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Case Report

Clinical and Histological Features of Chorda Tympani Tumors

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Tumors of the chorda tympani are extremely rare. To date only 15 cases have been reported in the literature. Slow growing rate and nonspecific clinical features allow these lesions to reach a large size and resemble more common otological entities such as cholesteatoma. Diagnosis is usually established after surgical resection. However, lack of specifity of conventional pathological studies emphasizes the importance of immunohistochemical studies for final diagnosis. It is advisable to use the correct terms when reporting the histology of chorda tympani tumors in literature.

KEY WORDS: Schwanomma, neurinoma, neuroma, chorda tympani, middle ear, S-100

INTRODUCTION

Primary involvement of the chorda tympani is very rare in benign facial nerve tumours. Tumours of the facial nerve may be clinically suspected when they cause irreversible functional deficits (hearing loss, facial palsy, disbalance, etc.) due to compression of nearby structures. Slow growth and benign characteristics make it possible for these tumours to become large while staying clinically silent. Isolated chorda tympani schwanommas usually present with conductive hearing loss and tinnitus, and rarely facial palsy. Although they may affect sensitive fibres they rarely cause any taste dysfunction, so they demand a high clinical suspicion.

Occasionally, nervous tumours may be mistaken for primary or secondary cholesteatomas due to their ability to erode the surrounding bone. Therefore, diagnosis is often made after the surgical procedure. Immunohistochemical techniques are necessary in atypical cases and when conventional histology is nonspecific.

A review of the literature showed 15 cases of primary chorda tympani tumours reported until date (Table 1). Seven of them were described as neuromas, 6 schwannoma and 2 neurilemmomas. Patients presented with facial palsy only in 2 cases, one of them likely of perinatal origin [1, 2]. The aims of this work are to review clinical and immunohistochemical features of chorda tympani benign neoplasms and to emphasize the need for precise diagnosis and correct use of terminology when reporting about these tumours.

CASE REPORT

A 34-year-old man presented to our clinic complaining of aural fullness in the right ear following an episode of acute ear pain without otorrhoea 3 months earlier. It was treated with oral antibiotics by a general practitioner. Afterwards, he had attended another otological centre where he was diagnosed with chronic otitis media.

On examination, there was inflamed mucosa over the epitympanum, with a prominence close to the ear canal. The audiometry showed a mild shift in 4000 and 8000 Hz in the right ear, with normal thresholds in the rest of the frequencies. Incudostapedial reflexes were absent.

A computed tomography (CT) scan was performed, showing complete occupation of the middle ear cavity with extension to the antrum and mastoid cells and erosion of ossicles. There was thinning of the tegmen of the middle ear on both sides. No erosion of the scutum could be observed (Figure 1).

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Table 1. Comparative review of cases of chorda tympani tumors

Author, year	Preoperative facial palsy	Clinical presentation	Histological diagnosis	IHC
Nager, 1969 [14]	Not reported	Not stated	Neurinoma	Not done
Pou, 1974 [3]	Absent	Tinnitus EAC mass	Neuroma	Not done
Fuentes, 1983 [4]	Absent	Tinnitus. Conductive HL EAC mass	Neurinoma	Not done
Wiet, 1985 [5]	Case 1-Absent Case 2-Absent	Case 1-Conductive HL Case 2-Conductive HL and anterior tympanic mass	Case 1-Neurilemmoma Case 2-Neurilemmoma	Not done
Sanna, 1990 [1]	Present	Progressive facial palsy	Type A neuroma	Not done
Lopes Filho, 1993 [12]	Absent	Ear fullness Otorrhoea EAC mass	Neuroma	Not done
Saleh, 1995 [6]	Absent	Tinnitus. Conductive HL EAC mass	Neuroma	Not done
Browning, 2000 [7]	Absent	Hearing loss Otorrhoea EAC mass	Schwannoma	Not done
Magliulo, 2000 [8]	Absent	Tinnitus. Conductive HL EAC mass	Neuroma	Not done
Chai, 2000 [9]	Absent	Conductive HL	Schwannoma	Not done
Biggs, 2001 [15]	Absent	Otorrhoea Otalgia	Schwannoma	Not done
Hopkins, 2003 [10]	Absent	Vertigo Mixed HL EAC mass	Neuroma	Not done
Huoh, 2012 [2]	Present	Progressive HL Tinnitus	Neuroma	Not done
Undabeitia, 2013 [11]	Absent	Mixed HL Middle ear mass	Schwannoma	Not done
Present study	Absent	Otorrhoea Middle ear mass	Neuroma VS schwannoma	S-100+

HL: hearing loss; IHC: Immunohistochemistry; EAC: external auditory canal

These findings made it difficult to make a differential diagnosis between chronic suppurative otitis media and cholesteatoma. The patient underwent a right ear exploration in the form of canal wall up mastoidectomy. After opening the antrum, a flesh-coloured mass of elastic consistency was found. The mass filled the posterior epitympanus, aditus, and antrum and eroded the head of the incus. A fragment was sent for intraoperative pathological examination and no malignancy was identified. The tumour was removed *en bloc* in continuity with the chorda tympani nerve and sent for pathological study. No reconstruction of the ossicular chain was performed. The patient did not complain of dysgeusia prior to or after the surgery. Facial function was normal. Audiometry obtained 1 month after surgery showed a mild loss (35-40 dB mean thresholds) that was similar to the results of the preoperative test.

The specimen was processed for routine histology. Macroscopically, the tumour had a whitish, pearly colour and rubbery consistency and measured 0.7×0.5×0.3 cm. Haematoxylin-eosin staining showed a spindle-shaped cell proliferation of scarce density, with no nuclear atypia, mitosis, or necrosis. The proliferation was well delimited but there was no capsule. Cell density was homogeneous. No specific growing pattern, palisading (Verocay bodies), Antoni zone A or B, nor hyalinised vessels could be identified. There were nonspecific lymphocyte aggregates at the periphery of the tumour as well as isolated histiocyte groups in relation to cholesterol crystals (Figure 2a).

Immunohistochemical staining for S-100 protein was positive (Figure 2b). Staining for cytokeratins (AE1-AE3), CD34, ALK1, beta-catenin, epithelial membrane antigen (EMA), and neurofilaments was negative. The features of the mass were consistent with spindle cell proliferation with peripheral nerve sheath differentiation and no malignant features, suggestive of the differential diagnoses discussed below (schwannoma vs. neurofibroma vs. neuroma).

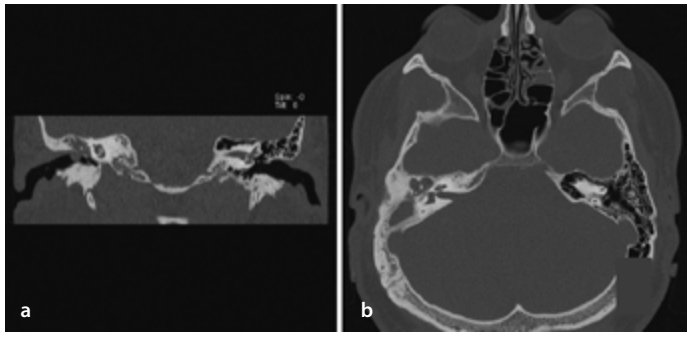

Figure 1. a, b. Coronal CT scan showing Prussak's space occupation with slight erosion of the ossicles and conservation of the scutum (a). Axial CT scan showing epitympanum and antrum occupation and erosion of ossicles heads. Mastoid cells were occupied (not shown here) (b)

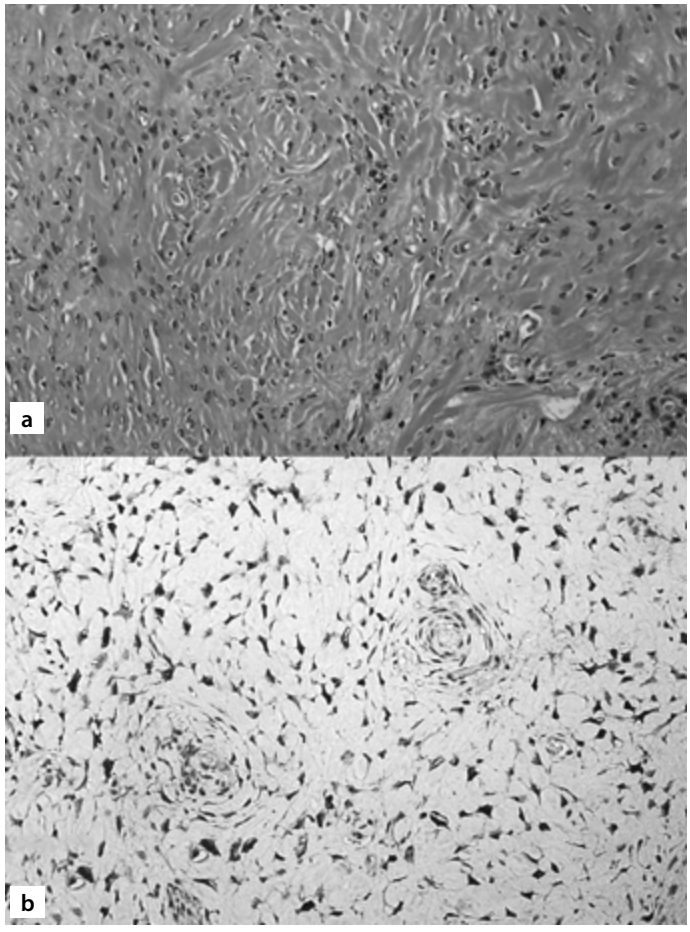

Figure 2. a, b. Microphotograph of the specimen showing a highly collagenised stroma with spindle-cell proliferation, low cellular density, and without an organised growth pattern. There is no palisading, which is typical in schwanommas. HEx20 (a). Positive nuclear and cytoplasmic staining for S-100 using the avidin-biotin-peroxidase technique (b)

DISCUSSION

Chorda tympani tumours reported in the literature usually grow for some time without causing facial paralysis. They are able to fill the middle ear air spaces and erode the bony margins by compression, sometimes mimicking other entities like cholesteatoma. This is why specific imaging such as magnetic resonance imaging (MRI) is not usually requested and diagnosis is made after surgical removal, such as in the case presented here.

Although a loss of gustatory sense might be expected, high variability of innervation from both sides could explain why these patients often do not complain of dysgeusia. Depending on the size and location of the tumour, there may exist hearing loss, as reported in 10 previous cases [2-11] and in our patient (Table 1). Interestingly, seven patients showed a mass in the external ear canal [3-8, 10]. Only two cases were associated with neurofibromatosis [2, 14].

Traditional diagnosis of these tumours has been made by means of conventional staining. However, in some cases the lack of specificity of the haematoxylin-eosin staining prompts an immunohistochemical study. In our case, the immunohistochemical study ruled out some possibilities (Table 2): fibromatosis (lack of expression of beta-catenin); fibroma (positive S-100 expression); perineuroma (lack of EMA expression); meningioma (lack of EMA expression); solitary fibrous tumour (lack of CD34 expression); and inflammatory myofibroblastic tumour (inflammatory pseudotumour; positive S-100 and negative ALK1 expression).

The conventional histology and the immunophenotypic findings suggested three possible diagnoses in the following decreasing order of probability: schwannoma; neurofibroma; and neuroma (Table 2). In our opinion, neuroma was the least probable diagnosis as neuromas have a typical pattern (irregular submucosal nerve bundles surrounded by prominent perineurial cells) that was lacking in our case. Also, there was no history of trauma in the middle ear and the site of origin is uncommon for spontaneous neuromas (which usually arise in the eyelids, tongue, lips, and intestinal mucosa). There are seven English papers describing 'neuromas' of the chorda tympani [1-3, 6, 8, 10, 12], but in all cases it was described as a tumour arising from Schwann cells, which would in fact correspond to a schwannoma and not a neuroma.

The morphological pattern was more suggestive of neurofibroma, although residual axons could not be seen. Neurofibromas as well as neuromas usually express S-100, CD34, and EMA (due to the presence of perineural and epineural cells). Therefore, the immunophenotype in this case ruled out a diagnosis of neurofibroma. No neurofibromas of the chorda tympani have been described to date.

The homogeneous and isolated expression of S-100 in this case can only be explained by the histological diagnosis of schwannoma (also called neurinoma or neurilemmoma). The fact that the morphological pattern was atypical (absence of capsule and Antoni A and B patterns) suggested a low cellularity form (collagenised form).

It is scientifically accepted that immunohistochemistry is essential for pathological diagnosis of peripheral nerve tumours [13]. Only one of the previously published cases employed these methods [7]. It is remarkable that the terminology is often confusing. Nonetheless, treatment of benign nervous tumours of the ear depends more on the site and size than the histology with respect to the approach and expected sequelae. In future, new therapies may need a more complete assessment of the cellular components of each type of tumour.

In conclusion, tumours of the chorda tympani nerve are extremely rare. All the cases described to date (15 unilateral cases) are described as originating from Schwann cells, although the terminology

Table 2. Differential diagnosis of bening tumors of peripheral nerves with immunohistochemical techniques

	S100	EMA	NF	CD34	B-CAT	ALK1
Fibromatosis	-	-	-	-	+	-
Fibroma	-	-	-	-	-	-
Solitary Fibrous Tumor	-	-	-	+	-	-
Inflammatory Miofibroblastic Tumor	-	-	-	-	-	+
Perineuroma	-/+	+	-	-	-	-
Meningioma	-	+	-	-	-	-
Neuroma	+	-/+	+	-/+	-	-
Schwannoma	+	-/+	-	-	-	-
Neurofibroma	+	-/+	+	-/+	-	-
Present Case	+	-	-	-	-	-

S100: protein S100; EMA: epithelial membrane antigen; NF: neurofilaments; B-CAT: beta-catenin (nuclear staining); ALK-1: anaplasic lymphoma kinase 1

employed has been varied (schwannoma, neurinoma, and neuroma). We advocate the use of common terminology to facilitate future research.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

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