Supratentorial neuroepithelial cysts

Report of two cases

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Two cases of supratentorial neuroepithelial (ependymal) cysts, one parasagittal and the other located over the parietal convexity, are reported. Comparable reported cases are discussed and a theory of pathogenesis presented.

Keywords: neuroepithelial cysts • ependyma • developmental anomalies

Intracranial ependymal cysts that do not communicate with the ventricles or with the subarachnoid spaces are rare (Table 1).1,8–11,13,16,17,21,23,24,26,27,32,33 Most of these cysts are in the midline. The rare supratentorial lateral cysts have always been found near the Sylvian fissure. This is a report of two such cases.

Case Reports

Case 1

This 34-year-old woman was admitted on May 20, 1973, because of two episodes of loss of consciousness in the past 2 months; the second was immediately preceded with dysarthria and tonic rotation of the head and eyes to the right. Neurological examination showed only a slight increase of the deep reflexes on the right. Electroencephalogram, skull films, and brain scan (Tc 99m) were normal. Left carotid angiography showed a non-vascular mass in the parasagittal frontal region (Fig. 1).

Operation. A cyst approximately 2 × 3 cm resting in a depression of the parasagittal cortex was found in front of the motor area. It contained a fluid whose macroscopic and laboratory characteristics were identical with those of the cerebrospinal fluid (CSF). The roof of the cyst was removed, leaving the rest of the wall adhering to the surface of the cortex. The gyri underlying the cyst bed were barely perceptible; they were compressed and had small rudimentary sulci.

Histological Examination. The cyst wall consisted of cuboidcolumnar epithelium, mostly in a single layer and supported by loose connective tissue often rich in blood vessels (Fig. 2 left). At one or two circumscribed points small formations resembling rudimentary choroid tufts could be discerned (Fig. 2 right).

Case 2

Eleven years before admission this 34-year-old woman had had epileptic seizures with brief interruptions of consciousness several times a day for a period of 2 to 3 months. An electroencephalogram (EEG) then was described as normal. The patient had no treatment and the attacks disappeared spontaneously. One year before the second admis-
TABLE 1
Site of neuroepithelial brain cysts

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>No. of Cases</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitek, et al. 1929</td>
<td>1</td>
<td>right thalamus</td>
</tr>
<tr>
<td>Dandy 1933</td>
<td>1</td>
<td>quadrigeminal plate</td>
</tr>
<tr>
<td>Hamby &amp; Gardner 1935</td>
<td>1</td>
<td>quadrigeminal plate</td>
</tr>
<tr>
<td>Zehnder 1938</td>
<td>1</td>
<td>deep Sylvian fissure</td>
</tr>
<tr>
<td>Beckett, et al. 1950</td>
<td>1</td>
<td>mesencephalon</td>
</tr>
<tr>
<td>Kahn, et al. 1955</td>
<td>1</td>
<td>upper cerebellar vermis</td>
</tr>
<tr>
<td>Handa &amp; Bucy 1956</td>
<td>1</td>
<td>upper cerebellar vermis</td>
</tr>
<tr>
<td>Ziilch 1956</td>
<td>1</td>
<td>quadrigeminal plate</td>
</tr>
<tr>
<td>List &amp; William 1961</td>
<td>1</td>
<td>interhemispherical fissure</td>
</tr>
<tr>
<td>Rand, et al. 1964</td>
<td>2</td>
<td>frontal lobe, parasagittal</td>
</tr>
<tr>
<td>Jakubiak, et al. 1968</td>
<td>2</td>
<td>frontal lobe, parasagittal</td>
</tr>
<tr>
<td>Argyopoulos &amp; Heppner 1970</td>
<td>1</td>
<td>deep temporal lobe</td>
</tr>
<tr>
<td>Harrison 1971</td>
<td>3</td>
<td>chiasmal region</td>
</tr>
<tr>
<td>Patrick 1971</td>
<td>1</td>
<td>Sylvian fissure</td>
</tr>
<tr>
<td>Tandon, et al. 1972</td>
<td>2</td>
<td>frontal lobe, near Sylvian fissure</td>
</tr>
</tbody>
</table>

Histological Examination. The cyst wall consisted chiefly of a few lamellae of connective tissue, partly dense and partly loose, on which rested a single layer of epithelial cells, some flat, some cuboidal or columnar, and some ciliated (Fig. 4).

Discussion

These cysts are usually considered to be a developmental anomaly, but different hypotheses have been offered to explain the primary mechanism. The rare condition described by Yakovlev and Wadsworth under the name of "schizencephalie" may be excluded from discussion of these cases because it is a symmetrical cystic lesion of the cerebral hemispheres resulting from failure of primitive development of the cortex, associated with microgyria and severe mental deficiency. One theory is that an extraventricular neuroepithelial cyst may develop from an island of ependymal cells segregated during embryonal development; this is usually adjacent to corners of the ventricles or in the zones where considerable tissue activity takes place during the formation of the choroid plexuses or the primitive cerebral fissures. In fact, small cavities or islands of ependymal cells in the white substance near the occipital horn or the Sylvian aqueduct quite often appear in otherwise normal anatomical specimens. This concept suggests a possible relationship to the colloid cysts of the third and fourth ventricles, particularly in the region of the lamina quadrigemina. This same concept of embryonal pinching off of ependymal tissue has also been invoked to explain the pathogenesis of similar cysts found in the spinal canal. Another hypothesis suggests that neuroepithelial cysts originate from primitive ectopic glial tissue in the subarachnoid spaces; these tissues often include ependymal canaliculi and choroid tufts. A variation of this theory suggests that the ectopia occurs because of a primary lesion of the cerebral cortex. According to the experimental work of Birge, the presence of choroid tufts shows that the cysts are secondary to a circumscribed encephaloclastic lesion. This lesion occurs early in embryonal development.
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Fig. 1. Case 1. Left carotid angiogram showing a nonvascular mass in the parasagittal frontal region.

Fig. 2. Case 1. Left: Photomicrograph showing columnar epithelium resting on a large blood vessel. H & E, × 60. Right: Photomicrograph showing small papillary projections resembling rudimentary choroid tufts. H & E, × 200.
when the primitive ependymal spongioblasts of the germinal zone surrounding the lesion site not only reconstruct the ependymal investment of the neural tube, but can apparently still differentiate into choroid tissue when they contact the leptomeningeal germ layer.

In our Case 1 the cyst was located far from the ventricle, but the underlying cortex looked like a focus of microgyria. In Case 2 of Tandon, et al.\textsuperscript{26} part of the floor of the cyst was in non-communicating contact with the wall of the ventricle, while the remainder rested on cortex in which “it was hard to see any sulci.” In both cases the cyst’s wall contained small papillary projections like rudimentary choroid tufts (Fig. 2 \textit{right}). It seems reasonable to suppose that a circumscribed embryonal cortical lesion causes this pattern.

In our Case 2 the cyst compressed the parietal cortex, ventricular walls, and Sylvian fissure far from the midline. We believe this cyst was really secondary to a small lesion of the primitive cortical mantle that had been totally repaired in the later development in accordance with the concept of Rand, et al.\textsuperscript{23} The absence of choroid tufts might simply be due to non-inclusion in the specimen biopsied.

Thus we believe that the origin of neuroepithelial cysts compressing the cortical convexity, with or without microgyria, is a primary ependymal ectopia, or an early embryonal lesion with secondary ectopia.

References

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