

# 抗デスモグレイン3抗体価が高値・蛍光抗体間接法が陰性を示す尋常性天疱瘡寛解例2例の血清解析

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授与機関名 順天堂大学

学位記番号 乙第 2357 号

抗デスモグレイン 3 抗体価が高値・蛍光抗体間接法が陰性を示す尋常性天疱瘡寛解例 2 例の血清解析

(A high anti-desmoglein 3 antibody ELISA index and negative indirect immunofluorescence in two patients with pemphigus vulgaris in remission)

中原 とも子 (なかはら ともこ)

博士 (医学)

### Abstract

Pemphigus vulgaris (PV) is a disease that features blistering of the skin and mucous membranes caused by autoantibodies directed against desmoglein 3 (Dsg3) and/or desmoglein 1 (Dsg1). Indirect immunofluorescence (IIF) and enzyme-linked immunosorbent assay (ELISA) are two methods that are widely used to measure Dsg3 or Dsg1 antibody titers in PV. Although the titers of these autoantibodies are generally correlated with disease activity, it is known that some PV patients with a high ELISA index do not necessarily have severe symptoms. The autoantibodies of such patients could display interesting characteristics, and many studies of nonpathogenic autoantibodies have been performed in order to understand more about the pathogenesis of PV. We encountered two patients with PV in remission who had a high anti-Dsg3 antibody ELISA index while IIF was negative and we report these cases here focusing on the autoantibody profile.

The anti-Dsg3 antibodies of our patients mainly recognized Ca<sup>2+</sup>-dependent conformational epitopes and targeted mature Dsg3 protein. We report these cases focusing on the discrepancy between ELISA and IIF findings, as well as on the specific characteristics of the patient's autoantibodies evaluated by newly developed methods. These cases emphasize that a discrepancy between disease activity, the ELISA index for Dsg3, and/or IIF findings can occur in PV. Further research on similar patients will be required to elucidate the pathogenic mechanisms in PV patients who have nonpathogenic antibodies and show a discrepancy between ELISA and IIF.