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ORIGINAL ARTICLE

Labyrinthitis as a Presentation of Middle Ear Salivary Gland Choristoma

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A 44 year-old woman referred to our ENT DEPT for the study of a progressive hearing loss in right ear (RE) and dizziness. Otoscopy showed by transparency a pinkish mass located on the bottom back of the tympanic cavity.

After imaging studies, a retroauricular surgical approach was performed identifying a mass that occupied the attic and it was closely related with facial nerve, promontory and oval window. Histopathology diagnosis was a salivary gland choristoma of the middle ear, a rare disease whose treatment depends on the degree of facial nerve compromise with the tumour. To our knowledge, this is the first report of labyrinthitis as a presentation of a middle ear choristoma.

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Introduction

Middle ear salivary gland choristoma is a tumour formed by heterotopic glandular tissue from normal characteristics. Since 1961, when Taylor and Martin[1] described it, there have been reported about 25 cases in the literature. It occurs usually in women (2:1) during the first two decades of life with an age range between 3-52 years, as a unilateral conductive or mixed hearing loss in the left ear. It's a benign lesion, whose treatment is complete or partial excision in cases where tumour is closely related with facial nerve, that in most cases will present anatomical alterations associated[2].

Case Report

The patient was a 44 year old woman with a progressive hearing loss in right ear (RE) of one year of evolution. Tinnitus and dizziness were associated.

Otoscopy revealed by transparency a pinkish mass located behind the tympanic membrane. No associated head and neck malformations were found. PTA showed profound RE hearing loss with absence of stapedius reflex and normal hearing of the left ear (LE). Computed Tomography (CT) study showed a soft tissue mass that occupied the attic and covered the ossicular chain (Figure 1). In enhanced Gadolinium MRI an increase of signal level in semicircular canals and in tympanic and mastoid portions of facial nerve canal was observed, with a 5 mm tumor inside the middle ear (Figure 2).

As a complementary study a Videonystagmography (VNG) revealed a hypoexcitability in the right ear.

A surgical retroauricular approach identifying a pinkish mass that occupied the attic closely related with facial nerve, promontory and oval window was performed. We observed a damage of the stapes superstructure and a facial nerve dehiscence. Lesion was completely removed preserving facial nerve function. Microscopic study described a connective tissue, covered by cylindrical ciliated epithelium. In the stroma there was a focus of seromucinous glands (Figure 3). No clinical evidence of recurrence after ten months follow up has appeared.

Discussion

The choristoma is a proliferation of histologically normal tissue that occurs in an ¨unusual region¨. It seems that it appears more often in women and left ears.

Our case is not typical as compared to other cases of salivary gland coristoma: it debuted as a hearing loss in the right ear and dizziness although there have been described all kinds of hipoacusia[3], the most common symptom of this entity are unilateral conductive hearing loss with integrity of the tympanic membrane[4]. Otorrhea, tinnitus, associated malformations of the external auditory canal, seromucous otitis and otalgia have also been described[5]. The structures most frequently affected

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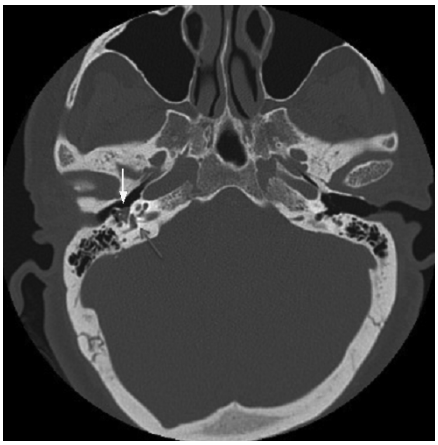

Figure 1. CT axial slide showing a middle ear mass in contact with inner ear (yellow arrow: choristoma, red arrow: vestibule).

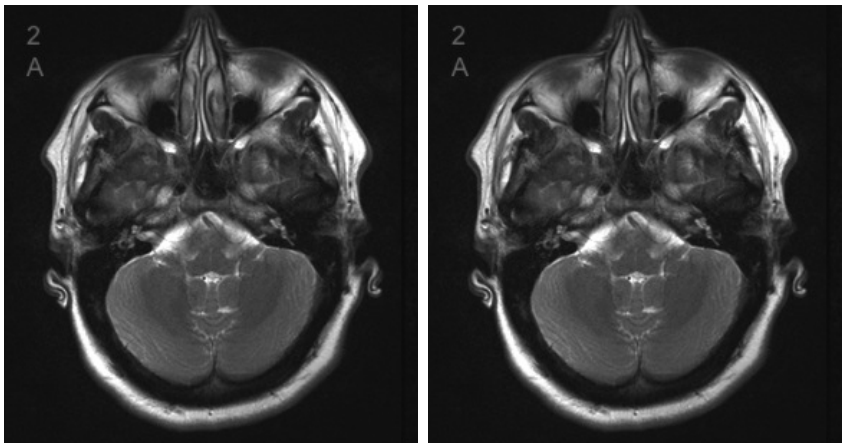

Figure 2. MRI T1 axial slide (2A) MRI T2 (2B) showing an increase of signal level in semicircular canals and facial nerve canal in its tympanic and mastoid portions.

are the long crus of the incus, which may be absent, hypoplastic or fixed and the horizontal portion of the facial nerve, which may have an abnormal (inferolateral) position or be dehiscent. Alterations of the malleus, stapes, oval or round window and even the absence of any muscle of the ossicular chain have also been described[6]. Our case had a compromise of the ossicular chain and labyrinthitis because of the involvement of the round and oval window, vestibule and –semicircular -canals.

The etiology is still unknown. It has been associated with abnormalities at the first and second branchial arches that would generate alterations in the otic capsule and auricular tubercles[7]. Others[8], have related to the development with a congenital cholesteatoma. Choristoma histopathology can present serous and/or mucous tubuloacinar glands forming a homogeneous tissue4 as in this case (Figure 3).

High-resolution CT is preferred and MRI also is a useful imaging tool[9]. Differential diagnosis should be done with paragangliomas, schwannomas, rhabdomyosarcoma, dermoid cysts, congenital cholesteatoma and other teratogenic lesions[5]. The definitive diagnosis will be done with histologic study of the surgical specimen. Resection is the election treatment and will be total or partial depending on the relationship with the seventh pair[10]. Facial nerve monitoring is advisable in order to achieve better facial nerve preservation. Laser excision, if available may be helpful in dissection. Ossicular reconstruction can be accomplished and usually improves hearing[11]. There has not been evidence of malignant transformation in the long term follow up[12].

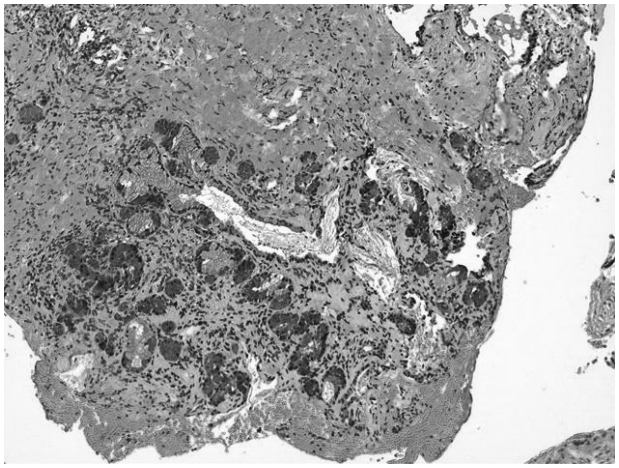

Figure 3. H&E stain at 60x magnification, showing a connective tissue, recovered by cylindrical ciliated epithelium. In the stroma there is a focus of seromucinous glands.

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