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Endodermal Cyst Ventral to the Lower Brain Stem

—Case Report—

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Abstract

A 28-year-old woman presented with a rare case of endodermal cyst located ventral to the lower brain stem manifesting as recurrent aseptic meningitis. Computed tomography and magnetic resonance imaging demonstrated a cystic mass located ventral to the lower brain stem and extending from the prepontine to the upper cervical (C-2) cistern. The lesion was totally removed through a retrosigmoid craniotomy and C1–2 hemilaminectomy. Histological and immunohistochemical examination showed the cyst was derived from the endoderm. The histological diagnosis was endodermal cyst. Early diagnosis and surgical removal are important for patients with these cysts.

Key words: endodermal cyst, brain stem, meningitis

Introduction

Intracranial endodermal cysts are rare, accounting for no more than 0.01% of central nervous system (CNS) tumors. Endodermal cysts can occur anywhere in the CNS,[1,4,12] but are usually located in the posterior fossa, especially on the ventral surface of the brain stem.[1,21] The embryogenesis of the cysts is generally considered to be disorder of gastrulation in the 3rd week of the embryonic life. Incomplete regression of the foregut (the endoderm) and the notochord (the ectoderm) occurring before regression of the primitive streak leaves endodermal remnants in the caudal neurenteric canal.[1,3,8,9,11,12,20,21,25,26] This theory can explain the occurrence of the cysts in the spinal canal and the basal cranial cavity. Therefore, cysts in the cranial cavity tend to occur in the posterior fossa, especially on the ventral surface of the brain stem.

We report a rare case of endodermal cyst located ventral to the lower brain stem.

Case Report

A 28-year-old woman presented with nuchal and occipital pain and general fatigue. She had a past history of meningitis at age 8 years. Her cerebrospinal fluid showed an elevated white blood cell count of 256/3 cells/mm^3 with 45.3% monocytes, a normal protein concentration of 45 mg/dl, and a reduced glucose concentration of 4 mg/dl. Therefore, the diagnosis was aseptic meningitis of unknown origin. Though she had no neurological abnormality, neurological screening including computed tomography (CT) and magnetic resonance (MR) imaging was performed to exclude CNS organic lesions as a probable cause of meningitis.

CT revealed a slightly high density lesion located ventral to the lower brain stem with no definite rim enhancement (Fig. 1). MR imaging showed a cystic lesion with irregular walls resembling a bunch of grapes. The upper pole of the cyst was located in the mid-portion of the prepontine cistern, and the lower pole at the C-2 level. The contents appeared as high intensity on T_1-weighted MR imaging and iso–low

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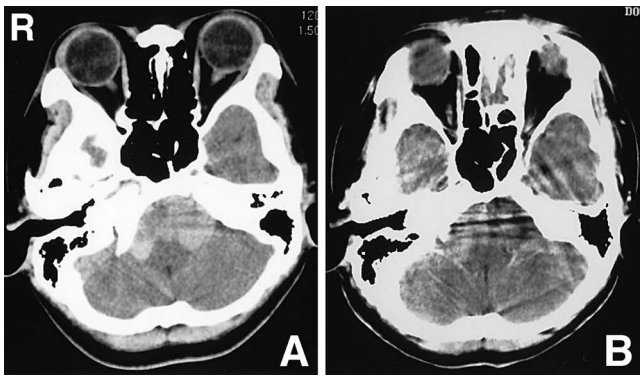

Fig. 1 Computed tomography scans showing a slightly high density mass ventral to the lower brain stem (A) with no definite enhancement of the rim of the tumor (B).

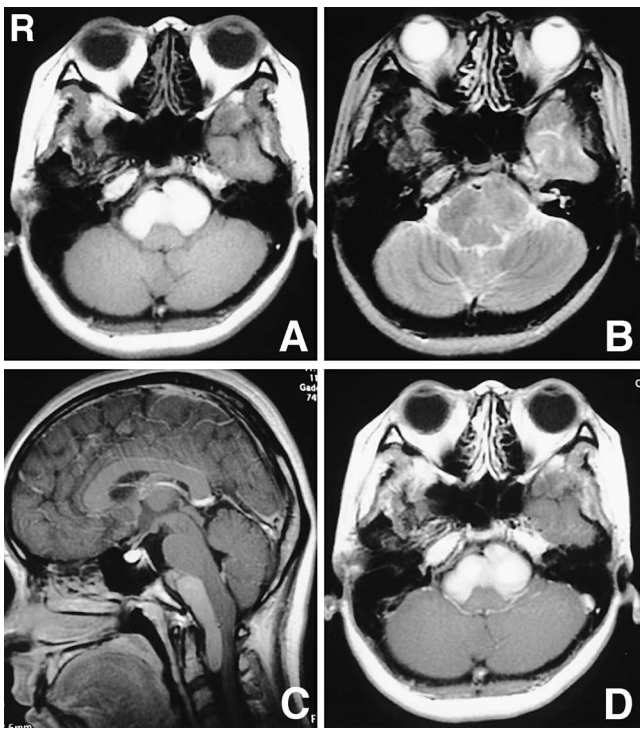

Fig. 2 Magnetic resonance images demonstrating a lobulated cystic lesion located ventral to the lower brain stem and the upper cervical cord, with homogeneous high intensity contents on the T_1-weighted image (A) and iso–low intensity contents on T_2-weighted image (B), and no rim enhancement after gadolinium administration (C, D).

intensity on T_2-weighted MR imaging. Diffusion-weighted MR imaging showed no positive findings, and gadolinium administration caused no enhancement of the cyst or the contents (Fig. 2). Vertebral angiography demonstrated ventral deviation of the bilateral vertebral and proximal basilar arteries, but no distinct tumor stain.

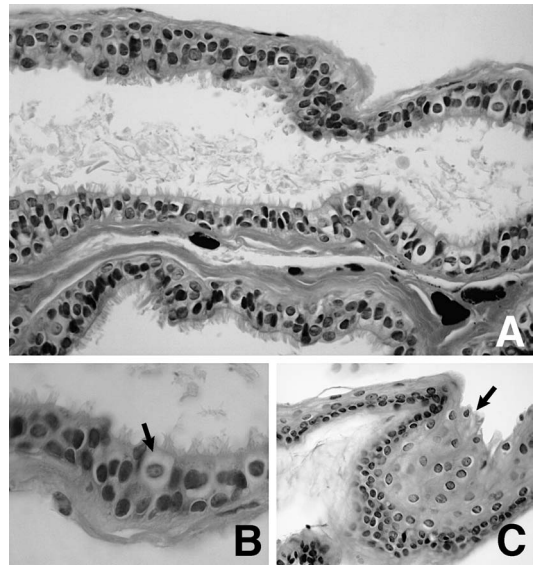

Fig. 3 Photomicrographs of the surgical specimen showing the cyst wall lined with a layer of cuboidal and columnar ciliated cells (A, B) with ciliated epithelium (B: *arrow*), and stratified squamous cells containing basal cells indicating squamous metaplasia (C: *arrow*). Hematoxylin and eosin stain, A: × 10, B: × 100, C: × 40.

A right retrosigmoid approach was performed through a C1–2 hemilaminectomy. The multi-cystic yellowish mass was observed between the vertebrobasilar artery junction and the brain stem. The tumor had a thin, translucent, slightly elastic membrane, and contained creamy, yellowish, homogeneous soft material. The membrane of the tumor was weakly adherent to the surrounding neurovascular structures. The cyst contents were substantially removed by suction, and the membranous portion of the tumor was totally removed without damage to the surrounding brain and nerves. The postoperative course was uneventful and she was discharged without neurological deficits or complaints. Postoperative MR imaging showed complete removal of the tumor.

Histological and immunohistochemical examinations showed the cyst wall lined with a layer of cuboidal and columnar cells (Fig. 3A, B), mostly positive for low molecular weight cytokeratin (simple type, CAM5.2) (Fig. 4A). Squamous metaplasia was observed in places (Fig. 3C), with positive staining for high molecular weight cytokeratin

Table 1 Cases of endodermal cyst ventral to the brain stem

Author (Year)	Age	Sex	Symptoms	MR imaging (T_1/T_2)	Cyst content	Origin
Small (1962)[28]	ND	ND	ND	ND	ND	ND
	ND	ND	ND	ND	ND	ND
	ND	ND	ND	ND	ND	ND
Hirai et al. (1981)[12]	30 yrs	F	headache, visual blurring, hearing loss	ND	clear fluid	ED
Yoshida et al. (1983)[31]	2 mos	M	megacephaly, nystagmus	ND	ND	ED
Chavda et al. (1985)[4]	26 yrs	M	III–IV cranial nerve pareses, motor weakness, dysphasia	ND	ND	ED
Schelper et al. (1986)[26]	38 yrs	F	headache, meningitis	ND	amorphous, soft white cheesy material	ED
van der Wal and Troost (1988)[30]	4 yrs	F	oculomotor nerve palsy	low/ND	yellowish fluid	ED
Hasegawa et al. (1988)[10]	42 yrs	M	headache, rectovesical disturbance, sensory disturbance, motor weakness	low/ND	ND	ED
Fukushima et al. (1988)[7]	23 yrs	M	headache, vertigo, visual blurring	ND	crystal clear fluid	ED
Kak et al. (1990)[15]	21 yrs	M	numbness and weakness of upper extremities	high/high	viscous milky opalescent fluid	ED
Breeze et al. (1990)[3]	37 yrs	M	neck pain, dysarthria, rectovesical disturbance, sensory disturbance	low/ND	straw-colored fluid	ED
Koksel et al. (1990)[16]	40 yrs	M	neck pain, motor weakness	low/ND	clear yellow fluid and cholesterol crystals	ED
Boyar et al. (1991)[2]	23 yrs	M	motor weakness, nystagmus	ND	colorless cystic fluid	ED
Hirai et al. (1991)[13]	53 yrs	M	V–VIII cranial nerve pareses, motor weakness, sensory disturbance	high/high	creamy viscous fluid	ED
Fleming et al. (1991)[6]	33 yrs	F	headache, meningitis, hearing loss	high/high	viscous, cloudy, yellow fluid	ED
Harris et al. (1991)[9]	30 yrs	F	headache, nausea, visual blurring	ND	clear fluid	ED
	58 yrs	F	dizziness, vertigo, headache, nausea, tandem gait	low/ND	turbid fluid containing white mucoid particles	ED
Malcom et al. (1991)[20]	57 yrs	F	headache, vertigo, diplopia	ND	viscous yellow material	ED
	31 yrs	M	III–VI cranial nerve pareses, ataxia	high/low	dark brown fluid with recent hemorrhage	ED
Lee et al. (1992)[18]	34 yrs	M	headache, central cord syndrome	low/high	thick, grayish, gelatinous material	ED
Del Bigio et al. (1992)[5]	8 yrs	F	meningitis	ND	creamy white material	ED
Hirabayashi et al. (1992)[11]	31 yrs	M	motor weakness, facial nerve paresis, dysarthria	high/low	white cheesy	ED
Menezes and Ryken (1995)[21]	8 yrs	F	meningitis	high/ND	ND	ED
Gao et al. (1995)[8]	5 yrs	F	ND	low/low	clear or proteinaceous fluid	ED
	8 yrs	M	ND	iso/iso	clear or proteinaceous fluid	ED
	15 yrs	M	ND	low/high	clear or proteinaceous fluid	ED
	58 yrs	F	ND	iso/high	clear or proteinaceous fluid	ED
Shimizu et al. (1996)[27]	51 yrs	F	tinnitus, dysphagia	low/high	yellow material	ED
Bejjani et al. (1998)[1]	31 yrs	F	meningitis	high/high	clear fluid	ED
	21 yrs	F	nausea, vomiting	high/low	thick, greenish, and inspissated solid material	ED
	21 yrs	M	headache, ptosis, dysphagia, oscillopsia, ataxia, lower cranial nerve pareses	high/low	green mucoid material	ED
Ray et al. (2000)[25]	13 yrs	M	hearing loss, headache, dizziness, nausea, ataxia	low/high	ND	ED
Lin et al. (2004)[19]	45 yrs	F	headache, nausea, vomiting, diplopia, stiff extremities	low/high	gelatinous whitish material	ED
Present case	28 yrs	F	headache, meningitis	high/iso–low	creamy, yellowish, homogeneous soft material	ED

ED: endodermal, MR: magnetic resonance, ND: not described.

(stratified type, 34βE12) (Fig. 4B). Squamous epithelia was also positive for carcinoembryonic antigen (CEA) (Fig. 4C). Some of the lining cells of the cyst wall contained periodic acid-Schiff (PAS)-positive secretory granules (Fig. 4D). Basal cells (reserve cells) abutting on the basement membrane in some places were clearly shown to cover these lining cells. Given the immunohistochemical find-

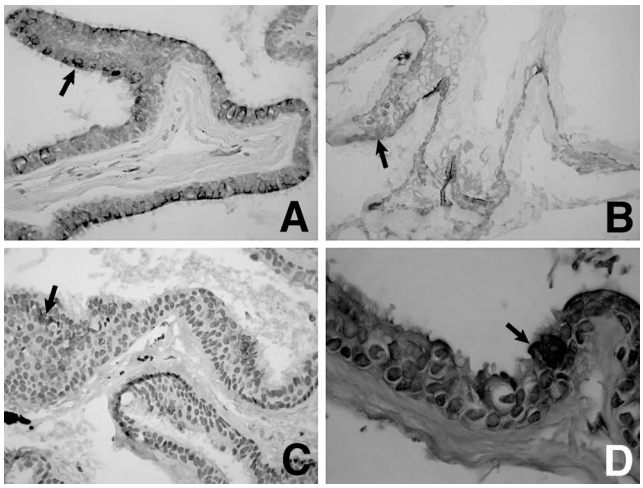

Fig. 4 Photomicrographs of immunohistochemical staining revealing that many cuboidal and columnar cells in the epithelial layer of the cyst are positive for cytokeratin (CAM5.2) (A: *arrow*), cells in the area of squamous metaplasia are positive for cytokeratin (34βE12) (B: *arrow*) and carcinoembryonic antigen (C: *arrow*), whereas most cells in the epithelial layer contain secretory granules positive for periodic acid-Schiff (D: *arrow*). A: ×10, B: ×100, C: ×40.

ings of the lining cells of the cyst wall, electron microscopy was not needed to make the histological differential diagnosis. According to these findings, we concluded that the cyst was derived from the mesoderm. The histological diagnosis was endodermal cyst.

Discussion

In the present case, the cyst was considered to be derived from the endoderm based on the histological and immunohistochemical findings. Review of the histological description of 31 reported cases[1–13,15,16,18–21,25–27,30,31] (excluding 3 cases[28] in which histopathological examination was not described) of epithelial cyst ventral to the lower brain stem showed that all originated from the endoderm (Table 1).

The histological classification of intracranial epithelial cysts is controversial. Generally speaking, intracranial epithelial cysts can be divided into two types, endodermal and neuroepithelial cysts.[6,7,12,17,20,22–26,31] Cysts originating from the endoderm contain two types of lining cell, ciliated or nonciliated, and basal cells. The inner surface of the cyst has a layer of cuboidal or columnar cells, which sometimes contain secretory granules positive for PAS and mucicarmine. Basal cell hyperplasia is sometimes followed by squamous metaplasia, but always rests on a basement membrane. Positive immunoreactivity for epithelial membrane antigen, cytokeratin, and CEA suggests an endodermal origin for the cyst.[3,6,7,14,16–18,20–23,27,29] In contrast, neuroepithelial cysts have an inner lining layer of emboliform cells. The basement membrane is often absent, and instead, the neuroepithelial cysts are directly adjacent to the glial cells which show positive immunoreactivity for glial fibrillary acidic protein.[6,7,29] Therefore, electron microscopy is not always necessary to make the histological differential diagnosis between the two types of the cyst.

The most common clinical features of patients with endodermal cyst ventral to the brain stem are headache, nausea, and vomiting caused by raised intracranial pressure or meningeal irritation following leakage of the intracystic material.[1,21] The next most common manifestations are various cranial nerve pareses and motor weakness, probably caused by the mass effect of the cyst.[1,11] Recurrent aseptic meningitis is the most characteristic symptom of this pathology and was observed in six cases,[1,5,6,21,26] including the present case. Our patient suffered at least two episodes of aseptic meningitis. Screening with CT and MR imaging are recommended to exclude CNS epithelial cyst as a possible cause of aseptic meningitis of unknown origin.[1,6,17] Microsurgical total removal without neurological deficits is usually easy for these epithelial cysts.[1,13,19,23,25] Early diagnosis and surgical removal are important for patients with these cysts.

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