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Case report

Supratentorial neurenteric cyst—A case report

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ARTICLE INFO

Article history:
Received 24 February 2014
Accepted 4 April 2014
Available online 18 April 2014

Keyword:
Supratentorial neurenteric cyst
Cystic lesion of the central nervous system
Wilkins–Odom classification

ABSTRACT

Supratentorial neurenteric cyst is a rare congenital lesion. We report here a case of a 33-year-old female who presented with seizures. A multicystic lesion in the left premotor cortex with moderate contrast enhancement was demonstrated with MRI. Microscopically, the lesion showed small cystic structures filled with a proteinaceous fluid. The wall of the cysts was lined with a single layer of ciliated columnar or cuboidal epithelium on a basement membrane. Glandular structures resembling gastrointestinal glands were also present. The cells of the cyst lining and glandular structures revealed strongly positive immunoreactions for epithelial markers (cytokeratin and epithelial membrane antigen).

1. Introduction

Neurenteric cyst, also known as endodermal, enterogenous or epithelial cyst, is a rare developmental lesion found mostly in the ventral cervicothoracic regions of the spine where it appears as an intradural extramedullary lesion [1]. It is commonly associated with congenital defects of the overlying skin and/or vertebral bodies. Intracranial localization is very uncommon and, when present, the cyst occupies the fourth ventricle or cerebellopontine angle regions. Supratentorial location of a neurenteric cyst is exceptionally rare. We reviewed the literature and found 24 cases of a supratentorial neurenteric cyst reported so far [1,2].

The clinical presentation of a neurenteric cyst results from compression or irritation of the surrounding neural structures [3]. Patients present most often with sensory and motor deficits, partial and generalized seizures and signs of raised intracranial pressure [1]. Facial spasm was a major clinical feature in four of the reported cases [4,5]. On magnetic resonance imaging (MRI) neurenteric cysts are well-demarcated usually non-contrast-enhancing lesions. Computed tomography (CT) may demonstrate calcifications within the cyst wall [6].

2. Case report

2.1. *Clinical presentation*

A 33-year-old female experienced three partial and one secondary generalized motor seizures. MRI demonstrated a multicystic lesion of approximately 35 mm in the longest dimension in the left premotor cortex with moderate contrast enhancement and no other pathologies (Fig. 1A). CT angiography of the brain revealed calcifications and excluded the diagnosis of a hemangioma.

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http://dx.doi.org/10.1016/j.pjnns.2014.04.001

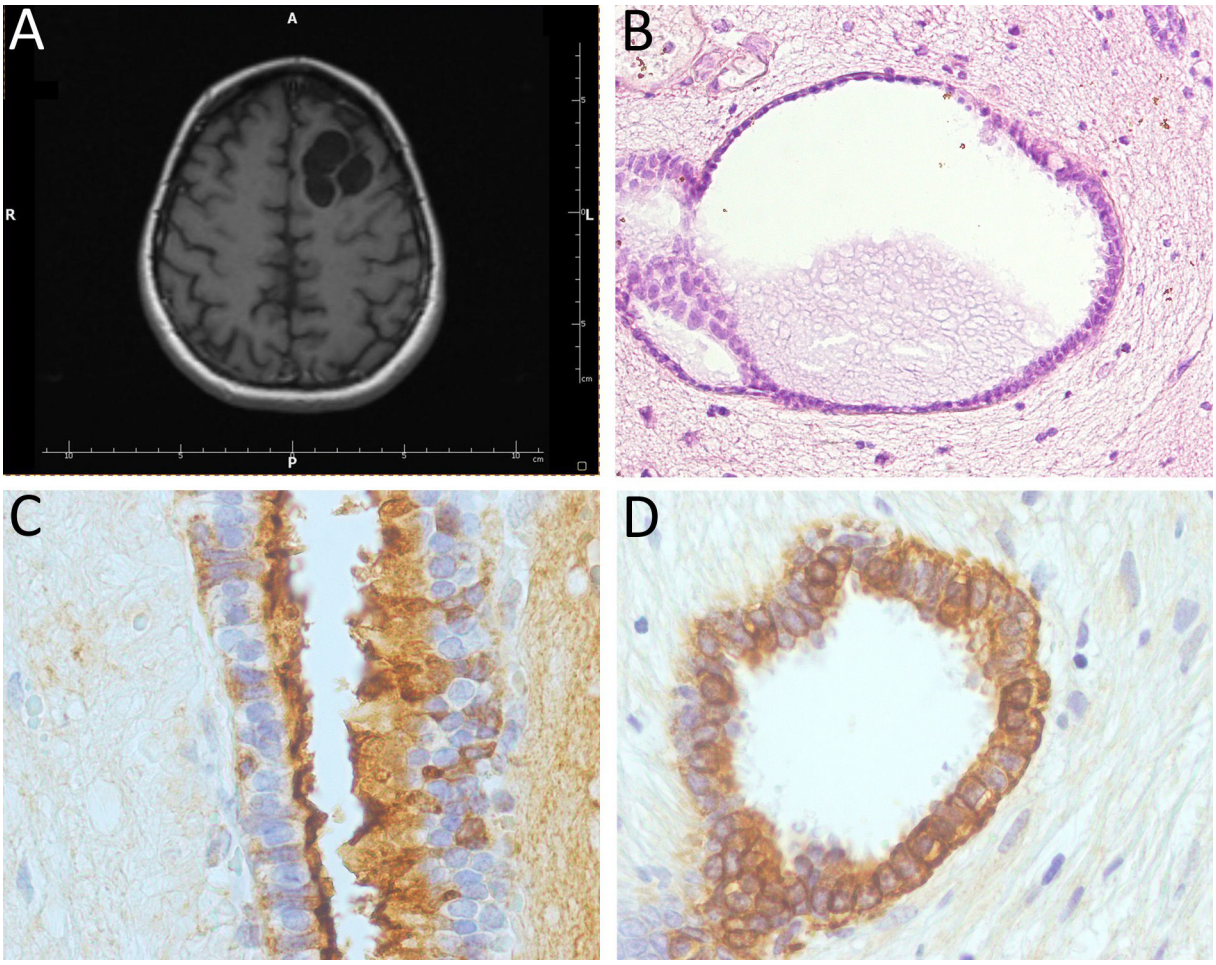

Fig. 1 – A multicystic lesion in the left premotor cortex on MRI (A). Cystic spaces filled with a proteinaceous fluid, lined with a single layer of epithelium on a basement membrane (B). H&E staining. Strong EMA (C) and cytokeratin (D) expression by cells of the cyst lining. Peroxidase immunohistochemistry. B and C original magnification 200× and D 400×.

The patient underwent a left frontal parasagittal craniotomy. Intraoperatively, a multicystic lesion macroscopically resembling thickened arachnoid was revealed and subtotally resected. The cerebral cortex beneath the lesion was compressed.

The postoperative course was unremarkable with no further seizure episodes.

3. Results

Microscopic examination of the postoperative material revealed multiple cystic spaces lined by a single layer of ciliated columnar or cuboidal epithelium mounted on a basement membrane (Fig. 1B). The cysts were filled with a proteinaceous fluid. In addition, glandular structures resembling gastrointestinal glands were present but no goblet cells were identified. There was no evidence of cellular atypia in the epithelial cells. In the surrounding brain reactive astrocytes and small foci of calcification were demonstrated. The immunohistochemical examination showed that the cells of the cyst lining and glandular structures were strongly positive for epithelial markers such as epithelial membrane antigen (EMA) and cytokeratin (Fig. 1C and D), but negative for carcinoembryonic antigen (CEA). There was no staining with a glial marker glial fibrillary antigen (GFAP) except for reactive astrocytes. The expression of Ki-67 antigen was observed in 1% of cells.

These morphological features and immunophenotype are consistent with a diagnosis of a neurenteric cyst. Glioependymal elements, mucous glands, serous glands, smooth muscle, connective tissue components, lymphoid tissue or ganglia were absent, indicating a type A neurenteric cyst according to Wilkins–Odom classification [7].

4. Discussion

The exact etiology of neurenteric cysts remains incompletely understood. Numerous hypotheses were proposed to explain the embryopathogenesis of these lesions. Most of the postulated mechanisms provide an explanation for the development of neurenteric cysts located in the spine and the posterior fossa but fail to explain the origin of neurenteric

cysts found supratentorially [1,8]. Recently, supratentorial neurenteric cysts were postulated by Mittal et al. to be a result of a dorsal migration of endodermal cells through the primitive neurenteric canal into the ectoderm, in the developing neuroectoderm near the primitive pit [1]. This hypothesis is supported by the relatively most frequent incidence of intraspinal cysts, while lower incidence of posterior fossa and especially supratentorial neurenteric cysts.

The neuroradiological imaging modality of choice in the diagnosis of a neurenteric cyst is MRI but the signal intensities are variable depending on the protein content of the cyst. Calcifications in the wall of a supratentorial neurenteric cyst may be encountered [6].

Wilkins and Odom divided neurenteric cysts into three types depending on histopathological features [7]. Type A cysts are lined by a single-layered or pseudostratified, cuboidal or columnar, ciliated or non-ciliated epithelium mounted on a basement membrane. Presence of mucous or serous glands, smooth muscle, connective tissue elements, lymphoid tissue and ganglia is characteristic for type B endodermal cysts. In type C endodermal cysts glioependymal elements are identified.

Immunohistochemically, the neurenteric cysts are positive for cytokeratins, EMA and usually for CEA. They are negative for markers such as GFAP and S-100 protein. However, there is no specific marker for neurenteric cysts since the immunoexpression of CEA, ferritin and cytokeratins is not a universal finding in all lesions of this type.

The differential diagnosis of a supratentorial neurenteric cyst includes any well-demarcated cystic lesions such as a meningeal arachnoid cyst, true neuroepithelial cyst, ependymal cysts, teratomatous cysts and metastatic carcinomas.

The difference in signal intensity on imaging between meningeal arachnoid cysts and neurenteric cysts is helpful in distinguishing those lesions preoperatively (the meningeal arachnoid cysts are isodense with cerebrospinal fluid on CT) [6]. The histopathological and immunohistochemical findings in cases of Rathke's cleft cysts, colloid cysts and neurenteric cysts are similar. Therefore, the location is important for the differential diagnosis. Histologically, the arachnoid cysts are lined by meningothelial cells.

Presence of a basement membrane, ciliated cells and structures resembling gastrointestinal glands differentiates neurenteric cysts from true neuroepithelial cysts. The positive immunohistochemical staining for cytokeratin and negative staining for neuronal and glial markers (e.g. neuronspecific enolase, synaptophysin, GFAP and S-100 protein) favor the diagnosis of a neurenteric rather than a neuroepithelial cyst.

In contrast to neurenteric cysts, the ependymal cysts lack basement membrane, the cells express GFAP and are negative for cytokeratin. The absence of tissues such as muscle or nervous system elements helps to exclude the diagnosis of a teratoma.

Metastatic carcinomas should also be included in the differential diagnosis of neurenteric cysts. Apart from clinical findings, low mitotic activity, benign appearance of the epithelial component, only a slight gliosis and lack of brain edema in the adjacent tissue help in distinguishing neurenteric cyst from a metastatic tumor.

Proliferation index was only rarely measured in the reported cases of intracranial neurenteric cysts [9–14]. In a case described by Sahara et al. the recurrence of neureneteric cysts after total resection was associated with an increase of MIB-1 positive cells from 0% in the first surgical specimen to 6.7% in the second [11].

In our case described here the percentage of cells positive for Ki-67 antigen was low (1%).

The recommended clinical approach in symptomatic patients with neurenteric cyst is a complete resection [15]. The risk of recurrence in cases when biopsy or incomplete resection is performed is high [16,17]. Perry et al. documented a craniospinal dissemination of a neurenteric cyst of the cerebellum after an incomplete resection in a 63-year-old female [9]. It is worth to stress that none of the reported supratentorial neurenteric cysts have recurred.

Summarizing supratentorial neurenteric cyst is a rare entity that should however be considered in adult patients with supratentorial non-contrast-enhancing or only moderately-contrast-enhancing cystic lesion.

Conflict of interest

None declared.

Acknowledgement and financial support

None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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