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CASE REPORT

Supratentorial neuroenteric cyst in children: a case report and brief literature review

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Abstract

Neurenteric cysts are rare developmental lesions typically found outside the central nervous system but when they do, we most likely find them in a spinal (cervical or dorsal) intradural extramedular location, often associated with dysraphism. The more unusual intracranial cases have been published because of its rarity, occurring mostly as a posterior fossa extra-axial cyst, in adults. Supratentorial cases are distinctly infrequent, especially in children, resulting in few case reports and even fewer case reviews. We describe a case of a child with a supratentorial neurenteric cyst and present a brief review of the literature about these cysts in children, a noticeable gap in the literature.

Keywords Intracranial neurenteric cyst · Endodermal cyst · Enterogenous cyst · Neuroenteric cyst · Supratentorial neurenteric cyst

Introduction

Neurenteric cysts (NEC) are uncommon congenital malformations of endodermal origin that result from displaced elements of the alimentary canal. They are usually located in the posterior mediastinum and only represent 0.01% of all CNS tumors. NEC can be found at any level of the neuraxis but they occur most likely as spinal (lower cervical or upper dorsal) intradural extramedular cysts, often associated with dysraphism [9]. Intracranial cases, predominately located in the posterior fossa in adults, are further unusual, what makes supratentorial ones the most infrequent [13, 14, 23], especially in children. In this report, we describe a rare case of a child with a supratentorial NEC and review the literature in pediatric population.

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Case report

A 14-year-old previously healthy boy with a history of chronic progressive headaches presented with a first complex partial seizure. He had no physical or neurological examination abnormalities. Cranial computed tomography (CT) showed a right parasagittal frontal cystic lesion and associated bone remodeling. Brain magnetic resonance imaging (MRI) revealed a well-defined thin-walled cyst with a total volume of 136 cc, slightly more hyperintense than CSF on T1, hyperintense on T2-weighted images, and hypointense in fluid-attenuated inversion recovery (FLAIR), with no fluid restriction on diffusion-weighted (DWI) and no contrast-enhancement (Fig. 1). Thereafter, the patient was placed on levetiracetam and became seizure-free. A whole spinal MRI was also performed and no pathological findings were found.

He underwent a right frontal craniotomy, maximum possible excision of the cyst wall and marsupialization of the cyst to the interhemispheric fissure. After the dura mater was opened a translucent thin-walled little-vascularized cyst was immediately exposed. Cystic fluid was whitish and opaque, with mucinous deposit, which was collected for culture and pathology (Fig. 2). The wall of the cyst was sent for histopathological examination.

There were no postoperative complications and the patient was discharged home 3 days later.

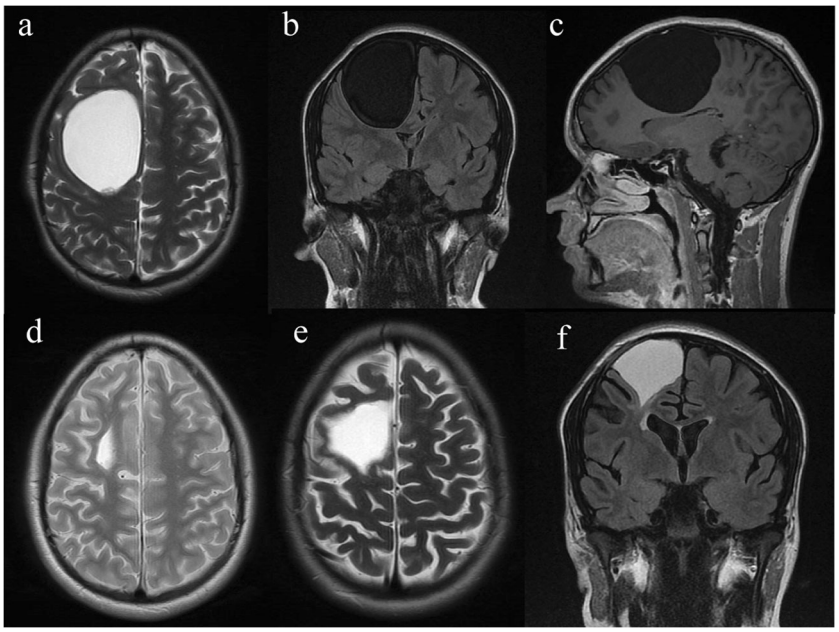

Fig. 1. Preoperative MRI: **a** axial T2, cystic intracerebral lesion with a posterior deposit; **b** axial post-contrast T1, cystic intracerebral lesion without contrast enhancement and ventricular deformity; **c** sagittal post-contrast T1, cystic intracerebral lesion without contrast enhancement and ventricular and corpus callosum mass effect and deformity. Postoperative MRI: **d**, **e** axial T2, clear reduction of the cystic lesion without mass-effect and **f** coronal FLAIR, clear reduction of the cystic lesion without ventricular deformity

Histology revealed a thin fibrous cyst wall composed of pseudostratified ciliated columnar epithelium, a typical respiratory epithelium. There were no signs of cytologic atypia. The immunohistochemistry showed positivity for cytokeratin 7 (CK7). The collected mucinous deposit was composed of devitalized cells. The diagnosis of NEC was done (Fig. 3).

At follow-up of 3 months, the patient is fine with no symptomatic or radiological recurrence (Fig. 1). He was gradually tapered of antiepileptic drugs.

Discussion

Since the first description of the NEC in 1934 by Puusepp [22], even today, there are still some controversial aspects regarding these rare congenital malformations and their origin. To date, several theories about with their pathogenesis and location along the CNS (extracranial more than intracranial, infratentorial more than supratentorial, and medial more than lateral lesions) have been proposed but they all remain to be

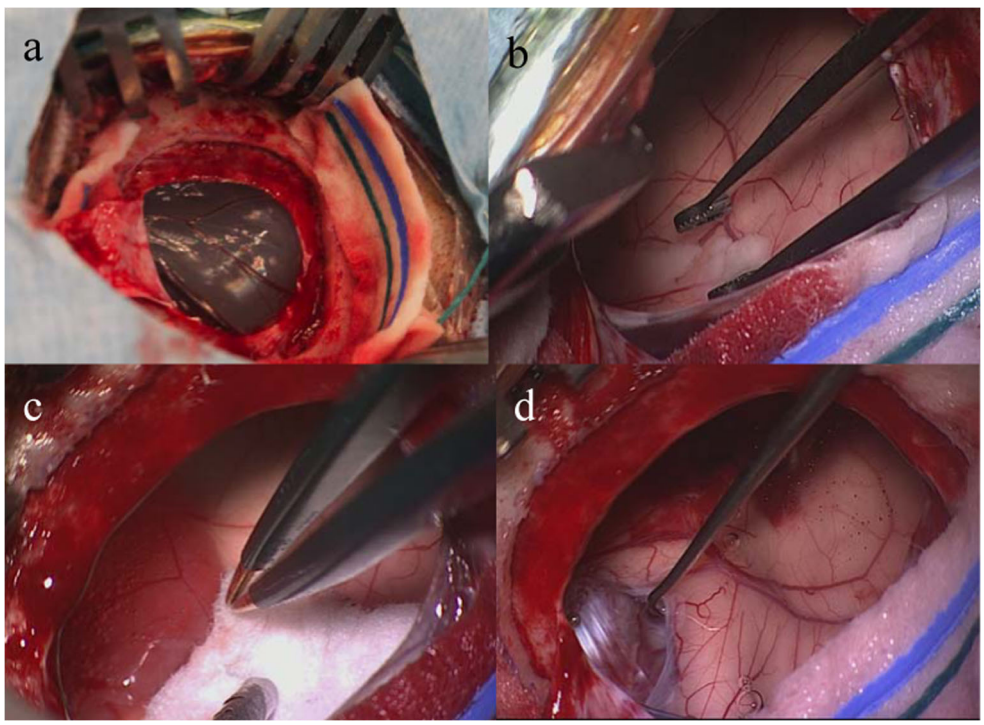

Fig. 2 Surgical images. **a** Cyst wall after dura mater opened. **b** Mucinous deposit inside the cyst. **c** Arachnoid membrane which was fenestrated **d** for comunication with the interhemisferic fissure space

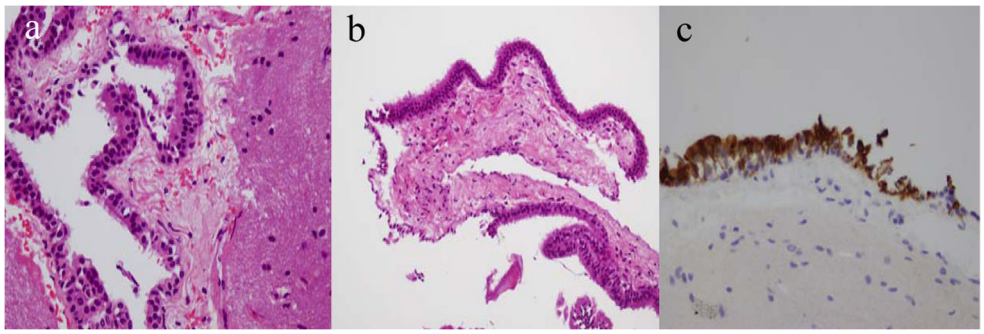

Fig. 3 Histology analysis: **a**–**b** hematoxylin-eosin stain showing a ciliated pseudostratified epithelium. Immunohistochemistry analysis: **c** cyst wall cells showing positivity for CK7

validated. Some authors suggested a failure of separation between the notochord and the foregut with the incorporation of primitive endodermal cells in the notochord. The "Seessel's pouch origin" hypothesis supported a common embryological origin for medial NEC, Rathke's cleft and colloid cysts [11, 18]. Mittal et al proposed an anomalous endodermal cell migration through the primitive neuroenteric canal into the ectoderm [18], fact that explains the existence of non-midline supratentorial NEC.

Histologically, NEC are lined by a mucin-secreting cuboidal or columnar epithelium, with or without cilia, similar to the gastrointestinal, respiratory epithelium or both of them, descending in order of frequency [18]. In 1976, Wilkins and Odom proposed a histological classification for spinal NEC based on cyst's wall histological features. Type A NEC are composed of a single layer of epithelium lining on a basement membrane and a layer of vascular connective tissue, type B NEC contain mesodermal elements such as mucous or serous glands, smooth muscle, fat, cartilage, bone, lymphoid tissue, or exceptionally nerve, and type C cysts exhibit glial or ependyma tissue [28].

Supratentorial NEC most frequently present with neurological deficits from local mass effect, signs, or symptoms of intracranial hypertension or with an inaugural seizure. Ongoing mucin secretion can lead to cyst growth and symptomatic worsening. Resorption or leakage may lead to a fluctuating clinical picture.

After a MEDLINE/Pubmed search for key words as "supratentorial," "neuroenteric," or "neurenteric," "endodermal," "enterogenous," and "cyst," with or without "child/children," we found that intracranial cases represent 25% of all NEC [24] and they have been mostly diagnosed in adult patients, involving the ventral posterior fossa [26]. There are only two reviews of NEC in children [1, 7] with few intracranial cases. Supratentorial reviews also include a scarce number of children patients [2, 4, 10, 16]. Table 1 summarizes the cases of supratentorial NECs previously reported in children. The most common position seems to be parasagittal (36.4%), and then other medial locations such as sellar/parasellar/suprasellar/cavernous sinus. Apparently, there is no sex or age predominance.

As in our case, NEC may be difficult to diagnose preoperatively because their radiological similarities with other more common pathologies such as arachnoidal cysts, neuroepithelial cysts, dermoid/epidermoid cysts, parasitic cysts, or even tumors with large cystic component such as pleomorphic xanthoastrocytoma or metastasis. On MRI, the intensity patterns of this well-demarcated lesion are highly

Table 1 Cases of supratentorial neuroenteric cysts in children

Authors and year	Age	Sex	Location
Oliveira et al. [7]	2 years	F	Cavernous sinus
Chen et al. [5]	Neonate (38 w)	M	Sellar, parasellar, right anterior and middle fossa
Pulido-Rivas et al. [21]	Neonate (38 w)	M	Left parasagital posterior
Pulido-Rivas et al. [21]	Premature (34 w)	M	Right parasagital posterior
Breshears et al. [3]	10 years	F	Third ventricle
Chen et al. [6]	6 years	M	Left parietotemporal
Chen et al. [6]	5 days	M	Right frontotemporal base
MacMahon et al. [17]	3 years	F	Right parasagital
Noureldeian et al. [20]	14 years	F	Left hemisphere
Kim et al. [15]	3 years	F	Suprasellar, left middle fossa
Our case, 2019	14 years	M	Right parasagital frontal

depending on protein content or presence of hemorrhage; however, they regularly show hypointensity on T1-weighted signal and hyperintensity on T2-weighted signal/FLAIR sequences, without surrounding edema and mild restriction on DWI images. Contrast-enhancement is rare on both MRI and CT [9], and bone remodeling can be rarely seen.

Surgery is thought to be the treatment of choice for NEC with the goal of gross-total resection. Incomplete resection or cyst fenestration has been associated with higher probability of cyst recurrence and seeding into the arachnoid space. Other surgical risks are fluid leakage, aseptic meningitis, and hydrocephalus. Age < 30 years and cyst's size > 30 ml have been associated with higher risk of recurrence, especially during the first 2 years [6]. Tumor coexistence [19] and malignant transformation [8, 12, 25, 27] were also reported.

Conclusions

Despite the rarity of this entity, it should be considered in the differential diagnosis of supratentorial cyst lesions in children. Although MRI remains the diagnostic modality of choice, the radiological findings may be similar in both arachnoid cysts and NEC. However, unlike arachnoid cysts, surgery should include the complete excision of the cyst wall.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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