208. GALACTOSE DEFICIENT IGA1 (GD-IGA1) IN SKIN AND SERUM FROM PATIENTS WITH SKIN-LIMITED AND SYSTEMIC IGA VASCULITIS

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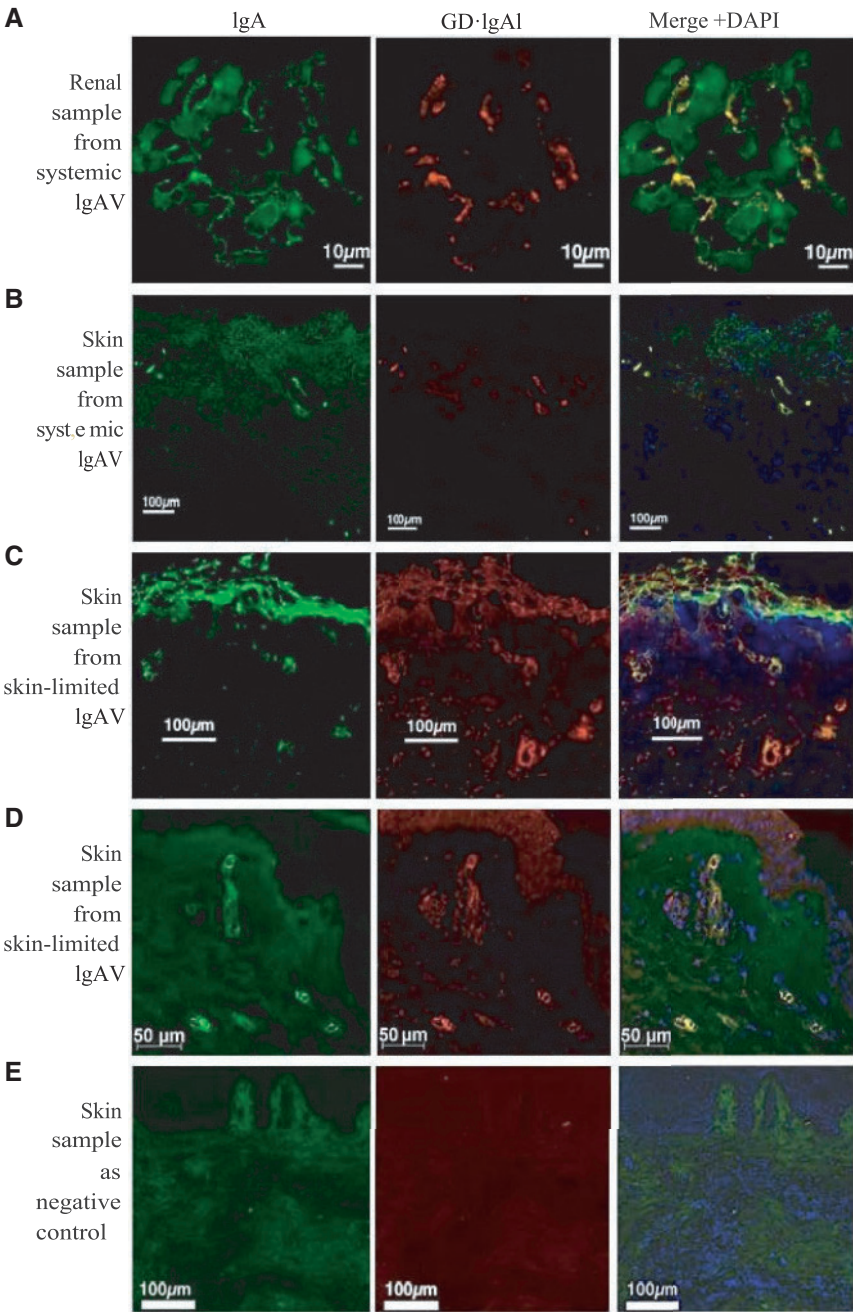
Background: IgA vasculitis (IgAV) in the skin is characterized by perivascular IgA deposition. Besides the systemic form of IgAV with IgA nephritis (IgAVN, formerly referred to as Henoch-Schönlein purpura), there is a skin-limited form of IgAV without clinical involvement of other organs. The reasons for this restriction to the skin are unknown. Systemic IgAV is associated with deposition of galactose deficient IgA1 (GD-IgA1) in glomerular capillary walls and the mesangium, as in IgA nephropathy. We investigated if GD-IgA1 is also detectable in cutaneous vessels in patients with IgAV and if there is a difference between skin-limited IgAV and systemic IgAV with regard to dermal deposition and serum levels of GD- IgA1.
Methods: Immunohistochemical detection of GD-IgA1 using a specific antibody (KM55) was performed on skin biopsies from 12 patients with skin-limited IgAV and 4 patients with systemic IgAV. Serum GD-IgA1 concentrations in 15 patients each with skin-limited and systemic IgAV and in 11 healthy subjects were measured using a KM55-based ELISA.
Results: All skin samples from 12 skin-limited IgAV as well as from 4 systemic IgAV patients showed deposition of GD-IgA1 in postcapillary venules. In serum, average levels of GD-IgA1 were significantly higher in patients with systemic IgAV and renal involvement than in patients with skin-limited IgAV. The latter had higher levels than healthy subjects, but the difference was not significant in our sample of 15 and 11 subjects.
Conclusion: Restriction of vasculitis to cutaneous vessels in skin-limited IgAV cannot be explained by an absence of GD-IgA1 in perivascular IgA deposits. However, the significantly higher average serum levels of GD-IgA1 in systemic IgAV indicate that high concentrations of serum GD-IgA1 in serum may be causally related to extracutaneous involvement and could potentially be used as a predictive marker for renal involvement in patients presenting with cutaneous IgA vasculitis. Skin-limited IgAV, systemic IgAV and IgAN (which is not associated with cutaneous vasculitis) appear to share a GD-IgA1-mediated pathogenesis and may be variants of the same disease.
Disclosures: None



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