were observed in MEC matrix and perimatrix. There was no meaningful difference between congenital and acquired MEC with respect to p21 contrary to p53. A statistical significance was obtained for APO2.7-positive cells in MEC epithelium (43.23 ± 4.8%) as compared to CS (29.89 ± 6.2%).

More extensive positive immunohistochemical reaction with anti-TGF-alpha, Ki67 and PCNA was observed in MEC matrix and perimatrix compared with CS.

RAGE expression levels was present in all cholesteatoma tissues (strong in 86 %) vs skin 25% (weak) respectively ($p < 0.0001$).

Conclusion: Selected markers of apoptosis, proliferation, angiogenesis and inflammatory response are associated with cholesteatoma development. The co-expression of HMGB1 and RAGE in MEC may result in activation of the intracellular signaling pathways. This process may be responsible for faster accumulation of keratin debris, more invasive process, and affect the clinical course and the treatment outcome.

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Congenital Cholesteatoma (R634)

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Congenital cholesteatoma of the middle ear: a report of 62 cases

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Introduction: Congenital cholesteatoma (CC) of the middle ear is a rare clinical entity that classically presents as a white mass situated in the anterior-superior quadrant of the middle ear behind an intact tympanic membrane (TM). Derlacki and Clemis established the diagnostic criteria for CC: 1) A pearly white mass medial to an intact TM, 2) Normal Pars Tensa and Pars Flaccida, 3) No history of otorrhea, perforation or previous otologic procedures. CC is seen far more frequently in children, but House and Sheehy remarked adult patients with cholesteatoma behind an intact TM.

Materials and Methods: A retrospective analysis was conducted of the clinical charts of all patients with CC in both children (n = 56) and adults (n = 6) from 1992 to 2015. CCs of the petrous apex (n = 15) were excluded. 1445 cases of acquired and congenital cholesteatomas were treated, therefore, the prevalence of CC should be 4.3% (62/1445).

Results: Based on the staging system by Potsic 54 patients were classified into stage1–4 according to the surgical findings: 11 cases in stage1, 7 in stage 2, 24 in stage3, and 20 in stage4. It was suggested that most CCs could be derived from the epidermoid formation (EF) in 53 cases. A planned two-staged surgery was conducted in 54 cases (87%), while one-stage surgery was adopted in 8 cases. The residual cholesteatoma at the time of second stage surgery was detected in 19 out of 48 cases (40%). The most common residual sites were at oval window (n = 7). Hearing assessment was done in 55 cases: success in 46 cases (84%), moderate in 8 cases, and failure in one.

Discussion: As the stage of CC advanced, the area of its invasion could be enlarged, which should result in a higher risk of CC residual. Considering that CC is usually discovered in its advanced stages (stage 3–4), the establishment of a screening program including otoscopic and CT examinations and hearing tests for early CC diagnosis should be required.

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Back to the future: the evolution of cholesteatoma diagnosis and management (N635)

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Back to the Future: The Evolution of Cholesteatoma Diagnosis and Management

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Confucius once said, “Study the past if you would define the future.” As an introduction to the 10th International Conference on Cholesteatoma and Ear Surgery, the American Neurotology Society has assembled panelists (Mr. David Moffat (Addenbrooks Hospital), Dr. Jack Lane (Mayo Clinic), Dr. Clough Shelton (U. of Utah), Dr. Moises Arriaga (LSU) and Dr. Dennis Poe (Harvard) to discuss the evolution of cholesteatoma diagnosis and management. Dr. John McElveen (Carolina Ear Research Institute) will moderate the panel.

Mr. Moffat will trace the history of the diagnosis of cholesteatoma from ancient times to the present. Based on the research in 1967 by McKenzie and Brothwell, the existence of chronic suppurative otitis media in prehistoric times has been clearly documented. It was the French anatomist Joseph-Guichard Du Verney who in 1683 first described a temporal bone tumour which was probably a cholesteatoma. However, the term, “cholesteatoma”, was first used by Johannes Peter Muller in 1838. Although a misnomer, it has continued to be used to describe “keratomas” involving the temporal bone and skull base. Abramson *et al* in 1977 provided a more detailed definition of cholesteatomas at the First International Conference on Cholesteatoma.

The classification of cholesteatoma into congenital and acquired and the latter’s subdivision into primary and secondary acquired was the natural sequel of refinements in diagnostic capability which accompanied the use of the microscope both in histopathology and in the clinical examination of the ear (Nylen, 1921).

Since the dawn of medical imaging, radiographic examination of the temporal bone has been used in the evaluation and management of cholesteatoma. X-ray modalities have evolved from plain radiographs (1900–1940s) to polytomography (1950–60s) to single slice Computed Tomography (CT) acquired separately in the axial and coronal planes (1970–1980s) to multislice CT with multiplanar

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