Bilateral congenital cholesteatoma – Review of literature and report of a case complicated by acute mastoiditis

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Introduction
Bilateral congenital cholesteatoma are very rare. Especially in children the role of unnoticed congenital cholesteatoma should not be underestimated. Complications such as hearing loss impairing acquisition of speech, vestibulopathy, mastoiditis and even sinus venous thrombosis can lead to a severe outcome for the young patient. Therefore, quick diagnosis, sufficient therapy and prevention of long term impairment are important. A recent case of bilateral genuine cholesteatoma in our clinic complicated by unilateral acute mastoiditis led to this review of literature. This is the first case ever reported with acute mastoiditis.

Results by review of literature
33 cases (ours included) have been found, 23 male, 4 female cases, 6 gender unknown. Peak age was between 4 to 6 years. Whenever a CT-Scan was performed (16 times), it showed a pathology on at least 1 side (3 cases, one with later-onset cholesteatoma on the other side). One time, CT showed cholesteatoma on the contralateral side 15 months before it was seen in ear examination, in one case there was a finding on both sides with normal ear-examination. Ossicular erosion could be seen in 6 cases, 3 had more extensive lesions such as erosion of the labyrinth and petrous bone.
5 of 18 patients had an otitis, 4 bilateral, 2 with otorrhea. There were 2 cases with asymptomatic effusion. Hearing loss was found in 11 of 17 cases, mainly conductive and no sensorineural hearing loss only.

Ear examination showed a white mass on both sides in 12 of 18 patients, 1 time unilateral mass.
There were 2 patients with auricular fistulas (one with branchio-oto-renal syndrome, one with mental retardation). 5 patients carried comorbidities combined with more extensive findings (in ear examination as well as in CT Scan) and a severe hearing loss.

Case Report
A 1.5 year old boy was referred as an emergency with right sided otitis media for 2 weeks. 1 day ago, he developed otorrhea and a protuberant ear.
A CT scan showed masses filling up both mastoidal cavities by whole with bony erosion of the right lateral petrous bone as well as ossicular erosion right more than left. (Fig. 1a-d)
Surgery was immediately performed, revealing a cholesteatoma filling up the whole tympanon as well as the mastoid cavity and erosion of all ossicles. The chorda could not be identified. Ear examination showed an intact tympanic membrane left with signs of effusion or cholesteatoma behind it. There was no adenoid hypertrophy to be found.
Ear surgery on the left side shortly performed thereafter revealed cholesteatoma filling up the tympanon and mastoidal cavity as well as ossicular erosion.

2nd-look-surgery (+BERA indicating a conductive hearing loss of 30dB right) on the right presented a big relapse cholesteatoma filling the whole mastoidal cavity again. (Fig. 1a-d) Hence, 2nd-look-surgery to the other side was preponed also presenting a relapse less extensive.

Right now, waiting for re-2nd-look-surgery, ear examination presents intact and clear tympanic membranes on both sides, the boy has been supplied with hearing aids and seems to be developing well.
There were no known other diseases except for a non-relevant ventricular septal defect and no history of oitis with effusion before. On a side note, the child had a dacrycotic twin, ear examination showed no retraction pockets or white masses.

Key Points / Conclusion
- Rapid growth of cholesteatoma in young patients should not be underestimated, 2nd-look-surgery therefore under circumstances preponed
- CT Scan is a superior option in diagnosing uncertain findings in ear examination, especially for congenital cholesteatoma even in young children
- Follow-up ear examination is very important to identify relapse or contralateral later-onset cholesteatoma
- The major symptom seems to be hearing loss more than oitis, effusion or otorrhea
- Comorbidities are linked to more extensive findings, later-onset diagnosis and should be examined carefully
- Male subjects tend to be affected far more often than female ones

References

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