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EDITORIAL

Primary Immunodeficiency and Related Diseases

In Allergology International (AI) Vol. 61 No. 2, we have 6 Review Articles, 11 Original Articles, and 1 Letter-to-the-Editor report. AI is publishing theme issues that include reviews in important fields of current research activity written by eminent experts. The theme of this issue is "Primary Immunodeficiency and Related Diseases".

Cutaneous manifestations are displayed as both concomitant or more rarely main symptoms in immunological disorders such as immunedeficiency disease, autoimmune disease, or autoinflammatory disease. Patients with immunedeficiency often have cutaneous abnormalities (eczema, telangiectasia, molluscum contagiosum, warts, etc.) and chronic eczema is a common problem, including atopic dermatitis-like skin lesion. On the other hand, autoinflammatory disease, which is categorized as a group of inflammatory disorders that are non-infectious, non-allergic, non-autoimmune and non-immuno-deficient, also displays distinctive cutaneous manifestations.[1]

Tadashi Ariga discusses about Wiskott-Aldrich syndrome (WAS), including the renewed clinical and basic researches for WAS. WAS is characterized clinically with the triad; immunodeficiency, bleeding tendency with microthrombocytopenia and severe eczema. Topical tacrolimus would be effective in some WAS patients with severe eczema. The pathogenesis of eczema in WAS remains unknown; however, the similar basis for atopic dermatitis could be considered, because high level of IgE and inbalanced cytokine production patterns (Th1 < Th2) are generally observed.[2]

Minegishi and Saito discusses comprehensively reviewed hyper-IgE syndrome (HIES). HIES is a complex primary immunodeficiency, characterized by atopic dermatitis-like skin lesion associated with extremely high serum IgE levels and susceptibility to infections with extracellular bacteria and fungi. Yoshiyuki Minegishi is an excellent pioneer for clinical and basic researches of HIES and identified that most of the patients with HIES have dominant negative mutations in STAT3.[3]

Nobuo Kanazawa is also an excellent pioneer for Nakajo-Nishimura syndrome. Nakajo-Nishimura Syndrome is distinct inherited inflammatory and wasting disease originally reported from Japan. This disease usually begins in early infancy with a pernio-like rashes, especially in winter. The patients develop periodic fever and nodular erythema-like eruptions, and gradually progress lipomuscular atrophy in the upper body. Kanazawa identified homozygous mutation of PSMB8 gene in this disease, which encodes β5i subunit of immunoproteasome.[4]

Tomoki Kawai *et al.* discussed about anhidrotic ectodermal dysplasia with immunodeficiency (EDA-ID). Skin manifestations of EDA-ID include decreased skin pigmentation, periorbital wrinkling and hyperpigmentation, sparse to absent hair, and hypoplastic to absent sweat glands. NF-κB signaling, through nuclear factor-κB (NF-κB) essential modulator (NRMO) or IκBα, is impaired in this disease.[5]

More than 20 million people suffer from cedar hay fever in Japan. In original articles, Makihara *et al.* have added solid evidence regarding the efficacy and safety of intranasal steroid spray for treating Cedar / Cypress pollenosis.[6] Topical steroids cause more undesired adverse reactions to the skin and the eyes compared to the lung and the nose. Ebihara *et al.* have reported[7] that Tacrolimus ophthalmic suspension was safe and effective for severe allergic conjunctivitis and did not increase its concentrations in blood. Genetic variations of IL-33 and its receptor ST2 are almost always significantly associated with asthma in the recent large scale genome-wide association studies.[8] Morita *et al.* found the expression of IL-33 and ST2 mRNA upregulated in the skin after tape stripping in mice and revealed that IL-33 play a role in antigen-induced airway eosinophilic inflammation when the lung had been epicutaneously sensitized with antigen.[9] This report suggests the importance of eczema-related IL-33 for preventing asthma development.

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