Giant petroclival endodermal cyst with xanthogranulomatous changes

Case report

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Endodermal cyst is a rare developmental cyst of the CNS, such as a Rathke cleft and colloid cyst lined by columnar epithelium of presumed endodermal origin. Intracranial endodermal cysts are rare, and most are found in the posterior fossa. The authors report a case of petroclival endodermal cyst with extensive bone destruction. A 12-year-old boy presented with transient facial weakness and headache. Imaging revealed a 3 × 3 × 4-cm, partial rim, enhanced cystic lesion in the petroclival area that was isointense on T1-weighted imaging and hyperintense in T2-weighted imaging. The cyst wall was partially removed and the cyst was obliterated using a lateral approach. Histological examination revealed ciliated, simple-to-pseudostratified cuboidal epithelium with a basement membrane that was consistent with an endodermal cyst, with the rare finding of xanthogranulomatous changes. (http://thejns.org/doi/abs/10.3171/2013.6.PEDS1362)

**KEY WORDS** • endodermal cyst • petroclival cyst • neurenteric cyst • xanthogranulomatous change • oncology

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**Case Report**

**History and Examination.** A 12-year-old boy presented with a 3-month history of right facial weakness and headache. No tinnitus, dizziness, or other neurological symptoms were reported by the patient. The neurological examination revealed a mild facial paresis of House-Brackmann Grade II and a mild sensorineural hearing loss of the right ear, with poor SDS (Fig. 1).

A skull base CT scan revealed a homogeneous, low-density, space-occupying lesion in the right petroclival area extending into the cerebellopontine angle. The mass had a well-defined and lobulating contour with destruction of clivus and petrous apex. Findings on MRI studies also suggested that the mass mainly involved the right inferior pons at the origin of the seventh and eighth cranial nerves. The mass showed hyperintensity on the T2-weighted image and isointensity on the T1-weighted image, with a thin marginal enhancement (Fig. 2). Because of the pattern of bone destruction and the location of the mass, the preoperative diagnosis of neurogenic tumor or congenital cystic lesion was made.

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Abbreviation used in this paper: SDS = speech discrimination score.
Atypical case of endodermal cyst with a rare location

**Intraoperative and Pathological Findings.** Because the jugular bulb was positioned very high in this patient, the mass could be approached between the internal carotid artery and the jugular bulb only after canal wall down mastoidectomy and anterior rerouting of the facial nerve. The mass was cystic and its contents were yellow, gelatinous, and semisolid (Fig. 3). After drainage of the contents, the cyst wall was carefully peeled from the adjacent structures and removed as much as possible. The cavity was filled with small pieces of abdominal fat. The mastoid cavity was also obliterated with abdominal fat and the external auditory canal was closed in a blind-sac fashion. Pathological examination with H & E staining of lesion sections revealed ciliated, simple-to-pseudostratified cuboidal epithelium with basement membrane and xanthogranulomatous changes. This confirmed the diagnosis of endodermal cyst (Fig. 4).

**Postoperative Course and Follow-Up.** The postoperative course was uneventful. A postoperative audiogram showed a conductive hearing loss, with improvement of SDS. A postoperative MRI study of the internal auditory canal revealed a well-obliterated cavity without accumulated fluid 3 months after the operation (Fig. 5).

**Discussion**

Endodermal cysts in the CNS are rare benign lesions that are lined by columnar or cuboidal epithelium of a presumed endodermal origin.2–4,6,10 Endodermal cyst nomenclature has been confusing because of the poorly defined pathogenesis; the variety of names includes neurenteric cyst and enterogenous cysts.3,4,6,10 These lesions are most frequently encountered in the lower cervical and upper thoracic spine.2–4,6,10 Intracranial endodermal cysts are even rarer. When this occurs, the cysts are frequently located in the posterior fossa.3,4,6,7,10,12,13 The most common locations are anterior to the brainstem or within the fourth ventricle. The cysts have also been found in the cerebellopontine angle and clivus, but endodermal cysts in the petroclival area are extremely rare.2,4,7,10,13

Embryogenesis of the endodermal cyst is not known with certainty. The common belief is that they originate from the failed dissolution of the transient neurenteric canal between the foregut or the respiratory buds and the notochord during notochordal development. Because the rostral closure of the notochord by the mesenchyme forms the clivus, it is possible to explain the pathogenesis of posterior fossa endodermal cyst occurring at the midline. The other theory is that supratentorial endodermal cyst of the midline may originate from the Seessel pouch, which occurs transiently between the rostral end of the notochord and the Rathke pouch at 42 days of fetal life. Although this pouch, which is composed of endodermal cells, regresses in most cases, remnants may give rise to endodermal cyst according its location.1,2–5 If they are present in the intrasellar region, Rathke cleft cysts can develop; if they are present in the suprasellar region, ectopic Rathke cleft cysts or suprasellar endodermal cysts can form; and in the third ventricle, colloid cysts may occur.2,3 There are no pathological or immunohistochemical criteria for a distinction between Rathke cleft cyst, colloid cyst, and neurenteric cyst; this is why the initial pathological report of the presenting case was ectopic Rathke cleft cyst.4,5

Diagnosis of an endodermal cyst is based on the histological findings. The wall of the cyst consists of a single layer of ciliated or nonciliated columnar or cuboidal epi-
epithelium, with a basement membrane and mucin-produc-
ing droplet. In our case, the results of pathological in-
vestigation revealed ciliated, simple-to-pseudostratified
cuboidal epithelium with xanthogranulomatous changes.
A xanthogranulomatous reaction occurs very rarely in
gen benign, noninflammatory cystic lesions. Preoperative
diagnosis of intracranial cyst is important, because the
operative management may vary from complete excision

to simple drainage. However, it is difficult to differentiate
endodermal cyst from other cystic lesions of the petro-
clavial fissure in making a correct preoperative diagnosis.

Imaging of an endodermal cyst usually reveals a cystic
mass with no enhancement after the addition of contrast
medium. The signal intensity characteristics vary depend-
ing on the protein content of the cysts. Most cysts are pro-
teinaceous, are iso- to slightly hyperintense on T1-weight-
ed images, and are iso- to hyperintense on T2-weighted
images. In addition, they are hyperintense on FLAIR im-
egages and may show mild restriction on diffusion-weighted
images. In general, the absence of contrast enhance-
ment of the cyst wall and of bone destruction is useful for
distinguishing endodermal cysts from other cystic lesions
such as cystic schwannoma and cystic meningiomas.

Therefore, a cystic tumor was included in the differential

Fig. 2. Preoperative skull base axial CT scan (A) revealed a ho-
mogeneous, low-density, and well-defined petroclival mass with bone
destruction, and the mass involved the right inferior pons at the origins
of the seventh–ninth cranial nerves. The tumor size was approximately
30 × 30 × 40 mm. Isointensity was revealed on the axial T1-weighted
image (B), with peripheral thin rim enhancement on the coronal T1-
weighted image (C), and hyperintensity on the T2-weighted image (D).

Fig. 3. The cystic mass was drained and the contents consisted of
yellow, gelatinous, semisolid material. The cyst wall opening was lo-
cated between the internal carotid artery (arrowhead) and the jugular
bulb (asterisk).

Fig. 4. Photomicrographs showing columnar epithelium with base-
ment membrane and xanthogranulomatous changes with cholesterol
clefs. H & E, original magnification x400 (left) and x200 (right).

Fig. 5. Postoperative MRI and audiogram data. The scans reveal
fat obliteration of the cyst without fluid collection on both the T1- and
T2-weighted images. Postoperative audiogram revealing a conductive
hearing loss with improvement in SDS.
Atypical case of endodermal cyst with a rare location
diagnosis because our case showed bone destruction and rim enhancement, which are rare in intracranial cysts. A thin peripheral rim of enhancement could be attributed to inflammatory change or squamous metaplasia of the cyst wall.

The differential diagnosis includes epidermoid cyst and arachnoid cyst. Epidermoid cysts are usually restricted on diffusion-weighted images, whereas arachnoid cysts are not. On FLAIR images, arachnoid cysts are typically suppressed completely. In this case, both FLAIR images and diffusion-weighted images show hyperintensity.

Surgical treatment of an endodermal cyst is widely accepted, but the goal of excision has been controversial. Complete excision of the cyst wall has been emphasized to prevent recurrence, but this approach may cause surgical difficulties and tissue scarring. Subtotal resection of the cyst wall may be a reasonable approach if there is a significant risk for damage of vulnerable structures. Because the access to the cyst was limited in our case due to the high jugular bulb, total resection of the cyst wall was not possible with the lateral approach. Careful follow-up with imaging is essential because there is risk of cyst recurrence.

Conclusions
We report an endodermal cyst located in the petroclival area accompanied by bone destruction and rim enhancement of the cyst wall. The atypical location and rare presentation of this case may be helpful when making the differential diagnosis of intracranial cysts.

Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Choi. Drafting the article: Choi. Critically revising the article: Choi. Reviewed submitted version of manuscript: Cho. Seol. Approved the final version of the manuscript on behalf of all authors: Cho.

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