Neuroepithelial Cysts of the Lateral Ventricles: MR Appearance

The MR imaging appearance of neuroepithelial cysts in the lateral ventricle is reported. Two cases of proven and two of presumed intraventricular neuroepithelial cysts are presented. In one case, MR observations documented spontaneous regression of a large intraventricular cyst. Theories regarding the origin of neuroepithelial cysts are briefly reviewed. Standard T1- and T2-weighted spin-echo pulse sequences were used to study cysts in the lateral ventricles in axial, coronal, and sagittal planes. The cyst wall can be demonstrated reliably with MR images, eliminating the need for CT and/or contrast ventriculography. MR may also be useful in monitoring cyst size on serial examinations.

The most commonly known intraventricular neuroepithelial cyst is the so-called colloid cyst of the third ventricle. Symptomatic neuroepithelial cysts of the lateral ventricles are rare, with only 10 cases reported [1–9]. One of these is described as an arachnoid cyst arising in the lateral ventricle. Several epithelial cell types or connective tissue may constitute the cyst membrane, accounting for the variety of names used to describe these cysts, including epithelial cyst, ependymal cyst, choroid plexus cyst, choroidal-epithelial cyst, and subarachnoid-ependymal cyst [1–16]. Controversy still exists over the precise origin of these cysts. The most unifying theory suggests that all developmental intraventricular cysts, including the colloid cyst, arise from choroid plexus tissue derived from primitive neuroepithelium [17–19]; hence, the term neuroepithelial cyst is perhaps the most appropriate.

The MR imaging appearances in two cases of proven and two cases of presumed neuroepithelial cysts arising in the lateral ventricles are reported; two patients were symptomatic. A case of spontaneous regression of a large intraventricular cyst is also described. Intraventricular cysts are difficult to detect with CT because the cyst wall cannot be seen unless contrast material is instilled into the ventricles. The cyst wall is demonstrated clearly by MR, making it the procedure of choice for evaluating these cysts.

Materials and Methods

Between September 1985 and September 1986, four patients (three males and one female) with prior CT scans showing left lateral ventricular trigone dilatation were studied by MR scanning. Images were generated by a General Electric Signa MR system operating at 1.5 T. Multislice spin-echo (SE) pulse sequences were obtained with a short repetition time (TR) of 600 or 800 msec and an echo time (TE) of 25 msec, two or four excitations, and a long TR of 2500 msec and a TE of 25 and 80 msec, two excitations. Slice thickness was 10 mm with a 2-mm gap, and matrix size was 256 × 128 or 256 × 256. Patients were studied in axial, sagittal, and coronal planes.

Case Reports

Case 1

A 28-year-old man presented with headaches and bilateral extremity numbness associated with prolonged walking. He had occasional episodes of vomiting and blurred vision. Neurologic
Case 3

A 3-year-old girl was followed since birth for a large "intraaxial cyst" of unknown origin discovered initially on prenatal sonography. She was completely asymptomatic and neurologic examination was normal. Normal growth and development were observed. Baseline CT at age 1 month suggested a large region of porencephaly or possibly an intraparenchymal cyst, 6 cm in diameter, located deep in the left hemisphere (Fig. 2A). The left temporal horn was markedly dilated. MR at 3 years of age revealed a cyst 2 cm in diameter with a well-defined wall entirely within the atrium of the left lateral ventricle (Fig. 2B). Spontaneous regression of the cyst had occurred, with only mild residual obstruction of the temporal horn. On long TR images, the intensity of the cyst was greater than that of the CSF (Figs. 2C and 2D). While no pathologic confirmation was obtained, this is presumed to represent a neuroepithelial cyst in the left atrium because of the location and MR characteristics.

Case 4

A 52-year-old man was seen with left optic atrophy. Bilateral lower extremity weakness had become progressively worse since closed head trauma 8 years before. There was no history of headache or seizures, but the patient described episodes of intermittent vomiting over the previous 2 years. Neurologic examination was normal. MR demonstrated an ovoid cyst, 5 cm in diameter, within the left lateral ventricular trigone that deformed the roof of the third ventricle (Fig. 3). The cyst membrane was well demarcated in axial, sagittal, and coronal planes, and the cyst contents had signal intensity identical with that of CSF. The left temporal horn was not dilated. Because the patient's symptoms were minimal, histologic confirmation was not obtained. Owing to the MR features, the presumptive diagnosis of neuroepithelial cyst was made.

Discussion

Neuroepithelial cyst was the term introduced by Fulton and Bailey [20] in 1929 to describe cysts of the central nervous system lined by ependyma (that is, neuroepithelium). Neuroepithelial cysts may be intraparenchymal [10, 11], extraxial [12, 13], intraspinal [21, 22], or intraventricular [1-9, 14-16]. Any ventricle may contain them. The most common intraventricular cysts are the small, asymptomatic cysts of the choroid

examination was normal. Cranial CT revealed focal dilatation of the left lateral ventricular trigone, which was 6 cm in diameter. A cyst membrane could not be identified by CT. Moderate dilatation of the left temporal horn was noted. Radionuclide cisternography failed to detect tracer activity in the trigone region. The patient was managed medically, but because of severe headaches, episodes of mild confusion, and transient global amnesia, he was reevaluated 2 years after initial presentation. MR revealed a large cyst in the left lateral ventricle with a distinct cyst wall. The cyst contents were isointense with CSF on all sequences. At surgery, a large cyst was found in the ventricle encased by a thin, transparent membrane. A window was made in the cyst wall; biopsy was performed, and the liquid contents were evacuated. Several nearby cysts in the adjacent choroid plexus were also removed. The cyst was lined with ependyma, consistent with the diagnosis of neuroepithelial cyst. The protein concentration of the cyst fluid was 32 mg/dl (normal CSF protein, 5-15 mg/dl). Despite surgical decompression, the patient's headaches persisted.
Fig. 2.—Case 3: spontaneous regression of intraventricular cyst in asymptomatic 3-year-old girl.
A, Contrast-enhanced CT scan at age 1 month. Large, low-density lesion initially believed to represent porencephalic or parenchymal cyst.
B and C, Axial SE 2500/25 (B) and SE 2500/80 (C) MR images 3 years after CT show marked reduction in cyst size. Cyst (arrows) has higher signal than CSF, possibly because of higher protein content of cyst fluid (unproven).
D, Left parasagittal MR image (SE 300/25) clearly shows cyst wall (arrows) within atrium and dilated temporal horn.

Fig. 3.—Case 4: presumed neuroepithelial cyst, left lateral ventricle.
A, Axial MR image (SE 800/20). Large cyst in left trigone (small arrow). Glomus displaced laterally (large arrow).
B, Axial MR image (SE 2500/80) shows thin cyst wall (arrow). Signal intensity of cyst identical to CSF.
C, Sagittal MR image (SE 600/20). Cyst (black arrows) deforms root of third ventricle, inferiorly displacing internal cerebral vein (white arrow).

plexus that occur in more than 50% of some autopsy series [22]. The majority of these cysts measure less than 1 cm in diameter and occur in any part of the choroid plexus, most often in the glomi of the lateral ventricles. Occasionally, these small choroid plexus cysts are discovered incidentally during fetal or neonatal sonography and may exist transiently [23, 24]. Schnopfhagen [25] believed they were secondary to cystic degeneration of the choroid plexus related to aging.
In 1936, McLean [26] distinguished epithelial-lined, developmental cysts of the third ventricle (colloid cysts) from connective-tissue-lined cysts of the choroid plexus. In 1965, Shuangshoti et al. [17] proposed a common origin of so-called “colloidal” and “noncolloidal” intraventricular cysts, because the choroid plexus, ependyma, and paraphysis are all derived from primitive neuroepithelium. The paraphysis is an embryologic tissue now believed to represent extraventricular choroid plexus; it forms along the roof of the primitive third ventricle, and some colloid cysts arise from it.

Shuangshoti and Netsky [18] asserted that during maturation of the choroid plexus, cysts may develop that are lined by either ependyma or fibrous connective tissue, and they preferred the term neuroepithelial cyst in describing these developmental intraventricular cysts. This is an attractive theory since it explains why a variety of epithelial cell types are found in the cyst lining. It does not explain the greater tendency of colloid cysts to possess an epithelial lining, in contrast to choroid plexus cysts, which are usually lined by connective tissue with only sparse epithelial cells present. Colloid cysts differ in appearance both histologically and radiographically.

Radiographic findings in neuroepithelial cysts, regardless of type, reflect the secretions and breakdown products of the epithelial lining. For example, colloidal neuroepithelial cysts contain dense mucoid material, CSF, fat, and desquamated cells in various proportions [17]. Therefore, the CT appearance is variable. On noncontrast CT, colloid cysts are most often hyperdense relative to brain parenchyma, but some are isodense or even hypodense [27–30]. On MR, colloid cysts reported thus far [31] have short T1 and long T2. By comparison, noncolloidal neuroepithelial cysts differ from colloid cysts in appearance and content. On CT, intraventricular neuroepithelial cysts have a density similar to CSF, and the cyst wall is generally not seen. With MR, the cyst wall is easily detected, even though the cyst wall is very thin and closely applied to the ventricular lining. T1-weighted SE images in axial and coronal planes are usually sufficient to demonstrate the cyst wall. If these are equivocal, sagittal T1-weighted images are also obtained. T2-weighted images are not necessary to detect the cyst membrane but may be useful in further characterizing the cyst fluid and screening for other intracranial disorders. Before MR, air or water-soluble contrast ventriculography in conjunction with CT was usually required to detect the cyst wall. In this regard, MR has a distinct advantage over CT.

These developmental cysts contain clear, serous liquid resembling CSF both on gross inspection and with MR. In the surgically proven cases, the protein concentration of the cyst fluid was mildly elevated relative to CSF, but there was no observable difference between the signal intensity of the cyst fluid and CSF. It is likely that at these protein levels there is insufficient T1 shortening to produce a detectable increase in signal. This is in agreement with others [31, 32], who have suggested that only highly proteinaceous cystic lesions have higher signal intensity than CSF. In case 3 (Fig. 2), the signal intensity of the cyst fluid was higher on T1- and T2-weighted images. While this could be explained on the basis of high protein content, the presence of nonpulsatile fluid within the cyst could also account for its higher signal intensity. It has been shown that nonpulsatile CSF has higher signal intensity on T1- and T2-weighted images [33], and this feature may aid in the diagnosis of some arachnoid and intraventricular cysts [32]. However, in the absence of a significant signal intensity difference between CSF and cyst fluid, demonstration of the cyst wall is necessary to make a definitive diagnosis.

The natural history of these cysts is unknown. Small cysts of the choroid plexus may involute spontaneously [25]. In case 3, the cyst size diminished significantly over a 3-year period without treatment. This is the only case known to us of a large intraventricular cyst regressing spontaneously.

While the MR appearance of intraventricular neuroepithelial cysts seems likely to be quite characteristic, several abnormalities should be considered in the differential diagnosis. Porencephaly of the lateral ventricle can be confused with a large intraventricular cyst on CT. Ventricular porencephaly can be secondary to a variety of insults including developmental defects, trauma, and cerebrovascular disease [34].

Arachnoid cysts typically arise in the cisterns. Only one reported case of an intraventricular arachnoid cyst is known [9], and on CT it appears identical to the lateral ventricular cysts presented here. In contrast, a variety of neuroepithelial cysts may arise anywhere within brain substance, ventricles, or subarachnoid space [13, 35–37]. Arachnoid cysts containing ependymal elements have been reported [12].

Epidermoid tumors most often arise in extradural locations. Intraventricular epidermoid tumors are very rare, but may occur in the fourth ventricle or temporal horn of the lateral ventricle [38, 39]. Depending on the ratio of lipid to keratin present, the signal intensity of epidermoid tumors on MR is variable [40]. If they contain mostly cholesterol, the T1 and T2 values are short, and therefore they will not resemble a neuroepithelial cyst.

Intracranial echinococcal cysts occur in only 2% of those affected. When the brain is involved, the cyst is nearly always intraparenchymal, growing to large size and compressing the lateral ventricles [41]. Intraventricular echinococcal cysts are exceedingly rare. However, a large echinococcal cyst positioned in the trigone could mimic a neuroepithelial cyst on both CT and MR [42].

Cysticercosis is a common parasitic disease affecting the brain in endemic areas. Intraventricular cysts occur in 11–17% of those with CNS involvement and may produce acute, fatal ventricular obstruction [43, 44]. The cysticercal cyst is usually 1–2 cm in diameter and contains a mural nodule (scolex) that can be identified on MR [45]. In addition, the parasitic cyst wall typically has high signal intensity on long TR images [45]. Hence, it has an MR appearance distinctly different from a neuroepithelial cyst.

In summary, neuroepithelial cysts arising in the lateral ventricles can be diagnosed reliably with MR by demonstrating the cyst wall with conventional SE pulse sequences. The ability to diagnose these cysts with MR makes the use of CT and/or contrast ventriculography unnecessary. MR may also be used to monitor cyst size on serial examinations.
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