

5th International Conference on

Clinical & Experimental Dermatology

July 13-15, 2015 New Orleans, USA



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Dermolysis bullosa aquisita: A novel auto-immune disease characterized by presence of auto-antibodies to skin specific chains of type IV collagen

We describe a novel autoimmune disease characterized by severe subepidermal bullous eruptions and renal insufficiency with IgG and/or autoantibodies directed against the NC1 domain of the $\alpha 5(\text{IV})$ and $\alpha 6(\text{IV})$ collagen. *In vivo* deposits of IgG and C3 were found along the dermal-epidermal junction of skin lesions. The identity of the target antigen was determined by immunochemical analyses of candidate antigens using the patients' autoantibodies. The patients' IgG autoantibodies reacted with a 185-kDa polypeptide that was distinguished from the known autoantigens of the extracellular matrix including type XVII collagen, type VII collagen, or the $\alpha 3$, $\beta 3$, and $\gamma 2$ chains of laminin 5. Preincubation of the serum with recombinant $\alpha 5(\text{IV})$ NC1 domain of type IV collagen abolished immunoreactivity with the 185-kDa antigen. The serum reacted specifically with the $\alpha 5(\text{IV})$ NC1, among the six NC1 domains of type IV collagen, by Western blot and enzyme-linked immunosorbent assay analyses. The patients' autoantibodies reacted with normal skin and renal glomerulus but not with skin and glomerulus of a patient with Alport syndrome in which the basement membranes are devoid of the $\alpha 5(\text{IV})$ collagen chain. This study provided for the first time unambiguous evidence for the $\alpha 5(\text{IV})$ and $\alpha 6(\text{IV})$ collagen as the target antigen in a novel autoimmune disease characterized by sub-epidermal blisters and glomerulonephritis.

Biography

Reza F Ghohestani completed his internship in surgery at Penn and derm residency at Thomas Jefferson University. He received his Master's in Cutaneous Biology & Pharmacology, and PhD. in Skin Immunobiology from Claude Bernard University, Lyon. Dr. Ghohestani served as the Principal Investigator and team leader for many years at various Academic Institutes. He is a former Editor of Eur J Derm and Chief of Dermatology and Cutaneous Surgery at the University of Texas at San Antonio. Dr. Ghohestani's outstanding work and dedication to excellence have earned him numerous honors and peer recognitions including the American Skin Assoc. & Derm Found Awards.

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