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Congenital Cholesteatoma*

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IN almost 40 years of otology, I have frequently operated on cases of cholesteatoma with a small attic perforation and have thought that the perforation was produced by the cholesteatoma growing from within out, and not from without in. Now I have been able to obtain a small cholesteatoma tumour complete and have been able to have it sectioned serially. Many years ago Dr. Dan McKenzie expressed the view that a third of all cases of cholesteatoma were of congenital origin but this suggestion received little support, but I think that perhaps a true congenital origin of the disease is much more common than many of us are aware.

A small healthy little boy, aged 5½ on first going to school was examined by the School Medical Officer, who found what he thought was a small pearl bead in the left ear. He referred him to hospital and at first it was thought that he had a pearl bead in the external meatus but Sister could not remove the bead by syringing. When the boy was examined under general anaesthesia and with the operating microscope, this small bead was found to be deep to the drum in the anterior part of the middle-ear cavity. It appeared to be in contact with the inner surface of the drum over a small area (Fig. 1). A small incision was made over the bead, but on touching it, it appeared to fall into the lower part of the tympanum and was then removed through another incision by means of the suction tube. It was a small tumour. The tumour was a little too large to enter the suction tube and we were able to recover it. It was only 2 mm. in diameter

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FIG. 1.

and must have been attached by a small stalk in the region of the anterior ligament. The boy had had no previous history of any trouble with his nose or throat or ears. X-rays of the sinuses were normal and X-rays of his mastoid processes showed normal pneumatization. The perforation in the drum healed quickly and there is only very slight residual loss of hearing.

Sections of this tumour consisted almost entirely of laminated keratin. Dr. Angus Stewart of the University Pathological Department had serial sections made and only at one part could be found the lining membrane; a single layer of cuboidal cells. I had expected to see a nice layer of squamous epithelium. How has this keratin been produced? As far as I am aware no one has suggested that cuboidal cells produce keratin and yet these are the cells which are lining the tumour. At one part can be seen a foreign body giant cell. I think it possible that the whole of the tumour was lined by this single layer of cuboidal cells but that most of the lining layer was removed by the sucker. There is no doubt that if this tumour had been allowed to grow it would eventually have given rise to a perforation in the anterior part of the drum, and that perhaps some of our colleagues would have suggested that the cholesteatoma had been produced by squamous epithelium growing inwards from the meatus. Yet this tumour does not appear to be lined by squamous epithelium.

This was then a small cholesteatoma occurring in a young boy in the anterior part of the tympanic cavity. I can see no explanation other than that it has been due to epithelial cells which have been displaced during

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development and that it is of congenital origin. Once perforation has occurred in cholesteatoma, it is very difficult to prove that the condition has not been due to meatal epithelium growing inwards. It is probable that the congenital origin of cholesteatoma is more common than many of us think. Sir Terence Cawthorne has described a number of these cases and I feel sure that many of you may have similar cases. Perhaps it may yet be shown that Dan McKenzie's view that a third of all cholesteatomas are of congenital origin is more accurate than we have thought.

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