**INTRODUCTION**

Neurenteric cysts are rare benign neoplasms with a mucin-secreting intestinal or respiratory epithelium, thought to originate from embroyologic dysgenesis of the notochord or neurenteric canal. These neoplasms most commonly present in the mediastinum. They also occur in the spine (0.3-0.5% of spinal tumors) and CNS (0.1% of CNS tumors), and rarely in the clivus.¹

Neoplasms of the clivus most frequently represent metastases, with breast carcinoma metastases more common than lung or prostate. Other clival lesions include chordoma, chondrosarcoma, lymphoma, nasopharyngeal carcinoma, cholesteatoma, fibrous dysplasia, and hemangioma.²,³

This report summarizes a case of clival neurenteric cyst with systematic review of the literature.

**RESULTS**

Including the present case, five cases of clival neurenteric cyst were identified.²⁻⁵ Two patients were female and three were male. Patients ranged in age from five to 79 years, with a mean of 33.8 years; two were under twelve years of age.

Adult presentations included several weeks of headache, new onset nausea and vomiting, and chronic daily headache with an asymptomatic clival lesion. One child presented with visual change, the other with recurrent retropharyngeal abscess.

All patients underwent surgical resection of the cyst, one by direct transnasal approach, two by endoscopic transnasal approach, and two by transoral approach. Our patient experienced an asymptomatic recurrence. Follow up ranged from 12-48 months, with a mean duration of 22.3 months.

**DISCUSSION**

The broad differential diagnosis of a clival mass includes metastasis, most commonly breast and less frequently lung and prostate; primary malignancy, such as chondrosarcoma, lymphoma, and nasopharyngeal carcinoma; benign neoplasms including chordoma and hemangioma; and cholesteatoma. Neurenteric cysts are a rare congenital benign neoplasm of the clivus, and more commonly present intracraniially or in the spine.

Neurenteric cysts appear on MRI as smooth, well-demarcated lesions, typically low-intensity on T1 and high-intensity on T2, and do not enhance with contrast.²,⁴

The majority of patients presented with acute visual symptoms or headaches. One pediatric patient presented with a retropharyngeal abscess. The current patient presented with 10 years of headache; the lack of acute symptoms and the failure of symptoms to resolve with surgery suggests that his neurenteric cyst was a truly incidental finding on imaging, rather than the etiology of his headaches.

The reasons for recurrence in our patient are unclear, but could be perhaps attributed to insufficient stripping of the cyst membrane. No further intervention has been planned due to lack of symptoms.

**CONCLUSIONS**

Neurenteric cyst of the clivus is a rare benign neoplasm occurring in both pediatric and adult populations. Presentations vary from acute onset of neurologic or infectious symptoms to incidental findings on imaging. Prognosis following surgical resection is excellent. Although patients with clival neoplasms are more likely to present to a neurosurgeon, the otolaryngologist plays a role in transsphenoidal approaches to clival lesions. These findings should inform the differential diagnosis of these lesions.

**METHODS**

A systematic literature review was performed using the PubMed database, with search terms “neurenteric cyst,” “neurenteric cyst [MeSH],” “neuroenteric cyst,” “neuroenteric cyst [MeSH],” “enterogenous cyst,” “enterogenous cyst [MeSH],” “clivus,” “clival,” “posterior cranial fossa,” and “skull base” and time period 1950 through December 31, 2014. The search was limited to English peer-reviewed literature.

Inclusion criteria required pathologic description consistent with a neurenteric cyst, and anatomical position within the clivus. The bibliographies of relevant papers were further reviewed for other cases. Variables extracted are listed in Table 1.

**REFERENCES**