Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.elsevier.com/locate/mjafi

Case Report

Atretic parietal encephalocoele – An unusual diagnosis

Col C.G. Muralidharan^{a,*}, Wg Cdr Rohit Aggarwal^b, Col Dalbara Singh^c^a Senior Advisor (Radiodiagnosis), Military Hospital, Jalandhar Cantt, Punjab, India^b Classified Specialist (Radiodiagnosis), AIIMS, New Delhi 110608, India^c Senior Advisor (Radiodiagnosis and Interventional Radiology), Military Hospital, Jalandhar Cantt, Punjab, India

ARTICLE INFO

Article history:

Received 6 September 2011

Accepted 7 February 2012

Available online 24 August 2012

Keywords:

Straight sinus

Encephalocoele

Flow void

Spinning top

Peaking

Introduction

Atretic encephalocoeles are small, skin covered subscalp lesions that contain meninges, neural and glial rests. They have been termed atypical or rudimentary meningocoeles, meningeal heterotopias and meningocoele manqué.¹ These rare lesions generally occur within a few centimetres of the lambda and may be associated with intracranial anomalies. Approximately half the reported cases are parietal in location.² Although their pathogenesis has not been definitively elucidated, they are thought to lie in the spectrum of defects of neural tube closure. It has been proposed that they represent remnant nuchal blebs that may relate to transient over-distension of the rhombencephalic vesicle.² We report a case of atretic parietal encephalocoele with characteristic imaging features.

Case report

A 22 month old male child presented with history of painless, cystic swelling in the parietal region gradually increasing since birth. There was no history of trauma or pus discharge. The parents did not notice any increase in the size of swelling with crying. On examination a cystic swelling was seen in the superior parietal region. Underlying bones appeared normal on palpation. The swelling did not show transillumination. The overlying skin was normal and could be moved over the swelling. He is the first child of his parents, born at term, from a non consanguineous marriage, without any remarkable prenatal or natal history.

Multiplanar imaging was done using T1 FLAIR, T2 FLAIR, and T2W FSE sequences on a 1.5T high definition magnetic resonance (HDMR) imaging system (GE Signa Excite 1.5T HD, USA). The study revealed a well defined, midline, spherical, cystic, subscalp lesion in the parietal region measuring 23.5 mm × 14.4 mm × 19.2 mm (APXTRXCC). The lesion was isointense to brain on T1W and FLAIR images and hyperintense on T2W images (Fig. 1a,b). Sagittal T1W and T2W images demonstrated a bony defect involving the underlying parietal bone. A linear flow void on T2W sagittal images was seen extending posterosuperiorly into the interhemispheric fissure representing the embryonic positioning of the straight sinus (Fig. 2). This was hyperintense on T1W and on sequential images this could be traced to the base of the cystic swelling (Fig 3). Prominence of superior cerebellar and suprapineal recess of third ventricle was noted (Figs. 2 and 3). Coronal images were remarkable for superior peaking of the posterior tentorium with elevated straight sinus identifiable as a linear flow void at the apex of the tentorium (Fig. 4a). The axial images revealed a spinning top configuration of the tentorial

* Corresponding author. Tel.: +91 9915161745.

E-mail address: murali.cg@rediffmail.com (C.G. Muralidharan).

0377-1237/\$ – see front matter © 2012, Armed Forces Medical Services (AFMS). All rights reserved.

<http://dx.doi.org/10.1016/j.mjafi.2012.02.005>

incisura (Fig. 4b) and a cigar shaped cerebrospinal fluid tract could be sequentially followed superiorly in the interhemispheric fissure upto the base of the cystic swelling. No other intracranial abnormality was seen. Based on these features a diagnosis of atretic parietal encephalocele was made. The patient is being managed conservatively as the parents have refused surgery.

Discussion

Atretic encephalocele was first reported by James and Lassman in 1972 who defined it as a degenerative form of encephalocele.³ Its reported incidence is 4–17% of all the encephaloceles⁴ and parietal location comprises 40–50% of cases. It has been rarely reported in adults.⁵ The embryological defect in these lesions is faulty neural tube closure as in conventional encephaloceles. The solid components of these lesions have been proposed to originate from arachnoid cell rests displaced during embryogenesis into cutis or subcutis or a hamartoma with ectopic component of meningotheial cells.⁶ Associated intracranial venous anomalies have been reported in literature.^{7,8} Causation of abnormal venous drainage in the great vein of Galen, the straight sinus and the superior sagittal sinus may be secondary, through interaction with a developmentally pre-existing encephalocele. Based on pathological features atretic encephaloceles are classified into two categories.² In Type I encephalocele the lesion consists mainly of arachnoid tissue found at the lesion stalk and is accompanied by a cluster of anomalous blood vessels. The dome consists of dermal tissue with normal hair follicles. In Type II encephaloceles the blood vessels extend towards the lesion dome and in this variety ectopic focus of neural/glial elements are also seen.

Parietal location is related to the prevalence of additional intracranial anomalies as compared to other locations.⁴ Atretic parietal encephaloceles have a very high incidence of associated midline anomalies, including porencephalies, interhemispheric cysts and callosal agenesis which affect prognosis of the patient.^{9,10} Other associated malformations include ventriculomegaly, cystic malformations, heterotopias

Fig. 1 – T2W image (1a) and T1W image (1b) shows a parietal encephalocele hyperintense on T2W images and hypointense on T1W.

Fig. 2 – T2W sagittal image shows flow void extending posterosuperiorly representing embryonic position of the straight sinus, also note prominence of superior cerebellar and suprapineal recess.

of the posterior fossa. In our case no concomitant anomaly was seen despite the parietal location and typical imaging findings. The present case and that of Martinez-Lage et al² dispute the importance of location of the encephalocele as determinant of clinical outcome.

Clinical presentation of atretic encephalocele is variable. Patients may develop painless or painful subcutaneous nodules, masses or swelling. Severe mental retardation may

Fig. 3 – T1W sagittal image shows hyperintense straight sinus (white arrows) extending upto the base of encephalocele. Also note the bony defect in the parietal bone.

Fig. 4 – T2W coronal image (4a) shows the superior peaking with flow void of straight sinus at the apex. T1W axial image (4b) shows spinning top configuration of the tentorial incisura (white arrow).

occur in patients with associated anomalies such as Walker-Warburg syndrome, or ventriculomegaly.¹ The differential diagnosis of painless swelling in midline parietal location in an infant include cephalhaematoma, traumatic cyst, dermoid cyst, sinus pericranii and vascular malformations.¹¹ In the current case cephalhaematoma and traumatic cysts were excluded by absence of history of injury. Presence of embryonic straight sinus and absence of fat on MRI excluded the possibility of dermoid cyst. Sinus pericranii is filled with venous blood and is usually reducible.

Embryonic positioning of the straight sinus, prominence of superior cerebellar and suprapineal recesses, peaking of the posterior tentorium and spinning top configuration of the tentorial incisura are characteristic of atretic encephalocoeles.¹²

The presence of embryonic straight sinus, as seen in our case deserves a mention both as a marker of the timing of the embryologic insult and as a clue to radiologic diagnosis.^{2,10} Concomitant venous anomalies in these patients are better delineated by 3D-High Definition MR.⁸ Surgical treatment is offered in symptomatic cases and those with concomitant anomalies. Conservative approach may be followed in

asymptomatic cases and those akin to our case without any associated anomalies.⁵

Conflicts of interest

All authors have none to declare.

REFERENCES

1. Patterson RJ, Egelhoff JC, Crone KR, Ball WS. Atretic parietal cephalocoeles revisited: an enlarging clinical and imaging spectrum? *AJNR*. 1998;19:791–795.
2. Martinez-Lage JF, Sola J, Casas C, Poza M, Almagro MJ, Girona DG. Atretic cephalocele: the tip of the iceberg. *J Neurosurg*. 1992;77:230–235.
3. James CC, Lassman LP. Spina bifida occulta. In: James CC, Lassman LP, eds. *Spinal Dysraphism*. 1st ed. London: Butterworths; 1972:89–97.
4. Yokota A, Kajiwaru H, Kohchi M, Fuwa I, Wada H. Parietal cephalocele: clinical importance of its atretic form and associated malformations. *J Neurosurg*. 1988;69:545–551.
5. Ahmet Ş, Miktat K, Can HY, Erol T. Atretic cephalocele in adults. *Acta Neurochir*. 2011;153:2275–2277.
6. Suster S, Rosai J. Hamartoma of the scalp with ectopic meningotheial elements: a distinctive benign soft tissue lesion that may simulate angiosarcoma. *Am J Surg Pathol*. 1990;14:1–11.
7. Hsu SW, Chaloupka JC. Atretic parietal cephalocele associated with sinus pericranii: embryological consideration. *Brain Dev*. 2011 Jul 6.
8. Morioka T, Hashiguchi K, Samura K, et al. Detailed anatomy of intracranial venous anomalies associated with atretic parietal cephalocoeles revealed by high-resolution 3D-CISS and high-field T2-weighted reversed MR images. *Childs Nerv Syst*. 2009;25(3):309–315.
9. Yamazaki T, Enomoto T, Iguchi M, Nose T. Atretic cephalocele- report of two cases with special reference to embryology. *Child Nerv Syst*. 2001;17:674–678.
10. Drapkin AJ. Rudimentary cephalocele or neural crest remnant? *Neurosurgery*. 1990;26:667–673.
11. Massimi L, Debonis P, Esposito G, et al. Vertex scalp mass as presenting sign of a complex intracranial vascular malformation. *J Neurosurg Pediatrics*. 2009;3:307–310.
12. Saliha S, Umit MS, Nilgun E, Aysel Y, Can DK. A case of atretic parietal cephalocele. *New J Med*. 2007;24:237–238.