

Endodermal cyst in pineal region: Rare location

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Abstract

Background: Pineal tumors are very uncommon intracranial lesions, and endodermal cysts in this location are extremely rare.

Case Description: A 49-year-old right-handed female presented with 3 weeks history of progressive dizziness and imbalance. Imaging studies showed 1.8 cm × 1.7 cm × 1.8 cm pineal lesion with small enhancing mural component displacing ventrally the quadrigeminal plate and narrowing of aqueduct of Sylvius without hydrocephalus. In addition, she was found with small interhemispheric lipoma, and small posterior falx possible meningioma. Cerebrospinal fluid markers obtained by lumbar puncture were all negative. She underwent tumor resection, and final pathology reported endodermal cyst. No new deficits were encountered, and her gait imbalance improved significantly by 3 months follow-up.

Conclusions: With evidence of enlargement or symptomatic pineal lesions, surgical consideration is necessary. Among pineal lesions, endodermal cysts are extremely uncommon and although benign pathology, long-term follow-up is advised due to unknown chronic behavior.

Key Words: Endodermal cyst, pineal region, pineal tumor

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INTRODUCTION

Pineal region tumors are very uncommon, accounting for <1% of adult brain tumors. To the best of our knowledge, among the multiple lesions in the differential diagnosis, endodermal cyst has not been reported only within the pineal region.^[7,8,10] In this paper, we intend to describe the first case of a pineal region endodermal cyst in addition to its clinical and surgical management. Endodermal cysts are rare benign lesions, also called neurenteric cyst or enterogenous cyst, lined by columnar or cuboidal epithelium of primitive endodermal cells.^[3] These lesions are more frequently reported in the cervical and thoracic spine, and less frequently in intracranial locations, accounting for about 0.01% of central nervous system tumors.^[11] They can produce a local mass effect and become symptomatic depending on their location. The intracranial locations reported are more

frequently within posterior fossa in fourth ventricle, cerebellopontine angle, ambient and quadrigeminal cistern, around brainstem, and less frequently in supratentorial compartment.^[1-5,9,11] The embryogenesis of endodermal cyst is not well-understood, and, like Rathke cleft and colloid cysts, they all are considered the result of failed separation between notochord and neurenteric canal.^[6]

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CASE DESCRIPTION

Case presentation

A 49-year-old right-handed female, chronic and heavy smoker, with high blood pressure who was admitted to medicine service with 3 weeks of progressive dizziness and gait imbalance confirmed on physical examination. Rest of her neurological examination was intact. A lumbar puncture was obtained, and then she was managed with a low dose of steroids, and acetazolamide for 2 weeks until obtaining cerebrospinal fluid (CSF) results. She continued to be symptomatic, and CSF results showed chorionic gonad beta-Human Chorionic Gonadotropin (bHCG), carcinoembryonic antigen, and alpha-fetoprotein within normal limits.

Imaging findings

The head computed tomography (CT) scan showed hyperdense pineal lesion. The brain magnetic resonance imaging (MRI) showed a partially cystic pineal region mass of 1.8 cm of major diameter containing T1 hyperintense proteinaceous debris, and T1 isointense/T2 hyperintense enhancing mural nodule within the right lateral aspect of the lesion, no restricted diffusion, and increased signal on fluid-attenuated inversion recovery (FLAIR). The lesion splayed the internal cerebral veins laterally, and displaced tectum and superior colliculi inferiorly, producing a narrowing of aqueduct of Sylvius [Figure 1]. In addition, small (0.7 mm) homogenous enhancing midline mass in posterior interhemispheric fissure was found (possible meningioma), and small interhemispheric lipoma.

Surgical procedure

Surgery was recommended due to worsening of gait imbalance, lack of improvement with steroids and acetazolamide, relatively large size of the lesion with mass effect on midbrain, and for diagnosis due to atypical imaging features. A right frontal external ventricular drain was placed in anticipation of possible postoperative hydrocephalus. The patient was placed in a semi-sitting position and midline suboccipital craniotomy was performed using burr holes above the transverse sinus exposing torcula. The dura was opened in a Y fashion, and 4-0 nylon suture were placed to retract transverse

sinus and torcula superiorly. The favorable tentorial angle facilitated the supracerebellar infratentorial approach, using microsurgical techniques, stereotactic navigation and electrophysiology monitoring (somatosensory evoked potentials and brainstem auditory evoked potentials). The capsule of the tumor was solid, yellowish, and brown with thick and yellow gel-like internal component. The capsule was subtotally resected, leaving a small segment with tight adhesions to the right internal cerebral vein. After lesion resection, the anterior and superior aspect of the third ventricle was visualized. The dura was closed in a watertight fashion with 5-0 prolene and surgical glue using the surgical microscope.

The postoperative MRI showed gross total resection [Figure 2] of the lesion and final pathology reported endodermal cyst [Figure 3]. The diagnosis was based on histopathology findings with epithelial lining in the cyst wall varying from cuboidal to pseudostratified, composed with cells with prominent cilia and with immunohistochemistry with the presence of periodic acid-Schiff (PAS) positive diastase resistant material, and negative for glial fibrillary acidic protein and S100.

The ventricular drain was kept clamped without any signs of hydrocephalus on postoperative images, removed on postoperative day 2, and discharged home on a postoperative day 4. No new deficits were encountered, and her gait imbalance improved significantly by 3 months follow-up.

DISCUSSION

The preoperative imaging findings in this patient as described above are not specific, and atypical for conventional cystic lesions in this location due to the enhancing mural component. The typical endodermal cyst shows hyperdensity on the head CT scan, as well as mixed iso- and hyper-intensity on T1 and T2 MRI, which are considered caused by high content of proteinaceous debris. FLAIR images show hyperintensity, and mild or no restriction on diffusion weighted images, which differs from significantly restricted diffusion of epidermoid cysts. There is usually no contrast enhancement on endodermal cysts which differs from our present case.

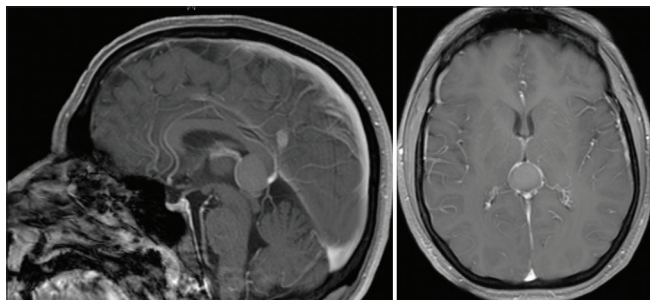


Figure 1: Preoperative brain magnetic resonance imaging

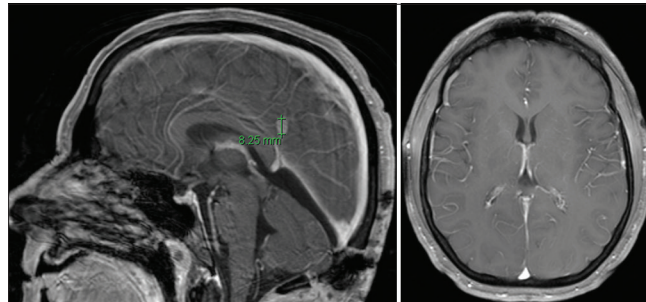


Figure 2: Postoperative brain magnetic resonance imaging

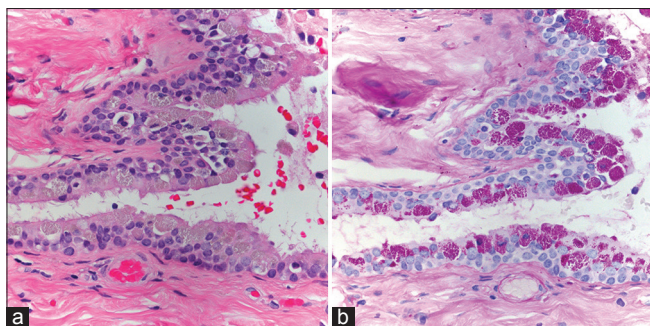


Figure 3: (a and b) Collapsed cyst wall with pseudostratified epithelial lining composed of ciliated cells (a) with periodic acid-Schiff positive diastase resistant inclusions (b) (a: H and E, 400; b: PAS-D, ×400)

In general, lack of contrast enhancement of the cyst wall, and of bone destruction is useful for distinguishing endodermal cysts from other cystic lesions such as schwannomas or cystic meningiomas.^[4,6] In retrospect, the concomitant presence of a small interhemispheric lipoma, and an additional small posterior falx possible meningioma could have raised the possible diagnosis of a developmental lesion. There are no histopathological or immunohistochemical criteria for a clear distinction between Rathke cleft cyst, colloid cyst, and endodermal cyst.^[6] In general, endodermal cyst has a wall of single layer of ciliated or nonciliated columnar or cuboidal epithelium, with basement membrane and PAS diastase-resistant material that breaks down the high proportion of carbohydrates macromolecules in mucin component. The findings may resemble colloid cyst although the location in the pineal region excludes the diagnosis. The histological features are not consistent with a pineal cyst, which typically is a glial cyst without epithelial lining. The present case differs from previously reported cases because the lesion is located only within pineal region, mimicking an intrinsic pineal tumor, and not arising from quadrigeminal cistern and compressing pineal area. The relevance of current report resides that differential diagnosis in pineal region lesions should also include endodermal cysts. Given the rarity of these lesions and unknown lesion behavior in the long-term, with either complete or partial resection, subsequent imaging follow-up is advised. Postoperative MRI at 3 and 6 months did not show recurrence, and follow-up plan will include imaging studies 1 year after surgery, and subsequently every 2 years.

CONCLUSIONS

Endodermal cyst is another differential diagnosis that should be considered within the pineal region. The ideal treatment of symptomatic endodermal cyst is a total resection of the lesion, including all cyst contents to decrease the possibility of chemical meningitis, as well as the complete capsule resection to avoid the risk of recurrence. In locations where the capsule is densely adhered to important neurovascular structures, a partial resection is reasonable. Long-term follow-up is advised.

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Conflicts of interest

There are no conflicts of interest.

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