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Cholesteatoma in children

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Abstract
Of 325 previously untreated cholesteatomas, 109 were found to be in children 13 years old and younger—however, these 109 ears were not found to consist of one clinical entity. Sixty-three of the 109 ears presented a marginal perforation or a retraction pocket, at the level of Shrapnell's membrane or beyond the postero-superior quadrant. In this group the cholesteatoma was distributed mainly in the attic and mastoid and was associated with a non-cellular mastoid. A second group comprising 31 ears presented with cholesteatoma behind an intact drum and were considered to be primary cholesteatomas. These were distributed mostly in the tympanic cavity as cystic epidermoid formations—their mastoid was usually pneumatized. Eight cholesteatomas were related to a central perforation. These ears presented features very similar to the primary cholesteatomas i.e., a pneumatized mastoid and tympanic cavity distribution. The similar features of this group make us think that they may have also originated as primary cholesteatomas which eventually perforated and bring the percentage of primary cholesteatomas in children to 38.3%. Seven of the 109 ears with cholesteatoma were of an indeterminate character.

Cholesteatoma in children is customarily viewed as a separate and special clinical entity. It is supposed to be more aggressive than in adults (Baron, 1969) and to be usually associated with a pneumatized mastoid. However, no detailed qualitative or quantitative data characterizing these presumed specific features of cholesteatoma in children have so far been presented. This study was prompted by our clinical observations that cholesteatoma in children may actually be *a non-homogenous entity*. A cholesteatoma was defined by us as the presence of stratified squamous epithelium in the middle ear which cannot clear itself. This will include the following entities:

A. Deep-seated retraction pockets whose fundus, when containing non-self cleansing keratin, cannot be visualized—and cannot be cleansed by suction.
B. Attic and postero-superior perforations leading to deep-seated cholesteatomas which in all probability are a more advanced stage of a deep-seated retraction pocket (Figs 1 and 2).
C. Epidermoid cysts behind an intact drum which mostly appear as a cystic formation (Fig. 3). At times stratified squamous epithelium was found behind an intact drum, proliferating freely over the promontorium in place of the normal middle ear mucosa (Fig. 4).

It should be emphasized that while it is usually easy to recognize a cholesteatoma—some cases are borderline and are termed by some as retraction pockets and as cholesteatomas by others.

Material and methods

During a 10-year period (1976–86) 325 cholesteatomatous ears which had not previously been treated surgically, were operated at the ENT Department of the Meir General Hospital, Kfar Saba, Israel. Two hundred and sixteen (67%) of these ears belonged to patients 14 years and older; these were considered as the adult population. One hundred and nine ears (33%) belonged to infants and children younger than 13 years; these were considered as the infant and childhood population and comprised the material for this study.

The 109 children with cholesteatoma were subclassified into four categories according to the pathology presented at the drum level as seen under the operating microscope:

Group A. Retraction pocket cholesteatoma (R.P. Cholest.): these presented deep-seated, attic or postero-superior retraction pockets, or alternatively perforations in one of these places leading to a deep-seated cholesteatoma.
Group B. Primary cholesteatoma (Prim. Cholest.) were defined as presenting a cholesteatoma behind an intact tympanic membrane with no atelectatic retraction or any other tympanic membrane abnormality.
Group C. Cholesteatomas associated with central perforations (C.P. Cholest.).
Group D. Undetermined situation—where it was impossible to describe with certainty the appearance of the tympanic membrane as being associated with one of the above situations. This was usually due to extensive destruction or swelling of the middle ear structures, which accompanied an acute exacerbation of the inflammatory process.

The first three groups were analyzed according to (1)

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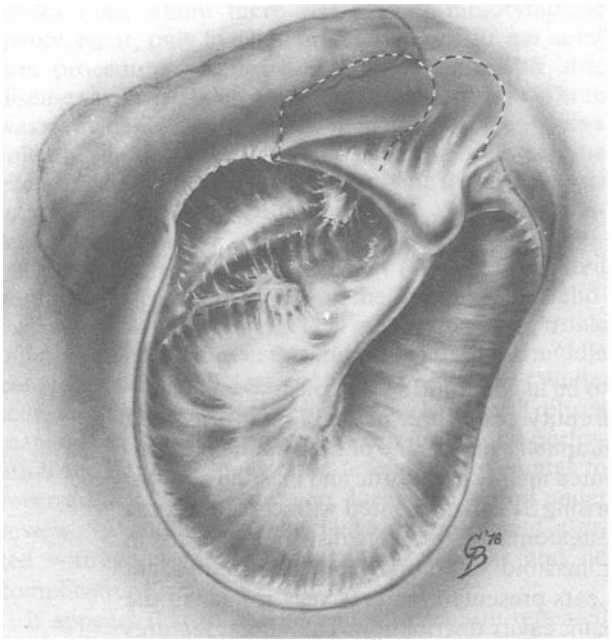


FIG. 1:
Postero-superior retraction pocket cholesteatoma.

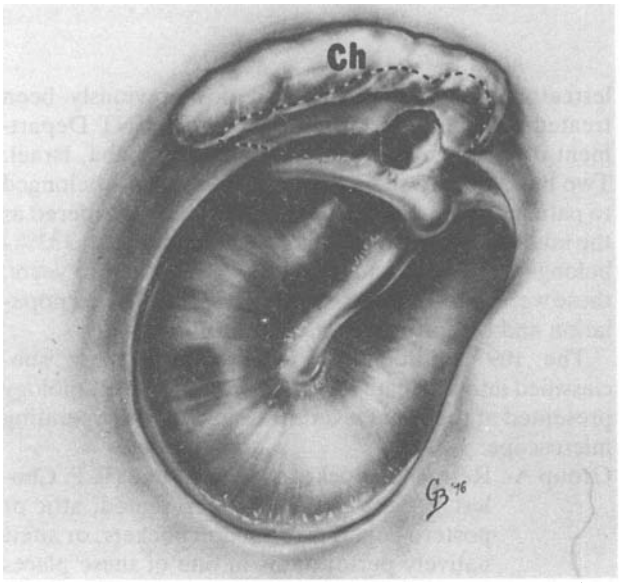


FIG. 2:
Retraction pocket in Shrapnell's membrane leading towards an attic cholesteatoma.

the distribution of cholesteatoma in the middle ear cleft. (2) The configuration of the cholesteatoma. (3) The degree of mastoid pneumatization. (4) The aggressiveness of the cholesteatoma; this was correlated with (a) the amount of ossicular destruction, (b) facial paralysis, (c) semicircular canal fistulas (d) other pre-operative complications e.g. acute mastoiditis.

The distribution, extension, configuration and aggressiveness (ossicular chain destruction etc . . .) of the cholesteatomas were obtained from the operative reports. The degree of mastoid pneumatization was obtained for each patient using the Schuller lateral projection X-rays of the mastoids. Measurements of the pneumatization were undertaken with the aid of computerized planimetry as described by us elsewhere (Sade and Schatz, 1988).

Results

A. *Types of cholesteatomas*

Group A comprised 63 ears (or 57.8%); of these 21 (33.3%) ears presented with an attic retraction pocket cholesteatoma and the remainder (42 ears; 66.6%) a postero-superior retraction pocket cholesteatoma.

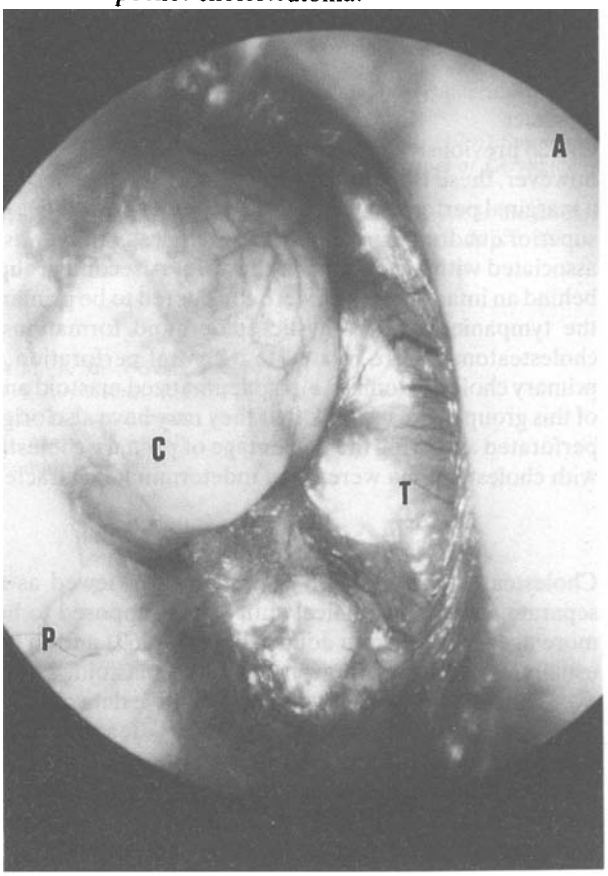


FIG. 3:
Large epidermoid (C) behind an intact drum of a 2-year-old child who was followed-up for recurrent secretory otitis media. At operation, the cystic cholesteatoma was found in the postero-superior quadrant. External ear anterior wall—A, posterior wall—P, tympanic membrane—T.

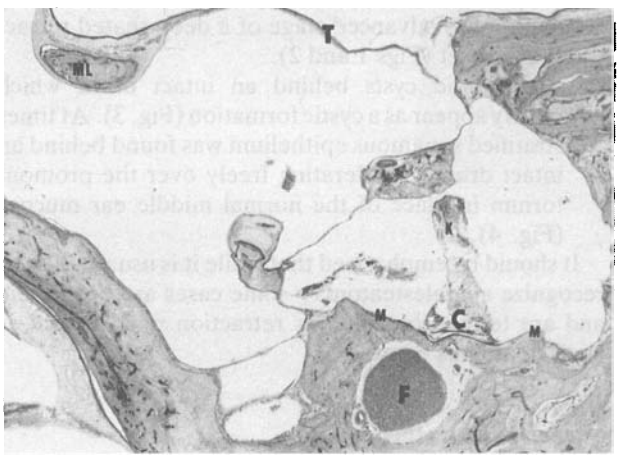


FIG. 4:
A primary cholesteatoma (C) growing from the medial wall of the middle ear. M—mucosa. M.L.—malleus handle. T—tympanic memberane. F—facial nerve.

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Group B comprised 31 ears (28.4%).
Group C comprised eight ears (7.3%).
Group D presented seven ears which were of an undetermined type.

B. *Age and sex*

The mean age for each group at the time of subjective appearance of the first symptoms was about the same in all four groups i.e. six years. The average age at arrival for the first surgical procedure was, Group A 7.4 years, Group B 8.4 years, Group C 8.6 years and Group D 7.7 years. The sex distribution was almost the same for all groups yet within each group males predominated comprising about 75% of all the children.

C. *Configuration of the cholesteatoma*

Group A showed either an infected retraction pocket or what seemed to be a penetrating and infiltrating cholesteatoma communicating with a marginal poster-superior or Shrapnell's membrane perforation which had the basic configuration of a deep retraction pocket. Group B showed a closed epidermoid cyst in 27 cases (87%), and in four cases (13%) a small diffuse tuft of squamous epithelium springing from the posterior superior medial wall of the tympanic cavity. Group C appeared in five as a cystic formation and spread out in three. Group D was impossible to characterize.

D. *Mastoid pneumatization*

Pneumatization was more developed in Groups B and C than in Group A. The mean area of the mastoid air cell system in Group A was 0.88 cm^2 (range 0–2.8cm^2); 26.4% of the mastoids in this group were sclerotic (Fig. 5), 41% diploic and 32% showed some pneumatization with a mean pneumatized area of 2.3 cm^2. The mean area of the mastoid air cell system for Group B was found to be 5.9cm^2 (range 0.1—18.6 cm^2), for Group C 4.4 cm^2 (range 0.6–6.9 cm^2)—no sclerotic or diploic mastoids were found in groups B and C where all ears showed pneumatized mastoids (Fig. 6).

E. *Location of the cholesteatoma in the middle ear cleft*

Atticoantral cholesteatoma distribution was frequent in Group A where it was present in 70% of the ears, and less common in Groups B and C (29% and 12% respectively). Seventy per cent of Group B and 87% of Group C presented with their cholesteatomas in the tympanic cavity compared with 28% in Group A.

F. *Ossicular destruction*

No significant difference in the ossicular lesions was seen between Groups A. B, C and D. These findings are not significantly different from those found in adult cholesteatoma (Sade *et al.*, 1981a) (Table I).

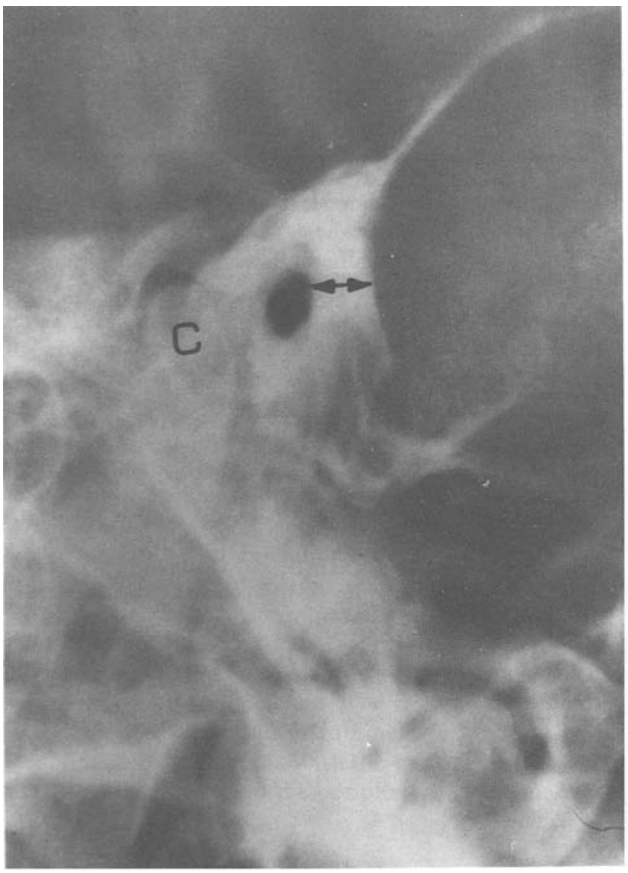


FIG. 5:
Sclerotic mastoid of a child with a retraction pocket cholesteatoma.

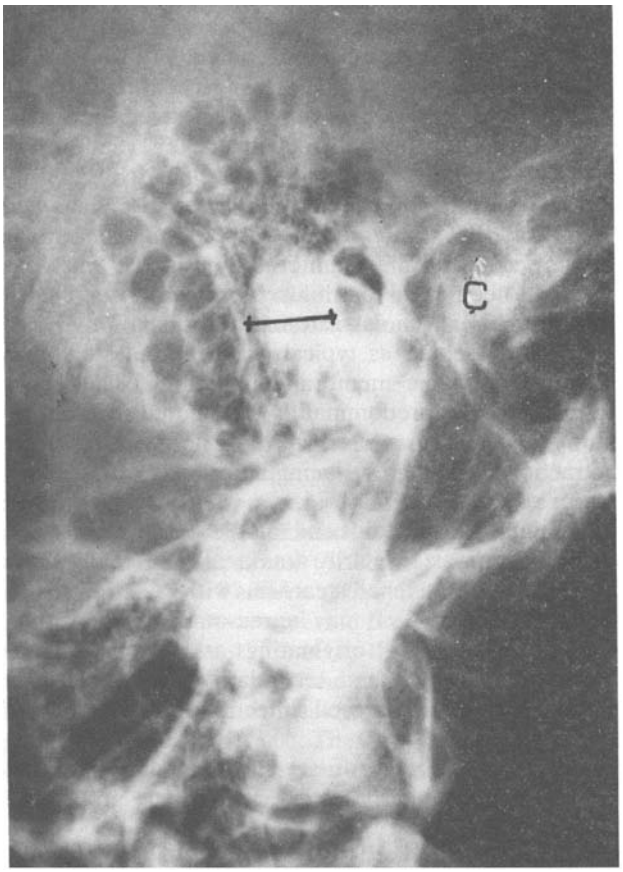


FIG. 6:
Pneumatized mastoid of a child with a primary cholesteatoma.

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TABLE I
OSSICULAR DESTRUCTION IN CHOLESTEATOMA OF CHILDREN AND ADULTS

	Retraction Pocket Cholesteatoma in Children	Primary Cholesteatoma in Children	Adult Cholesteatoma
Incus	56%	67%	78%
Stapes	44%	53%	39%
Malleus	26%	34%	32%

G. *Complications*

None of the 109 cholesteatomas in our children presented with a facial paralysis, one case in Group A showed a fistula in the semicircular canal and three cases in Group A presented as acute mastoiditis.

Discussion

From our material we learn that one-third of our cholesteatoma population appeared in children—the percentage of children with cholesteatomas varies between other reports: Palva *et al* (1977) and Cody Taylor (1977) mention 10% and 22% respectively as occurring under the age of 16, Sheehy *et al* (1977) reported 18% below the age of fifteen.

The groups of children which came under our care did not comprise a homogenous clinical entity. Two-thirds presented with cholesteatoma which appeared as retraction pocket cholesteatomas which had a character similar to those which was found in a population dominated by adult cholesteatomas Sade *et al*. (1981b). Most of the adult cholesteatomas are secondary to deficient middle ear aeration as seen by their presenting as retraction of the tympanic membrane and as has been shown by following-up these retraction pockets (Sade *et al*, 1981c and 1986). Adult cholesteatomas are typically associated with non-pneumatised mastoids (Gans and Wlodyka, 1966; Sade *et al*., 1981c)

The second form encountered by us is the type considered to be typical for children and comprized 28.4% of our childrens' cholesteatomatous population. These children presented as typical primary cholesteatomas with no tympanic membrane pathology. Their cholesteatoma was predominantly distributed in the tympanic cavity. These cases were also distinct from the retraction pocket group being associated with a much larger pneumatized mastoid.

Pneumatized mastoids and middle ear cholesteatoma distribution was similarily found also in those cases which presented as cholesteatomas with a central perforation, i.e. Group C. It may be reasonable to look upon this group also as originating as "primary" cholesteatomas—which had secondarily perforated their drum and which showed infection at the time of presentation.

If we combine the primary cholesteatomas with the central perforation cholesteatomas as one group, the primary cholesteatoma group increases from 28.4% to 37.3%.

However, the majority of cholesteatomas which appear in children originate in retraction pockets and are associated with a negative middle ear pressure. The treatment of choice is probably similar to adult cases whose mastoids are usually non-pneumatized, namely a small open cavity leaving no recesses in which a recurrence can occur. (Sade, 1982).

Primary cholesteatomas are usually considered to originate from congential rests—yet such sites have not so far been demonstrated though their presence was taken for granted. It is our contention that present evidence tends to favour their origin as being metaplastic in origin (Sade *et al* 1982). Their treatment in view of their extensive pneumatization may pose special problems. Some form of intact canal wall technique may be considered, especially if the cholesteatoma is detected in its initial stages. Under certain conditions this technique is much more appealing than the long-term problems of a large open cavity which may be present in a pneumatized mastoid.

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