Epithelial cyst of the fourth ventricle

Case report

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A case of epithelial cyst in the fourth ventricle of a 4-year-old child is described. A single epithelial layer with a clear basement membrane lining the cyst wall was observed. There were no prominent histological findings to suggest a pathogenesis for this cyst based on immunohistochemical or ultrastructural studies; however, the cyst fluid contained significant amounts of carcinoembryonic antigen. It is considered that the epithelial layer lining the cyst wall was possibly of endodermal origin.

KEY WORDS • epithelium-lined cyst • carcinoembryonic antigen • neuroepithelial cyst • enterogenous cyst

Epithelium-lined cysts of the central nervous system (CNS) are rarely encountered. Such benign cysts have been reported at autopsy in symptomatic or asymptomatic individuals. Since Parkinson and Childe described an epithelial cyst of the fourth ventricle, few reports have appeared in the literature.1,2,8,10,11,14,15,16,24 The origin and pathogenesis of the cyst remain controversial. Two main theories have been proposed: the first involves a neuroectodermal origin and the second an endodermal origin. In 1981, Afshar and Scholtz described a fourth ventricular cyst and suggested that the cyst wall was related to enterogenous epithelium, based on ordinary microscopic examinations.

The present report documents a patient with a fourth ventricular cyst and suggests the possibility of an endodermal origin due to the presence of a high concentration of carcinoembryonic antigen in the cyst fluid. We discuss the pathogenesis and origin of this cyst from the standpoint of detailed ultrastructural and immunohistochemical studies.

Case Report

This 4-year-old boy had a 1-month history of headache and poor balance. He was referred to the neurosurgical department of Nihon University School of Medicine in July, 1988. He had been completely well until the onset of these symptoms. There was no history of head trauma, infections, or disorders of the CNS.

Examination. The results of neurological examination revealed no symptoms of cerebellar disorder except for bilateral upper limb ataxia and a slightly broad-based ataxic gait. The result of Romberg's test was normal. The cranial nerves were intact. Routine laboratory examination data were within normal limits. A plain skull x-ray film revealed no remarkable changes except for elevation of the confluence. Computerized tomography (CT) demonstrated a low attenuation area in the dorsal region of the fourth ventricle, with contrast enhancement around the lesion (Fig. 1 left). The third

Fig. 1. Left: Computerized tomography (CT) scan demonstrating a well-demarcated low-density area in the dorsal region of the fourth ventricle (left), the rim of which is slightly enhanced after intravenous injection of contrast medium. Right: Metrizamide CT cisternography clearly reveals a cystic mass lesion. No communication between the low-density area and the fourth ventricle is observed.
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and lateral ventricles were slightly dilated. Vertebral angiography revealed an avascular area in the midline of the posterior cranial fossa and upward displacement of the branches of the posterior inferior cerebellar artery.

Intrathecal metrizamide-enhanced CT cisternography revealed no communication between the cyst and fourth ventricle (Fig. 1 right). Magnetic resonance (MR) imaging was performed in the axial and sagittal planes, producing T1- and T2-weighted and proton density images (Fig. 2). The axial spin-echo proton image revealed a cystic lesion as a hypointense area, and the T2-weighted axial image showed it as hyperintense. The sagittal T1-weighted image clearly showed the outline of the cyst and its relationships to the surrounding structures, from which hypoplasia or aplasia of the cerebellar vermis was suspected.

Operation. A midline posterior fossa craniectomy was performed in August, 1988. Following incision of the dura and arachnoid membrane, a translucent yellowish thin-walled cyst was found to protrude onto the cerebellar surface, shearing both hemispheres of the cerebellum. The cyst was punctured, and a white turbid fluid was aspirated. It contained a total protein level of 57 mg/dl, sugar content of 33 mg/dl, and a carcinoembryonic antigen value of 2886 ng/ml. A radioimmunoassay for carcinoembryonic antigen was performed by a one-step “sandwich” method using a carcinoembryonic antigen kit. The cyst wall was readily separated from the surrounding tissue and was totally removed. After removal of the cyst, it was noted that it had been attached to the fourth ventricle and had occupied the dorsal region of the fourth ventricle, extending from the aqueduct to the obex. The choroid plexus was not observed in the operative field, and hypoplasia of the cerebellar vermis was confirmed.

Postoperative Course. The patient was discharged from the hospital 3 weeks after the operation. His ataxic gait and dysdiadochokinesis in both hands gradually resolved over a period of 3 months.

Pathological Examination. The specimen consisted of an opened cyst, 2.5 cm in diameter, with a wall thickness of 0.1 cm. On light microscopy, the cells lining the inner aspect of the cyst wall were found to form a single columnar epithelium and cuboidal epithelium which was pseudostratified in places (Fig. 3). Periodic acid-Schiff (PAS) and mucicarmine stains were applied; positive reactions were observed in many cells which covered the epithelial surface. Argentophil cells were not noted on silver staining. Carcinoembryonic antigen staining of the cyst wall was carried out using an immunohistochemical technique, but was negative in the specimen; an immunohistochemical study by the peroxidase-antiperoxidase method was also performed.* An ultrastructural study (Fig. 4) revealed that the cuboidal and columnar epithelial cells lining the cyst wall had an oval- to irregular-shaped nucleus, well-developed Golgi apparatus, and endoplasmic reticula. The apical surface of the cells had numerous short microvilli without surface coating material. Ciliated cells and basal bodies were not observed. The lateral surface of the cells showed interdigitations with desmosomal junctional complexes. Some cells were filled with round osmiophilic secretory granules. The epithelial cells were bordered by a continuous basal lamina.

Discussion

Symptomatic epithelial cysts in the neuraxis are usually found in the third ventricle and have been reported as colloid cysts. Epithelial cysts are rarely encountered in the fourth ventricle. Parkinson and Childe first reported such a case in a patient with hydrocephalus who had two cysts involving the fourth ventricle. It was

* Rabbit anti-human carcinoembryonic antigen, carcinoembryonic antigen isolated from a hepatic metastatic tumor from a human colonic cancer, and rabbit immunoglobulin goat serum supplied by Cambridge Research Laboratory, International Reagent Co., Tokyo, Japan.
considered that these were identical with the typical colloid cysts of the third ventricle but had originated in the fourth ventricle. Subsequently, few other reports have appeared in the literature.\(^1\)\(^2\)\(^8\)\(^11\)\(^14\)\(^15\)\(^18\)\(^24\) Moreover, it has been difficult to clarify the histogenesis because of the simple epithelial cyst lining.

The origin of epithelial cyst of the fourth ventricle has been discussed on the basis of two theories. One theory proposes an origin from the neuroepithelial structures including: ependyma, paraphysis, and/or choroid plexus, which were related to the primitive neuroepithelium that lines the neural tube. Palma\(^16\) reported two cases of supratentorial neuroepithelial cysts, which had their origin in a primary ependymal ectopia or an early embryonal lesion with secondary ectopia. Hoenig, et al.,\(^8\) described multiloculated cysts associated with the choroid plexus of the fourth ventricle. The investigations of Shuangshoti and Netsky\(^19\) suggested that neuroepithelial cysts may develop in any part of the ventricle and brain tissue, arising from the ependyma and/or choroid plexus and the primitive neuroepithelium in a manner similar to the cysts found in the third ventricle. They postulated that the mechanism of formation of such cysts was a simple folding of the neuroepithelium into or out of the ventricle.

The second theory proposes an origin from the endoderm. Hirano and Ghatak\(^7\) reported a case with an epithelial cyst of the spinal cord. They stressed that the epithelial lining resembled respiratory epithelium, which to them suggested that the cyst had an endo-
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dermal origin based on detailed ultrastructural studies. Afshar and Scholtz\(^1\) described a case of epithelium-lined cyst of the fourth ventricle. The cyst was composed of simple or pseudostratified epithelium, with scattered argentophil cells in the region of the basement membrane. The cyst resembled intraspinal cysts lined with epithelium of intestinal origin which are called “neurenteric cysts” and result from embryonic anomalies. They concluded therefore that this cyst was probably of enterogenous origin. Miyagi, et al.,\(^12\) reported an enterogenous cyst in the cervical spinal canal which displayed a positive reaction to carcinoembryonic antigen immunohistochemical staining. Based on these reports, the origin of fourth ventricular cysts is thus still controversial.

The pathological findings in our case revealed the presence of epithelial mucin (due to positive reaction for PAS and mucicarmine staining) and a prominent basement membrane. Ciliated cells, blepharoplasts, and argentophil cells were not detected. These results were insufficient to support the possibility of an endodermal origin for the cyst or to exclude a neuroepithelial origin. In addition, we failed to detect carcinoembryonic antigen immunohistochemically in the cyst itself, even though it was present in the cyst fluid. We cannot yet offer a clear interpretation for this discrepancy, but it might involve a carcinoembryonic antigen variant such as nonspecific cross-reacting antigen.\(^3,4,9,23\) However, the presence of a high concentration of carcinoembryonic antigen in the cyst fluid strongly supports the idea that the cyst was composed of endodermal material, because carcinoembryonic antigen has been detected in colonic adenocarcinoma and in embryonic and fetal digestive tissues in the first two trimesters of gestation.\(^5,6,22\)

In summary, conclusive evidence concerning the pathogenesis of the cyst described in this report was not available from the findings obtained by light and electron microscopy. However, since the cyst fluid contained significant amounts of carcinoembryonic antigen, which is unusual in CNS diseases except for metastatic tumor, craniopharyngioma, or germ-cell tumor,\(^13,22\) we consider that the present cyst may have originated in the endodermal tissue.

References
6. Gold P, Freedman SO: Specific carcinoembryonic anti-

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