Case Report
Bilateral internal auditory canal gangliogliomas mimicking neurofibromatosis Type II
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Background: Gangliogliomas are rare low grade, typically well-differentiated, tumors that are composed of mature ganglion cells and neoplastic glial cells. These tumors can appear at virtually any location along the neuroaxis but classically occur in the temporal lobe of young patients. In a small number of cases, gangliogliomas have presented as masses in the brainstem or involving cranial nerves. With the exception of vestibular schwannomas, bilateral tumors in the region of the internal auditory canal (IAC) or cerebellopontine angle (CPA) are exceedingly rare.

Case Description: We report a case of a 58-year-old male who presented with hearing loss, tinnitus, and vertigo. Initial magnetic resonance imaging revealed bilateral nonenhancing IAC/CPA tumors. Based on this finding, a presumptive diagnosis of neurofibromatosis Type II was made, which was initially managed conservatively with close observation. He returned for follow-up with worsening vertigo and tinnitus, thus prompting the decision to proceed with surgical resection of the symptomatic mass. Intriguingly, pathological study demonstrated a WHO Grade I ganglioglioma.

Conclusion: This is the first reported case of bilateral IAC/CPA gangliogliomas. When evaluating bilateral IAC/CPA lesions with unusual imaging characteristics, ganglioglioma should be included in the differential diagnosis.

Key Words: Bilateral, cerebellopontine angle, gangliogliomas, internal auditory canal, tumors

INTRODUCTION
Gangliogliomas are relatively rare tumors accounting for only 1% of all intracranial neoplasms. These tumors are composed of a combination of both neuronal and glial cell types and are typically benign, low-grade, and well differentiated.\(^2\) Classically, these tumors occur within the temporal lobe of children and young adults and present clinically with seizures. However, gangliogliomas have also been reported throughout the central nervous system including the posterior fossa, brainstem, spinal cord, and cranial nerves.\(^1\)\(^,\)\(^10\)\(^,\)\(^13\)\(^,\)\(^22\)\(^,\)\(^24\)
for vestibular schwannomas in the setting of neurofibromatosis Type II (NFII). Descriptions of bilateral nonschwannomatous IAC/CPA tumors are limited to case reports. We present a case of a 58-year-old male with unusual bilateral IAC tumors who was initially referred to our practice carrying the diagnosis of NFII. To the best of our knowledge, this is the first reported case of bilateral gangliogliomas of the IAC/CPA region.

CLINICAL PRESENTATION

A 58-year-old male presented with new onset vertigo and chronic asymmetric hearing loss, which was worse in the left than the right ear. The patient also complained of mild left-sided tinnitus. Over 3 months he developed progressively worsening balance causing him to fall, typically to his right side. Neurological exam was unremarkable except for bilateral sensorineural hearing loss. His initial audiogram confirmed the finding of bilateral sensorineural hearing loss, with profound loss of hearing in the higher frequencies that were most prominent on the left side. Magnetic resonance imaging (MRI) of the brain revealed bilateral nonenhancing IAC/CPA masses [Figure 1a and b]. The lesion on the left measured 4.5 mm × 4.5 mm × 5 mm; the lesion on the right was slightly larger, measuring approximately 8 mm × 8.3 mm × 8.2 mm with a 4 mm intracanalicular portion. Given the small size of these masses at presentation, the patient was initially managed conservatively with close follow-up. A repeat MRI of the brain 3 months later revealed no change in the size of either mass. During this time, the patient underwent occupational therapy and tried multiple medications for symptomatic relief.

At his 1 year follow-up visit, the patient reported worsening left-sided hearing loss, tinnitus, and otalgia and had begun taking a benzodiazepine to alleviate his symptoms. Repeat MRI of the brain at this time demonstrated the modest growth of both IAC/CPA masses, which remained nonenhancing. The mass on the left measured 6 mm × 5 mm × 4 mm; the right-sided mass measured 10 mm × 8 mm × 7 mm with growth of the intracanalicular portion to 8 mm in length [Figure 2a and b]. Based on evidence of tumor growth and progressively worsening symptoms, the patient elected to pursue surgical resection of the left-sided tumor. Radiosurgery was not considered a viable option since the patient’s age and unusual radiographic findings made the tissue diagnosis uncertain.

INTERVENTION

The patient underwent a left retrosigmoid craniotomy for tumor resection. A small lesion involving the intracanalicular portion of the eighth nerve complex was encountered and completely resected.

The excised tumor was an oval, well circumscribed, gray-tan, nodule that measured 0.5 cm × 0.5 cm × 0.4 cm. One-half was submitted for intra-operative consultation, which yielded a diagnosis of “ganglioneuroma versus ganglioglioma.” Examination of formalin-fixed, paraffin-embedded tissue sections revealed circumscribed neuroglial tissue composed of mature ganglion cells existing in a background of low-grade astrocyte-like cells with coarse processes [Figure 3a-d]. Ganglion cells contained Nissl substance and vesicular nuclei with prominent nucleoli and were randomly distributed in a haphazard fashion without polarization of cell processes [Figure 3a]. Neurofilament protein was strongly immunoreactive in the ganglion cells and revealed un-oriented cells’ processes extending into the glial stroma [Figure 3b]. Glial fibrillary acidic protein (GFAP) was strongly reactive in abundant astrocytic cell processes [Figure 3c]. Only weak reactivity for neuronal nuclear antigen was observed in the ganglion cells while granular synaptophysin reactivity was associated with ganglion cells and the stroma. S-100 was strongly and diffusely reactive in the lesion. The background stroma was distinctly astrocytic as demonstrated by strong GFAP-immunoreactivity of the cell processes. There were no areas of schwannoma-like
The role for adjuvant therapy is not well defined for WHO Grade III and IV gangliogliomas but is generally recommended, particularly in the setting of subtotal resection. Radiosurgery for gangliogliomas has also been reported to be effective but has not been thoroughly studied as a primary treatment modality.

In this case, gross total resection was achieved for the right-sided tumor, the patient is being followed closely with regular hearing monitoring and serial imaging. For chronic and/or benign lesions, it is paramount to approach the patient conservatively, focusing on preservation of hearing, and facial nerve function.

CONCLUSION

Gangliogliomas are rare and typically benign tumors that may occur at any location along the neuroaxis. Clinical
course and MRI can aid the diagnosis of gangliogliomas affecting cranial nerves before surgery. When evaluating bilateral IAC/CPA lesions with unusual imaging characteristics, ganglioglioma should be included in the differential diagnosis.

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Conflicts of interest
There are no conflicts of interest.

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