Occipital Lobe Ependymal Cyst
—Case Report—

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Abstract

A 51-year-old female with an ependymal cyst in the left occipital lobe presented with headache, vomiting, dizziness, and right incomplete homonymous hemianopsia. Following a cyst-ventricular communication and cyst-peritoneal shunting procedure, the visual field loss improved markedly. On the basis of the visual field symptoms, computed tomographic findings, and intraoperative observations, the cyst was considered to have developed in the vicinity of the body and posterior horn of the left lateral ventricle, extending to the left occipital lobe.

Key words: ependymal cyst, intracerebral supratentorial cyst, computed tomography, choroid plexus, occipital lobe

Introduction

Arachnoid cyst is typical of intracranial cysts, but there have been few reports of ependymal cyst in the intracerebral or subarachnoidal space.41 We describe a case of an ependymal cyst in the left occipital lobe and discuss the relevant literature.

Case Report

A 51-year-old female had experienced episodes of dizziness, headache, and vomiting for a few years. Six months prior to admission, she had developed a visual field defect in which the right half of objects was obscure. This defect gradually enlarged and the headache, vomiting, and dizziness continued. Her history included extirpation of an ovarian cyst 10 years previously.

Physical examination revealed right incomplete homonymous hemianopsia. Visual acuity was 0.8 in both eyes, and opthalmoscopic examination revealed no papilledema or marked arteriosclerotic changes. There were no abnormalities in the motor, sensory, or reflex systems, and no cerebellar signs. Computed tomography (CT) revealed a low-density area with a partial septum extending from the body and posterior horn of the left lateral ventricle to the occipital lobe, and a shift of midline structures of about 1.5 mm to the right. No ventricular dilatation was observed. After intravenous administration of 100 ml of Angiographin, CT scans showed that the low-density area was anteriorly demarcated with an enhanced area, indicating that the cyst wall might be directly attached to the body and posterior horn of the left lateral ventricle (Fig. 1). Cerebral cisternography with intrathecal administration of 10 ml of a metrizamide solution (170 mgI/ml) revealed no communication of the cerebral cistern with the low-density area. Other CT findings included compression and deformation of the left cerebral peduncle of the midbrain (Fig. 2).

Surgery was performed through the left occipital approach. Incision of the dura mater revealed no defect or atrophy of the cerebral cortex, except for swelling. When the overlying cortex was resected,
a very thin, membrane-like structure was observed
5 mm beneath it. Incision of the membrane released
a watery fluid. The internal wall of the cyst exhibited
poor vascularity. The cyst wall was broken to allow
communication with the left lateral ventricle. Next,
cyst-peritoneal shunting was carried out. Specimens
for pathological examination were collected from
the border of the cyst facing the left lateral ventricle
and a portion of the peripheral cyst wall.

Samples of the cyst fluid were collected by direct
aspiration during surgery. Examination of the fluid
revealed it to be clear and watery. The cell count was
3/mm³ (mono : poly = 1 : 2). The protein concentra-
tion, at 190 mg/dl, was elevated. The glucose content
was 3 mg/dl and that of Cl, 126 mEq/l. The tripto-
phan test was positive.

Light microscopic examination of HE-stained
tissue specimens revealed columnar epithelial cells to
constitute one layer of the cyst wall. There were
cilia-like structures on parts of the cell surfaces. No
distinct basement membrane was identified beneath
the cells, which were directly attached to glial cells.
There was no choroid plexus tissue (Fig. 3). Electron
microscopic examination revealed large, spindle-
shaped nuclei in the round cells and a particularly
dense distribution of nuclear chromatin in the
margins. The cell bodies contained few mitochondria
or glial fibrils. Intermediate junctions and desmo-
somes were present among the cells. There were
many microvilli on the cyst surfaces of cells, and
opposing surfaces were directly attached to glial
cells, without an intervening basement membrane
(Fig. 4).

Within 7 days after surgery, the right incomplete
homonymous hemianopsia had improved to the
extent that it was almost unnoticed subjectively.
At present, 6 months postoperatively, no subjective
symptoms are present (Fig. 5). Follow-up CT dis-

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Fig. 1 upper: Precontrast CT scans showing a low-density area with a partial septum ranging from the
body and posterior horn of the left lateral ventricle to the occipital lobe. lower: Postcontrast CT
scans showing the low-density area to be clearly demarcated from the left lateral ventricle (ar-
rowhead).

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closed a marked decrease in the size of the low-density area, resolution of the compression of surrounding brain tissue, and definite separation of the left lateral ventricle and cyst (Fig. 6).

Discussion

There have been only 32 reported cases, including ours, of ependymal cyst in the supratentorial space of the cranium.1,2,5,6,10,12,14,16,22,23,26-30,32-34,37,40,43) The patients included 15 males and 17 females with a mean age of 32.8 years (range, 5 days–74 years). The cyst developed on the left side in 19, on the right side in seven, in the interhemispheric fissure in two, in the bilateral cerebral hemispheres in one, and within a suprasellar lesion in three. The most common point of origin (23 cases) was the central white matter of the frontoparietal lobe.41) In contrast, there were only three patients with cysts in the occipital lobe, one of whom was our patient (Table 1). The cyst fluid was clear and/or xanthochromic in 16 patients and turbid or milky in 10. The fluid protein level varied from 11 to 580 mg/dl. It is believed that the protein content depends on the amount of choroid plexus in cyst wall cells.4) In our patient, however,
the protein content was elevated despite the fact that no choroid plexus tissue was found in the cyst wall cells.

Ependymal cyst does not communicate with the ventricle or subarachnoid space, and the initial symptoms in most cases are indicative of space-occupying lesions, e.g., increased intracranial pressure and focal symptoms, as occurred in our patient. In some cases the lesion was asymptomatic and was discovered at autopsy.7 Certain intracranial deformities, particularly agenesis of the corpus callosum, have also been reported.25,28,38,39,44

Fig. 4 Electron microscopic examination disclosed large, spindle-shaped nuclei in the round cells, few mitochondria or glial fibrils in the cell bodies, and many microvilli on the cyst surfaces of the cells. The surfaces opposite the cyst surface are directly attached to glial cells, without an intervening basement membrane.

upper: Bar = 5 \mu m, lower: Bar = 1.5 \mu m.

Fig. 5 Pre- (upper) and postoperative (lower) visual field examinations. After surgery, the right incomplete homonymous hemianopsia improved markedly and was almost unnoticed subjectively within 7 days postoperatively.

Fig. 6 Postoperative CT scans revealing marked reduction of the low-density area, absence of compression of surrounding brain tissue, and definite separation of the left lateral ventricle and cyst.
The pathogenesis of ependymal cyst is believed to involve aberrant tissues of embryonic origin. There are various theories concerning the mechanism of such aberration which can be roughly divided into two types. One is the choroid plexus theory, according to which ependymal cyst develops as a result of an aberration in the site corresponding to the tela choroidea ("primitive ependymal lining") of the neural canal wall in the cortical mantle or perimedullary mesh, particularly near the midline. The other is the endoderm theory, which holds that the cells of the cyst wall are derived from endoderm. This theory is based on the observation that, other than in the central nervous system, ependymal cell characteristics are observed only in upper respiratory tract epithelial cells. However, there have been cases in which a basement membrane was composed of epithelial cells or of choroid plexus tissue. Thus, no definite consensus has yet been established concerning the pathogenesis of ependymal cyst.

Table 1 Reported cases of supratentorial intracerebral and subarachnoid ependymal cyst

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age, Sex</th>
<th>Location</th>
<th>Cyst fluid (protein content: mg/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zehnder (1938)</td>
<td>34 yr, M</td>
<td>rt. F</td>
<td>turbid, milky</td>
</tr>
<tr>
<td>Simk &amp; Gutmann (1949)</td>
<td>35 yr, F</td>
<td>lt. F</td>
<td>—</td>
</tr>
<tr>
<td>Jakubiak et al. (1968)</td>
<td>74 yr, F</td>
<td>lt. F-P</td>
<td>clear (280)</td>
</tr>
<tr>
<td>Argyopoulos &amp; Heppner (1970)</td>
<td>49 yr, M</td>
<td>lt. F</td>
<td>clear</td>
</tr>
<tr>
<td>Patrick (1971)</td>
<td>58 yr, F</td>
<td>rt. T</td>
<td>clear</td>
</tr>
<tr>
<td>Harrison (1971)</td>
<td>4 yr, suprasellar</td>
<td>cleat</td>
<td></td>
</tr>
<tr>
<td>Argyopoulos &amp; Heppner (1970)</td>
<td>20 mo, M</td>
<td>suprasellar</td>
<td>—</td>
</tr>
<tr>
<td>Tandon et al. (1972)</td>
<td>36 yr, F</td>
<td>lt. F</td>
<td>milky, opalescent (580)</td>
</tr>
<tr>
<td>Argyopoulos &amp; Heppner (1970)</td>
<td>35 yr, M</td>
<td>lt. F</td>
<td>milky white, opalescent (108)</td>
</tr>
<tr>
<td>Bhandari (1972)</td>
<td>49 yr, F</td>
<td>rt. P</td>
<td>clear (40)</td>
</tr>
<tr>
<td>Bouch et al. (1973)</td>
<td>64 yr, F</td>
<td>rt. F</td>
<td>clear</td>
</tr>
<tr>
<td>MacGregor et al. (1976)</td>
<td>52 yr, F</td>
<td>lt. centrum semiovale</td>
<td>—</td>
</tr>
<tr>
<td>Ghatak et al. (1974)</td>
<td>43 yr, F</td>
<td>lt. T-P</td>
<td>clear</td>
</tr>
<tr>
<td>Palma (1975)</td>
<td>34 yr, F</td>
<td>lt. F</td>
<td>clear</td>
</tr>
<tr>
<td>Aicardi et al. (1975)</td>
<td>34 yr, F</td>
<td>lt. P</td>
<td>milky</td>
</tr>
<tr>
<td>MacGregor et al. (1976)</td>
<td>5 d, M</td>
<td>lt. O</td>
<td>—</td>
</tr>
<tr>
<td>Friede &amp; Yasargil (1977)</td>
<td>1 yr, F</td>
<td>lt. P</td>
<td>—</td>
</tr>
<tr>
<td>Markwalder &amp; Zimmerman (1979)</td>
<td>43 yr, M</td>
<td>lt. F-T-P</td>
<td>brown turbid (50)</td>
</tr>
<tr>
<td>Haddad et al. (1982)</td>
<td>4 mo, F</td>
<td>rt. T-P-O</td>
<td>crystal-clear</td>
</tr>
<tr>
<td>Pearl et al. (1982)</td>
<td>38 yr, F</td>
<td>lt. P</td>
<td>cloudy, milky</td>
</tr>
<tr>
<td>Oro et al. (1983)</td>
<td>33 yr, M</td>
<td>bifrontal</td>
<td>—</td>
</tr>
<tr>
<td>Shelpet et al. (1985)</td>
<td>27 yr, M</td>
<td>lt. F</td>
<td>turbid, whitish (215)</td>
</tr>
<tr>
<td>Morimoto et al. (1986)</td>
<td>11 mo, M</td>
<td>interhemispheric</td>
<td>clear (13)</td>
</tr>
<tr>
<td>Inoue et al. (1987)</td>
<td>4 mo, M</td>
<td>interhemispheric</td>
<td>clear (63)</td>
</tr>
<tr>
<td>Present case</td>
<td>51 yr, F</td>
<td>lt. O</td>
<td>clear (190)</td>
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</tbody>
</table>


References


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