Lipochoristoma of the Internal Auditory Canal

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Abstract

Educational Objective: At the conclusion of this presentation, participants will understand the presentation, clinical course, and radiographic features of a rare benign tumor, lipochoristoma of the internal auditory canal.

Objectives: Lipochoristomas comprise 0.1% of all cerebellopontine angle tumors and may be misdiagnosed as vestibular schwannomas. We describe a lipochoristoma of the internal auditory canal and present the salient features of the evaluation, diagnosis and management.

Study Design and Methods: Retrospective case review.

Results (Case Report): A 51 yo woman presented with left sided severe hearing loss and tinnitus, mild episodic vertigo, and facial paresthesias progressive over one year. An MRI demonstrated a small (5mm x 4mm) T1 hyperintense lobulated lesion in the distal internal auditory canal. With fat suppressed images, there was no enhancement of the lesion. A diagnosis of IAC lipochoristoma was made. Conservative management was recommended and on 17 month follow up there has been no interval growth. The patient remains symptomatically stable with improved equilibrium but persistent left sided hearing loss.

Conclusions: Differential diagnosis of an enhancing internal auditory canal lesion includes acoustic neuroma, meningioma, epidermoid and arachnoid cysts, lipochoristoma and metastatic tumors. Fat suppressed MRI sequences aid in definitive diagnosis of lipochoristomas. Because lipochoristomas may have a tendency for more indolent growth and intimate involvement of the auditory nerve, conservative management with interval imaging is recommended. Surgical treatment is reserved for growing lesions or those with disabling vestibular symptoms.

Introduction

Asymmetric hearing loss in the absence of a known cause (meningitis, head injury, previous ear surgery, etc) should be considered clinically significant and warrant further investigation with imaging.

Neoplasm arising within the internal acoustic canal (IAC) and cerebellopontine angle (CPA) are most commonly of neuroepithelial origin. Vestibular schwannoma represents the most common lesion in the IAC (80-90%) with meningiomas comprising nearly all the rest (10%) of IAC tumors (1,2). Additional rare tumors include epidermoid and arachnoid cysts, lipochoristoma and metastatic tumors. Each of these lesions has typical imaging characteristics that frequently allow a diagnosis to be made with an MRI scan alone.

Lipochoristomas (lipomatous choristomas) comprise 0.1% of all CPA tumors, these rare tumors of the CPA and IAC are slow growing and are often discovered incidentally. Classically, these tumors were thought to arise from cells of the meninx primitiva, the mesenchymal derivative of the neural crest, and thus were referred to as lipomas of the IAC/CPA. However, research has since shown that these tumors arise from mesenchymal endogenous to the vestibulocochlear nerve, and thus are more appropriately characterized as lipomatous choristomas (4). This theory offers an explanation for the failure of hearing conservation reported after surgical resection of these lesions. Patients with lipochoristomas may present with hearing loss, vestibular symptoms, or tinnitus. In this report we examine the appropriate workup, diagnosis, and management of lipochoristoma of the IAC/CPA.

Case

A 51-year-old woman presented with one year of progressive left sided severe hearing loss accompanied by tinnitus and weekly episodes of mild vertigo. In addition, the patient reported chronic headaches and intermittent ipsilateral facial paresthesias. She did not report facial weakness and her past medical history was non-contributory. Her initial audiogram was notable for left sided severe sensorineural hearing loss with a word recognition score of 30%, while her right ear demonstrated normal hearing. An MRI obtained at an outside institution suggested the possibility of a “small enhancing intracanalicular acoustic neuroma on the left.” Initial review of this MRI demonstrated a small (5mm x 4mm) T1 hyperintense lobulated lesion in the distal IAC. It was recommended that the patient undergo an interval MRI with gadolinium. With fat suppressed images, there was no enhancement of the lesion and the diagnosis of IAC lipochoristoma was made. Conservative management was recommended and on 17 month follow up there has been no interval growth. The patient remains symptomatically stable with improved equilibrium but persistent left sided hearing loss. She is currently considering a bone anchored hearing aid.

Table 1. Imaging Characteristics of Cerebellopontine Angle Lesions

<table>
<thead>
<tr>
<th>MASS</th>
<th>T1</th>
<th>T1-contrast</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipochoristoma</td>
<td>Hyperintense</td>
<td>No Enhancement</td>
<td>T3 signal disappears with fat suppression (4)</td>
</tr>
<tr>
<td>Acoustic Neuroma</td>
<td>Isointense to brain</td>
<td>Enhances</td>
<td>Most sensitive sign is tumor extension along labyrinthine segment of facial nerve (5)</td>
</tr>
<tr>
<td>Facial Nerve Neurina</td>
<td>Isointense to brain</td>
<td>Enhances</td>
<td>Dural Tail, Rarely with IAC component, Can have calcification (6)</td>
</tr>
<tr>
<td>Meningioma</td>
<td>Isointense to brain</td>
<td>Enhances</td>
<td>Restricted diffusion on DWI (7)</td>
</tr>
<tr>
<td>Epidermoid</td>
<td>Hypointense</td>
<td>No Enhancement</td>
<td></td>
</tr>
<tr>
<td>Arachnoid Cyst</td>
<td>Hypointense</td>
<td>No Enhancement</td>
<td></td>
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T1 with fat suppression | T1 + Contrast | A mass within the left internal auditory canal that is T1 bright | The mass enhances with gadolinium contrast | T1 and T2 weighted images | On fat suppression imaging, the mass is dark | On high-definition T2 imaging (DRIVE), the mass is visible in the fundus of the IAC

Discussion

Tumors of the CPA and IAC frequently present with hearing loss, vestibular complaints and facial paresthesias. Distinguishing between the type of lesion present in the IAC/CPA is possible with MRI imaging and specific sequences. A comprehensive understanding of the differing imaging characteristics is of crucial importance to the otolaryngologist. Lipochoristomas may mimic a schwannoma, and care must be taken to look carefully at the T1-weighted images, which show a hypointense lesion. Lipochoristomas have an isointense appearance on T2-weighted images. Diagnosis is made with T1-weighted images that show fat suppression (3).

Lipochoristomas (lipomatous choristomas) classically were thought to arise from cells of the meninx primitiva, the mesenchymal derivative of the neural crest, which underwent abnormal differentiation (3). As such, they were classically and still are often referred to as “lipomas” of the IAC/CPA.

The clinical observation that these tumors are intimately associated with the auditory nerve was reported by Bigelow et al in 1998. This review of all 84 documented cases of lipomas of the IAC/CPA showed that while surgical resection had been performed in 62%, total tumor resection was accomplished in only 17/82. Furthermore, 68% of patients experienced postoperative complications, with additional hearing decrement the most common deficit (64%). Despite Bigelow’s report, a 2013 retrospective cohort study published in the Laryngoscope continued reference to these lesions as lipomas, despite the authors comments that these tumors exhibit a “unique behavior” by frequently engulfing coursing neuovascular structures (9).

In 2003, Wu et al from the House Ear Clinic presented a series of 11 cases which further elucidated the histogenesis of these tumors. By showing that mesenchymal elements of the tumor are integral components of the vestibulocochlear nerve, the paper’s authors argued that given the intimate relationship of the tumor to the nerve, “lipomas” of the IAC/CPA are most likely endogenous to the nerve, therefore suggesting that these tumors are better characterized as lipomatous choristomas (4). This theory offers an explanation for the failure of hearing conservation by surgical resection and is one of the main arguments for conservative management of these lesions. In addition, the natural history of lipochoristomas appears to be one of indolent growth as only three documented cases of growth have been reported in the literature.

Conclusions

- Recent histologic research has shown that fatty tumors of the cerebellopontine angle are more appropriately named “lipochoristomas” than lipomas.
- Because lipochoristomas may have a tendency for more indolent growth and intimate involvement of the auditory nerve, conservative management with interval imaging is recommended as the treatment of choice.
- Neurotologists should be aware that hearing preservation surgery is likely not to be successful given the intimate relationship between the lesion and the auditory nerve.
- Surgical treatment is reserved for growing and symptomatic lesions or those with disabling vestibular symptoms.

References