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CASE REPORT

Supratentorial endodermal cysts - Report of two cases

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ABSTRACT

We describe two rare cases of frontal cystic lesions presenting with symptoms and signs of raised intracranial pressure. Both had a preoperative diagnosis of an arachnoid cyst and were subjected to a craniotomy with marsupialization of the cyst. However, the histology confirmed them to be an endodermal cyst (EC) on both occasions. Both the patients have been closely followed with no recurrence of symptoms. ECs of the central nervous system are usually reported in the spinal canal, mid-line posterior fossa, and the suprasellar regions. Supratentorial and non-midline ECs are rare, with only about 22 cases previously reported in literature. We discuss both the cases and review the relevant literature.

Key words: Enterogenous cyst, frontal cyst, neurenteric cyst, supratentorial endodermal cyst

Introduction

Epithelial cysts are either neuroepithelial cysts or endodermal cysts (EC) based on their cell of origin. The neuroepithelial cysts are believed to arise from the heterotopic rests of primitive ependyma, whereas the ECs are believed to arise from the remnants of the transient neurenteric canal during notochordal development. This explains the fact that ECs tend to be axial in location while the neuroepithelial cysts tend to be more variable.[1,2]

Case Reports

Case 1

A 52-year-old female, with no known comorbidity, presented with complaints of episodic headache over the past 3 years. About a week back, she noticed right-sided limb weakness and difficulty in walking. On examination, higher mental function and cranial nerves were normal. Motor system examination revealed right-sided spastic hemiparesis with a power of grade 3/5. There were no cerebellar or meningeal signs.

A magnetic resonance (MR) image of the brain revealed a left frontoparietal lesion, hypointense on T1 and hyperintense on T2-weighted images, with no contrast enhancement, causing a mass effect [Figure 1a-e]. She underwent a left frontal craniotomy, maximum possible excision of the cyst wall and marsupialization of the cyst. Cyst fluid was xanthochromic. Wall of the cyst was sent for histopathological examination. She gradually improved in her hemiparesis over the next few months. Histology revealed the lesion to be an EC [Figure 1f]. At follow-up of 3 years, she is fine with no symptomatic or radiological recurrence.

Case 2

A 32-year-old male presented with an episode of generalized tonic-clonic seizure about a week back. He was admitted and started on antiepileptic medications. On examination, he had no neurological deficit.

An MR image of the brain revealed a bifrontal (left > right) lesion, hypointense on T1 and hyperintense on T2-weighted images, with no contrast enhancement, with a mass effect [Figure 2a-e].

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He underwent a left frontal craniotomy, maximum possible excision of cyst wall and marsupialization of the cyst. He did well postoperatively with no added deficit or fresh seizures. Histology revealed the lesion to be an EC [Figure 2f]. At follow-up of 2 years, the patient is fine with no symptomatic or radiological recurrence. He was gradually tapered of antiepileptic drugs.

Discussion

Epithelial cysts are either neuroepithelial cysts or ECs based on their cell of origin. Enterogenous, respiratory, and colloid cysts are of endodermal origin. The true neuroepithelial cysts are composed of choroid plexus or ependymal cysts which have neuroectodermal origin. Although the true incidence is still unknown, the retrospective studies suggest that intracranial ECs comprise approximately 0.15–0.35% of all intracranial neoplasms.[1,2] The predominant opinion is that these cysts form on account of the faulty separation of the ectodermally derived spinal canal and the endodermally derived foregut during the closure of the neurenteric canal in the 3rd week of embryonic life.[1,2]

ECs are usually found in the posterior mediastinum, abdomen, and pelvis, where these occur predominantly in males. These seldom occur in the central nervous system (CNS). EC originating in the CNS is most commonly located intraspinally, generally ventral to the spinal cord and is usually seen in the lower cervical and upper thoracic regions.[3-6] In the intracranial compartment, these are usually located in the posterior fossa, explained by the rostral closure of the notochord forming the clivus. Graziani *et al*. proposed that supratentorial EC originates from the Seessel's pouch, which in lower vertebrates give rise to the adenohypophysis. However, in humans, this may lead to the formation of EC along with Rathke's cleft cyst as well as colloid cysts, explaining their common immunohistochemistry. However, even this does not explain the extremely rare non-midline supratentorial cysts.[1,2,7]

ECs have been classified into three types by Wilkins and Odom. The simpler forms (Type A) are thin walled, with a layer of stratified or a pseudostratified cuboidal or columnar epithelium on a basement membrane. More complex varieties are less common and have additional mesodermal elements such as smooth muscle and fat (Type B) and sometimes also ependymal or glial tissue (Type C).[6]

Supratentorial locations were reported almost exclusively in adult cases, with about 22 cases reported until date.[6] These locations include the cavernous sinus, septum pellucidum, interhemispheric fissure, orbital apex, and the optic nerve.[4] Unlike spinal EC, where coexisting vertebral anomalies are seen in almost 50% of the cases, intracranial cysts are never associated with bony anomalies of the skull, clivus, or skull base. Solitary extra-axial supratentorial EC greatly outnumber the intra-axial localization. These occur in the 3rd to the 7th decade with a slight female preponderance. The clinical symptomatology appears to be secondary to

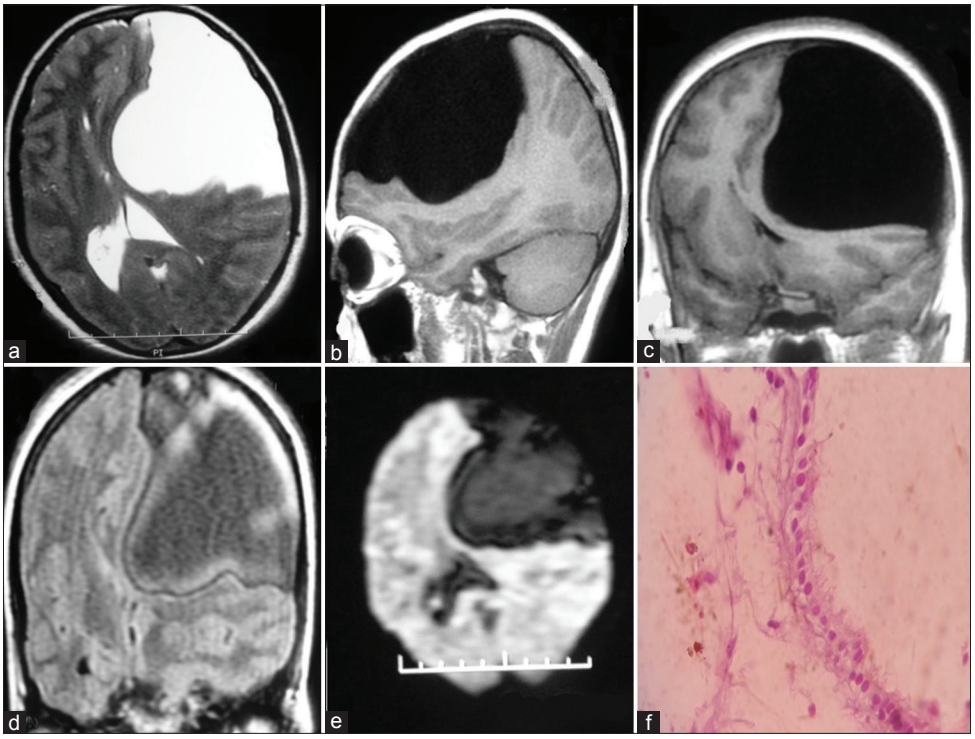

Figure 1: Left frontoparietal cystic lesion on T2 axial (a), T1 sagittal (b), T1 coronal (c), fluid-attenuated inversion recovery coronal (d) and diffusion (e) weighted magnetic resonance imaging. Histology of the cyst wall (f) microphotograph (oil immersion, ×100) showing ciliated cuboidal epithelium lining cyst wall

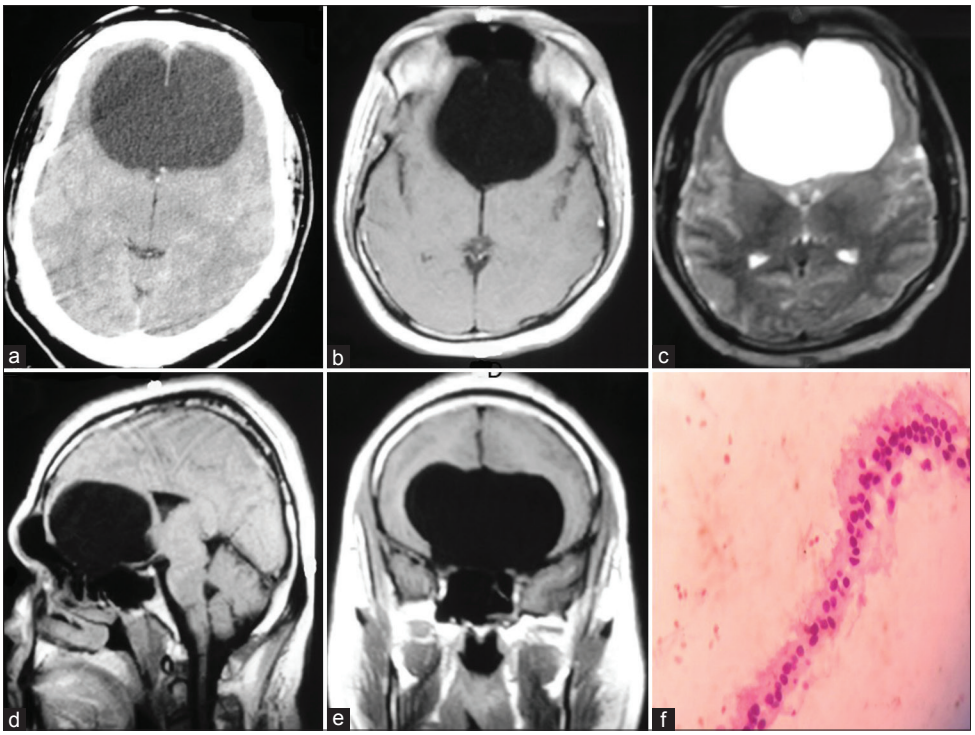

Figure 2: Bifrontal/anterior interhemispheric cyst on computed tomography (a), T1 axial (b), T2 axial (c), T1 sagittal (d) and T1 coronal (e) weighted magnetic resonance imaging. Histology of the cyst wall (f) Microphotograph (oil immersion, ×100) showing ciliated cuboidal epithelium lining cyst wall (arrow)

compression or irritation of surrounding neural structures and thus depend on their location, size, and content. Major clinical features include sensory and motor deficits, partial and generalized seizures, and manifestations of raised intracranial pressure.[6]

Computed tomography scan typically shows a low-density area with no contrast enhancement. Rarely, there may be rim enhancement.[6] On MR imaging, most ECs are isointense or hyperintense compared to cerebrospinal fluid in both T1- and T2-weighted images. Occasionally, they show homogeneous, very bright signal intensity on T1-weighted images and low signal intensity on T2-weighted images due to very high protein content. These cysts are hyperintense on fluid-attenuated inversion recovery and show no or mild diffusion restriction. EC is nonenhancing, but rare cases of rim enhancement and sometimes solid enhancement occur.[3] The differential diagnosis for intracranial EC on imaging include epidermoid/dermoid cysts, parasitic cysts, true neuroepithelial cysts, and arachnoid cysts.[6] Diagnosis of an EC is confirmed on histology.[8]

It is difficult to differentiate these various subtypes of ECs under a light microscope. Immunohistochemistry is a useful adjunct in these cases. However, in our case, immunohistological studies could not be done due to the poor financial status of patients.

The ideal treatment would be complete surgical excision, along with the cyst wall, with follow-up serial imaging for potential complications such as recurrence, dissemination, and malignant transformation.[3,6] However, the rarity of this location precludes a general consensus on the most appropriate line of management. We believe that the cysts which are symptomatic ought to be excised, the others can be closely observed. When surgically approached, these are best evacuated completely, and the wall excised in total. However, when this is not feasible as in our cases, it is best to attempt a maximal excision of cyst wall with marsupialization of the cyst and careful observation thereafter. Placement of a cyst-subarachnoid or cyst-peritoneal shunt or placement of an ommaya reservoir could be considered on recurrence.

Conclusion

ECs have been reported in the supratentorial region and ought to be kept in mind as a differential diagnosis of cystic lesions; whenever possible complete excision should be attempted.

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Conflicts of interest

There are no conflicts of interest.

References

1. Hasegawa M, Nouri M, Nagahisa S, Hayashi T, Adachi K, Hirose Y, *et al.* Neuroepithelial cyst of the fourth ventricle. Childs Nerv Syst 2015;31:155-9.

2. Cheng JS, Cusick JF, Ho KC, Ulmer JL. Lateral supratentorial endodermal cyst: Case report and review of literature. Neurosurgery 2002;51:493-9.
3. Arora R, Rani JY, Uppin MS, Ca R. An unusual case of large posterior fossa neurenteric cyst involving bilateral cerebellopontine angle cisterns: Report of a rare case and review of literature. Pol J Radiol 2014;79:356-9.
4. Al-Ahmed IH, Boughamoura M, Dirks P, Kulkarni AV, Rutka JT, Drake JM. Neurosurgical management of neurenteric cysts in children. J Neurosurg Pediatr 2013;11:511-7.
5. Chen PY, Wu CT, Lui TN, Jung SM. Endodermal cyst presenting as a prenatally diagnosed large intracranial cyst. Case report and review of the literature. J Neurosurg 2007;106 6 Suppl: 506-8.
6. Mittal S, Petrecca K, Sabbagh AJ, Rayes M, Melançon D, Guiot MC, *et al*. Supratentorial neurenteric cysts-A fascinating entity of uncertain embryopathogenesis. Clin Neurol Neurosurg 2010;112:89-97.
7. Graziani N, Dufour H, Figarella-Branger D, Donnet A, Bouillot P, Grisoli F. Do the suprasellar neuroenteric cyst, the Rathke cleft cyst and the colloid cyst constitute a same entity? Acta Neurochir (Wien) 1995;133:174-80.
8. Choi JE, Seol HJ, Cho YS. Giant petroclival endodermal cyst with xanthogranulomatous changes. J Neurosurg Pediatr 2013;12:284-7.