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Title: Distal Intracanalicular Lipochoristoma: A Rare Case Report

Running Title: Internal Acoustic Canal Lipoma

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ABSTRACT

Background

Lipochoristomas are benign fatty tumours with rare occurrence in internal acoustic canal and cerebellopontine angle (0.14%). Despite its benign nature and slow growth potential, it poses serious dilemma for neurotologists due to its intimate involvement with the auditory nerve.

Case report

We would like to present a case of distal intracanalicular lipochoristoma in a 60 year male with 5 years radiological follow up and a brief literature review of this rare entity.

Conclusion

Magnetic resonance imaging plays an important role in both diagnosing and surveillance of the disease. Due to limited surgical outcome, conservative surveillance is the best treatment till date.

KEYWORDS

Lipochoristoma; internal auditory canal; cerebellopontine angle; magnetic resonance imaging; hearing loss
INTRODUCTION

Lipomas are tumours which histologically contain mature adipocytes with variable quantity of fibrovascular tissue. The occurrence of lipoma in intracranium is rare, with an incidence of 0.08% in autopsy series, with majority found in corpus callosum (50%). However, the presence of lipoma is extremely rare in internal acoustic canal (IAC) and cerebellopontine angle (CPA), comprising 0.14% of all CPA/IAC tumours (1). Previously, CPA lipomas are thought to arise from mesenchymatous cells of neural crest by aberrant differentiation of meninx primitiva. However, it is now known to derive from endogenous mesenchyme of the vestibulocochlear nerve, hence appropriately named as lipomatous choristoma or lipochoristoma (2). It is often discovered incidentally and thought to have a slow growth. In this article, we would like to present a case of distal intracanalicular IAC lipoma (lipomatous choristoma), with 5 years MRI surveillance and would like to highlight on diagnosis and managing this rare entity, along with literature review.

CASE REPORT

A 60 year old man presented to our Otorhinolaryngology clinic with 2 years history of right sided hearing loss accompanied with unilateral non-pulsatile tinnitus. However, he denied episodes of vertigo nor headaches. There was no prior history of fall, trauma or otologic infections. On general examination, patient was comfortable and otoscopic examination demonstrated normal findings. Tuning fork test demonstrated Rinne test was positive in left ear but false negative in right ear. Weber test lateralizing to the left. Other cranial nerves examinations were normal. His pure tone audiometry (PTA) test revealed right profound sensorineural hearing loss, with normal hearing on the left ear. Auditory Brainstem Response (ABR) showed normal wave V at 90dBnHL on the right ear but no extension of wave V latency seen.
Magnetic resonance imaging (MRI) demonstrated small intracanalicular lesion in distal part of right internal auditory canal, adjacent to inferior vestibular/cochlear nerve, measuring 5mm x 2mm. The lesion hyperintense on T1w, isointense on T2 and suppressed on fat suppression sequence (Figure 1a,b). Besides, no significant enhancement seen post gadolinium contrast, which concludes the mass as lipochoristoma. Patient was subsequently managed conservatively with annual MRI follow up which showed no changes in lesion in 5 years. During surveillance, patient had denied worsening of symptoms and no deterioration of quality of life.

**DISCUSSION**

The more common lesions in IAC are vestibular schwannomas (80-90%) and meningiomas (10%). Other rare tumours include epidermoids, lipochoristomas and metastatic tumours consisting less than 1% (3). CPA lipochoristoma was first described by Klob in 1859, comprising only 0.14% of all lesions in CPA/IAC (4). Previously, it was erroneously referred as lipoma as it was believed to arise from cells of meninx primitiva of neural crest cell. However, Bigelow et al. demonstrated the intimate association with auditory nerve and the resulting failure of hearing conversation in his 15 patients despite complete surgical resection (3,4). Current theory, as supported by Wu et al (2), states that it arises from endogenous mesenchyme of vestibulocochlear nerve, hence the name lipomatous choristoma (lipochoristoma). However, unlike other intracranial lipomas, lipochoristomas lack cellular atypia and frequently entrap unmyelinated nerve fibers. It resembles intradural spinal lipomas and is not associated with developmental abnormalities of the central nervous system. Besides, CPA/IAC lipochoristomas are at times misdiagnosed as hamartoma as they may commonly feature high degree of vascularity (5).

The majority of patients have cochleovestibular symptoms such as hearing loss, tinnitus and vertigo, which lasts for a few years. Some authors elucidated hemifacial spasm due to facial
nerve involvement in 9% of the cases, and trigeminal signs like paresthesia or neuralgia in 14.4% of cases. Tankere et al (1) mentioned in his review, out of 98 total reported cases, 65 cases were histologically confirmed, showing 95.4% of tumour involvement of cranial nerve, dominantly vestibulococlear nerve (96.7%) and facial nerve (82.2%).

Currently, diagnosis is made based on imaging modalities, mainly MRI. Lipochoristoma has specific features on MRI studies that helps to differentiate from other CPA tumours, especially vestibular schwannoma. Lipochoristomas generally display hyperintensity on nonenhanced T1 weighted images, with iso- to hypointensity with increasing T2 weighting. Though this feature differentiates lipochoristoma from schwannoma, T1 weighted images alone is inadequate to complete the diagnosis. Lipochoristoma also shows absence of enhancement post gadolinium with missing signal in fat suppression sequences (1,6). These characteristic features allow us to differentiate from other CPA lesions as depicted in TABLE 1 (3,6).

Lipochoristomas generally have indolent growth potential and malignant degeneration is yet to be reported. Similarly to our patient, Wu et al (2) also reported no tumour growth in conservatively managed lipochoristomas in 7-year follow-up period. Surgical excision of lipochoristomas of CPA and IAM is difficult, owing to its close proximity to cranial nerves and its tumour hypervascularization may induce bleeding complications during surgery. This is shown by Bigelow et al, who documented a total of 84 cases of lipochoristomas in CPA and IAM but only 17 out of 52 had total tumour excision. However, 68% of those patients had postoperative sequelae, mainly hearing loss (64%) (1,3,4), facial palsy, transient dysphagia and uvula deviation (7). Considering the frequency and severity of postoperative sequelae, conservative management with close radiographic surveillance remains a better option, even in young patients (8). Tankéré et al (1) highlighted that surgical option should only be considered in patients with severe vertiginous syndromes resistant to both medication and rehabilitation, and also patients with severe trigeminal neuralgia and hemifacial spasms.
CONCLUSION

Lipochoristoma is rare and grows very slowly. The diagnosis of lipochoristoma is based on characteristic MRI findings. In view of its slow growth rate and limited surgical outcome, we believe that conservative radiological surveillance is the best treatment option for this rare lesion.

REFERENCES


**Legends**

Figure 1 a – T1 weighted MRI image showing hyperintensed lesion (red arrow) in right CP angle (axial plane).

Figure 1 b – T1 weighted MRI image showing hyperintensed lesion (red arrow) right CP angle (coronal plane).

Figure 1 c - Hypointensity of the same lesion in T2 weighted MRI image.
TABLE 1: Magnetic Resonance Imaging Characteristics of Tumours in Cerebellopontine Angle and Internal Acoustic Meatus

<table>
<thead>
<tr>
<th>LESION</th>
<th>T1 WEIGHTED</th>
<th>T2 WEIGHTED</th>
<th>FAT SUPPRESSION</th>
<th>GADOLINIUM ENHANCEMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipochoristoma</td>
<td>Hyperintense</td>
<td>Iso/Hypointense</td>
<td>Absent signal</td>
<td>Absent</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>Isointense</td>
<td>Iso/Hyperintense</td>
<td>Unchanged</td>
<td>Increased</td>
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<tr>
<td>Meningioma</td>
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<td>Isointense</td>
<td>Unchanged</td>
<td>Increased</td>
</tr>
<tr>
<td>Epidermoid cyst</td>
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<td>Iso/Hyperintense</td>
<td>Unchanged</td>
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</tr>
<tr>
<td>Arachnoid cyst</td>
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