Ectopic Lymphoid Structures Harbor Desmoglein-Specific B Cells in the Chronic Skin Lesions of Patients with Pemphigus

Jong Hoon Kim^{1*}, Dawoon Han¹, Ji Young Choi¹, Mi Yeon Cho¹, Sang Eun Lee¹, Soo-Chan Kim^{1*}

Department of Dermatology, Gangnam Severance Hospital, Cutaneous Biology Research Institute, Yonsei University College of Medicine, Seoul, Korea¹

*correspondence : jhkim074@yuhs.ac (Jong Hoon Kim) and kimsco@yuhs.ac (Soo-Chan Kim)

Introduction

Results

Ectopic lymphoid structures (ELSs) exist in chronic skin lesions of the patients with pemphigus and contain desmoglein-specific B cells and plasma cells.

A. H&E staining and immunohistochemical staining of CD4, CD8, CD20, and CD138 in a chronic skin lesion of pemphigus are presented. **B.** Immunohistochemical staining of follicular dendritic cell (FDC), CD11c, D2-40, and lymphotoxin β (LTβ) in a skin ectopic lymphoid structure of pemphigus skin is presented. **C.** The characteristics of the pemphigus patients having ectopic lymphoid structures (ELSs) in skin lesions are listed. **D.** Immunofluorescent staining of CD138, CD20, and recombinant desmoglein 1 and 3 proteins (rDSG1 and rDSG3) in skin ELSs from the patients with pemphigus vulgaris (PV), pemphigus foliaceus (PF), and paraneoplastic pemphigus (PNP) are shown.

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ELS-positive and ELS-negative chronic skins of pemphigus have distinct gene expression profiles

A few CXCR5⁺PD-1⁺CD4⁺ T cells and many CXCL13⁺CD4⁺ T cells exist in skin ELSs of pemphigus

Therapeutic effects of intralesional corticosteroid injection on chronic blisters having skin ELSs

A. A PV patient have chronic blisters on back for 18 months. Clinical pictures and immunofluorescence staining of CD20, CD4, and DAPI are shown before and after 2nd treatment of intralesional corticosteroid injection. **B.** The change of skin lesion area are measured during the treatment of intralesional corticosteroid injection in 6 pemphigus patients who have chronic blisters with ELSs. Treatment was performed every month.

Conclusion

- Skin ectopic lymphoid structures (ELSs) containing desmoglein-specific B cells are present in chronic lesions of the patients with pemphigus vulgaris, pemphigus foliaceus and paraneoplastic pemphigus
- Chemokine signature genes are differentially expressed in skin ELSs from the patients with pemphigus.
- Many CXCL13⁺CD4⁺ T cells are identified in skin ELSs in pemphigus.
- Intralesional corticosteroid injection is effective for the treatment of chronic blisters with ELSs in patients with pemphigus.