DERMATOLOGY
FOR THE HOUSE OFFICER

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Williams & Wilkins

Dermatology for the House Officer

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Dedication

To Rubin Bressler. You have taught me as a father would a son, guiding gently, directing firmly and excusing error. Through you I have grown. For all this, and your friendship too, I am grateful.

Foreword

In his preface, Dr. Lynch writes that this book has three unique features. But it has a fourth: the unique style of the author. No one else has done what he has accomplished with such flare. It is the first book written in a style which makes the study of skin disease both exciting and approachable for the neophyte.

The text is neither all-inclusive or artificially abbreviated, but contains a wealth of information written in a style which expresses the personal touch and philosophy of the author. Dermatology, like other medical and surgical disciplines, is an art and science of diminishing return: the more one learns the less use he or she has for that information; a truth which often discourages the beginner. Better than 95% of all skin disease for which patients encounter a physician is included in the 65 conditions in this book. But the student need not even learn 65 diseases as distinct entities; the problem oriented approach permits most of the diseases to be included in one of the ten groups. Once the clinician has grouped an unknown disease the possibilities are relatively few; usually a maximum of 11 or 12 distinct diseases. Thus, even the novice can make a correct diagnosis by using the information obtained from observing and palpating skin lesions.

The author states this book is written for the same purpose as training wheels are put on a bicycle: it is not intended for the developed free-wheeling clinician. But, in reality, it is written for a certain experienced dermatologist - that individual who wishes to teach skin disease to non-dermatologists and make it understandable, exciting and just plain fun.

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Preface

This is a "working" book. It is written for medical students and house officers who find that dermatology must be learned by caring for patients. It is designed to be carried to the clinical setting where it can be quickly used to establish a list of differential diagnoses, to confirm a suspected diagnosis or to formulate a therapeutic plan. In short, it is a practical book for the self-learner.

This book contains three unique features. First is the problem oriented approach to diagnosis. Most textbooks are organized such that all of the diseases which share a common etiology are grouped together. Thus there are chapters on infections, genetic diseases and metabolic diseases. To use these conventional books one must already know the diagnosis.

This book is organized so that all of the diseases which look alike are grouped together. For this reason there are chapters on each major, morphologic group. When faced, for instance, with a blistering eruption the clinician can turn to a single chapter to find a list containing all of the most likely diagnoses.

The second unique feature is the utilization of a diagnostic algorithm. An algorithm is a systematic procedure for finding an answer to a particular problem. In this case it is a logical roadmap for the clinician to follow as one goes from the presentation of an unknown skin disease to the recognition of an appropriate diagnosis. Decision making is carried out by way of simple yes or no answers to a series of questions based on the appearance of the patient's lesions. The details of this algorithm are covered in Chapter 6.

The third unique feature concerns the way in which diseases were chosen for inclusion in this book. Most textbooks cover far too much material. The problem of selection is minimized if one recognizes that a very small number of diseases account for a very large proportion of the diagnoses a clinician must make. It has been my experience that 50 dermatologic conditions account for 95% of all office visits for skin disease. I have included these 50 diseases and have added an additional 15 diseases which, even though rarely seen, are otherwise important because of their seriousness, contagiousness or treatability. Thus only 65 diseases have been selected for full coverage in this book. I would emphasize that this list, while short, covers all of the important conditions necessary to practice safe, thorough dermatology.

The book is arranged in three sections. The first covers fundamental material on terminology, anatomy, physiology and therapy. The

second explains the use of the diagnostic algorithm and covers the diagnostic features, prognosis, pathogenesis and therapy of the 65 preselected dermatologic diseases. The third covers important and difficult problems in dermatologic differential diagnosis and also reviews the cutaneous aspects of many systemic diseases.

Photographs of skin disease have not been included. Extensive use of color photographs would have raised the price of the book beyond the reach of its intended audience. Black and white photographs (which would have also raised the price appreciably) have considerably less teaching value. Moreover, the diagnostic algorithm around which this book is designed has been constructed for direct application to the patient's skin disease as it is encountered in a clinical setting. It does not depend on visual aids for its utility. However, for those who wish them, good color photographs can be found in the inexpensive color atlases which are listed in Appendix B. One or more of these is likely to be available through your medical library. Medical libraries also often have slide-tape presentations containing good color slides of dermatologic disease. A quick review of the pictures present in these sources will be all that is necessary in the event that large numbers of patients with skin disease are not readily available.

This is not a reference textbook and the references cited in this book are chosen accordingly. References prior to 1977 are best obtained by reading the appropriate chapters in the major dermatologic textbooks (see Appendix A); newer references, with an emphasis on review articles, are included in a general bibliography at the end of each chapter or section. Of major importance is the fact that by using camera ready copy, references as recent as 6 months old have been included.

Finally, this is a single author book. I hope (and expect) that the advantages of uniform style, better organization and reduced duplication outweigh the disadvantage of possible personal bias. To minimize deception caused by bias I have indicated within the text those few areas wherein I disagree with what appears to be the mainstream of American dermatologic thought.

I am very greatful for the encouragement and assistance offered to me by Sara Finnegan, Marjorie Nelson and their many co-workers at Williams & Wilkins. I would also like to acknowledge the important role Marie Jones played in the preparation of this manuscript. All of the typing and most of the proof reading was done by her. This work was done quickly, accurately and uncomplainingly. Finally, I would like to ask forgiveness from my wife, children and friends for my preoccupation with the task at hand. Their understanding and cheerfulness have carried me through the many dark and unproductive moments experienced during the preparation of this manuscript.

Peter J. Lynch, M.D.

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Chapter 1

Anatomy and Physiology

The skin is an impressively large and heavy organ. It occupies well over a square meter of surface area and accounts for about 20% of total body weight. As the boundary between our body and a hostile world it serves several functions. First, the skin acts as a barrier for fluid movement. Internal fluids are kept within the body and external fluids are withheld from penetration. Second, the skin serves as the major means of temperature control for the body. Conservation of body heat occurs both through vasoconstriction and through the insulating properties of the skin itself. Cooling of the body occurs by way of vasodilation and through evaporation of sweat. Third, the skin offers important protection from ultraviolet light. Both melanin production by melanocytes and keratin production by keratinocytes serve to decrease the amount of damage done to cellular DNA as a result of ultraviolet light penetration. Fourth, the skin operates as the source of sensory input to the body. Sensory nerve endings which terminate in the papillary dermis carry important information to the brain regarding our external environment. Fifth, the skin is an organ of metabolism for some important molecules. This role is exemplified by the ability of epithelial cells to synthesize vitamin D.

The skin is composed of three layers: the epidermis, the dermis and the fatty layer. Embryologically the epidermis is derived from ectodermal tissue whereas the other two layers are mesodermal structures.

EPIDERMIS (Fig. 1-1)

The epidermis is the outermost layer of the skin and it is the thinnest of the three layers. In practical terms it is no thicker than three or four pages in this book. The epidermis is responsible for the impervious nature of the skin. Fluid movement is restricted by the presence of a barrier area which occurs at the junction of living and non-living keratinocytes in the outer third of the epidermis. Penetration of ultraviolet light is greatly reduced by the presence of melanin and keratin within the epidermal cells. The epidermis is composed of three major cell types: keratinocytes, melanocytes and Langerhans cells.

Keratinocytes. Keratinocytes account for 95% of all of the cells of the epidermis. These cells are mainly responsible for the production of the fibrillar protein known as keratin but to a lesser degree

other proteins and sterols are also synthesized. Keratinocytes begin their life as germinative, undifferentiated cells at the dermal-epidermal junction. These cells are known as basal cells. Cells in the basal layer and in the layer immediately above it are continuously dividing in such a way that one-half of the cells remain in place to provide cells for replenishment while the other half begin a progressive journey to the skin surface where they will be exfoliated. These basal cells (which already show some degree of perinuclear keratin formation) are attached to one another and to the underlying basement membrane in a series of junction points known as desmosomes. These desmosomal connections are continuously resorbed and rebuilt as cells move by one another during their outward journey. By way of these desmosomes and other connecting points known as gap junctions, a rather constant 200-angstrom distance is kept between adjacent cells. This intercellular space allows for the diffusion of nutrients to the cells as they move outward.

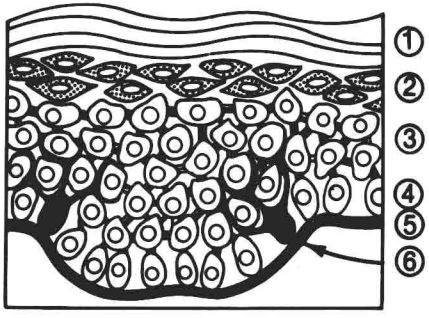

Figure 1-1. The structure of the epidermis. The epidermis is made up of several layers: 1, the stratum corneum; 2, the granular cell layer; 3, the prickle cell layer; 4, the basal layer; and 5, the basement membrane layer. 6 points to a dendritic melanocyte found in the basal layer.

As cells migrate outward from the basal layer the desmosomes, as a result of histologic fixation artifact, become visible as prickles or spines. Thus the mid layer of the epidermis has become known as the spinous or prickle cell layer. Cell activity in this layer is dominated by the production of keratin, the fibrils of which are now seen to extend to the cytoplasmic wall where they connect to the membrane at the site of desmosome formation.

In the outermost portion of the spinous layer, the keratinocytes begin to change in rather noticeable ways. Granules of two types

are now found in the cytoplasm and the shape of the cells becomes increasingly flattened. The larger of the two types of granules, the keratohyalin granule, develops within and around the fibrillar protein contributing in some unknown way to the functional maturity of keratin. The smaller granules, membrane coating granules or Odland bodies, attach to the cytoplasmic membrane where their contents convert the already present intercellular material to a "cement" which is impervious to the movement of fluid. At the same time cell membranes become closely opposed with the formation of "tight" junctions. Further out, the flattened keratinocytes die, losing their keratohyalin granules and nuclei and converting the granular layer to the stratum corneum layer. The dead cells in this layer gradually exfoliate at a rate which causes a steady-state equilibrium and a constant thickness of the epidermis.

Studies of cell kinetics suggest that, in normal epidermis, mitotically active basal cells have a cell cycle time of 200 to 400 hours and further suggest that a basal cell moves to the stratum corneum in an average time of approximately 2 weeks. The control for these kinetics depends on the presence of a number of molecules such as epidermal growth factor, chalones, prostaglandins and polyamines. All of these molecules probably affect the cell machinery by way of the cyclic AMP-cyclic GMP system. Disturbances in epidermal cell kinetics occur in a variety of skin diseases and **have been especially well studied** in psoriasis.

Melanocytes. Melanocytes account for about 1% of the cells of the epidermis. These dendritic, pigment producing cells are derived from the neural crest and are first found in the epidermis in 8-week-old embryos. They are found exclusively in the basal layer where they are interspersed among the basal keratinocytes. Each melanocyte, by way of its dendritic process, is in contact with 30 to 40 surrounding and overlying keratinocytes. Melanin pigment, which occurs through polymerization of tyrosine-derived indole quinones, develops in membrane-bound vesicles known as melanosomes in the cytoplasm of these cells. These pigmented granules are then delivered to the dendritic processes of the melanocytes and are transferred by way of a phagocytic process into the cytoplasm of the multiple keratinocytes which surround each melanocyte.

Variation in normal skin color, including that due to racial differences, is determined not by the number of melanocytes but rather by the number and size of the melanosome granules which are produced and transferred to keratinocytes.

Ultraviolet light, primarily that of 290- to 320-nanometer wavelengths (UVB), darkens the skin both through immediate photooxidation of preformed melanin and, more importantly, through new melanin production in the delayed process known as tanning. An increased amount of melanin pigmentation in the epidermis, whether due to natural coloring or to tanning, decreases the amount of ultraviolet light allowed to pass through the epidermis, thus protecting the cellular DNA of deeper structures from ultraviolet light damage. Individuals such as albinos or those of Celtic background with absent or limited ability to produce melanin are at considerably increased risk for the development of skin cancer.

The skin darkens in response to other stimuli besides that of ultraviolet light irradiation. Endocrine changes associated with

increased elaboration of melanocyte stimulating hormone (MSH) and adrenal corticotropic hormone (ACTH) cause darkening as does the presence of inflammation through the process known as postinflammatory hyperpigmentation.

Nevus cells are pigment producing cells that are almost certainly derived from melanocytes. These rounded up cells lack dendritic processes and occur in nests or clusters. Pigmented lesions containing such clusters are termed nevi. Lesions containing such clusters only at the dermal-epidermal junction are known as junctional nevi; lesions containing clusters both at the dermal-epidermal junction and within the dermis are known as compound nevi; lesions containing clusters only within the dermis are known as intradermal or, more simply, dermal nevi. The nevus cells in the deepest portions of intradermal nevi lack the enzyme(s) necessary to produce melanin from tyrosine. It is not certain whether this simply represents disappearance of the enzyme system due to aging or whether these latter cells have an embryologic derivation from neural cells other than melanocytes.

Langerhans Cells. Langerhans cells account for 3 to 5% of the cells in the epidermis. They are dendritic cells and thus superficially resemble melanocytes. They do not, however, produce pigment and they possess cell surface markers which identifies them as being of monocyte-macrophage lineage. They probably originate in the bone marrow and from there move in and out of the epidermis as required by their role as processors of antigens which adhere to the skin. After appropriate processing, Langerhans cells present the altered antigens to immunocompetent T cell lymphocytes which in turn trigger the inflammatory reaction of allergic contact dermatitis. Langerhans cells are also found in other benign and malignant inflammatory infiltrates such as occur in lichen planus, mycosis fungoides and diseases of the histiocytosis X group. Finally, Langerhans cells are unusually susceptible to deactivation or destruction by ultraviolet light irradiation. This property may be important in the induction of immune tolerance and in the pathogenesis of sunlight induced cutaneous malignancy.

DERMAL-EPIDEMAL JUNCTION

This undulating junction separates the ectodermally derived epidermis from the mesodermally derived dermis (Fig. 1-2). This basement membrane layer or zone presumably serves to attach the epidermis to the dermis and probably also plays a role in supporting the shape of the plasma membrane of the basal cells. From the top downward this basement membrane junction zone is made up of 1) the plasma membrane of the epidermal basal cells together with their hemidesmosome, 2) an electron lucent layer and 3) an electron dense band (the basal lamina) to which are attached connective tissue fibrils (anchoring fibrils) from the underlying dermis. These features can be recognized only on electron micrscopy. Fluids and other small to medium sized molecules pass easily through these layers but cells and very large molecules can only pass through disrupted areas.

The dermal-epidermal junction is important for a number of reasons: 1) it is the site of immunoglobulin and complement depositions in lupus erythematosus and pemphigoid, 2) it is the site of blister formation

for many of the vesiculobullous diseases, and 3) it is a "resistant factor" in the prevention of dermal invasion during the transformation of in situ to invasive basal and squamous cell carcinoma.

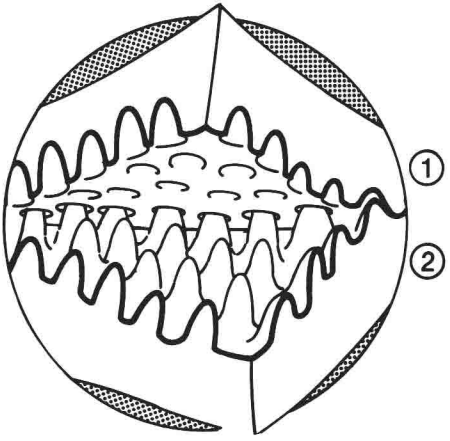

Figure 1-2. This drawing demonstrates the undulating interface between the overlying epidermis and the underlying dermis. The basement membrane layer occurs at this epidermal-dermal junction. 1, the dermis with its irregular surface known as the rete ridges. 2, the dermis with its irregular surface caused by upward projections of the papillary dermis.

DERMIS

Three somewhat indistinct compartments can be described within the dermis. The uppermost is relatively thin and consists most of the loose, areolar connective tissue which lies between the rete ridges of the epidermis. This compartment, known as the papillary dermis, is distinguished by its lighter staining and by the vertical orientation of the connective tissue fibers.

Just below the papillary dermis lies the thicker, major compartment of the dermis known as the reticular dermis. The collagen and elastic fibers found in this section are densely packed and are oriented in a horizontal fashion. It is this tissue that is responsible for the amazing strength and elasticity of the skin.

The lowest of the three compartments consists of the dermal and subdermal fat. Fingers and islands of fat cells are found intermingled with sections of connective tissue at the junction between the two layers but even in the deepest layers of fat, thin septa of connective

tissue outline larger globules of fatty tissue. The fatty layer, by way of its cushioning effect, protects the body from mechanical trauma. Fat cells may also be important as storage and metabolic units. The most important cells and structures found in these three compartments are discussed in the following paragraphs.

Fibroblasts. These mesenchymal, spindle shaped cells are responsible for the synthesis of the collagen, reticulin and elastic fibers of the dermis. The initial construction of all three fibers occurs within the fibroblast but the determination of the final structural morphological features occurs in an extracellular location where enzymes clip unnecessary terminal portions from both ends of the molecule. Elastic fibers are branched fibers which are folded extensively in their relaxed state. This configuration allows for great distensibility and excellent rebound when the skin has been deformed through stretching. Collagen fibers constitute the bulk of the dermis. They are unbranched, long, thin fibers made up of a triple helix of polypeptide chains. Variation in the amino acids of these polypeptide chains accounts for the five or more types of collagen known to be present in the human body. Collagen fibers are responsible for the leather-like toughness and flexibility of skin. Reticulin fibers are closely related to collagen fibers but their exact role is not known. All of these dermal fibers lie in a fluid or semi-solid matrix of mucopolysaccharides which collectively are known as the ground substance. Presumably ground substance, which is elaborated by fibroblasts, serves as a lubricant which allows for smooth, easy movement of the fibers against one another during bending and twisting of the skin.

Blood Vessels. Small arterial blood vessels (arterioles) occur throughout the mid dermis. They are connected at their proximal end to muscular arteries of the subcutaneous tissue and at their distal end to capillary loops. These capillary loops, together with their associated venules, form a complex network in the papillary dermis where they carry nutrients both to the upper dermis and to the epidermis. The blood vessels of the skin, by virtue of dilation or contraction, also serve an important role in heat discharge and heat conservation. Finally, blood vessels carry leukocytes to the skin where they participate in the defense mechanism of inflammatory response.

Nerves. Both myelinated and non-myelinated sensory nerves are found within the dermis. The non-myelinated nerves terminate for the most part as small free twigs at, or just above, the dermal-epidermal junction. Some of the myelinated fibers end in specialized nerve endings. The less well myelinated fibers are primarily responsible for itching and light pain whereas the myelinated fibers are the principal conveyors of deep pain, pressure and temperature. Motor nerves to the skin are entirely autonomic in type. They are sympathetic nerves responsible for the function of eccrine sweat glands, for the determination of blood vessel flow rates and for the contraction of arrectores pilorum muscle fibers.

Miscellaneous Cells. Fat cells are found in clusters within the lower portion of the dermis and constitute the majority of the cells between the lower dermis and the fascia of the underlying muscles. Muscle cells as part of the arrectores pilorum muscles are responsible for the tenting of hair follicles ("goosh flesh") which occurs under

some traumatic environmental conditions. A few other smooth muscle cells are found around the nipples and in the scrotum. Striated muscle is present in the skin only as part of the platysma muscle of the anterior chest. Mast cells are present as scattered isolated cells throughout the superficial dermis. These cells release vasoactive and chemotactic mediators when IgE molecules present on their outer surfaces are bridged by appropriate antigens. These mediators play an important role in all inflammatory diseases of the skin.

ADNEXAL STRUCTURES

Hair Follicles. Hair follicles (Fig. 1-3) are composed primarily of epidermally derived cells. Early in embryogenesis clusters of pluripotential epidermal cells "bud" down from the basal layer and extend into the dermis where they form the hair follicle. A cup shaped indentation at the base of the epidermal bud accepts a specialized dermal papilla with its contribution of blood vessels. The epidermal cells which surround the dermal papilla then differentiate into matrix cells responsible for the construction of hair keratin. Hair keratin because of its greater sulfur content is "harder" than regular epidermal keratin and thus a solid, tough hair shaft is formed. Hair follicles are found everywhere on the skin except on the palms, soles and mucous membranes. However, ectopic follicles composed mainly of sebaceous glands occasionally do appear on mucosal surfaces as small yellow papules. Such lesions are known as Fordyce spots when they occur in the mouth.

The varieties of hair produced by hair follicles differ greatly depending on the type and location of the follicles. Thus thickness, length and curliness are different for scalp hair, eyebrows, eye lashes, vellus hair, beard hair, pubic hair and trunkal hair.

Hair grows in a cyclical fashion. Scalp hair follicles, for example, produce a continuous strand of hair at a rate of about 0.4 mm per day. This continuous growth goes on for several years when, for unknown reasons, the actively growing follicle (anagen phase) rapidly converts to the resting stage (telogen phase). At this point hair construction stops and the epidermal bud of the hair follicle retreats toward the basal layer. During this retreat the hair shaft loosens in the follicle and is soon shed as a "club" hair. Then, after a resting stage of several months, the epidermal bud spontaneously relengthens and hair construction begins all over again. This cyclical process is carried out in a random, non-synchronous fashion in each of the 100,000 or so follicles of the scalp. As a part of this cycle, 50 to 100 follicles switch to telogen phase each day. The resultant loss of 50 to 100 hairs per day should be considered as a normal event since it is balanced by the simultaneous entery into anagen phase of a roughly similar number of previously resting follicles.

When the hair follicle shifts to the growing phase, protein production starts slowly. This results in a hair shaft with a tapered tip. These thin tips are flexible and soft in comparison to sharply edged, non-tapered hairs that are shaved. Thus shaved hair as it regrows looks and feels coarser than uncut hair. In actuality shaving has no other effect on shaft diameter or hair growth rates.

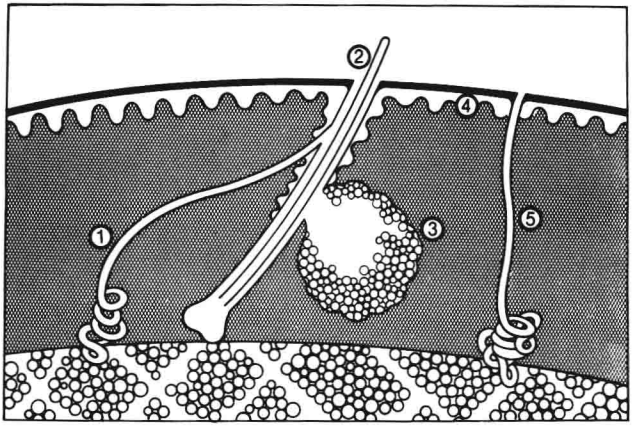

Figure 1-3. The adnexal structures of the skin. 1, an apocrine sweat gland connected by its duct to a hair follicle; 2, a hair shaft lying within a hair follicle; 3 a sebaceous gland attached to a hair follicle; 4, the epidermis, the pluripotential cells of which form the anlage from which hair follicles and eccrine sweat glands develop; and 5, an eccrine sweat gland and duct which opens directly onto the skin surface.

Nail Growth. In a manner of speaking the finger and toenails can be viewed as arising from a "follicle" analogous to that of the hair follicle (Fig. 1-4). The orientation of this "follicle" of course lies parallel instead of vertical to the surface of the skin. The nail matrix from which the nail plate grows lies deep to the posterior nail fold. The nail plate, which is made up of hard keratin very similar to that of the hair shaft, grows outward from the matrix. During this outward growth it rests on and is supported by the nail bed. Epidermal cells from the nail bed contribute only a very thin layer of cells to the ventral surface of the nail plate as it grows outward. Keratin produced from the epidermal cells of the posterior nail fold attaches to the dorsal surface of the nail plate to form the cuticle. Damage to the cuticle such as occurs with "hang nail" formation or overly vigorous manicuring breaks the seal between the cuticle and the nail plate. This creates a blind pocket under the nail fold and allows for the development of paronychial infection.

The lunula of the nail is the crescent shaped whiteness found at the proximal end of the nail plate. It is a reflection of the underlying nail matrix as it extends distally from under the posterior nail