Michael Hertl, editor. Autoimmune Diseases of the Skin. Pathogenesis, Diagnosis, Management. Wien, NewYork: Springer; 2005. Second, revised and enlarged edition. Format: hard cover, one volume. Pages 469, chapters 15.

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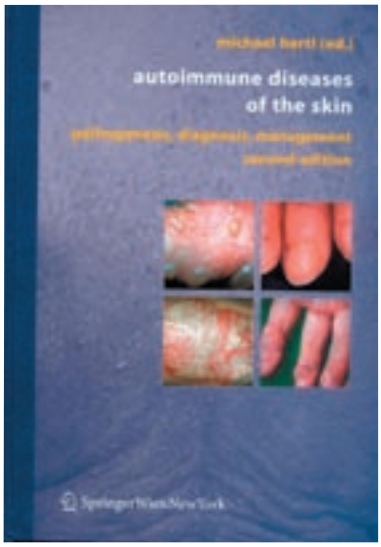

Here is the second, revised and enlarged edition of the book on autoimmune skin diseases, written by 36 authors from Europe, USA, Japan and Israel. Editor of the first and second edition is Dr. Michael Hertl from Marburg, who dedicated the second edition to his teacher, Professor Gerd-Klaus Staigleder, on the occasion of his 80th birthday.

The first, introductory chapter provides an overview of the latest concepts on the pathogenesis of autoimmune diseases and the phenomenon of autoimmunity in general. This complex issue is clearly and concisely presented, making it very convenient for the reader by three excellent schematic presentations. A total of 106 mostly recent references are listed at the end of the section. It should be noted that all chapters of the book are accompanied by a hundred or more references.

Second chapter deals with the epidemiology of autoimmune diseases. The authors stress that epidemiology is an important science that might lead to relevant information on the frequency of autoimmune diseases and the risk factors associated with the disease, thus allowing appropriate distribution of health care resources and planning of preventive mesaures.

The chapter on bullous autoimmune diseases of the skin provides latest data on the etiopathogenesis of pemphigus, with special reference to autoantibody reactivity against desmogleins. The chapter is accompanied by a very convenient and clear table listing autoantigens in various forms of pemphigus. This is continued by a review of animal models for the study of pemphigus, with special reference to desmoglein 3-deficient mouse model. Mention is made of all known clinical variants of pemphigus, and some novel possibilities for an early diagnosis of pemphigus are highlighted. Immunoblot and ELISA analysis of pemphigus sera provides a unique possibility to establish the diagnosis of pemphigus even without histology and direct immunofluorescence of skin biopsy. Some new adjuvant therapies such as immunoadsorption therapy are described. Adsorbents (protein A, tryptophan, phenylalanine, and dextran sulfate as ligands) have been introduced in clinical use. These adsorbents remove their targets through hydrophobic binding. The most important advantages of immunoadsorption over unselected plasmapheresis include higher selectivity in the removal of pathogens, reduced loss of essential plasma components, and no requirement of protein replacement with all its risks.

The chapter on bullous pemphigoid contains highly convenient schematic presentations. It is clear that patient's autoantibodies are directed against bullous pemphigoid antigens 230 and 180. These two autoantigens are components of hemidesmosomes, adhesion complexes in human skin that promote dermoepidermal cohesion.

The review of bullous dermatoses offers recent data on dermatitis herpetiformis, enriched by high-quality immunofluorescence photographs. Epidermolysis bullosa acquisita is an extensively

described clinical entity. Therapeutic options for this dermatosis include colchicine, immunosuppressants, intravenous immunoglobulins, photopheresis, and immunoadsorption.

Fourth chapter of the book tackles scleroderma, i.e. both localized and progressive systemic scleroderma, with special attention paid to the latter. Despite intense research efforts and major advances in the understanding of particular aspects of the disease process, the etiology of systemic scleroderma remains unknown while its pathogenesis is only partly understood.

The chapter on erythematous skin disorders, a thorough account is given of chronic discoid lupus erythematosus, subacute and systemic lupus erythematosus. Thalidomide is mentioned as a therapeutic modality in systemic treatment of subacute lupus erythematosus.

Dermatomyositis is extensively described. Dermatomyositis in adults is associated with malignancy, thus careful evaluation of each patient should be part of the initial and follow-up assessments.

The chapter on mixed connective tissue disease describes it as a rather well defined group of diseases with a characteristic mixed clinical picture of several connective tissue diseases and autoantibody to U1snRNP. This is continued by thorough description of Sjögren's syndrome. The condition may exist as a primary entity or in association with other autoimmune disorders such as systemic lupus erythematosus, dermatomyositis, scleroderma, etc.

The chapter on psoriasis has been written by J.C. Prinz. He concludes that psoriasis has only gained acceptance as a T cell mediated disorder. Therefore, its presentation in a book on autoimmunity may still raise objections. At closer sight, however, psoriasis meets many criteria for autoimmune disease: it has a genetic background with a strong HLA-class I association; microbial infection contributes to the disease onset; and T cells apparently play an essential role in the disease manifestation. Yet, only the identification of the putative autoantigens will definitely prove its autoimmune nature.

Chronic urticaria as an autoimmune disease is described in detail in the tenth chapter of the book. Between 30 and 50 per cent of patients with autoimmune chronic urticaria have IgG autoantibodies directed against alpha-chain of the high-affinity IgE receptor (FcRIα) expressed on dermal mast cells or on blood basophils.

The chapter on lichen planus, lichenoid eruptions and cutaneous graft-versus-host reaction delineates some of the recent aspects of the etiopathogenesis, clinical manifestations and tretment modalities.

Small vessel vasculitides affecting the skin are seen in primary systemic vasculitides, i.e. ANCA-associated vasculitides (Wegener's granulomatosis, MPA-microscopic polyangiitis) and immune complex-mediated vasculitides (cryoglobulinemic vasculitis, HSP, CLA). Secondary immune complex-mediated, e.g., in SLE and other connective tissue diseases, paraneoplastic conditions and infectious diseases also frequently involve small vessels of the skin. Diagnosis is based on detailed patient history, physical examination, focused laboratory investigation, and demonstration of vasculitis by biopsy.

The present, second edition of the book for the first time addresses vitiligo and alopecia areata from the aspect of autoimmune diseases. Critical analysis is given of the hypotheses raised to date on the pathogenesis of vitiligo. While autoimmunity still is the favored hypothesis on vitiligo, the evidence collected to date do not support the idea that vitiligo belongs to the group of primary autoimmune diseases. In case of alopecia areata, a rodent model study has shown that it is an immune mediated disease, strongly suggesting that the mechanism is autoimmune in nature. It is likely that alopecia areata susceptibility and severity modifyng genes are primarily involved in the immune system, while other susceptibility genes may control hair follicle function. The authors conclude that they still cannot claim alopecia areata as an autoimmune disease with complete evidence, yet all evidence produced so far point in this direction and strongly support the hypothesis.

The last chapter is dedicated to novel therapeutic procedures for autoimmune diseases. Mention is made of many new immunomodulators (immunosuppressant macrolides, leflunomide, and antigen specific approach in the management of autoimmune diseases, therapeutic options with the use of autoantigens, antigen modification to induce tolerance, modification of the effector stage of autoimmune diseases via inhibition of tumor necrosis factor α synthesis). The concept of immunoadsorption is explained. The reader's attention is drawn to the use of high doses of intravenous immunoglobulins as a therapeutic option in the treatment of autoimmune diseases. The chapter is closed by a review of biologicals. They

represent a new group of substances including monoclonal antibodies, fusion and recombinant proteins possessing biological activity. In the past years, a major progress has been made in the understanding of etiologic and pathogenetic factors involved in the pathogenesis of human autoimmune diseases. Nevertheless, the majority od autoimmune diseases including those of the skin remain an enigma. This book provides latest information on the spectrum of cutaneous autoimmune disorders for many clinicians and scientists, parctitioners in dermatology, rheumatology, internal medicine, otorhinolaryngology, and pediatrics.

Aida Pašić, MD, PhD

Your skin needs Nivea cream to look young and well-cared for.
From the Nivea collection of Zlatko Puntijar (1936)