Neuroglial choristoma of the middle ear

Coristoma neurogliale dell’orecchio medio

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SUMMARY

Neuroglial choristoma is a rare cerebral heterotopia which can involve various locations. Even if its occurrence is more frequent in midline structures, it can also be found in the non-midline structure such as, for example, even if only rarely, the middle ear. The described case is that of a 74-year-old male who had been operated on for a neuroglial choristoma located in the tympanic cavity and in the mastoid bone. High resolution computed tomography and intra-operative findings did not reveal any connection with the upper cerebral structures, thus excluding the hypothesis of an encephalocele. After careful histopathological examination, the aetio-pathogenetic hypothesis are analysed. Only 8 similar cases have been reported in the literature.

KEY WORDS: Middle ear • Neuroglial choristoma • Encephalocele

INTRODUCTION

With the term choristoma, we usually indicate histologically normal heterotopic tissue. Extra-cranial cerebral heterotopia usually occurs along the midline structures, including the nose 1 and nasopharynx, oropharynx, lips, soft palate, tongue and tonsils 23 while non-midline extra-cranial localizations (such as eye, lung, skin) are rare 45. In the literature, only 8 very rare cases of neural choristoma of the middle ear have been reported 246-10. On the other hand, a salivary gland choristoma, in the same localization, is relatively more frequent (≤ 30 cases described) 11-12. Because of the rarity of these lesions, it is very important to differentiate them from the more frequent enchephaloceles by looking for any cerebral connection in order to avoid the post-operative risk of recurrent infections 5. Herein, a case of neuroglial choristoma of the middle ear is described.

CASE REPORT

A 74-year-old male presented with recurrent bilateral otitis which had been present since early childhood. There was no evidence of congenital anomalies, no previous trauma reported or surgery on the middle ear.

At otoscopic examination of the left ear, the patient showed a small attic retraction pocket of dubious interpretation (no clear evidence of cholesteatoma). On the right side, a wide retraction pocket was found, the end of which was difficult to detect.

An audiometric test of the left ear showed a mixed hypoacusia with a severe perceptive component; on the right side, a mixed hypoacusia with a perceptive component of 30-40 dB at medium frequency, and a sensori-neural component of 50-60 dB at 4 and 8 kHz.

High resolution computed tomography (HRCT) of the left ear revealed evidence of a soft mass which partially involved the left tympanic cavity, occupied the epitympanum, aditus ad antrum, antrum and the adjacent part of the mastoid with no evidence of ossicle erosion; tegmen tympani and antrum were not involved. There were no signs of erosion of the wall of the cavity or semicircular channels (Fig. 1).

HRCT of the right ear showed the outcome of a chronic otitis with spontaneous atticotomy; the anvil and the hammer head were absent while it was possible to recognize the superstructure of the stapes. There was also evidence of a fistula in the lateral semicircular channel.

Radiographic signs of a neoformation localized mainly in the aditus and the antrum of the left ear suggested cholesteatomatous chronic otitis, thus requiring surgical treatment.
The closed tympanoplastic technique was, therefore, carried out with the identification of the typical landmarks (dura mater of the middle cranial fossa, sigmoid sinus and the posterior wall of the auditory canal). Next to the antrum and the aditus ad antrum, a mass of taut elastic consistency was identified, which was well-cleavable from the surrounding bony structures which partially projected into the epitympanum and the adjacent portion of the mastoid. Localization in the antrum required removal of the anvil and the hammer head by means of disarticulation. The mass was pushed into the tympanic cavity and removed. In order to control the posterior recesses of the tympanic cavity, posterior tympanotomy was performed which confirmed the absence of residual disease. The epitympanic gap was rebuilt with tragal cartilage, and the attic retraction pocket was reinforced with tragal perichondrium. On account of the macroscopic aspect of the neoformation in the left ear, which was totally different from a cholesteatomatous form, histopathologic studies were carried out (Fig. 2).

The resected specimen was fixed in formalin and paraffin embedded according to routine procedures. From the paraffin embedded sections, serial sections were cut at 5 μm and stained with hematoxylin and eosin (H&E) for light microscopy. The glial component of the lesion was stained with GFAP antibody (Fig. 3).

**Fig. 1.** HRCT of left ear demonstrates how, the soft mass shadow occupying most of the middle ear, does not involve tegmen tympani and antrum.

**Fig. 2.** Trans-mastoid view of mass occupying the antrum.

**Fig. 3a.** Lesion is composed of glial tissue containing nodules of neuronal cells (H&E).

**Fig. 3b.** GFAP antibody strongly stained the glial tissue composing the lesion.
fin block, serial sections were cut and stained with haematoxylin-eosin (H&E) and then with an immunohistochemical method, applying the ABC method in an automatic stainer (Ventana, USA).

Macroscopically, the lesion presented as a whitish nodule with a smooth surface, with a maximum diameter of 0.8 cm. Histologically, the lesion was composed of glial tissue, containing clusters of neurons (Fig. 3a). The lesion was covered by a layer of epithelium. The glial nature of the lesion was confirmed by positivity with anti-Glial Fibrillary Acidic Protein (GFAP Dako, monoclonal, clone M761, diluted 1:1200) (Fig. 3b). Neurons were stained with an anti-NEU N antibody (Chemicon, Canada, monoclonal, clone MAB 377, diluted 1:500) (Fig. 4a). The anti-cytokeratin antibodies (Dako monoclonal, clone MNF116, diluted 1:200) showed the epithelium overlying the lesion and rare glandular structures entrapped inside the glial tissue (Fig. 4b).

No cytological atypia, necrosis or atypical mitoses were observed.

The post-operative course of the patient was good. Twenty-five months after surgery, outcome of tympanoplasty was stabilized. There were no signs of neurological complications, especially CSF leaks, in the post-operative period.

**Discussion**

Neuroglial heterotopies, so-called neuroglial choristomas, are usually located extra-cranially on the midline. The most frequent localization is at nose level (so-called nasal glioma) \(^1\) but it is also found in the pharynx, palate and tongue \(^2,3\).

Extracranial non-midline lesions are even more rare. Middle ear choristomas are mostly salivary and 25 cases have been described in the literature but only 8 glial or neuroglial choristomas of the middle ear \(^2,4,6-10\). In the case under examination, the absence of any alterations of the tegmen seen at HRCT or encountered during surgery and the absence of any predisposing factors and any cerebro spinal fluid (CSF) leaks confirm the lack of any communication with the cerebral structures. This also confirms the hypothesis of a real heterotopy. In order to avoid over-treatment due to an incorrect diagnosis, it is important to carry out a scrupulous histological examination which permits the surgeon to make a differential diagnosis between the most frequent space-occupying lesions of the middle ear: teratoma, ganglioma, meningioma, neuroma, schwannoma \(^5\), adenocarcinoma, cholesteatoma and hamartoma \(^13\).

During surgery, the lesion appeared as a small roundish mass, easily cleavable from the layer below without any relationship to the tegmen tympani and antri which appeared to maintain their integrity. Moreover, the lesion had no relationship with the facial nerve which was embodied in the bony channel.

In some cases described in the literature \(^9,14-16\), the choristomas were strictly adherent to the facial nerve; however, these were glandular forms the presence of which was aetiopathogenically associated with ossicular chain and facial nerve abnormalities. In these cases, probably caused by an error in the development of the second branchial pouch, it is advisable to perform an intra-operative exam and, if the result is confirmed to be benign, the lesion can be partially removed \(^13\). The
different aetiopathogenic origin of neuroglial choristomas does not foresee such anomalies, as confirmed by studies reported in the literature. Histologically, the lesion was composed of glial cells, also found in the other cases described in the literature, and, less frequently, of an abundant number of neural cells. Even rarer is the presence of ependyma and choroid plexus in glial heterotopies. According to Genut et al., the presence of neuronal cells depends upon the stage in which the separation of the embryonal tissue, from which the choristoma originated, took place. In fact, neuronal precursors appear in the developing brain at the 10th week of intra-uterine life so there would be no neuronal elements in the heterotopic brain if the embryonic tissue has been separated prior to the 10th week. On the contrary, if the separation occurs after the 10th week, a high neuronal content would be possible.

The most reliable aetiopathogenetic hypothesis to try to explain this eventual heterotopy is that of a small congenital defect in the overlying temporal bone, the tegmen tympani, which permitted herniation of the cerebral tissue. The successive closure of this defect would justify the absence of any connection with the central nervous system (CNS). A pedicle directly connecting the neuroglial tissue with the CNS may become detached, eventually absorbed or vestigial; this would justify the term of “heterotopy.” According to Plontke et al., another aetiopathogenic hypothesis could be that of neural crest remnants.

According to Heffner, in order to distinguish between a choristoma and an encephalocele, the rarity of a middle ear choristoma is explained by the following factors:

- embryologic considerations: there does not seem to be a connection in the development between ectoderm and neuroectoderm in the area of the middle ear as occurs in the midline area. The middle ear epithelium develops from the first branchial pouch and, during early embryogenesis, this tissue is notably displaced from the developing brain;
- average age of patients: it is usually found in older people. It seems unlikely that a lesion, which does not usually tend to grow significantly, would be symptomatic only after decades, especially when the middle ears of children are more prone to produce symptoms;
- the tegmen tympani is very thin and, for this reason, encephaloceles are not rare at all. In some cases, there are no surgically or radiologically recognisable bony defects or CSF leaks. The lack of evidence of a connection does not necessarily prove that it does not exist.

However, in our case, the HRCT, pre-operative and intraoperative findings can exclude any cerebral connection with great certainty. We are reasonably convinced that we are dealing with a rare case of neuroglial choristoma.

References


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