CASE REPORT

Chorda tympani schwannoma: one new case revealed during malignant otitis externa and review of the literature☆

Schwannoma da corda do timpano diagnosticado no decurso de uma otite externa maligna: relato de caso e revisão da literatura

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Introduction

Facial nerve schwannomas (FNS) are a rare entity that are poorly reported in literature. Although the estimated prevalence is extremely low, FNS are the most common primary tumor of the facial nerve. FNS occur throughout the course of the facial nerve and its branches, and chorda tympani schwannomas are quite rare. In 2010, Huoh and Cheung reported that seven additional chorda tympani schwannoma had appeared in literature since the first report by Nager in 1969.1,2 Most of the cases were isolated, with only two cases reporting multiple neuromas in patients with neurofibromatosis.1,2 The presenting symptoms of isolated chorda tympani schwannoma were conductive hearing loss, tinnitus, and facial palsy.2 Interestingly, taste disturbance was not documented in any previous case.2 All of the cases showed a mass obstructing the external auditory canal (EAC) or behind an intact tympanic membrane, thereby confirming a common clinical history of this rare entity.

This report relates the case of an 89-year-old male patient who presented a chorda tympani schwannoma disclosed during the management of malignant otitis externa (MOE). To the best of the authors’ knowledge, this is the first case in literature with incidental radiological finding of asymptomatic chorda tympani schwannoma.

Case report

An 89-year-old male patient, who had had hypertension and type 2 diabetes mellitus for an extended period, presented to this tertiary care center with a six-month history of right ear ache, otorrhoea, and hearing loss. He had received oral antibiotics (amoxicillin–clavulanate) and local antibiotic eardrops during this interval. He denied tinnitus and vertigo. Otoscopic examination of the right ear revealed otorrhoea and inflammation of the EAC. The tympanic membrane was thickened and somewhat hemorrhagic in appearance but was otherwise intact. Cranial nerve examination revealed normal facial function. The patient reported...
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no complaint of taste disturbance. Pure tone audiogram showed a right mixed hearing loss with air bone gap at 15 dB and symmetric bone curve by presbycusis. Laboratory signs of inflammation or infection were absent (no elevation of blood cell count, C-reactive protein [CRP], or erythrocyte sedimentation rate). Diabetes was considered to be well controlled (HbA1c = 5.6%). Cultures of otorrhea were positive for Pseudomonas aeruginosa. P. aeruginosa was sensitive to ciprofloxacin and ceftazidime, and these systemic antibiotics were prescribed. Computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone were performed. CT showed partial opacification of the right mastoid air cells and middle ear cavity, and thickening of the right tympanic membrane and skin of the right EAC. CT also revealed an osteolytic lesion of the mastoid along the vertical segment of the facial nerve, involving the floor of EAC, with bone destruction, decreased bone density, and the lake of continuity and wormy appearance of the cortical bone (Fig. 1). On MRI, multiple soft tissue signals were observed in the middle ear and mastoid region, displaying equal T1 and long T2 signals, with gadolinium contrast enhancement. MRI revealed EAC inflammation and infiltration of retrocondylar fat. Although, a soft tissue lesion of 6 mm equal T1 and long T2 signals with a homogeneous hyper-enhancement after intravenous contrast injection, involved the floor of EAC, and caused an osteolytic erosion of the mastoid along the

Figure 1  Radiological examination of chorda tympani schwannoma with associated MOE. (A) Axial CT scan demonstrating well-defined soft tissue lesion. (B) Reconstructed coronal CT image demonstrating above soft tissue lesion caused an osteolytic erosion of the mastoid along the vertical segment of the facial nerve. (C) T1 with gadolinium MRI image in axial projection: the chorda tympani tumor exhibits a homogenous hyper-enhancement. (D) The MRI axial T2-weighed image in axial projection shows the hyperintense schwannoma. (E) T1 with gadolinium MRI image in coronal projection: the chorda tympani tumor exhibits a homogenous hyper-enhancement. MOE, malignant otitis externa; CT, computed tomography; MRI, magnetic resonance imaging.
vertical segment of the facial nerve (Fig. 1). Under general anesthesia, with facial nerve neuromonitoring, the mass was accessed by the transmastoid approach to exclude a malignant tumor. A malignant tumor was first suspected, and the surgery aim was diagnostic before the therapeutic decision. Only a biopsy was performed, and the nerve was preserved during surgery. The biopsy revealed a tumor arising from the Schwann’s cells (Fig. 2). Histological examination revealed a lesion composed of spindle cells with wavy appearing nuclei. The nuclei were arranged in a palisading fashion. Mitotic activity was not present. Moreover, in this particular case, inflammatory cells were associated. Spindle cells were diffusely and strongly positive for S100 protein. The postoperative period was uneventful, with well-preserved facial nerve function and no taste disturbance. Systemic antibiotics (ciprofloxacin and ceftazidime) were prescribed during six weeks for malignant external otitis, and periodic follow-ups were recommended for chorda tympani schwannoma. No attempt was made to resect the represented chorda tympani tumor because the patient was symptom-free. Follow-up at six months showed the patient in good health, without recurrent disease. Pure tone audiogram showed a stable bilateral sensorineural hearing loss.

Discussion

This report presents an additional case of chorda tympani schwannoma. To the best of the authors’ knowledge, this is the first case in literature with incidental radiological finding of asymptomatic chorda tympani schwannoma. This is a very rare benign tumor for all authors, and Huoh and Cheung in 2010 reported only seven other cases in the English literature. However, a new review of the literature showed that 14 cases of chorda tympani schwannoma have been published in the English literature. Table 1 reports these cases, as well as this report’s new case of chorda tympani schwannoma. Of the 15 patients, predominance among women was noted; six patients were males (40%) and nine were females (60%), a sex ratio of 0.6. The mean age at diagnosis was 38.5 years (range: 12–89 years). The tumor was located on the right side in nine cases (60%), the left side in four cases (27%), and this information was not reported in two cases (13%). Most of the cases were isolated chorda tympani schwannomas (87%), with only two cases reporting multiple schwannomas (13%); one patient with neurofibromatosis Type 1 reported by Nager in 1969, and one patient with neurofibromatosis Type 2 reported by Huoh and Cheung in 2010. Presenting symptoms at diagnosis were conductive or mixed hearing loss (60%), tinnitus (27%), earache (20%), vertigo (13%), facial palsy (13%), and fullness (7%). In one case, vertigo may have been unrelated and suggested benign paroxysmal vertigo. In another case, right facial weakness was present early in childhood, possibly from birth trauma. In the present case, MOE may have caused right earache and mixed hearing loss. In all cases except the present study, otoscopic examination found a retrotympanic mass or a mass in the EAC. From the evolution of medical imaging techniques, all cases presented a soft tissue lesion along the chorda tympani. Preoperative diagnoses included cholesteatoma, glomus, rhabdomyosarcoma, facial nerve schwannoma, and malignant tumor in the present case. Diagnosis is usually made by biopsy and treatment is surgical, with preservation of facial and auditory function. Two patients did not have surgical resection (13%): one patient with neurofibromatosis, and one post-mortem case. Of the 12 patients operated on, postoperative clinical evaluation reported two cases of facial palsy (17%), one case of mixed hearing loss (8%), and one case of temporary taste disturbance (8%). The number of previous chorda tympani schwannoma cited by authors was wrong in all articles when these data were present. Review of the literature showed that 14 cases of chorda tympani schwannoma had been published in the English literature before the present case. Taste disturbance was not documented in any previous case, which is in accordance with this patient. The slow growing nature of the neuroma is likely to allow compensatory mechanisms to occur without the patient experiencing dysgeusia. Moreover, resection of the chorda tympani often causes no subjective change in overall taste sensation. The nerve is often sacrificed in middle-ear surgery; one study noted a 31% incidence of permanent taste alteration in cases of complete nerve resection.

To the best of the authors’ knowledge, this is the first case in literature with incidental radiological finding of asymptomatic chorda tympani schwannoma. In this case, MOE may have caused right earache and mixed hearing loss.

Figure 2  Histological examination of chorda tympani schwannoma with inflammatory tissue by associated MOE (200×). (A) Histological examination shows typical elongated spindle cells that tend to form palisades on hematoxylin–eosin safran (HES) coloration. (B) Spindle cells are diffusely and strongly positive for S100 protein. MOE, malignant otitis externa.
# Table 1  Literature review of patients with chorda tympani schwannoma.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age/sex</th>
<th>Side</th>
<th>Clinical presentation</th>
<th>Otoscopic examination</th>
<th>Imaging examination</th>
<th>Postoperative clinical evaluation</th>
<th>Previous cases described</th>
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</thead>
<tbody>
<tr>
<td>Undabeta et al.</td>
<td>45/F</td>
<td>Right</td>
<td>Mixed HL, vertigo, tinnitus, FP</td>
<td>Retrotympanic mass in the posteroquadrant mass in the tympanic mass</td>
<td>Mass in the tympanic cavity</td>
<td>FP HBII, mixed HL</td>
<td>NR</td>
</tr>
<tr>
<td>Huoh and Cheung</td>
<td>24/F</td>
<td>Right</td>
<td>HL, tinnitus, FP</td>
<td>Retrotympanic mass</td>
<td>Chorda tympani tumor</td>
<td>No surgery</td>
<td>7</td>
</tr>
<tr>
<td>Hopkins et al.</td>
<td>53/M</td>
<td>Right</td>
<td>Vertigo, mixed HL</td>
<td>Retrotympanic mass in the posteroquadrant Tumors in the hypotympanum</td>
<td>Mass in the tympanic cavity, extending into the attic</td>
<td>Temporary taste disturbance</td>
<td>5</td>
</tr>
<tr>
<td>Biggs and Fagan</td>
<td>26/F</td>
<td>Right</td>
<td>Life-long ear disease</td>
<td>Mass retrotympanic</td>
<td>Lesion in the EAC extending into the lower tympanic cavity, with osteolytic erosion along the vertical segment of the facial nerve</td>
<td>Normal, no recurrence (two-year postop)</td>
<td>5</td>
</tr>
<tr>
<td>Magliulo et al.</td>
<td>58/F</td>
<td>Left</td>
<td>Conductive HL, tinnitus</td>
<td>Mass in the EAC</td>
<td></td>
<td>Normal</td>
<td>5</td>
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<tr>
<td>Chai et al.</td>
<td>60/F</td>
<td>Left</td>
<td>Conductive HL</td>
<td>Mass retrotympanic</td>
<td></td>
<td>Normal</td>
<td>5</td>
</tr>
<tr>
<td>Browning et al.</td>
<td>51/F</td>
<td>Right</td>
<td>HL, earache</td>
<td>Mass in the posteroquadrant EAC and retrotympanic in the posteroquadrant</td>
<td></td>
<td>Normal</td>
<td>5</td>
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<tr>
<td>Saleh et al.</td>
<td>62/F</td>
<td>Left</td>
<td>Tinnitus</td>
<td>Mass in the posteroquadrant EAC</td>
<td></td>
<td>Normal, no recurrence (seven-year postop)</td>
<td>NR</td>
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<tr>
<td>Lopes Filho et al.</td>
<td>25/M</td>
<td>Right</td>
<td>Fullness, earache Progressive FP</td>
<td>Tumor in the posterior EAC</td>
<td></td>
<td>Normal</td>
<td>2</td>
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<tr>
<td>Sanna et al.</td>
<td>14/F</td>
<td>Right</td>
<td>Progressive FP</td>
<td>NR</td>
<td></td>
<td>Normal</td>
<td>NR</td>
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<tr>
<td>Wiet et al.</td>
<td>12/M</td>
<td>Left</td>
<td>Conductive HL</td>
<td>Mass in the posteroquadrant EAC</td>
<td>Mass in the EAC with erosive changes of the posterior wall</td>
<td>Normal</td>
<td>NR</td>
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<tr>
<td>Babin et al.</td>
<td>18/M</td>
<td>NR</td>
<td>Asymptomatic</td>
<td>NR</td>
<td></td>
<td>No surgery; post-mortem study</td>
<td>NR</td>
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<tr>
<td>Pou and Chambers</td>
<td>18/F</td>
<td>Right</td>
<td>Conductive HL, tinnitus</td>
<td>Mass in the EAC</td>
<td></td>
<td>Normal</td>
<td>0</td>
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<td>Nager</td>
<td>22/M</td>
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M, male; F, female; NR, not reported; HL, hearing loss; FP, facial palsy; EAC, external auditory canal; HB, House-Brackmann grade; postop, postoperative.
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Conclusion

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Conflicts of interest

The
authors
declare
no
conflicts
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interest.

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