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Case Report

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Congenital Mastoidal Cholesteatoma in an 87-Year-Old Woman Treated by Watchful Waiting

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Keywords

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Introduction

Congenital cholesteatoma (CC) of the ear is a benign, slow-growing epidermoid cyst consisting of keratinized squamous epithelium, which may expand and destroy surrounding tissues. For a clinical case to be considered as a CC, there should be no previous history of tympanic disease, such as tympanic membrane retractions or ear discharge, and no previous ear surgery. This occurs most commonly in the middle ear, and because it typically causes ossicular erosion with hearing impairment, detection tends to occur relatively early.[1]

Congenital mastoidal cholesteatomas (CMCs; ie, CC originating from the mastoid process) are rarer, and because they cause few early symptoms, they tend to go undetected for many years. Thus, they are often detected in adulthood.[2] The diagnosis is confirmed with radiology, which is conclusive when applying current technologies. This includes computed tomography (CT) and magnetic resonance imaging (MRI) with a cholesteatoma protocol. High-resolution CT displays a nonspecific soft-tissue mass without connection to the tympanic membrane or to the external ear canal. On MRI, the cholesteatoma appears hypointense on T1-weighted images and hyperintense on T2-weighted images.[3] The current standard of treatment is surgical eradication.

Case Report

An 87-year-old woman with a history of hypertension and polycytemia vera was admitted to the hospital after a fall at home. A CT of her skull was made to exclude a subdural hematoma, with only degenerative cerebral findings normal for her age seen. As an incidental finding, there was an expansive mass in the posterior portion of her left mastoid process, which resulted in her being referred to the ear, nose, and throat department for further evaluation.

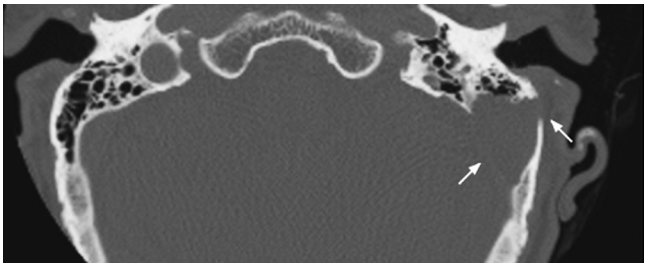

Figure 1. Computed tomography showing an expansive lesion in the left mastoid with large bony erosions toward the middle and posterior cranial fossae in the area of the sigmoid sinus and, laterally, toward the skin.

Otomicroscopic examination showed bilateral normal tympanic membranes, and tone audiometry demonstrated bilateral symmetric sensorineural hearing loss of high frequencies with no air-bone gap. The patient's clinical record was reviewed, ensuring no history of aural symptoms or surgery. A high-resolution CT was done, showing an expansive and destructive lesion measuring 18 × 20 × 30 mm located posteriorly in the mastoid process (**Figure 1**). The anterior mastoid air cells, the middle ear, and the antrum were unaffected, and there was no exposure of the facial nerve, cochlea, or vestibular aqueduct. On MRI, the lesion appeared hypointense on T1-weighted images and hyperintense on T2-weighted images, confirming the presence of a

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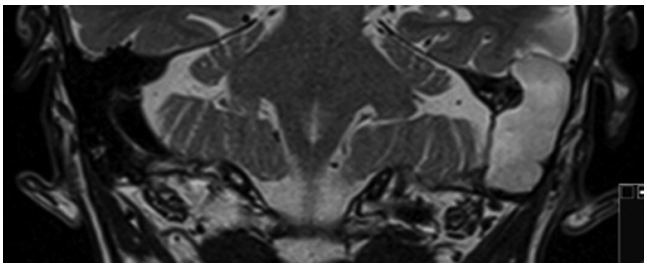

Figure 2. T2-weighted coronal magnetic resonance imaging shows a hyperintense lesion in the left mastoid.

cholesteatoma (**Figure 2**). At 1-year follow-up after watchful waiting, repeated clinical and radiologic examinations revealed no further progress of the cholesteatoma.

Discussion

This is the oldest case of a CMC reported in the literature. It demonstrates that a CMC may grow to a large size with extensive bony erosions without impairing the patient's hearing or quality of life. If a CT had not been performed on this woman, her disease would have gone undiagnosed. One can therefore suspect that many other similar cases escape detection during a lifetime.

The present case demonstrates that the growth rate of cholesteatoma may vary considerably. From clinical experience, we know that the growth rate may vary between different cases of cholesteatoma, and several factors have been suggested as promoters for fast growth. Young age and the presence of infection are most often discussed. In this case, the cholesteatoma was likely present at both a young and old age. One may speculate that the CMC may have had a faster growth period in the patient's youth, while slowing down later on. The absence of ear infections and the localization of the present CMC in a mostly sterile environment may have also contributed to a slow progress.

Finally, the present case inspires the idea that the diagnosis of CMC per se is not a compelling indication for surgery, especially in the absence of troublesome symptoms. Two groups have suggested that the radiologic and intraoperative findings alone are an indication for surgery.[4,5] For example, Giannuzzi et al[4] presented a case of a 60-year-old man with an incidental discovery of a CMC on MRI. The patients' symptomatology was not described. Because the finding was incidental, our interpretation is that the authors recommended surgery entirely based on radiologic findings. Our case confirms that radiologic findings of CMC, even when combined with bony erosion, do not per se indicate for surgery. A watchful waiting strategy may sometimes be a better option.

Ethics committee approval is not required according to Swedish law for case reports. The patient has approved with informed consent the anonymized publication of her medical record.

Author Contributions

Elnaz Sepehri, data analysis, PubMed search and drafting, followed up on the case, imaging of the case, final approval, accountable for text; **Magnus von Unge**, found the case in hospital, read through data analysis and drafting, contributed with changes in text, sources, and final approval, accountable for final text.

Disclosures

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References

1. Mevio E, Gorini E, Sbrocca M, et al. Congenital cholesteatoma of the mastoid region. *Otolaryngol Head Neck Surg*. 2002;127:346-348.
2. Nager GT. *Pathology of the Ear and Temporal Bone*. Baltimore, MD: Williams and Wilkins; 1993.
3. Thakkar K, Djalilian H, Mafee M. Congenital cholesteatoma isolated to the mastoid. *Otol Neurotol*. 2006;27:282-283.
4. Giannuzzi A, Merkus P, Taibah A, Falconi M. Congenital mastoid cholesteatoma: case series, definition, surgical key points, and literature review. *Ann Otol Rhinol Laryngol*. 2011;120:700-706.
5. Luntz M, Telischi F, Ress B, Bowen B, Balkany T. Congenital cholesteatoma isolated to the mastoid. *Ann Otol Rhinol Laryngol*. 1997;106:608-610.