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An epileptogenic intracranial cystic lesion lined with fallopian tube-type epithelium: illustrative case

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BACKGROUND Intracranial cystic lesions are often a trigger for epileptic seizures. However, there has never been a report of a cystic lesion lined with fallopian tube-type epithelium.

OBSERVATIONS A 48-year-old female presented with a cystic lesion in the right occipital lobe, which gradually grew over 8 years. Right occipital lobe epilepsy was diagnosed based on visual aura, convulsive seizures, and electroencephalogram findings and the cyst was surgically removed. Further examination revealed the cyst was lined with ciliated cells, which had morphological and immunohistochemical features similar to those of fallopian tube epithelium.

LESSONS The characteristics of the cyst did not conform to any known types of benign cystic lesion. To the authors' knowledge, no such cyst has been reported before. The authors discuss the origins and pathogenesis of this unfamiliar cystic lesion.

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KEYWORDS epileptogenic; intracranial cystic lesion; fallopian tube-type epithelium; ciliated epithelium

Intracranial cystic lesions are often a trigger for epileptic seizures. However, intracranial lesions, especially cystic lesions, rarely grow rapidly and often have no clear imaging effects on the brain, especially in benign cystic lesions.[1–3] Therefore, these types of lesions are seldom suspected, treated, and examined in detail using immunohistochemical methods. Here, we present a case of an epileptogenic intracranial cystic lesion lined with ciliated cells that showed the characteristics of fallopian tube-type epithelium. This is an unfamiliar histopathological feature that, to our knowledge, has not been reported before.

Illustrative Case

A 48-year-old female presented to a regional hospital with a primary complaint of headaches. She had given birth to a child and had no other medical history. A cystic lesion in the right occipital lobe was noted at the time. The cyst gradually grew over the course of 8 years and the patient developed seizures, which became frequent. As a result, the patient was admitted to our hospital for examination and treatment. No neurological deficits were observed on admission.

The patient's convulsive seizures were characterized by a rainbow-colored wispy visual disturbance (visual aura) followed by generalized tonic-clonic convulsions. The interictal electroencephalogram showed a focal epileptiform discharge in the right parieto-occipital region. Computed tomography revealed a low-resolution cystic lesion in the right occipital lobe (Fig. 1A). T1-weighted magnetic resonance imaging (MRI) showed a low-intensity cystic lesion, and T1-weighted gadolinium-enhanced MRI showed no enhancement in the lesion or the surrounding tissue (Fig. 1B). There was no hyperintensity area, such as perifocal edema, on T2-weighted imaging (Fig. 1C). Eight years later, the cystic lesion was clearly enlarged (Fig. 1D). On the basis of these findings, we diagnosed symptomatic epilepsy caused by arachnoid cysts. The lesion was excised under general anesthesia after craniotomy. The shiny cystic lesion was almost completely removed, including

ABBREVIATIONS CEA = carcinoembryonic antigen; MRI = magnetic resonance imaging; PAX8 = paired-box gene 8; WT1 = Wilms tumor 1.

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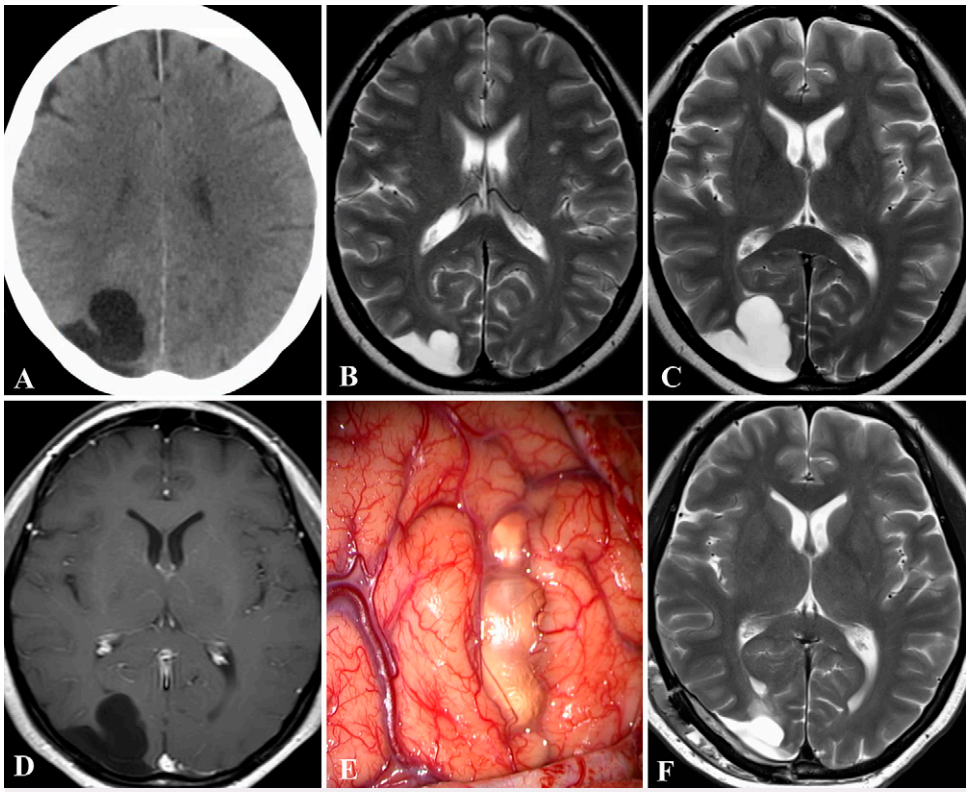

FIG. 1. Computed tomography image showed a low-resolution cystic lesion in the right occipital lobe (**A**). Initial T2-weighted MRI showed a hyperintensity area (**B**). The lesion grew over the course of 8 years (**C**). T1-weighted gadolinium-enhanced MRI showed no enhancement in the lesion or the surrounding tissue (**D**). An intraoperative photograph showed the cyst covered by a shiny membrane containing a clear, colorless fluid (**E**). Postoperative MRI showed that the cyst with high intensity on T2-weighted images had shrunk (**F**).

the cyst wall observed during surgery (Fig. 1E). Postoperative MRI showed that the cyst had disappeared, and the morphological changes in the brain caused by the lesion were reduced (Fig. 1F). The patient was discharged 10 days postoperatively, without neurological deficits. One year after the surgery, the patient was seizure-free and there was no recurrence of the cystic lesion.

Pathological Findings

The cyst wall consisted of thin fibroconnective tissue with focal calcifications (Fig. 2A). Approximately 50–60% of the cyst was lined with a single layer of bland cuboidal or columnar cells with occasional ciliated cells. The remaining cells were flattened or nondescript (Fig. 2B). The former was composed of a fallopian tube-type epithelium (comprising three types of cells; Fig. 2C). Pseudostratified ciliated and goblet cells mimicking the bronchial epithelium, observed in endodermal and colloid cysts, were not identified. Elements imitating the mucosa and muscularis mucosa of the gastrointestinal tract, which are characteristic of endodermal cysts, were also absent. Unlike the walls found in ependymal cysts, the wall of this cyst did not contain glial tissue.

Immunohistochemically, all cells from the lining of the cyst were strongly positive for cytokeratins AE1/AE3 and CAM5.2 (Fig. 3B–F). Both the fallopian tube-type and flattened epithelia were partly positive for epithelial membrane antigen and focally positive for carcinoembryonic antigen (CEA) (Fig. 3D–M). Wilms tumor 1 (WT1), estrogen receptor, and paired-box gene 8 (PAX8; a sensitive and specific marker for tissues of Müllerian origin) tests were predominantly positive for fallopian tube-type epithelium and partly positive for flattened epithelium (Fig. 3F–P).

The final pathological diagnosis was benign epithelial-lined cyst with ciliated epithelium of fallopian tube-type. The diagnosis was descriptive as the characteristics of the cyst did not conform to any of the established characteristics of benign cystic and hybrid lesions.

Discussion

We report an intracranial cystic lesion lined with fallopian tube-type epithelium, which caused epilepsy in the concerned patient. Generally, intracranial cysts are ectodermal (dermoid or pemphigoid cysts), neuroectodermal (arachnoid or choroid plexus cysts), or endodermal (neurenteric, colloid or ependymal cysts); cystic lesions with ciliated cells are mainly endodermal cysts. The definitive diagnosis should be based on anatomical location and the histopathological findings of light microscopy, immunohistochemistry, and electron microscopy.[1–3]

Observations

In the case we report here, approximately half of the cells lining the cyst were similar in appearance to fallopian tube-type epithelium. Moreover, many of these cells were immunohistochemically positive for the WT1, estrogen receptor, and PAX8 tests, similar to

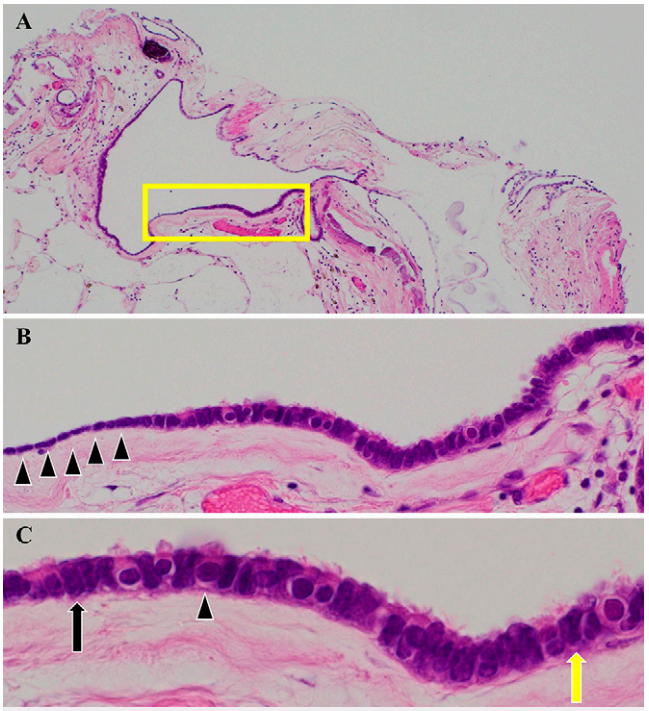

FIG. 2. Microscopic appearance of the cyst (**A**) lined with cuboidal or columnar cells with occasional ciliated cells, and flattened cells (*black arrowheads*, **B**). Yellow square–highlighted region (B) from panel A. Note the fallopian tube-type epithelium: ciliated cells (*black arrowhead*), nonciliated secretory cells (*black arrow*), and so-called peg cells (*yellow arrow*, **C**).

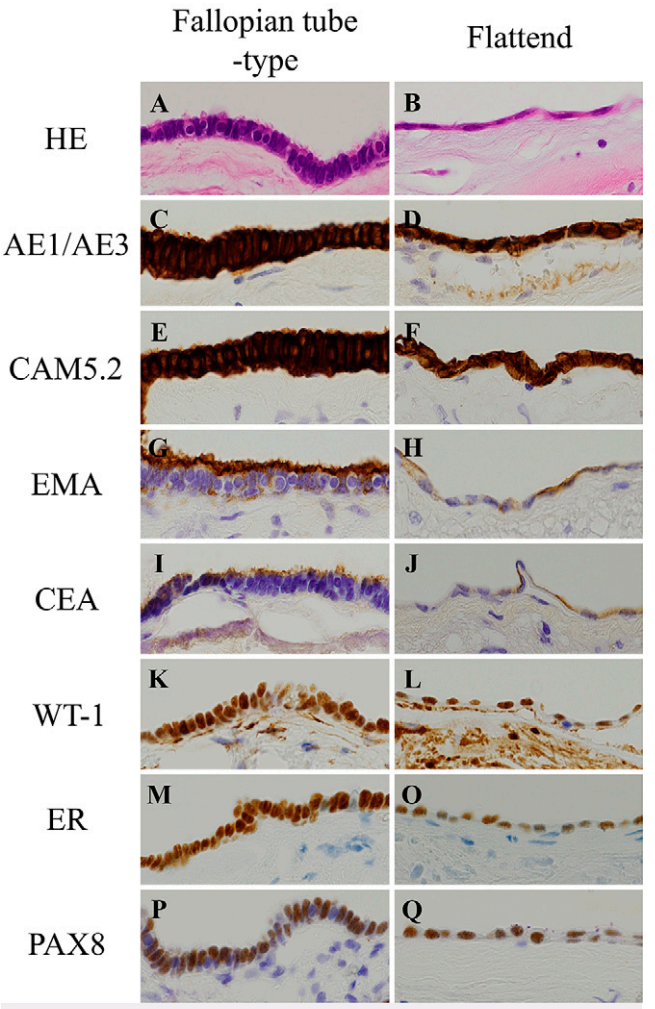

FIG. 3. The immunohistochemical profile of the cystic lesion. Both fallopian tube-type (**A**) and flattened (**B**) epithelia were positive for cytokeratins AE1/AE3 and CAM5.2, epithelial membrane antigen, and CEA.

fallopian tube epithelium. The literature shows no reports of such a cystic lesion, and we discuss the possibility of the following pathogenesis.

First, we considered the possibility of variants of endodermal cysts with marked tubal metaplasia that accompanied metaplastic changes in the fallopian tube-type epithelium. The cystic lesion reported here is consistent with an endodermal cyst as it tested positive for both cytokeratin and CEA; however, the cells that lined the cyst had features of Müllerian epithelium, which is of mesodermal, rather than endodermal, origin. Because of the difference in the presumed origins of the mesoderm and endoderm, it is highly unlikely that tubal metaplasia could occur in endodermal cysts.

Second, we considered the possibility that components derived from mature teratomas could differentiate into triple germ layers. However, this is unlikely because imaging and histological findings revealed no evidence of teratomas.

Third, we considered the possibility of endosalpingiosis—a tumor-like lesion of Müllerian duct origin—that is defined as the presence of glands lined with benign fallopian tube-type epithelium outside the fallopian tube. Endosalpingiosis is usually observed in the abdominal cavity, is rarely observed beyond the diaphragm, and has not been reported in the intracranial space.[4] Although cyst-lining cells are not found in endosalpingiosis, some cyst-lining cells were found in the cyst in our case. Furthermore, the immunohistochemical features of the cyst showed partly positive WT1, estrogen receptor, and PAX8 findings for flattened cells: in endosalpingiosis, most of the epithelial cells are positive for these markers, even if the epithelium is flattened. For these reasons, there was a small possibility of this being a case of intracranial endosalpingiosis.

Endometriosis occasionally presents with tubal metaplasia.[4] However, intracranial endometriosis is extremely rare,[5–7] and endometrial stroma was not identified in this case. Therefore, intracranial endometriosis was excluded.

Lessons

In summary, we report a case of an epileptogenic intracranial cystic lesion with fallopian tube-type ciliated epithelium, which was possibly derived from Müllerian-type or mesodermal epithelium. Because the fallopian tube is not routinely examined for intracranial cystic lesions, we suspect that similar cases might have occurred and that they may have been overlooked. We believe that further studies will elucidate the incidence of intracranial cystic lesions with ciliated epithelium that resembles that of the fallopian tube. A case series study is warranted to clarify whether the histopathological features of intracranial cysts cause differences in

recurrence rate and prognosis. These data would be important for determining treatment strategies and patient management.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Takemoto, Dekita, Ozono, Hamasaki, Tsubota, Mukasa. Acquisition of data: Takemoto, Dekita, Ozono, Mikami, Kuroda, Mukasa. Analysis and interpretation of data: Takemoto, Dekita, Ozono, Yamada, Mikami, Mukasa. Drafting the article: Takemoto, Dekita, Ozono, Mikami. Critically revising the article: Takemoto, Dekita, Ozono, Hamasaki, Mukasa. Reviewed submitted version of manuscript: Takemoto, Mukasa. Approved the final version of the manuscript on behalf of all authors: Takemoto. Statistical analysis: Takemoto. Administrative/technical/material support: Takemoto, Hamasaki, Kuroda. Study supervision: Takemoto, Hamasaki, Mukasa.

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