Introduction

Ependymal cells are neuroglial cells that line the ventricular system and central canal of the spinal cord. Glioependymal cysts lined by ependymal cells are postulated to arise from ectopic rests of primitive neuroglial tissue. These cysts can occur anywhere along the neuraxis; however, the cerebellopontine angle is not a usual site for these. We present an unusual case of a glioependymal cyst in the cerebellopontine angle in an adolescent boy presenting with hearing loss and facial palsy.

Objectives

Present an additional differential diagnosis to consider when evaluating pediatric patients presenting with cerebellopontine angle mass.

Methods

Retrospective review of patient’s clinical, radiographic, and histopathologic records as well as literature review.

Clinical Presentation

A 13-year-old boy presented to our tertiary neurotology clinic with progressive left-sided hearing loss for one year and three months of progressive left-sided facial weakness (House Brackmann IV). Otolologic exam was unremarkable. Audiogram showed severe to profound sensorineural hearing loss.

Radiologic Studies

MRI

MRI of the Brain and IAC identified a large extra-axial cystic septated mass measuring 5.1 x 3.6 x 4.3 cm at the left cerebellopontine angle involving the internal auditory canal with enhancement of the margins of the mass with posterior dural tail.

Operative Technique

The patient underwent translabyrinthine approach for excision of the mass. Beyond the porus, the facial nerve was splayed in the substance of the mass and was unidentifiable. The trigeminal was found to be significantly compressed but remained intact. He had gross total removal of the mass which was comprised of a number of cysts contained within a fibrous capsule.

Pathology Findings

Intraoperative frozen sections were suggestive of meningoia or schwannoma. Histo-pathology results confirmed the mass to be a glioependymal cyst composed of epithelial and spindle cells around a cyst that stained positively for GFAP and S100. Since there was disagreement on the final histopathological diagnosis the biopsy has been sent to an outside institution for further analysis.

Conclusion

The presentation of glioependymal cysts in the posterior fossa is rare, and their presentation in the pediatric population is exceptional. To our knowledge, there is only one case reported of a cerebellopontine angle glioependymal cyst occurring in an adolescent, and our case is the first presenting with facial paralysis. Although extremely rare, glioependymal cyst should be considered as a differential diagnosis in patients presenting with cystic mass at the cerebellopontine angle.

References