Unexpected Ear Disease in Infants with Cleft Palate

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The high incidence of middle ear disease in cleft palate children has been well documented; however, neither the age of onset nor the pathogenesis has been established. Accordingly, for the past two years the otolaryngology and plastic surgery services at Children's Hospital of Philadelphia have cooperated in a study which allowed systematic examination of tympanic membranes and middle ears of infants prior to reconstructive surgery for cleft lip and cleft palate. Shortly after this program began, we were impressed with the high incidence of pathological conditions found in these infant ears. The purpose of this report is to present these unexpected findings with the hope that others will be stimulated to examine their patients in a similar manner.

Method

The tympanic membranes and middle ears were examined bilaterally in 25 infants ranging in age from nine days to 12 months. The entire procedure was carried out with the aid of a Zeiss binocular operating microscope at magnifications of 6x, 10x, and 16x. The usual approach to the middle ear was employed. The external ear canal was cleaned with a curette, but no attempt was made to sterilize the canal because this is time-consuming and we did not wish to add to the duration of the procedure. Cultures were not taken routinely. As shown in Figure 1, the tympanic membrane of the infant is almost horizontal and normally has a dull appearance with prominent vasculature. In contrast to the normal infant, the tympanic membrane in the cleft palate infant typically bulges slightly at its inferior portion. An incision was made through the ear drum to allow small suction tips to be introduced for aspiration of the middle ear. When technically feasible, a piece of the middle ear mucosa was removed with suction or a small cup forcep. All patients

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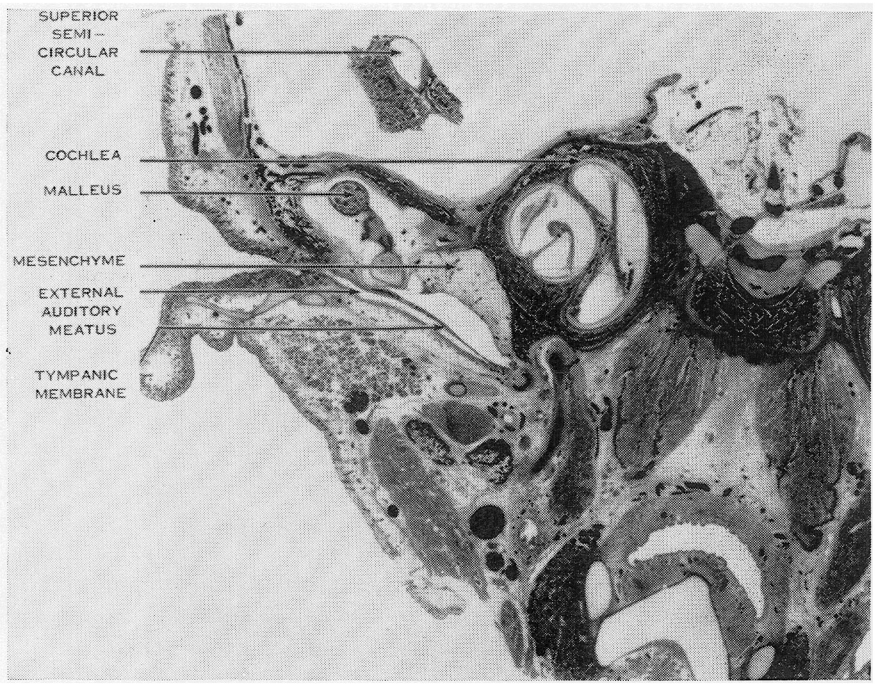

FIGURE 1. Coronal section of 19-week human fetal temporal bone. The tympanic membrane, middle ear, and inner ear have achieved near adult size. Because the external canal is extremely small and the tympanic membrane is oriented tangentially to the external auditory canal, examination of these structures in the infant is difficult without the benefit of anesthesia and the operating microscope. This specimen provided through the courtesy of Dr. Bertram Kraus, Cleft Palate Research Center, University of Pittsburgh, Pittsburgh, Pennsylvania.

received tetracycline for at least five days following the procedure. All the incisions healed without perforation or scarring.

Results

The normal middle ear is an air-filled cavity without fluid. But, in the present series of 25 infants, appreciable amounts of mucoid material were extracted from 47 ears. The consistency of this material ranged from very thin to highly viscid. Ten biopsy specimens were obtained and each revealed the presence of abnormal tissue within the middle ear. The most prevalent finding was that of poorly-organized granulation tissue similar to that shown in the photomicrograph in Figure 2. The specimen shown in Figure 2 was removed from a four-month-old patient. Ten infants without clefts of the palate but with clefts of the pre-palate were similarly examined as 'controls'. Three ears in this group showed similar findings.

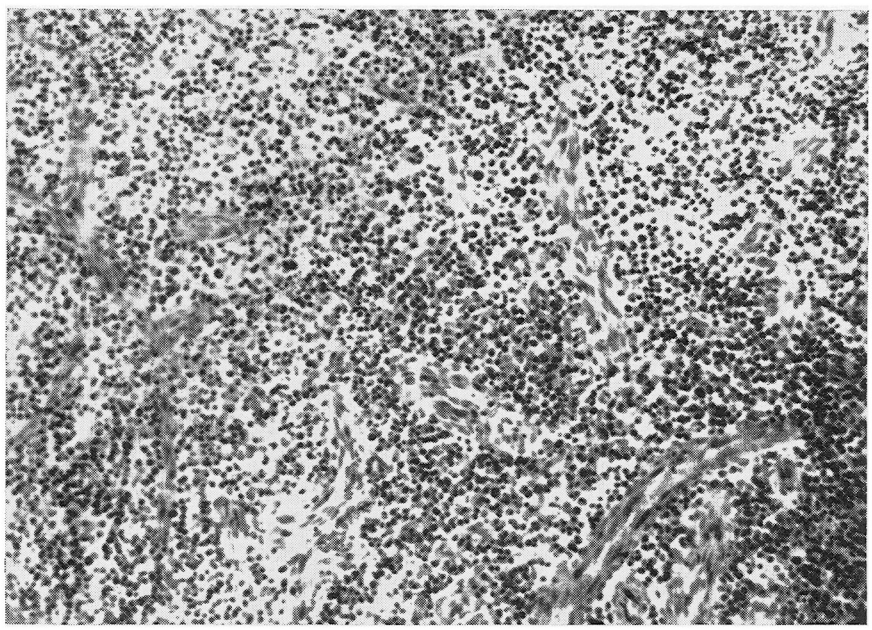

FIGURE 2. Granulation tissue from the middle ear of a four-month-old cleft palate patient. This patient had no symptoms referable to this pathogenic process.

Comment

In order to understand the significance of these findings, it is necessary to appreciate the normal development of the ear. Figure 1 shows a section of the normal ear of a 20-week fetus. The nerve and auditory ossicles are almost adult size. The middle ear space, however, is filled with an embryonic connective tissue. This tissue regresses after birth and the middle ear space enlarges. Beginning at the middle ear space, pneumatization of the temporal bone proceeds in an orderly manner. The eustachian tube connects the middle ear with the nasopharynx. This structure changes during life from a short wide tube to a longer, narrower, tortuous structure. Anything that interferes with this normal progression of events may result in disease and loss of function.

The infant ear is vulnerable to insult. The embryonic connective tissue provides an excellent media for bacterial growth and its presence makes drainage difficult. The tympanic membrane is thicker than that of the adult and will not perforate as easily to provide drainage. Also, the infant does not have a well-developed immunologic mechanism to resist infection. Considering these multiple factors, one can appreciate that an infant with a cleft who has an eustachian tube orifice exposed in the oropharynx may develop neo-natal otitis. This otitis may influence the subsequent pneumatization of the middle ear and temporal bone and

may produce changes in the tympanic membrane. These changes may result in fibrosis, fixation of the ossicular chain, and atrophy of the tympanic membrane. The detailed physical and functional findings of these changes are not appropriate to this paper but will be discussed in a subsequent report.

There exists within the literature little mention of ear disease in cleft palate infants. Skolnick (*5*), in 1958, stated that the incidence of pathology was 6% in children with clefts younger than one year and increased to 60% in preschool children. It should be noted, however, that these ears were examined without the aid of a microscope and myringotomies were not performed. The infant tympanic membrane is difficult to visualize and, unless magnification is used, subtle signs of pathology are easily missed. Indeed we have seen many children who, at the time of admission to the hospital, were diagnosed by competent physicians as having normal ears. Yet pathological conditions were found when the anesthetized patient was examined with magnification.

House (*4*) states that when eustachian tube function is impaired by scar tissue resulting from cleft palate repair, there is a high incidence of middle ear disease. Many otologists feel that fracture of the hammulus is responsible for ear pathology in these patients. In our group of patients, we have demonstrated that there is a high incidence of pathology long before repair of the palate is begun (since these children were examined before surgery). Homes and Reed (*3*) reviewed the findings in 26 postoperative palate repair patients and found a high incidence of hearing loss. They could not establish a correlation with factors such as age at palate closure, nasopharyngeal occlusion, removal of tonsils and adenoids, or type of cleft. In their comments (which seem quite appropriate), they conclude that 'the only practical manner of controlling and preventing hearing loss is that of prompt recognition and early treatment of all ear infections in this group of patients.' On the basis of a longitudinal study conducted in Iowa, Graham (*2*) concluded that careful otologic evaluation and therapy decreased significantly the chronic ear infections in cleft palate children. That method of therapy did not include the use of ventilation through the tympanic membrane, although Donaldson (*1*) recently reported good results with such a procedure.

The findings of the present study suggest that a reasonable approach to the problem of otitis media in the cleft palate infant is to examine these patients with magnification whenever they are having reconstructive surgery. The removal of abnormal material from the middle ear and the ventilation of this cavity should allow pneumatization of the ear to proceed in a more normal manner and should aid significantly in preventing subsequent hearing loss. The procedure is safe, requires only a few minutes, and does not add to the length of hospitalization.

Summary

Under general anesthesia, the middle ears of 25 cleft palate infants were examined bilaterally through a binocular operating microscope after myringotomy. Ninety-four per cent of these ears contained mucoid material and each of 10 biopsy specimens revealed the presence of granulation tissue. This incidence of pathological findings suggests that a more aggressive method of therapy should be developed to prevent the sequelae of infantile otitis.

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