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Image Report

Recurrent neurenteric cysts compressing the brainstem

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ABSTRACT

Background: Neurenteric cysts, also referred to as endodermal cysts and enterogenous cysts, are rare lesions of the neuroaxis occurring most frequently within the spinal cord and rarely intracranially. In the literature describing these lesions, examples of intraoperative imaging and cytology preparations are rare to non-existent.

Case Description: Here, we describe a case of a recurrent posterior fossa neurenteric cyst compressing the brainstem in a 47-year-old female and causing incontinence and progressive quadriparesis. Intraoperative findings and cytologic and histologic features are presented.

Conclusion: Neurenteric cysts are generally considered to be benign and slow-growing though recurrence is common. This case of a recurrent neurenteric cyst is illustrated by intraoperative macroscopic and cytologic images.

Keywords: Cytology, Endodermal cyst, Enterogenous cyst, Neurenteric cyst, Posterior fossa epithelial cyst

INTRODUCTION

Neurenteric cysts, also referred to as endodermal cysts and enterogenous cysts, are rare lesions of the neuroaxis occurring most frequently within the spinal cord and rarely intracranially. In the literature describing these lesions, examples of intraoperative imaging and cytology preparations are rare to non-existent.

CASE REPORT

A 47-year-old female with a history of posterior fossa neurenteric cysts presented with incontinence and progressive quadriparesis. In 1994, she experienced imbalance and falls, and head imaging revealed a 7 cm posterior fossa cyst which was resected. Another resection was performed in 1997. She developed hydrocephalus, resulting in a ventriculoperitoneal shunt placement with subsequent revisions due to infections and clotting. In the 2 months before presentation, she developed incontinence and progressive quadriparesis. Magnetic resonance imaging showed multiple, expanding, and recurrent neurenteric cysts in the left cerebellomedullary angle and left preportine cistern compressing the medulla [Figure 1a]. Surgical resection of the cysts was performed, requiring dissection of cyst capsules from the posterior inferior cerebellar artery and cranial nerves. The cysts were clustered like grapes and

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Figure 1: Neurenteric cysts compressing the brainstem. Magnetic resonance imaging demonstrated multiple, nonenhancing, multiolobulated, expanding, and extra-axial recurrent neurenteric cysts compressing the medulla (a). An intraoperative image showing the cysts which were adherent to but separable from the medulla and upper cervical spinal cord (b).

were pale, white, and adherent to but separable from the medulla and upper cervical spinal cord [Figure 1b].

Intraoperative squash preparations demonstrated sheets of epithelial cells with honeycomb architecture, distinct cell borders, and abundant cytoplasm within a background of mucin [Figure 2a]. Permanent sections revealed neurenteric cysts lined by columnar cells with basally located nuclei with underlying basement membrane and connective tissue stroma. Abundant colloid material was present [Figure 2b]. The patient was later discharged to a skilled nursing facility in stable condition.

DISCUSSION

Neurenteric cysts, also referred to as endodermal cysts and enterogenous cysts, are rare lesions comprising 0.7–1.3% of all spinal cord tumors and occurring less frequently within the posterior fossa and cerebral hemispheres.^[1,3] Headache is the most common presenting symptom for intracranial neurenteric cysts with extremity weakness and gait disturbance seen less frequently.^[1] These cysts are lined by columnar or cuboidal epithelium with goblet cells. The treatment often involves resection and recurrence is common.^[4] While neurenteric cysts are generally considered to be benign and slow-growing, rare reports of posterior fossa neurenteric cysts exhibiting rapid expansion or malignant transformation often in the form of adenocarcinoma have been reported.^[2,5]

CONCLUSION

Neurenteric cysts are generally considered to be benign and slow-growing though recurrence is common. This case of recurrent intracranial neurenteric cysts includes illustrations of intraoperative macroscopic and cytologic images.



Figure 2: Cytology and histology of the neurenteric cysts. Intraoperative squash preparation stained with hematoxylin and eosin revealed sheets of epithelial cells with honeycomb architecture, distinct cell borders, and abundant cytoplasm within a background of mucin, $\times 200$ (a). Hematoxylin and eosin-stained permanent sections showing cyst wall lined by columnar epithelium with underlying basement membrane and connective tissue stroma, $\times 200$ (b).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Conflicts of interest

There are no conflicts of interest.

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