Pathology of the Ear

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BLACKWELL

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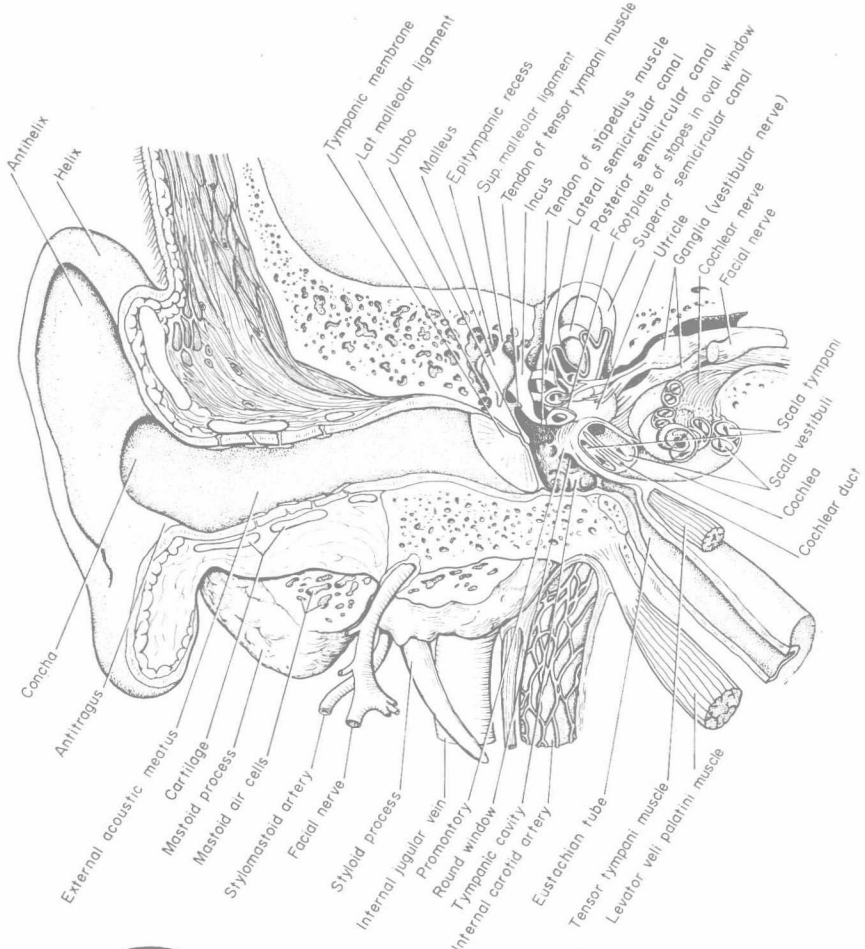

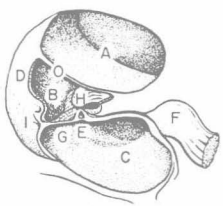

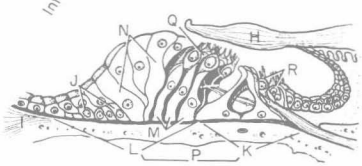

SECTION THROUGH SPIRAL ORGAN OF CORTI

CROSS SECTION OF THE COCHLEA

A Scala vestibuli
B Cochlear duct
C Scala tympani
D Stria vascularis
E Basilar membrane
F Cochlear nerve
G Organ of Corti
H Tectorial membrane
I Spiral ligament
J Cells of Claudius
K Arcuate zone
L Pectinate zone
M Cells of Deiters
N Cells of Hensen
O Reissner's membrane
P Basilar membrane
Q Outer hair cells
R Inner hair cells

Preface

In writing this book, I had, in common with other authors, two objectives constantly before me; to provide for the advanced student, and in particular the postgraduate who is training in otorhinolaryngology or pathology, a textbook outlining the pathological basis of diseases of the ear. I hope that the book will also be of use to teachers of pathology and otorhinolaryngology.

Since it is primarily concerned with human disease, no more than an outline of recent experimental studies can be included in it, fascinating and significant though they may be. It is based on a chapter forming part of the textbook *Systemic Pathology*, edited by the late Professor Payling-Wright and Professor W. St. C. Symmers and published by Messrs. Longmans Green & Co., London.

I am indebted to Professor Symmers and to his publishers for agreeing to the use of the chapter I wrote.

The pathologist called on to advise on the diseases of the ear – or any aspect of otorhinolaryngology – has a difficult path to tread. He can provide some additional information which, if appreciated, may assist the surgeon during an operation when he meets something abnormal. It is our duty to question dogma: but the pathologist has also to recognize the limitations of his own expertise. At the same time, the specialist pathologist has a duty 'to bring to the notice of his surgical colleagues any advances in pathology, and to draw the attention of his colleagues in pathology to the particular problems of the ear' (Milligan and Wingrave, 1923).

Biology and pathology have reached an interesting phase in their development, marked by the blending of various disciplines of research in the study of fundamental processes and in the re-investigation of vast morphological fields. Our field lies in otorhinolaryngology, considered by many, both inside and outside the specialty, rather a limited one.

It has not been fully appreciated that there exists a great variety of diseases which may affect, or are influenced by, the ear, nose and throat. Furthermore, there are the senses of smell, taste, hearing and equilibrium located here, which present challenging problems both in health and disease.

As a pathologist, for many years closely associated with otorhinolaryngology, I am well aware of the pioneering work of otorhinolaryngologists in the study of the pathology of the ear, nose and throat. Otologists have pioneered also the electron microscopic study of the inner ear. Moreover, microscopy and otorhinolaryngology have, in my view, certain interests in common, such as the study of small areas of diseased tissue and the application of more or less complicated optical instruments in which otorhinolaryngologists have always excelled.

The introduction of electron microscopy into otological research has been rewarded by a good deal of success in spite of the fact that there has been little or no opportunity to examine so-called control material from the human ear. Most of the investigators in this field have based their studies on tissues obtained from animals. Scandinavian, American, Swiss and Italian workers have shown the way to follow.

With the introduction of electron microscopy and the perfection of other methods and techniques, it has often been said, a new golden era of cytology has begun. As in the analogous period of the development of light microscopy, a flood of new discoveries has been reported concerning the fine structure of the protoplasm, called the physical basis of life by the great Czech anatomist Jan Evangelista Purkině. The long-drawn-out controversy over the very existence of the cytoplasmic organelles such as the mitochondria, the Golgi apparatus and the endoplasmic reticulum has been settled and the existence and structure of these essential organelles is no longer in doubt.

There is, however, the danger that once again we may be misled in matters of interpretation, because of the fixation artifact now being observed at a hundred times the magnification available to the microscopist of the last century.

Despite the difficulties in obtaining and preparing adequate material, an extensive literature exists on the pathology of deafness

present from birth or originating in early childhood, which is dealt with in the second part of this volume. One of the hindrances upon investigation of the pathology of deafness is that is is not a fatal disease, and deaf persons, at the time of death, are seldom under the care of physicians interested in temporal bone pathology. Furthermore, the ear has not attracted the attention of general pathologists, who appear to have fought shy of the apparently barren regions of the temporal bone.

The practical importance of regular pathological examination of all specimens from operations on the ear has not been appreciated. The so-called aural polyp or aural 'granulations' have been among the commonest specimens sent for histological examination; and even those commonly continue to be dismissed as of little account: for it is not always realized how often they may hide more than they reveal. Only when aural specimens are sectioned as a routine is it possible to detect an early stage of carcinoma, or glomus jugulare tumour, or of tuberculosis of the middle ear, which otherwise are liable to continue their course, unrecognized and untreated.

There remains much to be done in elucidating the problems met within this field, some of which are, in fact, peculiar to it. From time to time, otologists have raised certain questions as a result of clinical observation, e.g. has the pathology of otitis media changed since the advent of antibiotics? What is the origin of squamous epithelium occurring in the middle ear cleft and producing the so-called cholesteatoma of the ear? In relevant chapters of this book I have attempted to answer some of these questions and others like them.

This volume is divided into two main parts:

1. The general pathology of the ear, containing the chapters on inflammatory, neoplastic and bone diseases.
2. The pathology of the inner ear – or deafness. There is an appendix of methods.

Some embryological and anatomical aspects are included, but the reader is advised to consult the textbooks mentioned in the list of references. Normal light and electron microscopic cytology of the inner ear is described in greater detail. It was necessary to reduce the number of illustrations of anatomical and embryological relevance, which may be found in the excellent textbooks referred to.

It is difficult to be all-inclusive in a book, which, it is hoped, will appeal to a wide variety of readers and students. Furthermore, it is not an easy task for a single author to cover every detail of his subject, even in an apparently limited field.

The author is, therefore, grateful to all those authors whose work, listed in the references, has greatly contributed to his knowledge and has stimulated his interest.

It is with a deep sense of gratitude that I thank my technical staff, and in particular Mr. E. S. Bird, F.I.M.L.T., Chief Technician, and the late Miss V. Shepherd, Chief Technician, for their invaluable assistance and work with both light microscopic and electron microscopic preparations. Their excellent sections of biopsy specimens and temporal bones (experimental and human) form the subject of the illustrations which are the splendid work of Mr. D. Connoly, Clinical Photographer, all on the staff of the Institute of Laryngology and Otology, and my special thanks are due to them all.

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The Publishers have been extremely patient and helpful and I am grateful to Mr Per Saugman, Managing Director, in particular and Mr J. L. Robson, Production Manager, for their understanding interest.

Contents

PART ONE

Pathology of the External Ear The Middle Ear Cleft

CHAPTER ONE

Gross anatomy and development of the external ear, middle ear and the ossicles

Anatomically, the ear is usually divided into:

The external ear consisting of the auricle or pinna and the external auditory meatus (Figs. 1.1. and 1.2).
The middle ear comprising the tympanic cavity, which communicates with the mastoid antrum and through it with the intricate and important air-cell system of the mastoid process (Fig. 1.2).
The inner ear which harbours the sensory apparatus of hearing and equilibrium (Fig. 1.3).

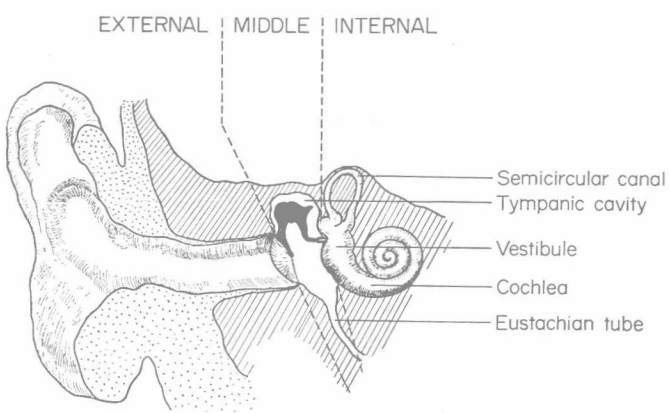

Fig. 1.1 Schematic diagram of the external, middle and internal ear.

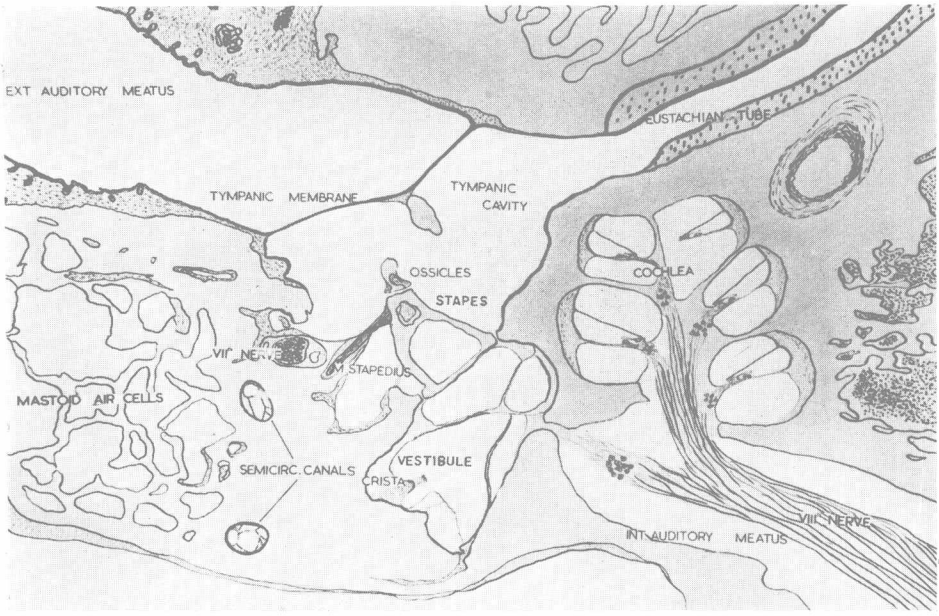

Fig. 1.2 Diagram of section of normal temporal bone (human). Note tympanic membrane, stapes and labyrinth. ($\times 5{\cdot}2$)

Certain aspects of the microscopical anatomy of the external and middle ear will be mentioned in conjunction with the pathology of the relevant parts.

The inner ear is located in the petrous portion of the pyramidal bone and is protected by the toughest part of the skull, the otic capsule, which encloses the cochlea, the vestibule and the three osseous semicircular canals (Fig. 1.4). The inner ear is described in detail in Part 2.

The development of the external ear and of the middle ear

The inner ear of the lower vertebrates and some other species is an organ concerned with equilibrium, but in higher vertebrates the latter function has led to the establishment of a special transmitting apparatus to convey the air vibrations to the internal ear, and this apparatus is situated in a diverticulum of the pharynx – the middle ear or tympanum.

The external ear

The external ear is derived from the first and second branchial arches. During the fourth week three tubercles appear on the

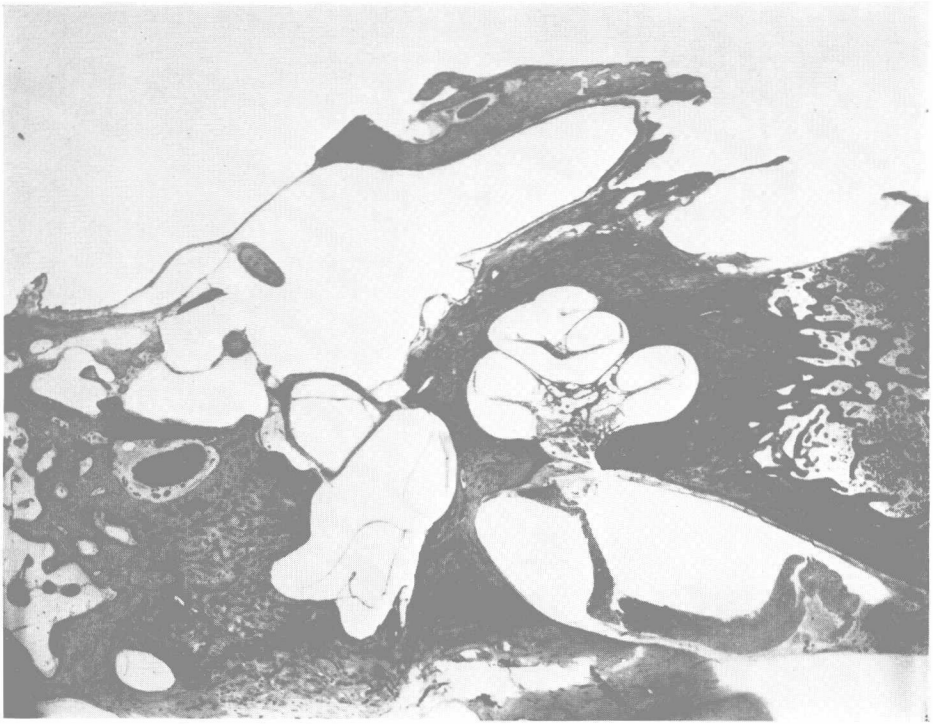

Fig. 1.3 Midmodiolar section of human temporal bone. Note tympanic membrane, ossicles (stapes!), facial nerve, cochlea. (× 3·9)

mandibular arch and another three and a ridge on the hyoid arch from which the auricle is formed.

A pitting of the ectoderm denotes the site of the external meatus which deepens by active proliferation of its ectoderm, thus forming a temporary epithelial plug, later breaking down or canalizing in its central portion (Figs. 1.5 and 1.6). A failure of the ectodermal core to canalize results in abnormal bone formation and obliteration of the external auditory meatus.

The attic links up with the external auditory meatus, without the interposition of a mesenchymatous layer, to form the pars flaccida (Shrapnell's membrane) which, under the light microscope, appears to consist of only two epithelial layers. Recent electron microscope investigations by Lim (1968) have revealed a thin connective tissue layer in the pars flaccida separating the epithelial layers. The pars tensa of the tympanic membrane is partly ectodermal in origin; an ectodermal layer joins the entodermal mucosa of the tubotympanic recess, from which it is separated by a thin mesenchymatous layer.

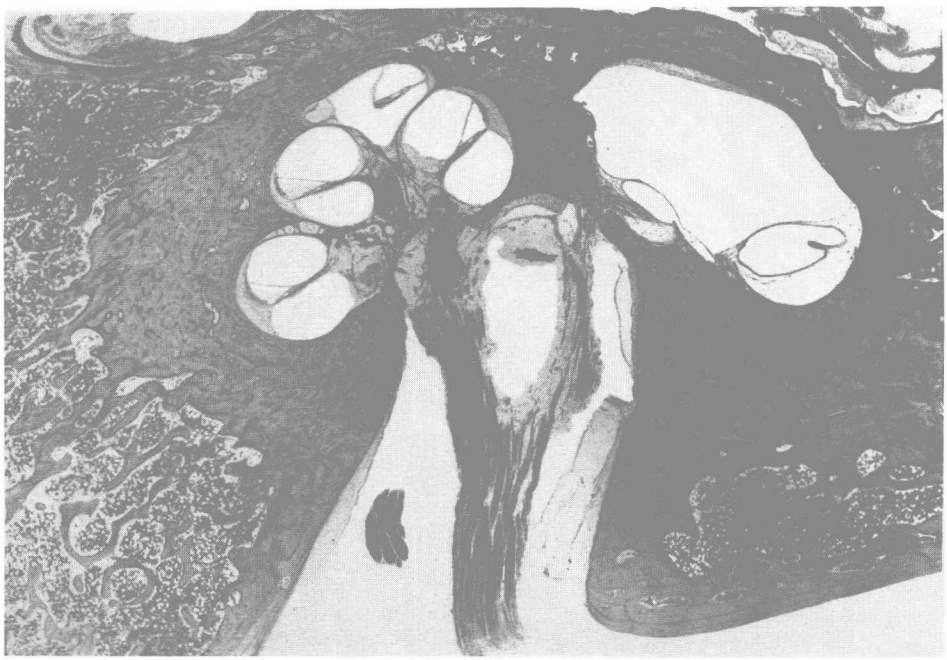

Fig. 1.4 Human labyrinth. Note cochlea, vestibule, auditory nerve, cochlear and vestibular branches, Scarpa's ganglion. (× 5·7)

Middle ear

The tympanic cavity and the Eustachian tube are derived from the tubotympanic recess formed from the first entodermal pharyngeal pouch. The expansion of the entodermal mucosa of the tubotympanic recess results in the formation of the tympanic cavity at about the fifth week. The epithelium gradually envelops the ossicles, their tendons and ligaments, as well as the chorda tympani nerve; so that all these structures receive a complete epithelial investment.

The medial part of the tubotympanic recess becomes elongated to form the pharyngo-tympanic tube (Eustachian tube).

The posterior expansion of the tympanic cavity gives rise to the tympanic antrum and mastoid cells. The cavity of the antrum and attic begin to develop separately towards the tenth week. Their pneumatization and that of the mastoid process results from the resorption of the mesenchymatous tissue and the lining of the so-formed cavities by the tubotympanic epithelium

The development of the ossicles

The auditory ossicles in man are of mesodermal origin and are