-FLAIR sequences in the axial, T2-W sequences in the sagittal and coronal images was obtained.

A cystic appearance in size of 13 x 10 mm was extending with the cerebrospinal fluid tract, under to skin, via sagittal suture, in posterior parietal subscalp region, T1hypointense, T2W hyperintense, on MR imaging (Figure 1).
Atretic parietal cephalocele is a benign subcutaneous lesion which presented with swelling in the scalp that covered with skin. Parietal cephalocele, constitute approximately 10% of cephaloceles. At first it was thought as a degenerative form of encephalocele and has been called as ‘meningocel manque’, by James and Lassmann in 1972.

Discussion

Atretic parietal cephalocele is a benign subcutaneous lesion which presented with swelling in the scalp that covered with skin. Parietal cephalocele, constitute approximately 10% of cephaloceles. At first it was thought as a degenerative form of encephalocele and has been called as ‘meningocel manque’, by James and Lassmann in 1972.

A brain CT examination with 16-detector Computed Tomography was performed to reveal relationships with bony structures. On CT examination a lesion in diameter of 13 mm with smooth margins, located in the posterior parietal subscalp region. It was extending from quadrigeminal cistern with CSF tract through sagittal suture to subscalp area (Figure 3).

With CT examination we understand that there was a diastasis at sagittal suture rather than a bone defect. CT Volume images of skin lesions in three-dimensional evaluation was reconstructed (Figure 4).

On MR and CT examinations, any obvious pathology in midline structures and other locations wasn’t detected. The radiological appearance was consistent with “Atretic parietal cephalocele”.

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Discussion

Atretic parietal cephalocele is a benign subcutaneous lesion which presented with swelling in the scalp that covered with skin. Parietal cephalocele, constitute approximately 10% of cephaloceles. At first it was thought as a degenerative form of encephalocele and has been called as ‘meningocel manque’, by James and Lassmann in 1972.
These cystic structures may contain meningeal, neural or glial residues. Although the etiology is unknown, the neural tube closure defects considered for pathology. Generally it occur as sporadic. In “Type 1 cephalocele”, arachnoid tissue and abnormal blood vessels cluster present in the lesions. In “Type 2 cephalocele” the vessels extends to lesion dome and neural-glial remnants can be found in.

Patients with atretic cefalosel not only have normal physical and neurological development, but also may possess with clinical findings which arise from different intracranial anomalies. Mental retardation is a often visible finding. Intracranial abnormalities such as Walker - Warburg syndrome, holoprosencephaly, Dandy-Walker syndrome, gray matter heterotopia, corpus callosum abnormalities, retrobulbar cysts may be present with atretic cephalocele.

Atretic parietal cephalocele and concomitant pathologies can be set forth in a detailed manner with radiological imaging techniques. As the best modality for soft tissue with multiplanar and high-resolution images. MR is very valuable in the diagnosis of cephalocele. CT can be used to put forward lesion's relationship with bone structure when needed. Curnes and Oakes defined three parietal cephalocele cases by using current version, old -tech CT and MRI in 1984-87. All patient has hydrocephalus, two had Dandy Walker Malformation, and in one the Chiari type 2 malformation was present.

CSF tract that extends through under the skin via calvarial defects and distal cystic lesions were radiographic findings in the diagnosis of cephalocele. Vertical course of straight sinus in the embryonic trace is seconder finding in majority of patients. Also, other intracranial malformations may be set forth with MR examination in a successful way.

After excision of the cyst, atretic cefalosel can be treated successfully with sutured dural defect and skin. Generally, the prognosis of patients with atretic cephalocele is good. As far as we know, in the literature, only one atretic cephalocele patient died because of multiple anomalies. Other patients, either asymptomatic or present with symptoms of related to associated anomalies. In our patient, any accompanying symptoms or abnormalities were not observed, except swelling.

As a result, the calvarial subcutaneous swellings, may caused by simple lesions like lipoma or sebaceous cyst or it can be a sign of intracranial pathology. After clinical evaluation, reason of swelling and existing complex developmental malformations can be demonstrated by radiological imaging techniques, in the early stages.

Declaration of Interest

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References