An Adult Patient with Initial Presentation of Myxopapillary Ependymoma at the Cerebellopontine Angle

Marcin Marciniak, BS[1]; Khatcher O. Margossian, PhD[1]; Akram Al-warqi, MD[1]; Kenneth Aldape, MD[2]; Mehmet Kocak, MD[1]; Lorenzo Munoz, MD[1]; Elias Michaelides, MD[1]; Michael S. Marshall, MD, PhD[1]

[1]Rush University Medical Center, Chicago, IL, USA

[2]National Cancer Institute, Bethesda, MD, USA

Background

Ependymomas are generally benign tumors of the central nervous system (CNS) that arise from ependymal cells, which are found lining the ventricles of the brain and the central canal of the spinal cord. Myxopapillary ependymomas (MPEs) are a subtype of ependymomas that most commonly occur in the lumbosacral spine and filum terminale. These tumors are not regularly found in the cerebellopontine angle (CPA). This study presents a rare case of a myxopapillary ependymoma presenting at the cerebellopontine angle in an adult male and reviews the current literature on this atypical finding.

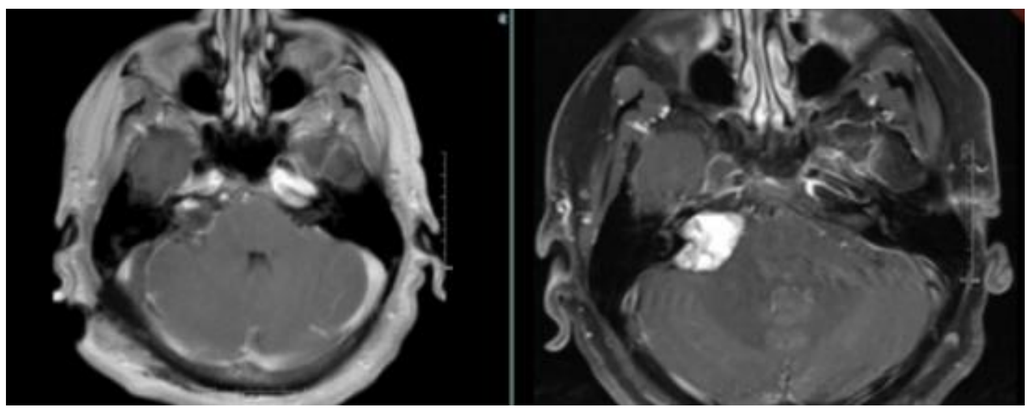

Figure 1. Axial MRI T1 Brain with contrast showing a right CPA mass post-operatively (left) vs pre-operatively with extension into the internal auditory canal (right).

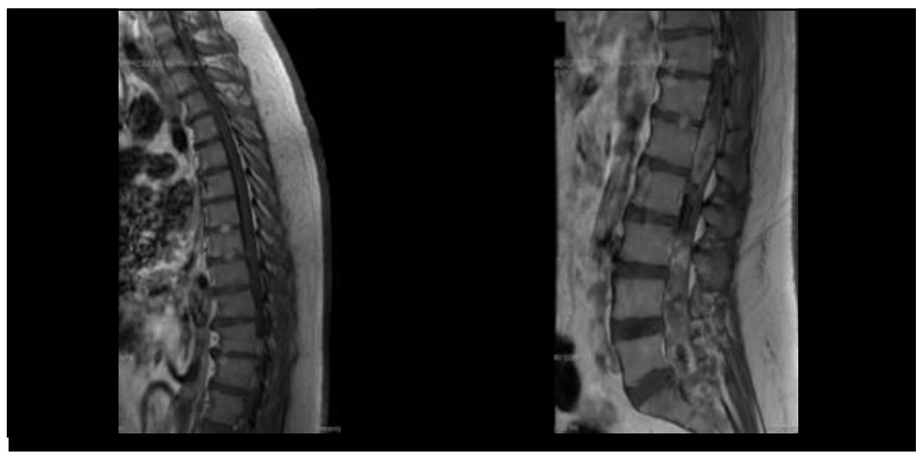

Figure 2. Sagittal MRI thoracic spine (left) and lumbar spine (right) showing enhancing lesions.

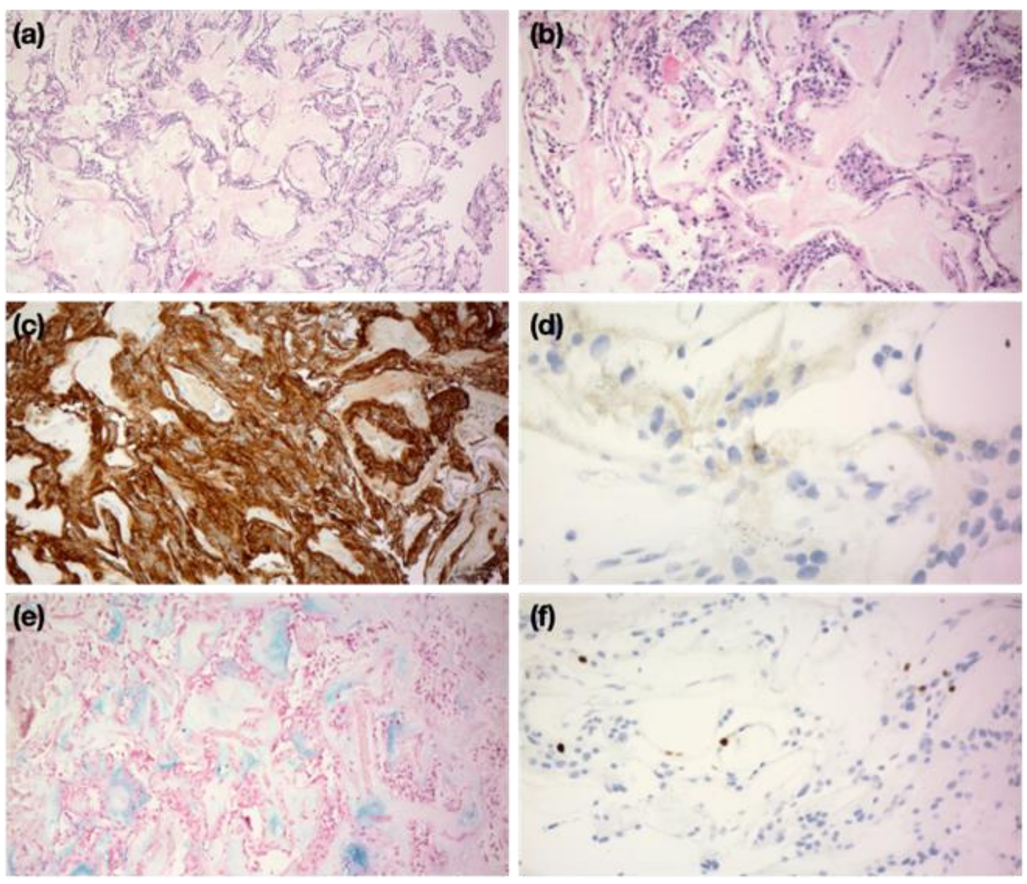

Figure 3: Histologic findings. Low power (a) and higher-power (b) views reveal a benign papillary neoplasm with a prominent myxoid appearance. GFAP (c) staining is diffuse positive in the tumor cells. EMA staining (d) reveals rare dot-like perinuclear positivity. Alcian blue staining (e) highlights the myxoid areas. MIB/Ki-67 staining (f) reveals a proliferative index of approximately 1-2%.

Case Description

A 44-year-old male originally presented to his primary care provider with the complaint of a clogged sensation in his right ear that began in April 2023. Notably, the patient also has a history of right tinnitus since 2020 and bilateral hearing loss. Magnetic resonance imaging (MRI) revealed a lobulated mass lesion at the right cerebellopontine angle measuring 2.3 x 2.2 x 2.0 cm and extending into the internal auditory canal. Based off the imaging, the leading diagnoses were a schwannoma or a meningioma. The tumor was excised through a right retrosigmoid resection in May 2024. Frozen section analysis intraoperatively showed findings consistent with vestibular schwannoma. Post-operatively, full histological analysis revealed that the tumor was a myxopapillary ependymoma, WHO grade 2. Due to the tumor's atypical location, there was concern for metastasis and a total spine MRI was performed which showed a heterogeneously enhancing lobulated lesion with cystic internal changes and a T2 hypointense rim occupying the spinal canal from T12 to S2, which was suggestive of myxopapillary ependymoma. There were additional enhancing areas at the C7 and T8-9 levels without significant cord compression. At post-op follow up visits, the patient noted slight right facial asymmetry, which was confirmed on physical exam. He also reported improvement in his right tinnitus and retained some right sided hearing. He denied lower extremity numbness, tingling, weakness, imbalance, or pain. He noted occasional urinary dribbling but no incontinence. The patient elected to treat the spinal lesions with proton therapy.

Conclusion

This case demonstrates the importance of maintaining a broad differential diagnosis when approaching skull-based tumors. The patient's myxopapillary ependymoma, first thought to be a schwannoma or meningioma based on imaging, highlights the need for careful evaluation of tumors in the CPA. Despite their rarity, myxopapillary ependymomas should be considered in the differential diagnosis for CPA lesions. If such a lesion is found in the CPA, we recommend a full CNS imaging study to rule out metastatic origins.

© Poster Template by Genigraphics® 1.800.790.4001 www.genigraphics.com