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Case Report

A Case of Occipital Rudimentary Cephalocoele

Abstract

We report a case of a 1-month-old boy with a cystic swelling in the occipital region without intracranial communication, called atretic cephalocoele. We discuss clues to the diagnosis of atretic cephalocoeles. We also discuss common clinical findings and a possible mechanism by which these lesions develop.

Keywords: *Atretic, cephalocoele, occipital*

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Introduction

Encephalocele is a congenital malformation of the central nervous system. It is the result of failure of the surface ectoderm to separate from the neuroectoderm. This results in a bony defect which allows herniation of the meninges or herniation of the brain tissue.[1-3] Various authors have reported small, skin-covered midline sub-scalp masses or cysts described as atretic cephalocoeles, atypical meningoceles, rudimentary meningoceles, and meningeal heterotopias.[1-7] Atretic cephalocoeles are different from common meningocele, in that they lack an intracranial communication.

We here report a 1-month-old boy who presented with a scalp mass, presenting since birth and was diagnosed as an atretic parietal encephalocele.

Case Report

A 1-month-old male child presented with a swelling in the occipital region, present since birth. The swelling, according to parents increased gradually to attain the present size. It was 14 cm × 12 cm in size. The area was marked by dysplastic skin with a ring of coarse hair surrounding the lesion. The lesion itself appeared cystic in nature and contained clear fluid. It was compressible, nonpulsatile, transilluminant, and did not appear to change in size or tension relative to position and crying. Computed tomography scan of the head revealed a 14 cm subcutaneous mass of soft tissue density similar to the brain tissue, with no intracranial communication. The brain parenchyma appeared unremarkable [Figure 1].

During surgery, an elliptical incision was made surrounding the lesion, which was removed in its entirety. The underlying pericranium and calvarium was completely normal without attachment to the overlying anomaly [Figures 2 and 3]. There was no identifiable fibrous stalk or bony defect communicating the lesion with the intracranial compartment.

Discussion

The lesion has been referred with varying nomenclature as type 1 primary cutaneous meningioma, hamartoma of the scalp with ectopic meningothelial elements, heterotopic meningeal nodules, sequestrated meningocoele of the scalp, rudimentary meningocoele, meningocoele manqué, arachnoid rest, extruded dura, or atretic cephalocoeles.[1-4] The various names in vogue reflect the confusion that prevails regarding the pathogenesis.

Sequestrated encephalocoele or rudimentary cephalocoeles are described as atrophic skin-covered scalp lesions that may be either solid or cystic, occur frequently in the posterior midline, may contain cerebrospinal-like fluid, and are often marked by alopecia or a dark "hair collar" sign.[1,2] Rudimentary cephalocoeles are distinguished from classic cephalocoeles by the absence of any direct communication between the extracranial meningothelial and/or neural elements with cerebral parenchyma; although the tract can be

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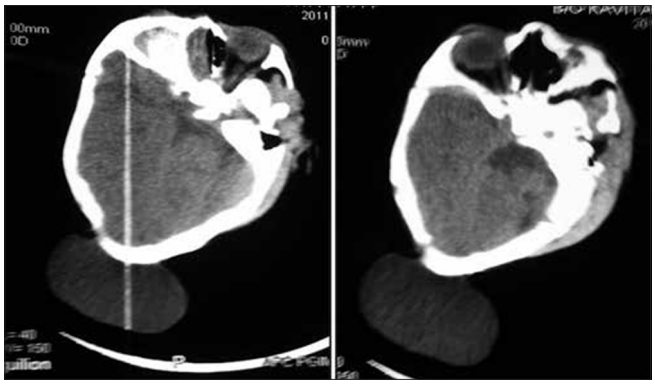

Figure 1: Computed tomography scan showing cystic occipital swelling

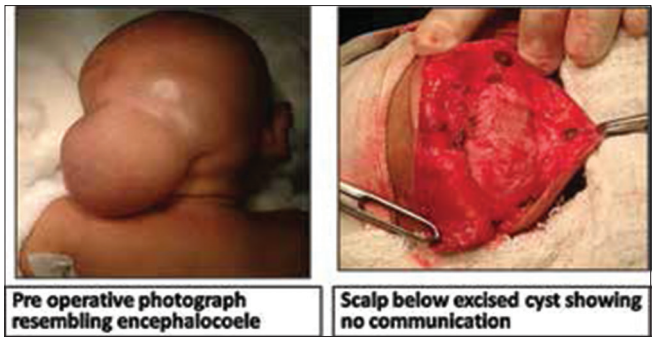

Figure 2: Intraoperative photographs

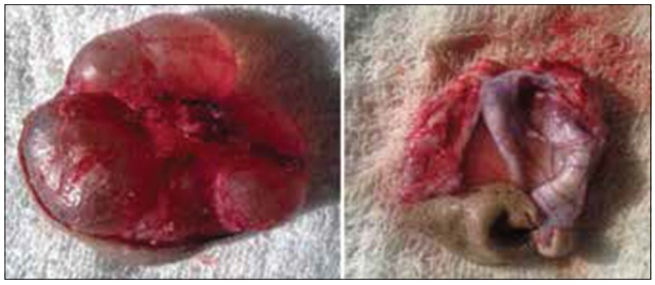

Figure 3: Photographs of excised cyst showing clear fluid-filled cysts

in continuity with the subarachnoid space.[1,3] When the rudimentary cephalocoele ends in a fibrous tract, the tract is devoid of any nervous tissue and lacks a discernible lumen.[1,4]

Lopez *et al*.[1] classified cutaneous meningiomas into three types:

Type I – Primary cutaneous meningioma; Type II – Ectopic meningioma of the soft-tissue with extension into the skin; and Type III – Central nervous system meningioma with extension into the skin.[1] According to Lopez *et al*.,[1] primary cutaneous meningiomas and classical meningocoeles represent two ends of a spectrum that includes solid meningeal hamartoma and rudimentary meningocoeles. Furthermore, rudimentary meningocoele varied from primary cutaneous meningioma by having very little hyperplastic cells and being predominantly cystic in nature.[1]

Sequestrated meningocoeles have predilection for the scalp, adjacent to cranial sutures,[5,6] however, they may also occur over the spine.[7] They have also been reported to occur along the peripheral nerves.[8] Based on this site distribution, various theories have been hypothesized. Sutural location is described by nonfusion of neuroectoderm with surface ectoderm with "trapping" or "isolation" of meningeal elements in the scalp that are "pinched off" during closure of the neural tube with or without skull bone defect or fibrous connection, that at spine is described by ectopic location of arachnoid rests. The location in peripheral nerves is described by the displacement of meningeal tissues along the cutaneous nerves leading to the ectopic location of meningeal tissues.

Atretic cephalocoeles mostly occur in parietal and occipital regions.[9,10] The incidence of atretic cephalocoeles is 4–17% of all cephalocoeles[9] and the parietal location compromises 40–50% of cases.

Yokota *et al*.[9] stressed the importance of the location of the atretic cephalocoele as related to the prevalence of additional cranial anomalies. He stated that atretic parietal cephalocoeles were much more related with extracranial anomalies and showed worse prognosis. However, in another study by Martinez-Lage *et al*.,[10] occipital cephalocoeles were more related with cranial malformations and showed a worse prognosis. In our case, however, there were no associated intracranial malformations.

Extracranial heterotopia is described if brain tissue is present inside meninges in the scalp with or without associated skull defect or connection to the brain. In the presence of a connection, it will be considered as a variant of encephalomeningocoele.[11] The term "atretic cephalocele" is for scalp nodules that contain meningeal and glial elements and have connection to the dura, falx, or tentorium through a skull defect with or without associated intracranial anomalies.[12,13] Unlike above, sequestrated meningocoeles are noncommunicating with the intracranial structures and not associated with bone defects; however, a few may have a pedicle extending intracranially that should be carefully evaluated on imaging before surgery.[5-7] The distinction between sequestrated meningocoele and other lesions may not be feasible all the time on an imaging basis. In the case of a scalp swelling, thus, the surgeon needs to consider the possibility of fibrous connection to the brain that might not be apparent on imaging.

Conclusion

With this case, we would like to emphasize that when a scalp mass is noted at the midline or near the vertex, the possibility of an atretic cephalocoele should be ruled out as a rare cause of cranial masses.

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Conflicts of interest

There are no conflicts of interest.

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