

# Endodermal Cyst of the Posterior Fossa

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We report a case of endodermal cyst of the posterior fossa. A 44-year-old man presented with headache for three months. Computed tomography and magnetic resonance imaging revealed a 6×2.5×2cm sized extra-axial non-enhancing cystic lesion on the ventral aspect to brain stem. To avoid retraction injury to brain stem, far lateral transcondylar approach was selected. Right suboccipital craniotomy and partial removal of occipital condyle with resection of C-1 and C-2 hemilaminae exposed the extra-axial cyst well. The cyst has a whitish thick membrane. It was not adherent to brain stem and lower cranial nerves. Total removal of the cyst was done without difficulty. Histological analysis disclosed a layer of pseudostratified columnar epithelium with basement membrane. The result of immunohistochemical study was consistent with endodermal cyst.

**KEY WORDS :** Endodermal cyst · Posterior fossa · Far lateral transcondylar approach.

## Introduction

Endodermal cyst is a rare developmental cyst lined by columnar epithelium of presumed endodermal origin<sup>7,15</sup>. It can develop anywhere along the neuraxis, but it is predominantly found in the spinal subarachnoid space of the cervical and thoracic spine<sup>1,16,17,21</sup>. Intracranial endodermal cyst is extremely rare, but when it occurs, it has a predilection for posterior fossa. There has been reported only 50 cases, so far<sup>2,3,5,6,9,11,12,15,17,18</sup>.

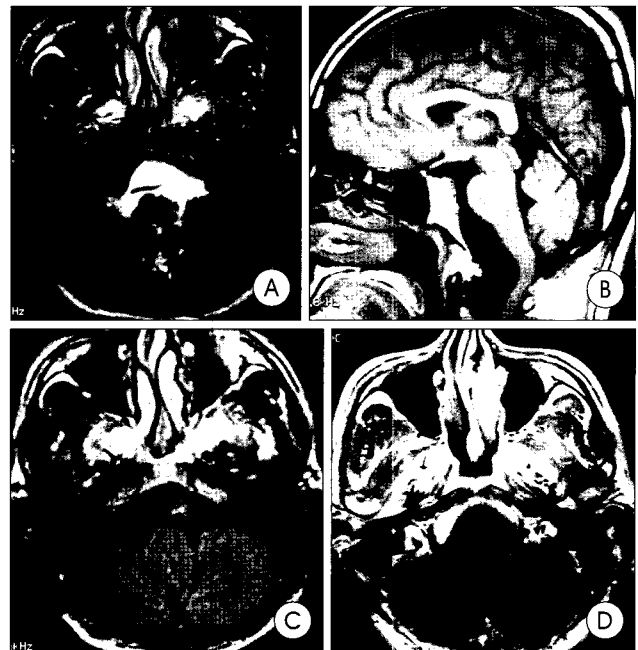
Endodermal cysts are also known by a variety of other names such as enterogenous, neurogenic, neurenteric, foregut, bronchogenic, respiratory, epithelial, epithelial-lined, teratomatous cysts and gastrocytoma<sup>4,7,13,17,20,21</sup>. Recently, we experienced a case of posterior fossa endodermal cyst, which was resected successfully via far lateral transcondylar approach.

## Case Report

A 44-year-old man presented with three months history of occipital headache extending to both shoulder. Headache was recently aggravated in its intensity and duration, especially in the early morning. He was alert and neurologically normal.

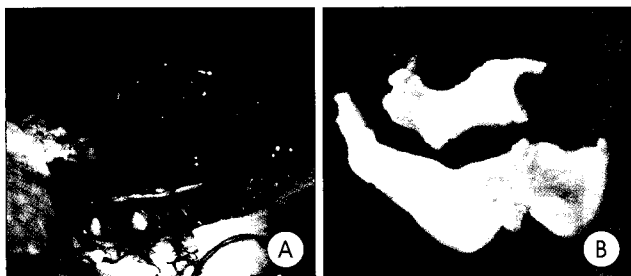
A magnetic resonance (MR) image of the brain demonstrated

a 6×2.5×2cm sized prepontine lesion extending downward to cervicomedullary junction. The lesion showed a high signal on T2 weighted image and a low signal on T1 weighted image. On fluid attenuated inversion recovery (FLAIR) image, it



**Fig. 1.** Magnetic resonance imaging sequence of the brain showing T2-weighted axial (A), T1-weighted sagittal (B), FLAIR axial (C), T1-weighted gadolinium enhanced axial (D) images. These studies reveal extra-axial cystic lesion at the ventral aspect of mid-pons extending downward to cervicomedullary junction. Fluid within the cyst has different signal intensity to that of cerebrospinal fluid on FLAIR image. Kinking of brain stem at cervicomedullary junction is evident on T1-weighted sagittal image. There is no associated hydrocephalus. The lesion is not enhanced after administration of gadolinium.

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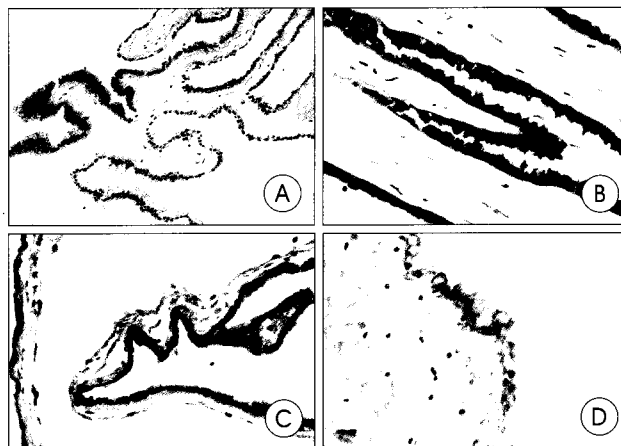


**Fig. 2.** Intraoperative photograph (A) showing an extra-axial cyst in front of the brain stem associated with multiple lower cranial nerves. Resected specimen (B) showing thick whitish cyst wall.

appeared to have different signal to cerebrospinal fluid (CSF). The lesion was not enhanced after administration of gadolinium. A sagittal T1 weighted image showed kinking of brain stem at the cervicomedullary junction, displacing the lower medulla posteriorly (Fig. 1). Preoperative vertebral angiogram demonstrated posterior displacement of basilar artery without tumor blush. The lesion was thought to be an epidermoid or arachnoid cyst preoperatively.

The patient underwent a right-sided far lateral transcondylar approach. Under general anesthesia, the patient is placed in a modified park bench position with 45 degree of head rotation to contralateral side and 15 degree of neck flexion. An inverted hockey stick incision begins at the mastoid process and courses inferiorly to the superior nuchal line as far as midline, continuing inferiorly down to the C-4 or C-5 vertebral level. The myocutaneous flap is elevated to expose the posterior craniovertebral junction. The ipsilateral laminae of the C-1 and C-2 vertebrae are also exposed in the subperiosteal fashion. Right suboccipital craniotomy with C-1 and C-2 hemilaminectomy is performed. One third of occipital condyle is drilled out. Curvilinear dural incision exposes whitish mobile cystic mass in front of brain stem and cervicomedullary junction. The tumor is not adherent to adjacent neurovascular structures (Fig. 2A). Cyst membrane is ruptured incidentally during the dissection of cystic wall. Shrinkage of cyst make it easy to dissect the cyst. The whitish cyst wall is relatively thick (Fig. 2B). Gross total resection is achieved. After watertight closure of the dura, bone plate from the suboccipital craniotomy are secured in their original location, as in the laminae of C-1 and C-2.

The patient had an uneventful recovery. His occipital headache disappeared after surgery. The cyst was lined by single-layered pseudostratified columnar cells without apparent cilia lying on a basement membrane. The lining epithelium was positive for cytokeratin, periodic acid-Schiff (PAS) and focally positive for carcinoembryonic antigen (CEA). There is no smooth muscle tissue in the cyst wall. No other element including ependymal cells and glial tissue was observed (Fig. 3). These findings



**Fig. 3.** Histologic and immunohistochemical findings of endodermal cyst. A : A simple columnar epithelium resting on a delicate fibrovascular tissue (H&E,  $\times 200$ ). B : The lining epithelium containing PAS-positive mucin material (PAS,  $\times 400$ ). C : The tumor cells show positive immunohistochemical reaction for cytokeratin ( $\times 400$ ). D : The tumor cells also show focally positive immunohistochemical reaction for CEA ( $\times 400$ ).

were consistent with endodermal cyst, type A, according to the Wilkins-Odom classification<sup>22</sup>.

## Discussion

Embryogenesis of the endodermal cyst is not known with certainty. A number of hypothesis have been postulated to explain the embryogenesis of endodermal cysts of the central nervous system<sup>3,6</sup>. The predominant proposal has been that endodermal cyst originate from the faulty separation of ectodermally derived spinal cord and endodermally derived foregut during closure of the neurenteric canal in the third week of embryonic life<sup>3,6,11,14,15,19</sup>. The rostral closure of the notochord by the mesenchyme forms the clivus, so it is possible to explain the pathogenesis of posterior fossa endodermal cyst by this concept. Therefore, most endodermal cyst located in the midline<sup>3,11,14,15</sup>.

Epithelial cyst of the CNS can be subdivided into endodermal and neuroepithelial cyst according to the proposed cellular origin<sup>10</sup>. Endodermal cyst can be differentiated from neuroepithelial cyst, histologically. In endodermal cyst, cyst wall is lined with simple columnar or cuboidal epithelium with mucin producing droplet<sup>7</sup>. Presence of the basement membrane is also the characteristics of endodermal cyst<sup>22</sup>. Positive immunoreactivity of a tissue for CEA suggest an endodermal origin. Immunoreactivity for S-100, GFAP favors the ectodermal origin. Epithelial markers such as cytokeratin and epithelial membrane antigen (EMA) have been shown to be present in endodermal cyst in contrast to those of neuroepithelial origin<sup>4,10,12,20</sup>.

Wilkins-Odom<sup>22</sup> classified intraspinal neurenteric cyst into

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three types : Type A cysts are lined by a single pseudostratified or stratified cuboidal or columnar epithelium with or without cilia lying on a basement membrane. Type B cysts have some of other elements found in the gastrointestinal or respiratory tracts (such as mucous gland, serous gland, smooth muscle, and striated muscle). Type C cysts have ependymal and glial elements.

In our case, the presence of basement membrane, positive staining for PAS, which was indicating secretory activity, and immunoreactivity for CEA and cytokeratin favor the diagnosis of the lesion as an endodermal cyst. In addition, there was no smooth muscle tissue, ependymal cells and glial tissue in the cyst wall. These findings were consistent with endodermal cyst, type A, according to the Wilkins-Odom classification.

Intracranial endodermal cyst is very rare, but when it occurs it has a predilection of posterior fossa. Benjjani, et al.<sup>3)</sup> reviewed 49 cases of posterior fossa endodermal cyst. It occurred in patient at all age (newborn infant to 77-year-old woman) with peak incidence in the third and fourth decades. The most frequent complaint of patients with endodermal cyst of the posterior fossa was headache, and followed by gait disturbance, upper extremity weakness and sensory complaints. Intermittent leakage of the cystic contents may cause recurrent aseptic meningitis. Although spinal endodermal cyst are frequently associated with congenital bony and CNS anomalies, intracranial endodermal cyst are rarely associated with them<sup>1,2,5,7)</sup>.

In our case, headache was the chief complaint. Even though the brain stem was compressed and kinked by cystic mass, neither any brain stem sign nor lower cranial nerve deficits were developed. And there was no associated bony and CNS anomalies. MRI is the imaging modality of choice, providing valuable information on the nature of the cyst<sup>9)</sup>. The cyst contains fluid that may be clear, mucoid, or brown as a result of previous hemorrhage<sup>14)</sup>. Depending on the protein content of the cystic fluid, the signal intensity can be variable<sup>17)</sup>.

In our case the cyst was avascular and showed low density on CT without contrast enhancement. MRI showed a similar signal intensity to CSF on T2-weighted and T1-weighted images. However, the FLAIR images demonstrated heterogeneous high signal intensity within the cyst, most likely because of the presence of the proteinaceous or other hydrophilic macromolecular materials interacting with cystic fluid. We should include endodermal cyst, in the differential diagnosis of epidermoid or arachnoid cyst.

## Conclusion

**I**ntracranial endodermal cyst is a rare congenital lesion. It can be removed successfully without difficulty if we choose

appropriate approach. Endodermal cyst should be included in the differential diagnosis of intracranial cyst.

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