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Middle Ear Adenoma, A Rare and Controversial Diagnosis: Case report and Discussion

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Learning Objectives: Learning Objectives Middle ear adenomas are exceedingly rare Pre-operative radiological workup does not always correlate with intraoperative findings and clinicians should keep an open mind Histological classification of middle ear adenomas remains controversial Surgical resection is the treatment of choice.

Introduction: Middle ear adenoma is a rare differential diagnosis for a middle ear mass. Histological classification of middle ear adenomas remains poorly defined. As well as presenting our experience of one such case, this study aims to assimilate findings from previously published cases in order to contribute to our knowledge of a rare diagnosis.

Method: We report the case of a 51 year old male who presented with symmetrical tinnitus and left sided hearing loss. Examination revealed a postero-inferior mass behind the left tympanic membrane. CT and MR imaging was consistent with a tumour arising from the chorda tympani nerve and a pre-operative diagnosis of chorda tympani neuroma was made. However, intra-operative findings revealed a well-defined mass, close to but distinct from the chorda tympani nerve. Histopathology of the tumour was consistent with a middle ear adenoma.

Results: A review of the literature shows 95 previous reported cases of middle ear adenoma with the entity first described in 1976. A common theme is the difficulty of making a correct pre-operative diagnosis when faced with a middle ear mass, despite modern imaging techniques. Histologically, middle ear adenomas continue to defy classification with their cell line of origin posing the main point of controversy: exocrine versus neuroendocrine cell types. This in turn poses controversies regarding treatment and follow up.

Conclusion: Middle ear adenomas are rare. Reported cases highlight the difficulty of interpreting preoperative imaging. The histological classification of these tumours poses a further difficulty. With the natural progression and prognosis if left untreated of these tumours remaining unknown, further studies and reports would be a welcome addition to the literature. A current literature review advocates a radiological workup and surgical excision where there is clinical suspicion of middle ear adenoma

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The cytokeratin pattern of congenital and acquired cholesteatoma, epidermoid, medial and lateral canal wall skin

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Learning Objectives: In this paper we present a study in which cytokeratins have been used to characterize congenital cholesteatoma and epidermoid (and we compared these patterns with previous data on acquired cholesteatoma and ear canal skin) in order to confirm or invalidate a developmental theory.

Introduction: Histologically ear canal skin, epidermoid, congenital and acquired cholesteatoma are indistinguishable. They all contain keratin, a matrix of keratinizing stratified squamous epithelium and a lamina propria-perimatrix. Nowadays still different theories on the development of congenital cholesteatoma and epidermoid are suggested.

Cytokeratins are intermediate filament proteins that are exclusively present in epithelial cells and can be used to study epithelial differentiation.

In this paper we present a study in which cytokeratins have been used to characterize congenital cholesteatoma and epidermoid (and we compared these patterns with previous data on acquired cholesteatoma and ear canal skin) in order to confirm or invalidate a developmental theory.

Methods: Cytokeratin Antibodies RCK103, RCK105, M20, CK18–2, LP2 K, AE14, RCK107, E3, KA12, LL025, RKSE60, 6B10 en 1C7 were used to characterize the cytokeratin pattern of congenital cholesteatoma and epidermoid of the cerebellopontine angle. These data were compared with previous patterns on acquired cholesteatoma and medial and lateral ear canal skin.

Results: Our results show that the cytokeratin pattern of congenital cholesteatoma and epidermoid differs. The cytokeratin expression of congenital cholesteatoma mimics the pattern of acquired cholesteatoma and medial ear canal skin: slightly positive LP2 K (Ck19), AE14 (Ck5), 6B10)Ck10) and 1C7 (Ck13). The pattern of epidermoid is comparable to that of normal skin: negative LP2 K, 6B10 and 1C7 and positive AE14.

Conclusions: The cytokeratin pattern of congenital cholesteatoma, acquired cholesteatoma and medial ear canal skin coincide. This may support the invasion theory as explanation of its development. Epidermoid and lateral ear canal

skin share the same cytokeratin pattern which may support the ectodermal rest theory.

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Novel human stem cell-like cells in middle ear cholesteatoma tissue and auditory canal skin

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Learning Objectives:

Being a potential life-threatening middle ear disease, cholesteatoma is an abnormal expanding cystic lesion leading to extensive tissue destruction in the temporal bone followed by conductive and sensorineural hearing loss and facial nerve palsy. Facilitating further infections beyond those of the middle ear, cholesteatoma may also result in meningitis or intracranial infections. Since surgical removal of cholesteatoma remains as the only therapeutical option, lack of non-advanced medical care results in increased pediatric morbidity, emphasizing the need of developing new treatment strategies.

Here we show for the first time the presence of a novel stem cell-like cell population in cholesteatoma tisse and auditory canal skin. Immunohistochemical analysis of cholesteatoma tissue revealed the presence of Nestin-expressing cells localized subepithelially within the matrix and perimatrix. Nestin-positive cholesteatoma-derived stem-like cells (CSCs) were successfully isolated and cultivated *in vitro* and showed the capability of neurosphere formation and clonal growth. CSCs were further successfully expanded within a human blood-plasma derived three-dimensional matrix. In accordance to the classification of cholesteatoma, proliferative Ki67-positive CSCs also showed a normal euploid DNA content and karyotype. We further observed no changes in proliferative capability and expression profile between CSCs and Nestin-expressing cells isolated from auditory canal skin (auditory skin derived cells, ASCs). In particular, cultivated CSCs and ACCs expressed epithelial and neural crest-specific stemness markers.

Our findings gain new insights in the complex biology of cholesteatoma and may thus broaden the range of treatment strategies for this severe lesion within the middle ear.

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The role of preoperative gadolinium enhanced magnetic resonance imaging (MRI) in anticipating postoperative middle ear aeration after canal wall up tympanoplasty for cholesteatoma

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Learning Objectives:

Objective: This study was designed to anticipate the postoperative middle ear aeration using preoperative gadolinium enhanced magnetic resonance imaging (MRI) after canal wall up tympanoplasty (CWU) for cholesteatoma.

Materials and methods: Retrospective review was performed on 56 patients with mastoid involvement undergoing CWU tympanomastoidectomy without mastoid obliteration at a single institution from 2010 to 2013. In all patients, the cholesteatoma was removed by a combined approach. The communication between the Eustachian tube and the attic was reestablished with a posterior and anterior tympanotomy. The attic bony wall defect was reconstructed using sliced auricular cartilage and fibrin glue (scutum plasty). Patients were classified into two groups according to the status of enhancement around the cholesteatoma sac using preoperative MRI: Group A (strongly enhanced) and Group B (weakly or no enhanced). In each groups, restoration of the middle ear aeration was assessed with high-resolution computed tomography (CT) before and after operation. Status of aeration was classified into 4 grades (no aeration, mesotympanum, epitympanum, mastoid).

Results: Although the middle ear aeration ameliorated in both groups, the range of re-aeration was much better in Group A (strongly enhanced group) than Group B. Re-aeration to the mastoid was achieved in 68% of the cases in Group A, 36% of the cases in Group B.

Discussion and Conclusion: The enhanced MR image was found to be related to postoperative middle ear aeration. These findings might be particularly useful for predicting re-aeration of acquired cholesteatoma.

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The results of obliteration technique in canal wall up and wall down tympanomastoidectomy in patients with acquired middle ear cholesteatoma

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Learning Objectives:

Introduction: The objective in the surgical management of acquired middle ear cholesteatoma is eradication of disease