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## Cerebellopontine angle astrocytoma producing Bruns nystagmus mimicking vestibular schwannoma



Cerebellopontine angle (CPA) extra-axial lesions (e.g., classically the vestibular schwannoma) may cause a characteristic neuro-ophthalmologic sign known as the *Bruns nystagmus*, a gaze-evoked horizontal nystagmus when looking away from the lesion and a gaze-paretic type nystagmus when looking toward the lesion. In contrast, intra-axial exophytic CPA astrocytomas are rare and, to our knowledge, have not been reported in the English-language ophthalmic literature previously as a cause of Bruns nystagmus. Clinicians should be aware that an exophytic astrocytoma involving the CPA may mimic the clinical and radiographic features of the more common CPA vestibular schwannoma.

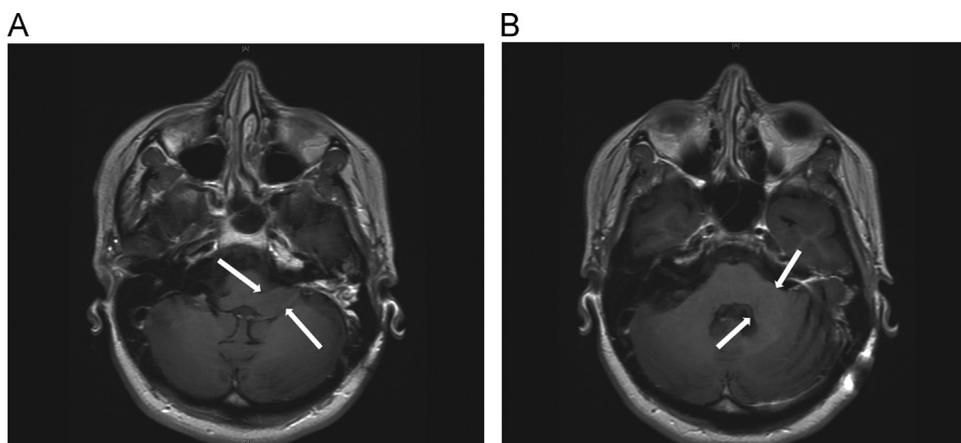
### CASE REPORT

An 18-year-old white female presented with progressive unilateral hearing loss and impaired coordination. Past medical, surgical, family, social, and medication histories were noncontributory. She first noted left-sided hearing loss a year prior to her presentation, but otolaryngology evaluation and

audiology testing were normal at that time. Progressive oscillopsia, diplopia, poor coordination, nystagmus, and imbalance then developed over the next several months. Magnetic resonance imaging (MRI) of the brain revealed a non-enhancing, hyperintense intra-axial mass in the left middle cerebellar peduncle approximately 1.8 cm in maximum diameter (Figs. 1A and 1B). Subtotal resection of the lesion confirmed the diagnosis of diffuse grade II astrocytoma.

Following surgery, the patient experienced complete left-sided hearing loss as well as some intermittent diplopia and gait disturbance. Proton therapy, with a margin of 50.4 Gy in 28 fractions, was initiated. The patient successfully completed therapy, with minimal complications. Follow-up imaging over 2 years continues to show a residual but stable lesion in the left middle cerebellar peduncle.

On neuro-ophthalmic examination, the visual acuity was 20/20 in each eye. The pupils were isocoric and reactive, and there was no relative afferent pupillary defect. Visual field testing, slit-lamp biomicroscopy, and fundus examinations were normal bilaterally. Extraocular muscle movements were full, but there was a large-amplitude, low-frequency nystagmus on left gaze and a small-amplitude, high-frequency nystagmus with right gaze consistent with a Bruns nystagmus.



**Fig. 1**—Brain magnetic resonance imaging (MRI) demonstrates focal swelling and subtle prolongation of T1 signal involving the left middle cerebellar peduncle (A), measured at a maximum diameter of 18 mm. The intra-axial mass is non-enhancing, but is infiltrative as denoted by the white arrows (B). There appears to be accentuated enhancement of the dura along the posterior superior surface of the petrous temporal bone as well as along the anterolateral tentorium on the left.

## DISCUSSION

CPA astrocytomas are rare tumors. Yilmaz et al. reported that the vast majority of CPA lesions were extra-axial tumors, with vestibular schwannomas comprising 80% of cases, followed by meningiomas and epidermoid cysts.<sup>1</sup> Other less common causes of lesions in the CPA include lipomas, teratomas, and cholesterol granulomas. There have also been reports of vascular-related lesions, such as giant aneurysms and hemangiomas. Intra-axial tumors, as was seen in our patient, are rarely found in this region.<sup>1,2</sup> The clinical and radiographic features of the more common extra-axial CPA tumors may be mimicked by an intra-axial, exophytic CPA astrocytoma.<sup>2-4</sup> In our case, the unique sign of Bruns nystagmus suggested a CPA mass. MRI imaging demonstrated increased T2 signal intensity in the region, suggestive of a glioma, but radiographic appearance alone could not be used for diagnosis. Tissue biopsy confirmed the diagnosis of low-grade astrocytoma.

Extra-axial lesions in the CPA (e.g., vestibular schwannoma) typically produce progressive ipsilateral hearing loss but may also cause diplopia (often resulting from sixth nerve palsy), and gait ataxia. Bruns nystagmus is a specific nystagmus that localizes to the CPA and is usually caused by a vestibular schwannoma.<sup>5</sup> Bruns nystagmus is characterized by large-amplitude, low-frequency beating toward the side of the lesion and a small-amplitude, high-frequency nystagmus when looking away from the side of the lesion.<sup>5</sup> The ipsilateral large-amplitude saccades that lessen exponentially are caused by compression of the ipsilateral neural integrator (primarily the cerebellar flocculus), which is responsible for maintaining eccentric gaze. The contralateral small-amplitude saccades, however, are constant in gaze and are caused by a lesion in the ipsilateral vestibular pathway.<sup>5,6</sup> Given our patient's presentation of oscillopsia prior to surgical resection of her tumor, the possibility of an iatrogenic etiology was ruled out.

In a retrospective study by Croxson et al., Bruns nystagmus in patients with advanced CPA tumors was found in 18 of 115 individuals (16%).<sup>7</sup> It was also noted that all 18 patients who had this type of nystagmus had tumors > 3 cm in diameter. Another study by Lloyd et al. suggested that Bruns nystagmus was associated with a larger tumor size, and of patients with CPA tumors > 3.5 cm in size; 92% had nystagmus, and 67% of those specifically had Bruns nystagmus (11% of the total patients).<sup>8</sup> In this series, none of the patients had a CPA astrocytoma. Our patient's tumor measured only 1.8 cm at its greatest diameter, and this may suggest that intra-axial tumors cause Bruns nystagmus through

a combination of both mass effect and direct invasion of the neural pathways and that the nystagmus may occur before the tumor reaches the larger size suggested by Lloyd et al.

Clinicians should be aware that intra-axial (but exophytic) as well as extra-axial CPA lesions may produce Bruns nystagmus, including CPA astrocytoma.

**Brian S. Park, BS,\* Stacy V. Smith, MD,†  
Ama Sadaka, MD,‡ Andrew G. Lee, MD<sup>‡,§,||,¶,\*,\*\*,+††</sup>**

\*Baylor College of Medicine, Houston, Tex; †Department of Neurology and Neurotherapeutics, UT Southwestern Medical Center, Dallas, Tex; ‡Department of Ophthalmology, Blanton Eye Institute, Houston Methodist Hospital, Houston, Tex; §Departments of Ophthalmology, Neurology, and Neurosurgery, Weill Cornell Medicine, New York, N.Y.; ||Department of Ophthalmology, The University of Iowa, Iowa City, Ia; ¶Department of Ophthalmology, Baylor College of Medicine, Houston, Tex; ††Department of Ophthalmology, University of Texas Medical Branch, Galveston, Tex; \*\*Section of Ophthalmology, UT MD Anderson Cancer Center, Houston, Tex; †††Department of Ophthalmology, University at Buffalo (SUNY), Buffalo, N.Y.

Correspondence to:

Andrew G. Lee, MD: alee@houstonmethodist.org

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## Case series: Merkel cell carcinoma of the eyelid



Merkel cell carcinoma (MCC) is a rare, aggressive cutaneous neuroendocrine malignancy hypothetically derived from a

precursor common to keratinocytes and Merkel cells.<sup>1</sup> Only 5% to 10% of MCCs involve the eyelids and the periorcular region,<sup>2</sup> of which fewer than 100 cases have been reported to date.<sup>3,4</sup> Here, we present 3 new patients with MCC of the eyelid and discuss diagnostic considerations.