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CASE REPORT – OPEN ACCESS

Distal Intracanalicular Lipochoristoma: A Rare Case Report

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ABSTRACT

Background: A lipochoristoma is a rare, benign, fatty tumor in the internal acoustic canal or 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: Presently described is a case of a distal intracanalicular lipochoristoma in a 60-year-old male, including 5 years of radiological follow-up and a brief literature review of this rare entity.

Conclusion: Magnetic resonance imaging plays an important role in both the diagnosis and surveillance of the disease. Due to limited surgical outcomes, conservative surveillance is believed to be the best treatment in most cases.

Keywords: Cerebellopontine angle, hearing loss, internal auditory canal, lipochoristoma, magnetic resonance imaging

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INTRODUCTION

Lipomas are tumors that contain mature adipocytes with a variable quantity of fibrovascular tissue. The occurrence of a lipoma in the intracranial structures is rare; an incidence of 0.08% has been observed in autopsy series, with most found in the corpus callosum (50%). The presence of a lipoma in the internal acoustic canal (IAC) or cerebellopontine angle (CPA) is extremely rare, representing only 0.14% of all CPA/IAC tumors (1). Previously, CPA lipomas were thought to arise from mesenchymatous cells of the neural crest through aberrant differentiation of the meninx primitiva. However, it is now known to derive from the endogenous mesenchyme of the vestibulocochlear nerve (2). It is often discovered incidentally and is regarded as having slow growth. This report describes a case of a distal intracanalicular IAC lipoma (lipomatous choristoma) with 5 years of magnetic resonance imaging (MRI) surveillance in order to highlight the diagnosis and management of this rare entity in the context of a brief literature review.

CASE REPORT

A 60-year-old man presented at the otorhinolaryngology clinic with a 2-year history of right-sided hearing loss accompanied by unilateral, non-pulsatile tinnitus. He claimed not to have had episodes of vertigo or headaches. There was no prior history of a fall, trauma, or otological infection. On general examination, the patient was comfortable and an otoscopic examination demonstrated normal findings. A Rinne test yielded a positive result in the left ear but a false negative in the right ear. A Weber test indicated lateralizing to the left. Other cranial nerve examination findings were normal. A pure tone audiometry test revealed profound sensorineural hearing loss on the right, with normal hearing in the left ear. An auditory brainstem response test showed a normal wave V amplitude at 90 dBnHL on the right, but no extension of wave V latency was seen.

MRI revealed a small intracanalicular lesion in the distal part of the right internal auditory canal, adjacent to the inferior vestibular/cochlear nerve, measuring 5 mm x 2 mm. The lesion was hyperintense on T1w, isointense on T2w, and suppressed on a fat-suppression sequence (Fig. 1a–c). There was no significant enhancement seen in a post-gadolinium contrast image, which provided confirmation that the mass was a lipochoristoma. The patient was subsequently managed conservatively with an annual MRI follow-up and for 5 years, no changes in the lesion have been observed. The patient has not reported any deterioration of symptoms or quality of life.

DISCUSSION

The more common lesions of the IAC are vestibular schwannomas (80–90%) and meningiomas (10%). Rare tumors include epidermoids, lipochoristomas, and metastatic tumors, but these make up less than 1% (3).

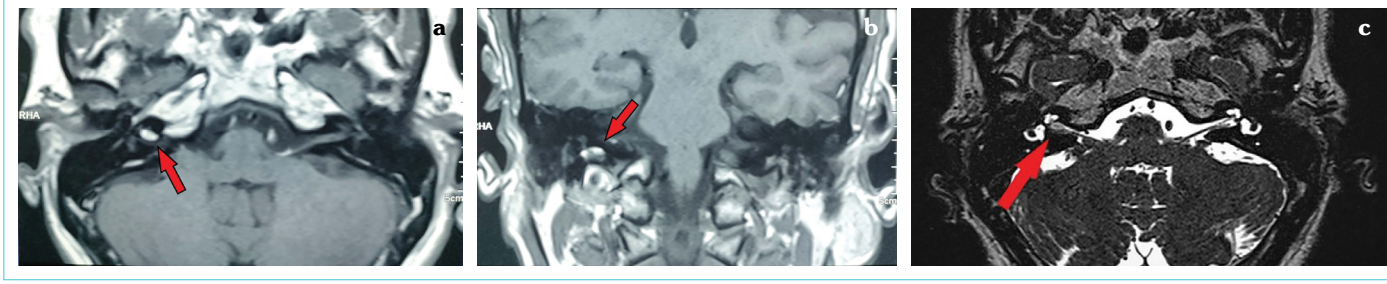

Figure 1. (a) T1-weighted magnetic resonance imaging (MRI) showing hyperintense lesion (red arrow) in the right cerebellopontine angle (CPA) (axial plane). (b) T1-weighted MRI showing hyperintense lesion (red arrow) in the right CPA (coronal plane). (c) T2-weighted MRI showing hypointensity of the same lesion

Table 1. Magnetic resonance imaging characteristics of tumors in the cerebellopontine angle or internal acoustic meatus

Lesion	T1-weighted	T2-weighted	Fat suppression	Gadolinium enhancement
Lipochoristoma	Hyperintense	Iso/hypointense	Absent signal	Absent
Schwannoma	Isointense	Iso/hyperintense	Unchanged	Increased
Meningioma	Isointense	Isointense	Unchanged	Increased
Epidermoid cyst	Iso/hypointense	Iso/hyperintense	Unchanged	Absent
Arachnoid cyst	Isointense	Hyperintense	Unchanged	Absent

A CPA lipochoristoma was first described by Klob in 1859, and represents only 0.14% of all lesions in the CPA/IAC (4). It was previously erroneously referred to as a lipoma, as it was believed to arise from cells of the meninx primitiva of the neural crest. However, Bigelow et al. (4) demonstrated the intimate association with the auditory nerve and the resulting failure of hearing conversation in 15 patients despite complete surgical resection (3, 4). Current theory, as supported by a study conducted by Wu et al. (2), is that it arises from the endogenous mesenchyme of the vestibulocochlear nerve, hence the more appropriate name, lipomatous choristoma (lipochoristoma). However, unlike other intracranial lipomas, lipochoristomas lack cellular atypia and frequently entrap unmyelinated nerve fibers. A lipochoristoma resembles an intradural spinal lipoma and is not associated with developmental abnormalities of the central nervous system. A CPA/IAC lipochoristoma can be misdiagnosed as a hamartoma, as they commonly feature a high degree of vascularity (5).

The majority of patients have cochleovestibular symptoms, such as hearing loss, tinnitus, and vertigo, which may have been present for some years. Authors have described hemifacial spasm due to facial nerve involvement in 9% of the cases, and trigeminal signs, like paresthesia or neuralgia, in 14.4% of cases. Tankéré et al. (1) mentioned in a review of 98 reported cases that 65 were histologically confirmed, showing tumor involvement of the cranial nerve in 95.4%, the vestibulococlear nerve in 96.7%, and a facial nerve in 82.2%.

Currently, diagnosis is made based on imaging modalities, primarily MRI. Lipochoristoma has specific features on MRI studies that help to differentiate it from other CPA tumors, particularly vestibular schwannoma. Lipochoristomas generally display hyperintensity on nonenhanced T1-weighted images, and iso- to hypo-intensity with increased T2 weighting. Though this feature differentiates lipochoristoma from schwannoma, T1-weighted images alone are inadequate to complete the diagnosis. Lipochoristomas also demonstrate an absence of post-gadolinium contrast enhancement, and a missing signal in fat suppression sequences (1, 6). These characteristic features allow us to differentiate it from other CPA lesions, as depicted in Table 1 (3, 6).

Lipochoristomas generally have a slow growth potential and malignant degeneration has not yet been reported. As in our patient, Wu et al. (2) also reported no tumor growth in conservatively managed lipochoristomas over a 7-year follow-up period. Surgical excision of lipochoristomas of the CPA or IAM is difficult, owing to its close proximity to cranial nerves and tumor hypervascularization may induce bleeding complications during surgery. Bigelow et al. (4) documented a total of 84 cases of lipochoristoma in the CPA or IAM. Surgical resection was performed in 52 lesions; however, total tumor removal was accomplished in only 17. Of those patients, 68% had postoperative sequelae. Cases have been reported with hearing loss, facial palsy, transient dysphagia, and uvula deviation (1, 3, 4, 7). However, given the frequency and potential severity of postoperative sequelae, conservative management with close radiographic surveillance remains the best option in most cases, even in young patients (8). Tankéré et al. (1) highlighted that a surgical option should only be considered in patients with severe vertiginous syndromes resistant to both medication and rehabilitation, and patients with severe trigeminal neuralgia and hemifacial spasms.

CONCLUSION

Lipochoristoma is rare and grows very slowly. The diagnosis is based on characteristic MRI findings. In view of its slow growth and limited surgical outcomes, conservative radiological surveillance is currently the best treatment option for this rare lesion in most cases.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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