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Case Report

Vestibular schwannoma or tanycytic ependymoma: Immunohistologic staining reveals

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Abstract

Background: The cerebellopontine angle (CPA) is a common location for primary tumors, most often vestibular schwannomas, and also meningiomas, dermoids, and a host of other neoplasms. Our case report illustrates how radiologic and histopathologic presentations of an unusual variant of ependymal neoplasm can be diagnostically challenging and how accurate diagnosis can affect treatment protocols.

Case History: Our patient had a CPA mass that was a variant of ependymoma known as tanycytic ependymoma that mimicked vestibular schwannoma radiologically and during intraoperative pathologic examination. Diagnosis as a World Health Organization (WHO) grade II tanycytic ependymoma was supported by its appearance on evaluation of the permanent sections, its diffuse immunoreactivity for glial fibrillary acidic protein (GFAP), and the perinuclear dot-and-ring-like staining for epithelial membrane antigen (EMA).

Conclusions: Our patient's CPA mass initially believed to be a vestibular schwannoma on preoperative evaluation, surgical appearance, and intraoperative pathologic consultation was then correctly diagnosed as a WHO grade II tanycytic ependymoma on permanent histologic sections with the assistance of immunohistochemical stains, including EMA. After this definitive diagnosis, our patient's adjuvant treatment was adjusted. Earlier diagnosis could have provided guidance for goals of resection and prompt initiation of adjuvant treatment.

Key Words: Cerebellopontine angle, diagnosis, epithelial membrane antigen staining, tanycytic ependymoma

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INTRODUCTION

The cerebellopontine angle (CPA) is a common location for brain tumors, which account for 6-10% of all intracranial masses.[12] The most common pathologies, vestibular schwannomas and meningiomas, make up 80% and 10% of tumors in this location, respectively; primary cholesteatomas constitute 4-6% of CPA pathologies while the remaining are a heterogenous group of tumors.[12,17] Of these, approximately 0.3-2% of lesions consist of primary central nervous system tissue including medulloblastomas, astrocytomas, and ependymomas.[17]

With vestibular schwannomas as the most common CPA pathology, patients typically present with progressive asymmetric hearing loss that occurs during the course of several years. Other types of tumors show a different progression of symptoms (e.g. nystagmus, altered facial sensation, gait ataxia) and develop in a sequential order not indicative of vestibular schwannomas;[17] these other symptoms that suggest alternative pathology include visual changes, facial weakness, or dysphagia.[12] In this case report, our patient had a tanycytic ependymoma that mimicked a vestibular schwannoma in symptom profile, radiography, and surgical pathology. We describe how further pathologic investigation, including immunohistochemistry, revealed it to be a tanycytic ependymoma, thus affecting changes to adjuvant treatment.

CASE REPORT

A 57-year-old male referred to an otolaryngologist for a 12-month history of left-sided hearing loss accompanied by episodes of tinnitus also noted intermittent headaches/migraines during the last several years. A magnetic resonace imaging (MRI) of the head showed a 3 × 3.5 cm homogenous contrast-enhancing CPA mass extending into the internal auditory canal with resultant mass effect and edema involving the cerebellum, pons, and medulla [Figure 1]. Ventricular size appeared normal and without hydrocephalus. Given the mass was believed to be a vestibular schwannoma, he was referred for a neurosurgical consult. After obtaining a history, physical examination, and preoperative workup, the decision was made to proceed with resection of the mass.

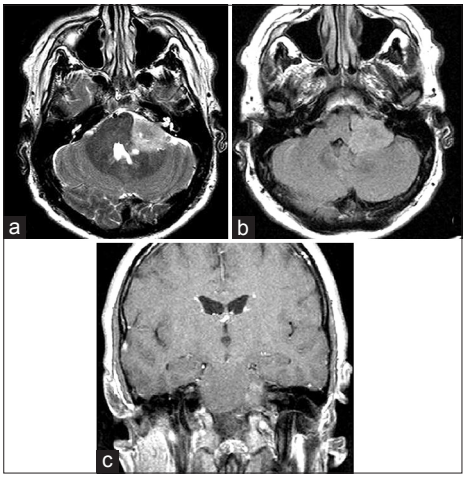

Figure 1: Tanycytic ependymoma. (a) T2-axial MRI showing a hyperintense lesion in the left CPA with extension into the internal auditory canal. The same lesion in axial (b) and coronal (c) views on T1-weighted MRIs. Postgadolinium enhancement showing a homogenously enhancing lesion extending from the ICA into the left CPA. There are clear mass effect shown on the brainstem and absence of hydrocephalus

The patient underwent a suboccipital transmeatal approach to expose the CPA and a left-sided partial C1 laminectomy to decompress a tonsillar herniation that became evident on opening dura. The tumor was identified and debulked. A specimen sent for intraoperative pathology consultation revealed cohesive groups of spindled cells on smear preparation, which was interpreted as consistent with a vestibular schwannoma. The remaining tumor was carefully dissected away from the facial nerve, trigeminal nerve, and brainstem superiorly, and the intracanilicular portion was removed from the internal auditory canal after drilling of the petrous ridge. The vestibulocochlear nerve was intentionally sectioned as a part of the resection of the tumor. The inferior margins of the tumor contacted the lower cranial nerves, and the anterior inferior cerebellar and posterior inferior cerebellar arteries were sharply dissected away sparing those nerves and vessels. The final portion in the pontomedullary junction was removed thus allowing cerebrospinal fluid (CSF) to flow from the fourth ventricle. Postoperatively, the patient developed left sigmoid sinus thrombosis, dysphagia, and urinary retention; all of these resolved by 1 month after surgery.

Permanent histologic sections showed areas of the tumor composed of spindled cells with an appearance of a schwannoma; however, for much of the tumor, the cells had a piloid appearance. Vague perivascular pseudorosettes were noted in some areas and no definitive Rosenthal fibers were seen. Based on the histologic appearance, the possibilities of a pilocytic astrocytoma and a tanycytic ependymoma were also considered. Immunohistochemical stains revealed the tumor to be diffusely positive for glial fibrillary acidic protein (GFAP) and S-100 protein. Staining for epithelial membrane antigen (EMA) revealed foci with dot- and ring-like perinuclear staining. A special stain for reticulin and an immunohistochemical stain for collagen IV did not show significant intercellular collagen within the tumor. Overall histologic appearance was most consistent with a tanycytic ependymoma (World Health Organization [WHO] grade II), and this finding was supported by immunohistochemical and special stains [Figure 2].

Before discharge, the pathologic diagnosis of a tanycytic ependymoma was finalized. Craniospinal imaging did not reveal any evidence of CSF dissemination of the tumor. The patient has since been followed by radiation/oncology team who is treating with fractionated radiation to the left CPA.

DISCUSSION

Our patient's CPA mass that was initially believed to be a vestibular schwannoma on preoperative evaluation, surgical appearance, and intraoperative pathology examination was later diagnosed as a WHO grade II

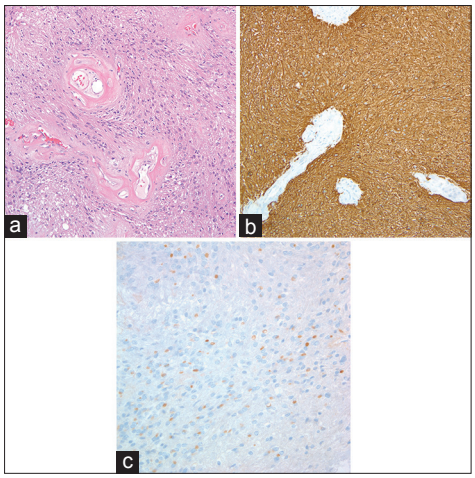

Figure 2: Histology provided accurate diagnosis of tanycytic ependymoma. (a) Permanent sections of the tumor show a piloid to spindled appearance with vague perivascular pseudorosettes (H and E, original magnification ×200). (b) Diffuse positive staining for GFAP (GFAP IHC; original magnification ×100). (c) Dot- and ring-like perinuclear staining for EMA (EMA IHC; original magnification ×400)

tanycytic ependymoma by permanent histologic evaluation, the assistance of immunohistochemical stains for GFAP, and EMA that showed dot-like and true ring-like perinuclear staining. Subsequently, our patient's adjuvant treatment was adjusted after the definitive diagnosis was obtained. However, earlier diagnosis could have provided guidance to goals of resection and prompt initiation of the adjuvant treatment.

Tanycytic-variant of ependymomas

The tanycytic-variant of ependymomas was first characterized in detail by Friede and Pollak as having an appearance more similar to a common ancestor of both ependymal cells and astrocytes known as ependymoglia or tanycytes.[6] Because of this derivation, tanycytic ependymomas were presumed to typically be found in the spinal cord where the raphe are abundant in ependymoglia and tanycytes. However, tanycytes have also been found scattered along the ventricular system and clustered in the area postrema, pineal gland, hypophysis, and lamina terminalis.[11] Case reports show that this variant, while rare, has appeared in various locations from the filum terminale to cervicomedullary junction as well as intracranially arising from locations in the parenchyma and intraventricularly.[4,6,9,14,15]

Radiologically, an ependymoma often appears as a heterogenous mass that is hypointense on T1 and hyperintense on T2, most often arising within the fourth ventricle.[1] Should the tumor enter the CPA, it is usually seen as a mass arising clearly from the fourth ventricle and exiting through the foramen of Luschka.[16] However, in several cases of ependymomas arising as extra-axial tissue, particularly within the posterior fossa,[3,7,18] the tumor often had a cystic component, which then appeared at the same intensity as the rest of the fluid in the image.[20] Tanycytic ependymomas can appear as homogenously enhancing with gadolinium administration.[10,15] This is a similar appearance to vestibular schwannomas, which tend to be homogenous appearing masses particularly after administration of gadolinium, and may also appear as hyperintense on T2-weighted imagery.[16] To further confound a preoperative diagnosis, vestibular schwannomas larger than 2-3 cm can also appear as a heterogenously intense cystic masses.

The histologic appearance of an ependymoma is typically depicted as a moderately to highly cellular tumor having true ependymal rosettes or, more commonly, perivascular pseudorosettes. These tumors have cells that may display either glial or epithelial characteristics.[11] In contrast, the tanycytic ependymoma has only low-to-moderate cellularity and is composed predominantly of bipolar cells with long processes that have a spindled to piloid appearance – characteristics that can mimic other tumor types.[10] For example, the piloid appearance of the bipolar cells with particularly long processes could lead to an inclusion of pilocytic astrocytoma in the differential.[5] Intraoperatively, diagnostic uncertainty can also occur secondary to the spindled-type characteristics of the cells reminiscent of schwannomas.[5] In these cases, immunohistochemical staining can help to provide a definitive diagnosis. Tanycytic ependymomas tend to have diffuse positive staining for GFAP and vimentin with variable S-100 staining, whereas schwannomas commonly have strong, diffuse S-100 staining, and are typically negative for GFAP.[10] As ependymal neoplasms, tanycytic ependymomas may display dot- and ring-like perinuclear staining for EMA and represent intracytoplasmic microrosettes.

Management

Surgery with a goal of gross total resection (GTR) has been the workhorse for managing both ependymomas and vestibular schwannomas for many years. However, refinements in stereotactic surgery have prompted revisiting which treatment course is best, including new data in recent years regarding management of pediatric ependymomas. In general, although GTR shows excellent outcomes for infratentorial or WHO grade III tumors, subtotal resection (STR) with radiation therapy can achieve even better outcomes than GTR alone.[2,19]

The role of radiation in management of vestibular schwannomas is highly debated, particularly for small tumors. Resection is undoubtedly the preferred primary modality for tumors >3 cm in diameter or 10 cm^3, particularly with brainstem mass effect or compression of the fourth ventricle.[8,13] However, debate continues about which treatment modality is best for tumors <3 cm given that stereotactic radiation offers similar progression-free survival with limited side effects

compared with surgery.[8,13] Yet, these data are limited by short follow-up and few studies of the long-term adverse effects of radiation exposure.[8]

CONCLUSIONS

This case illustrates an unusual variant of ependymal neoplasm, a tanycytic ependymoma, presenting as a CPA mass that was difficult to distinguish from a vestibular schwannoma on preoperative evaluation, surgical appearance, and intraoperative pathology. In our patient, a correct diagnosis was only reached on permanent histology with the support of special and immunohistochemical stains. This entity should be considered in the differential diagnosis of CPA masses because the potential goals of resection and need for adjuvant treatment may differ from that of a schwannoma.

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