Giant glioependymal cyst in an infant

Case report

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The authors report the case of an infant with a giant glioeependymal cyst. Although it has been suggested that these cysts originate from the tela choroidea, their origin remains controversial. This 35-month-old girl with truncal ataxia was referred to the authors’ hospital. Magnetic resonance imaging revealed a giant cystic mass extending from the anterior to the posterior cranial fossa. Hydrocephalus was caused by obstruction of the sylvian aqueduct. Endoscopic fenestration of the cyst wall was performed. Histochemical and immunohistochemical staining identified the lesion as a glioeependymal cyst. Magnetic resonance imaging performed 8 months later suggested that the cyst originated from the tela choroidea. At 5-year follow-up, there was no tumor recurrence and she had fully recovered. The origin of glioeependymal cysts is discussed, and the authors suggest that their origin is the tela choroidea. (DOI: 10.3171/2010.11.PEDS10270)

KEY WORDS • giant glioeependymal cyst • etiology • hydrocephalus • tela choroidea

Glioeependymal cysts are rare; they arise in para-ventricular or subarachnoid spaces2,5,6,9 and their walls present a heterogeneous histological appearance.2,3,7 Although the origin of glioeependymal cysts remains controversial, their histological variety supports the hypothesis that they originate from the tela choroidea.2,3,7 In the case presented in this paper, MR imaging indicated that the origin of our patient’s cyst was the tela choroidea.

Case Report

History and Examination. This 35-month-old girl was referred to our hospital for further examination of her gait impairment. Progressive gait disturbance was noticed 7 months earlier. Her mother’s pregnancy and delivery were uneventful, and there was no history of infections or CNS injuries. The child’s head circumference was slightly increased. Neurological examination disclosed mild truncal ataxia. There was no papilledema present. Magnetic resonance imaging demonstrated a well-defined homogeneous cystic mass located outside the brain parenchyma. The mass extended from the anterior and middle cranial fossa through the velum interpositum and into the ambient and supracerebellar cistern above a hypoplastic cerebellum. The cyst laterally compressed the trigone of the left lateral ventricle. The lesion was isointense with the CSF on both T1- and T2-weighted images. Magnetic resonance imaging also revealed obstruction of the sylvian aqueduct with hydrocephalus, compression of the cerebral and cerebellar cortices, and elevation of the corpus callosum (Fig. 1A–D).

Operation and Postoperative Course. The endoscope was introduced through a frontal bur hole into the left anterior horn of the lateral ventricle, and fenestration of the semitranslucent, elastic, soft cyst wall was performed. The cyst was filled with clear, colorless fluid. After cyst shrinkage was obtained, the third ventricle was entered via the dilated foramen of Monro, and opening of the sylvian aqueduct was confirmed. Inspection of the cyst cavity revealed that the communication in the cavity was preserved and that there was no communication with either the subarachnoid space or the ventricular system. The bilateral tentorium cerebelli were markedly deformed and displaced laterally; the cerebellum was symmetrically displaced downward. Additional fenestrations were placed at the right lateral ventricle. The continuity of the cyst cavity with the subarachnoid space and the ventricular system was confirmed by postoperative ventriculocisternography, which was performed using an Ommaya reservoir placed after the endoscopic procedure.

Abbreviations used in this paper: EMA = epithelial membrane antigen; GFAP = glial fibrillary acidic protein.
Pathological examination revealed that the cyst wall was partly composed of inner cuboidal ependymal cells with cilia resting directly on glial and outer connective tissue (Fig. 2A). The gliotic parenchyma was positive for GFAP and S100 protein but negative for EMA (Fig. 2B–E). The ependymal cells were positive for GFAP; faint, partial membrane immunoreactivity for EMA was confined to the luminal surface (Fig. 2D and E). These findings suggested that the cyst was of ectodermal origin; it was diagnosed as a glioependymal cyst.

The patient’s truncal ataxia gradually improved and postoperatively it subsided completely. An MR imaging study performed 8 months after the surgical procedure demonstrated a gradual collapse of the cyst and resolution of hydrocephalus (Fig. 1E–H). These images also showed that the cyst had continuity with the left septum pellucidum, internal cerebral veins, and the choroid plexus of the left lateral ventricle (Fig. 1H). The site of greatest expansion was between the left internal cerebral vein and the choroid plexus of the left lateral ventricle (the choroidal fissure). At the 5-year follow-up the patient showed no neurological deficits, mental retardation, or cyst recurrence.

**Discussion**

**Classification of Benign Intracranial Cysts**

There has been some confusion with respect to the naming and classification of benign intracranial cysts. There has been some confusion with respect to the naming and classification of benign intracranial cysts.2,5,7

**Fig. 1.** Preoperative (A–D) and postoperative (E–H) MR imaging obtained in the patient. A–D: Axial (A and B), sagittal (C), and coronal (D) images demonstrate a well-defined T2-hyperintense (A, B, and D) and T1-hypointense (C) homogeneous giant cystic mass lesion located outside the brain parenchyma from the posterior to the middle cranial fossa. E–H: Axial (E and F), sagittal (G), and coronal (H) T2- (E and F) and T1-weighted images (G and H) show cyst expansion between the internal cerebral vein and choroid plexus of the lateral ventricle. Note the internal cerebral veins (curved arrows), choroidal artery (arrows), and choroid plexus of the lateral ventricle (arrowhead).

**Fig. 2.** Photomicrographs showing the histological and immunohistological staining of the cyst wall. A: Staining with H & E demonstrated that the cyst wall was composed of 3 layers consisting of inner cuboidal ependymal cells (lower layer), glial tissue (center layer), and outer connective tissue (upper layer). B: Ependymal cells and glial tissue stained positively for GFAP. C: Glial tissue was stained positively for S100 protein. D: The GFAP-positive ependymal cells with cilia rested directly on glial tissue. E: Note the faint immunoreactivity of portions of the ependymal cell membrane, which was confined to the luminal surface after staining with EMA. Bar = 100 μm (A), 50 μm (B and C), and 20 μm (D and E).
Giant glioependymal cyst

TABLE 1: Proposed classification for intracranial epithelial cysts of ectodermal origin

<table>
<thead>
<tr>
<th>Epithelial Cyst (Epithelium-Lined Cyst) of Ectodermal Origin</th>
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<tr>
<td>A. neuroepithelial cyst</td>
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<tr>
<td>1. glioependymal cyst (neuroglial cyst)</td>
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<tr>
<td>2. ependymal cyst</td>
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<tr>
<td>B. choroidal epithelial cyst</td>
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<tr>
<td>C. dermal cyst</td>
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<tr>
<td>1. epidermoid cyst</td>
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<td>2. dermoid cyst</td>
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Epithelial cysts are synonymous with epithelium-lined cysts and are classified as having ectodermal and endodermal origin. There exists no widely approved and used classification with respect to the cysts of ectodermal origin. We suggest the classification system represented in Table 1. Among cysts of ectodermal origin, those with ependymal lining are called neuroepithelial cysts and derive from primitive neuroectoderm. Neuroepithelial cysts include ependymal cysts and glioependymal/neuroglial cysts. Ependymal cysts are primarily intraaxial and their walls consist of an inner single layer of ependymal lining resting on connective tissue; the outer brain parenchyma is normal.5,9,11 Glioependymal/neuroglial cysts are located in the paraventricular or subarachnoid space. Their wall consists of 3 layers and a single epithelium directly abuts the inner glial layer, which is not continuous with normal brain parenchyma and outer connective tissue.7,9

Origin of Glioependymal Cysts

The origin of glioependymal cysts is controversial. Previously suggested etiological factors include invagination or evagination of the neuroepithelial lining of the ventricular system in fetal life.8 Blake pouch persistence,4 and cystic generation of subarachnoid neuroglial heterotopia.1

Both the ependyma and choroid plexus develop from the primitive neuroectoderm. At 5 to 6 weeks of gestation, the vascular pia mater invaginates into a focally attenuated neuroepithelium, forming tufts of primitive choroid plexus.2 According to Shuangshoti and Netsky,8 ependymal cysts originate from primitive neuroepithelium lining the neural tube and can develop in any part of the ventricular system by either invagination or evagination, with a connective tissue layer inside or outside the neuroepithelial layer. Friede and Yaşargil3 postulated that glioependymal cysts originate from segments of the neural tube equivalent to the tela choroidea and that this accounts for the variance in their wall structure. In the tela choroidea, there is a transition between ciliated ependyma on glial tissue (G) and nonciliated epithelium on connective tissue (C). A basement membrane exists where the ependyma is adjacent to vascular structures such as the choroid plexus (CP). Glioependymal cysts may be lined by ciliated or nonciliated epithelium on glia without a basement membrane, or they may be located on connective tissue and manifest a basement membrane.3

Histologically, glioependymal cysts may be lined with a ciliated or nonciliated epithelium located either on glia without a basement membrane or on connective tissue with a basement membrane (Fig. 3). Glioependymal cysts arise around the tela choroidea (the velum interpositum), the ambient and supracerebellar cisterns, and the lateral ventricles. In our patient, MR imaging findings suggested that the glioependymal cyst originated from the tela choroidea by invagination. The cyst extruded through the
velum interpositum and extended as it grew posterior to the supracerebellar cistern via the choroidal fissure into the left lateral ventricle and anterior to the frontal horn of the left lateral ventricle. We believe that the cyst crossed to the right frontal horn and extended to the body of the right lateral ventricle through the septum pellucidum.

The Blake pouch is an ependyma-lined diverticulum that is formed beginning 7 to 8 weeks up to the 4th month of gestation at a time when the cerebellum is rudimentary. The formation takes place by posterior ballooning of the posterior membranous area of the medullary velum into the cisterna magna.4 Blake pouch persistence is probably due to evagination of the rudimentary tela choroidea of the fourth ventricle.10 The same hypothesis may also apply to supratentorial glioependymal cysts evaginating at the tela choroidea and the formation of glioependymal cysts around the tela choroidea of the fourth ventricle (the cerebellomedullary fissure); the cisterna magna and the fourth ventricle can be explained as Blake pouch cysts that have “pinched outward.”

Neuroglial heterotopia occurs at 7 weeks of gestation.9 Islands of heterotopic ependymal cells segregate during embryonal development; they are usually found adjacent to corners of the ventricle or in zones with considerable tissue activity during the formation of the choroid plexus or the primitive cerebral fissure.6 While this concept suggests a possible relationship with ependymal cysts, it fails to explain the histological wall variance of glioependymal cysts.

Disclosure

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 to the study and manuscript preparation include the following. Conception and design: Morigaki. Acquisition of data: Morigaki, Shinno, Pooh. Analysis and interpretation of data: all authors. Drafting the article: Morigaki. Critically revising the article: Shinno, Pooh, Nakagawa. Reviewed final version of the manuscript and approved it for submission: all authors.

References


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