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Case Report

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Blister as A Cutaneous Manifestation of Sjogren's Syndrome Determines IL-6 Antibody to Be A Potential Treatment: A Case Report

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A 64 years old female was admitted to hospital due to having repeated blister on skin. Her Dsg1, Dsg3, Bp180, Bp230, VII collagen antibodies were all negative while her ENA-SSA and anti-CCP were positive. Labial glands biopsy revealed that lymphocytes immersed around the glands and a diagnose of Sjögren's syndrome (SS) was made. Skin biopsy indicated subcorneal pustular with neutrophils immersed and direct immune fluorescence as well as blood and blister fluid culture were negative. Additionally, the patient showed a high expression of IL-6 (18371.09pg/mL) in peripheral blood and blister fluid (19997.89pg/mL).

SS is a chronic autoimmune disease characterized by eye and mouth dryness resulting from lymphocytic infiltration of the lacrimal and salivary glands. It has a variety of cutaneous manifestations, such as xerosis, purpura, Raynaud phenomenon, cutaneous vasculitis, annular erythema, etc [1]. Blister was first reported as one of SS's skin manifestation by Gyulai R in 2002 [2]. It was considered that VII collagen antibody took a place in the formation of the blister in SS. However, in our case the VII collagen antibody was not detected in the peripheral blood. Instead, significantly elevated IL-6 expression was observed in our case. Thus, the mechanism of blister as a skin manifestation of SS in our case could be associated with the high IL-6 level. High level of IL-6 was recognized as a promotor to the abnormal B cells, which is considered one pathogenetic reason of the appearance of blister in various autoimmune disease [3]. Elevated IL-6 recruited neutrophils under the connected keratinocytes which poses the foundation of subcorneal pustular existing.

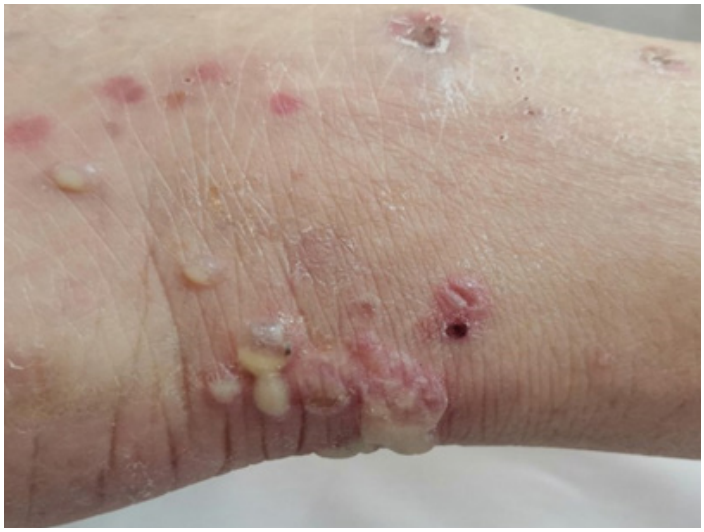

Figure 1

As a matter of fact, the monoclonal antibody targeting the IL-6 pathway has already been used in SS [4]. However, the proportion of patients who had a decrease in the systemic disease activity was 52.7% in the monoclonal antibody group and 63.6% in the placebo group, a non-significant difference. The possible reason why some patients were not sensitive enough to the IL-6 antibody could possibly because they could not be included in the "IL-6-elevated group". Then it is important to differentiate the SS patient who is in the IL-6-elevated group. Distinguishing these patients through the cutaneous manifestation could be most straightforward. In our case, the patient with blister had a high IL-6 expression in both peripheral blood and blister fluid which indicated that blister could be a sign of IL-6 pathway over activated in SS patients. Thus, skin manifestation could be a sign determine whether IL-6 antibody could be used in SS patients.

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