DISEASES OF THE SKIN

BY

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PREFACE

I HAVE tried in this book to provide a text that undergraduates and general practitioners may use for reference and that will serve as introductory reading for postgraduate students.

The basic scope of dermatological practice is much the same all over the world, and if I stress some conditions commoner in Southern Africa than elsewhere I do not think the balance has been affected thereby.

The art of recognition of the lesions and patterns of evolution in skin diseases can be learnt only by the observation of many cases, and I have not attempted to reduce it to writing.

The importance of histopathological examination in the investigation of skin disorders cannot be overestimated, and I have given detail enough to indicate the nature of the processes involved. Postgraduate students will need to supplement their studies in this, as in other subdivisions of the subject, in more specialized works. A bibliography of main references for further comparative and specialized reading is appended.

To avoid repetition, a list of prescriptions for topical applications and general instructions for the use of such remedies as corticosteroid hormones, antihistaminics, antimalarials and antibiotics are included in Chapter IV; and instructions for the use of the antifungous antibiotics will be found in the introduction to Chapter XVI.

The system of classification is largely orthodox, but I have occasionally considered some conditions out of context, e.g. epidermolysis bullosa next to porphyria, in order to stress a relationship. The nomenclature adopted is usually that which is at present most commonly used in the literature, but when common usage infringes on priority, as in some of the syndromes, the fact is stated.

J. M.

Cape Town, 1960.

ACKNOWLEDGEMENTS

A NUMBER of colleagues and institutions in Africa, Europe and the Americas have generously contributed photographs to fill the gaps in my collection. When an illustration has been borrowed its provenance is noted under the title, but I must also thank certain people who are not individually cited in this way.

Among these are Professor A. J. Brink, Dr. R. L. M. Kotzé and my colleagues at Karl Bremer Hospital and the University of Stellenbosch; Professor H. W. Synman, Dr. G. H. Findlay and Dr. I. K. Venter at the University of Pretoria; Dr. R. Lang and his colleagues in the Department of Dermatology at the University of Cape Town; Mrs M. E. Pfeiffer who lent the photographs from the collection of her late husband, Dr. D. H. Pfeiffer, who was Medical Officer of Health in Bloemfontein; Dr. Hugh Wallace of St. Thomas's Hospital, London; Dr. A. Touraine, Editor of the *Annales de Dermatologie et de Syphiligraphie*; Dr. F. W. F. Purcell who lent from his own collection and from that of the late Dr. D. C. McArthur; and Dr. A. P. Blignault, Editor of the South African Medical Journal, in which some of the illustrations have already appeared. The photographs lent by Drs. A. G. Shaper and P. W. Hutton come from the collection of the Makerere College Medical School, Kampala, Uganda. Drs. J. M. Martin, W. J. Pepler and H. I. Lurie collaborated with me in the investigation of some of the cases illustrated.

Some of the illustrations have already appeared in my earlier books, and I would like again to thank the lenders who were the following: The Curator of the Museum of the London Hospital Medical College; the Medical Committee of St. John's Hospital for Diseases of the Skin, London; the Director of the Wellcome Museum of Medical Science, London; and Drs. Sydney Thomson, J. E. Schneider, G. Leslie, S. R. A. Beckett and L. B. Bourne. Mr Norman K. Harrison took the original photographs used in these books.

Many of the photographs from my own collection were taken by Mr Robert Ellis, A.I.B.P., A.C.P.I.P., Department of Clinical Photography at Karl Bremer Hospital, some by his

predecessor, Mr P. de Chavigny, and others by Mr Theo Marais, Department of Clinical Photography, Pretoria General Hospital.

Only main subject references have been included in the bibliography, and I hope that those of my colleagues who recognize that I have quoted from their articles will accept this acknowledgement of my indebtedness.

I must, finally, thank Mrs F. J. F. Kellerman who typed the manuscript, my wife who read the proofs and my publisher, Mr Charles Macmillan of E. & S. Livingstone Ltd., for advice and instruction.

CONTENTS

CHAPTER I

ANATOMY AND PHYSIOLOGY

ANATOMY

THE skin is a dynamic organ composed of ectodermal and mesodermal components. The outer covering of epidermis (epithelium) is of ectodermal origin and lies upon the dermis or corium; beneath the dermis is the hypoderm or subcutaneous tissue. Both dermis and hypoderm arise from mesoderm.

The embryonic epidermis originally consists of a single layer of cells which, during the second intrauterine month, differentiates into two to form an outer *periderm* or *epitrichium* and an inner layer, the *stratum germinativum*; in the third month, a third layer, the *stratum intermedium*, appears between them. At the fourth month, the periderm separates to help in the formation of the *vernix caseosa* and the cells of the stratum intermedium multiply to develop into the *stratum Malpighii*.

From the stratum germinativum develop the basal cells of the adult epidermis, eccrine sweat gland germ cells and primary epithelial germ cells from which arise hairs and sebaceous and apocrine glands.

Melanocytes appear in the epidermis about the third month. The balance of opinion is now in favour of their origin in the neural crest and subsequent migration with the nerves to the epidermis. They are still, however, believed by some authors to be, simply, modified basal cells.

The foetal dermis consists at first of closely packed spindle-shaped mesodermal cells. In the third month, these cells develop into fibroblasts and reticulum fibres and later collagen fibres can be identified. Fat also develops in the third month, but elastin is seen only at the sixth month. Collagen and elastic fibres are probably precipitated out of the intercellular ground substance by some enzyme of mesodermal cells.

In the mesoderm develop the blood vessels and primitive

blood cells or haemocytoblasts, the bones and cartilages. Multipotent mesodermal cells remain in the adult dermis and other organs and may, if a suitable stimulus arises, take up again their primitive functions; this is the basis of the lesions of the diseases of the reticulo-endothelial and haemopoietic systems.

The epidermis

In sections of normal skin it is seen that the dermo-epidermal junction is a wavy line. This is due to the fact that

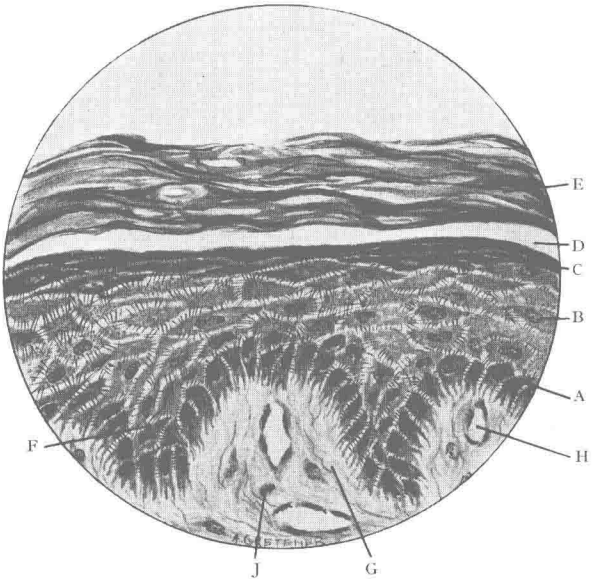

Fig. 1

Normal skin. A: stratum basale; B: stratum Malpighii; C: stratum granulosum; D: stratum lucidum; E: stratum corneum; F: rete peg; G: dermal papilla; H: capillary; J: connective tissue cell.

[*Henry Haber:* The Eczemas]

the under surface of the epidermis is studded with innumerable blunt digitate projections known as rete pegs; the corresponding upward projections of the dermis are known as dermal papillae.

In the adult epidermis four distinct layers of cells can be distinguished except in the palms and soles where a fifth layer is seen. The skin is not an inert structure and the various

epidermal layers represent a process of gradual maturation or evolution of the cells in the deepest layer and their upward growth until they are shed in an invisible desquamation. Mitotic figures are seen in the cells of the stratum basale and stratum Malpighii (Figs. 1 and 2).

The stratum basale, stratum germinativum or basal layer lies against the dermis and contains nucleated cells of two different types, basal cells and melanocytes. Basal cells are columnar or cylindrical and lie with their long axis perpendi-

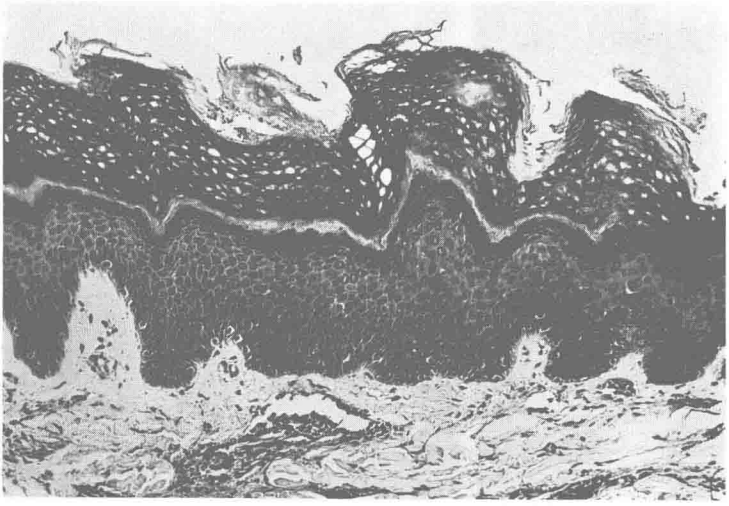

FIG. 2
Normal skin.
[*Henry Haber*]

cular to the dermo-epidermal line of junction. They are united by fine protoplasmic intercellular bridges to each other and to overlying cells. They may appear on routine staining to contain melanin granules, but special methods show that this is not so. Fine cytoplasmic processes from the basal cells interdigitate with reticulum fibres of the dermis, but a basement membrane is interposed.

In sections stained by routine methods some cells with clear cytoplasm and a small dark nucleus are seen between the

basal cells. These are melanocytes (clear or dendritic cells) and stain with Bloch's dopa stain (they can form melanin) and silver stains (they contain melanin). Silver staining shows that they send out long dendritic processes containing melanin granules.

The subepidemal basement membrane. In some routinely-stained skin sections a fine eosinophil membrane may be seen to separate the dermis from the epidermis. It appears from its special staining properties to consist of a mixture of a network of fibres (reticulin or degenerate collagen) and muco-polysaccharides. To its under surface are attached very numerous delicate fibres arising from the dermal elastic plexus;

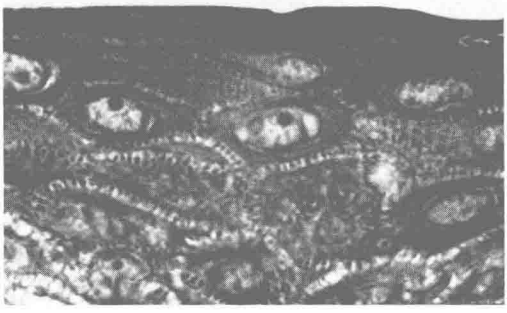

FIG. 3
Intercellular bridges.
[*M. Ruiter*]

its upper surface is invaginated by cytoplasmic digitations from the cells of the stratum basale.

The basement membrane completely divides the epidermis from the dermis while assuring their adherence, and it is lesions of the membrane which permit the formation of subepidermal bullae.

The stratum Malpighii (stratum mucosum, squamous or prickle-cell layer) lies above the stratum basale and consists of a mosaic of nucleated polygonal cells that become flattened in the upper layers. Across the spaces between the cells run intercellular bridges (prickles). Lymphatic fluid can circulate between the cells. Phase contrast and polarization microscopy have shown that tonofibrils (probably consisting of keratin) appear to run uninterruptedly across the cells and through

the intercellular bridges. Each intercellular bridge shows a small nodular thickening, the nodule of Bizzozero (Fig. 3).

The stratum granulosum or granular layer lies over the stratum Malpighii, is two or three cells thick, and consists of flattened nucleated cells whose cytoplasm is filled with coarse granules. The granules consist of *keratohyalin* which is probably identical with desoxyribonucleic acid. This layer and the stratum corneum are usually absent in the buccal mucosa.

The stratum lucidum is seen only in the palms and soles where it lies above the stratum granulosum. Its cells are not nucleated; they are flat and transparent because an oily substance, *eleidin*, is contained within them. Eleidin is believed to result from liquefaction of keratohyalin granules.

The stratum corneum, or horn layer, is the outermost layer of the epidermis and consists of closely-packed, flattened cells full of keratin and containing no nuclei. The thickness of this layer varies enormously in direct proportion to the amount of friction or trauma to which the part of skin is subjected. These cells are detached and shed.

The normal buccal mucous membrane possesses no granular or horny layers. The epidermal cells in their upward movement become vacuolated, then shrink and are shed.

Lymph circulates between the cells of the lower reaches of the epidermis, but there are no lymph or blood vessels; nerve fibres and Merkel's discs (touch receptors) are demonstrable.

The epidermal appendages

The sweat glands (eccrine glands) are present in all parts of the skin, but most abundantly in the palms, soles and axillae. They produce a liquid secretion containing no cellular substance. The sweat gland is a long blind tubule consisting of a spherical, coiled secretory part situated deep in the dermis or at the dermo-hypodermal junction and a duct which corkscrews upwards through the dermis and epidermis to open on the surface. The secretory portion of the gland is composed of an inner layer of cylindrical secretory cells and an outer layer of spindle-shaped myoepithelial cells which are contractile and drive the secretion into the lumen. The duct is composed of two layers of cuboidal cells and appears to maintain its identity even as it passes through the epidermis (Figs. 4 and 5).

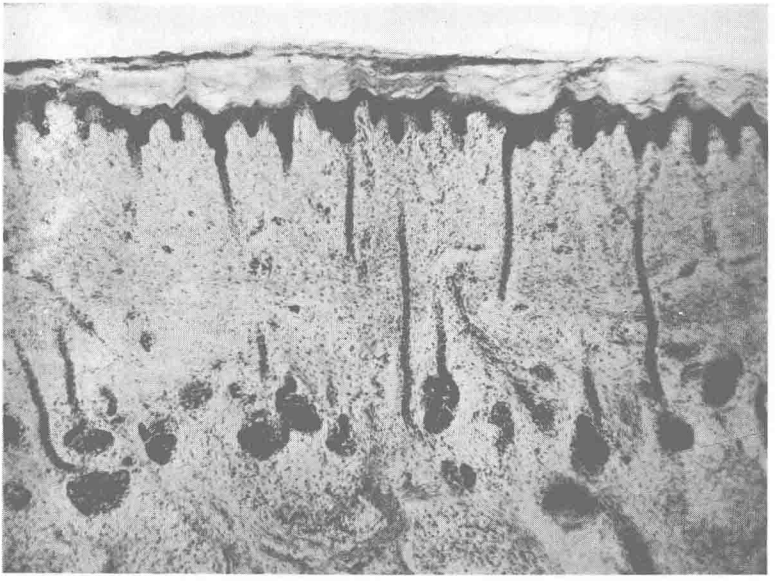

FIG. 4
Eccrine sweat glands and ducts.

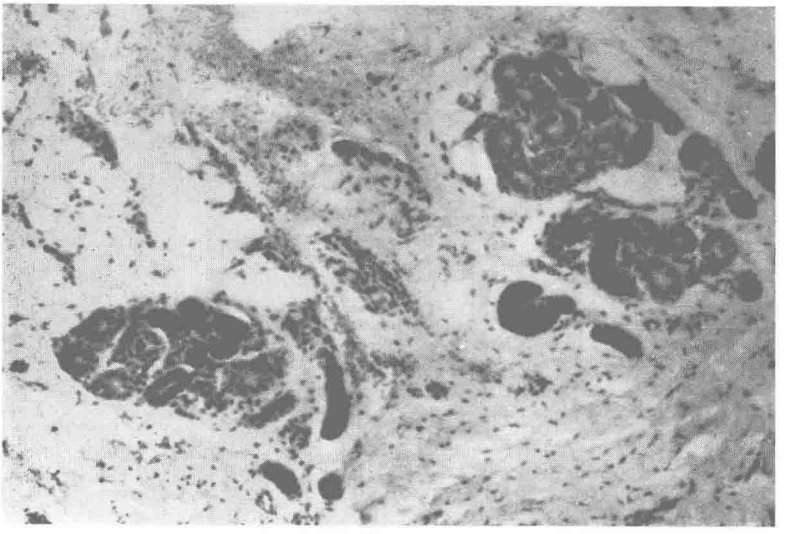

FIG. 5
Eccrine sweat glands.

[H. W. Weber]

The apocrine glands originate from the primary epithelial germ and their ducts lead into pilo-sebaceous follicles and not direct to epidermis like those of sweat glands. They are vestigial scent glands and are found only in the axillae, around the nipples and in the genital region; modified apocrine glands are the ceruminous glands of the ear, Moll's glands of the eyelids and the mammary glands.

Apocrine glands are tubular, but the lumen of the secretory portion is ten times larger than that of a sweat gland. The

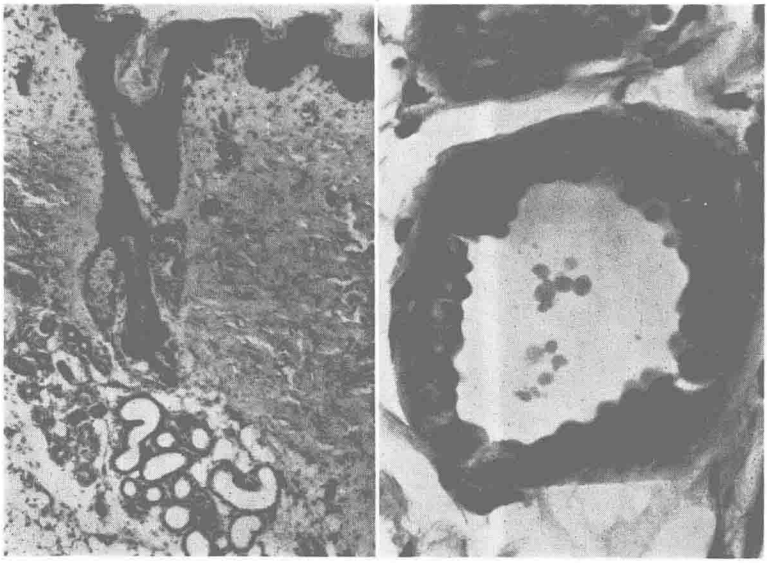

FIG. 6
Apocrine glands.
[*H. W. Weber*]

secretory portion is two layers thick, the outer layer being of myoepithelial cells. The inner layer is of cuboidal cells that protrude into the lumen and release part of their cytoplasm there (Fig. 6).

The sebaceous glands are alveolar, holocrine glands whose secretion, sebum, is formed by cellular decomposition. They are found all over the skin and genital mucosa except on the palms and soles, and modified forms are found in the lips

as Fordyce's glands and in the eyelids as the Meibomian glands. One to six glands are clustered round each pilo-sebaceous follicle into which they discharge. Each gland has several lobules with a peripheral layer of cuboidal generative cells that contain no lipids. The cells within the lobule are arranged in a delicate network and are filled with fat. These are the cells which finally break up into an amorphous fatty mass that diffuses upwards into the follicles and mixes with the eccrine sweat to form a surface film (Fig. 7).

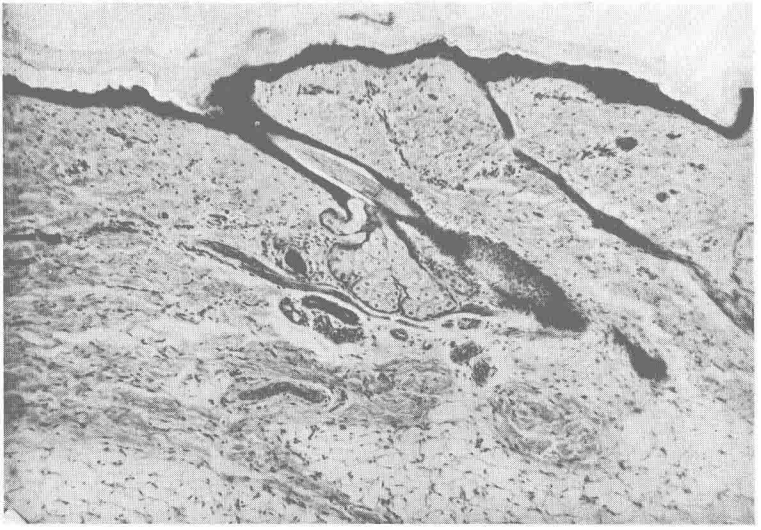

FIG. 7

Hair follicle, sebaceous gland and arrector pili.

The hair consists of a bulb of hair matrix cells lying in the dermis or hypoderm, a root consisting of non-keratinized cells and a projecting shaft of keratinized cells. Into the bulb goes a papilla of connective tissue containing blood vessels and nerves, and melanin in dark-haired people. A hair has two inner sheaths developing from the matrix cells and an outer sheath of downward-growing epidermis. Each hair projects from a follicle which is an invagination of the skin; sebaceous glands discharge into the depths of the open funnel-shaped part of the follicle.

The hair is constantly being renewed; soft lanugo hair and eyelashes have a short life of about five months, but that of scalp hair is three to five years. The old papilla ruptures and a new one forms and the new hair grows alongside the old one in the follicle until it is shed.

The nails are composed of keratin and are formed by the nail matrix which lies under the attached end of the nail and extends forward under the lunule. Neither the nail matrix nor the nail bed beyond it have any stratum granulosum. The growth of a nail from matrix to free end takes about six months.

The dermis

The dermis is a fibro-elastic layer that contains blood and lymph vessels, epidermal appendages, muscle and nerve

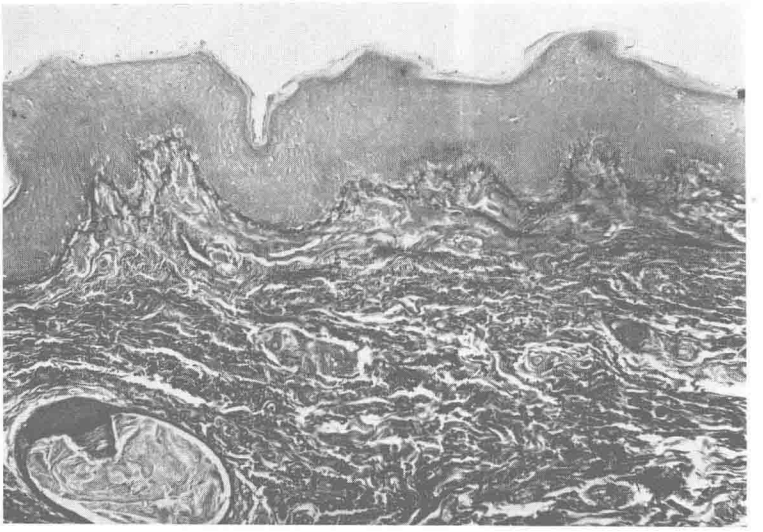

Fig. 8

Collagen and elastic fibres in normal skin. Elastic fibres are black.

[Henry Haber]

elements. Its uppermost part consisting of the papillae that project into the epidermis is known as the papillary layer; under this is the reticular layer. Collagen, elastic and reticulum fibres, enveloped in a ground substance, are the main elements of the dermis (Fig. 8).

Collagen fibres make up 98 per cent of the connective tissue and form bundles bound by ground substance. The bundles are irregularly disposed in the upper dermis, but lie parallel to the surface in the lower reaches. Fibroblasts are sparsely scattered between the collagen bundles.

Elastic fibres run mainly parallel to the surface in wavy lines among the collagen bundles and are most numerous in the lower parts of the dermis. They play no part in the dermo-epidermal attachment. Elastic tissue is ill-named as it is rigid; this rigidity prevents overstretching and helps to restore skin to normal after it has been stretched.

Reticulum fibres are revealed only by silver staining and are not present in large quantity in normal skin. They form a fine network (*gitterfasern*) running from the epidermis (where they interdigitate with cytoplasmic processes of the basal cells) to the glands and blood vessels. Reticulum fibres are seen in large quantities in chronic infective granulomatous diseases such as tuberculosis and in many of the reticulo-endothelioses. They probably arise from precipitation of an extracellular excretion of mesodermal cells such as reticulum cells, histiocytes, lymphocytes and endothelial, muscle and fat cells. Reticulum is a precursor of collagen.

Nerves. Sensory nerves and end-organs of the cerebrospinal system are found in the dermis and nerves of the autonomic system supply the blood vessels, smooth muscles and sweat glands. Sebaceous glands have no nerve supply and are under hormonal control.

Blood vessels. There is a deep plexus of arterioles at the lower border of the dermis and a superficial capillary plexus in the subpapillary region that sends branches up into the papillae.

A special structure concerned with temperature control by short-circuiting an arteriole to a venule, bypassing the capillaries, is known as the glomus. Glomus bodies are seen particularly in the toes, fingertips and nailbeds. An arterial segment, the Sucquet-Hoyer canal, branches off an arteriole and has a narrow lumen and a thick wall. The wall is lined by endothelial cells around which are large, clear, contractile glomus cells probably derived from pericytes. Among the cells goes a network of nerve fibrils. The venous segment has a wide lumen and leads to a subpapillary venule.