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Clinical and Autoantibody Associations in ANA-Positive Systemic Sclerosis Lacking Prototypic Autoantibodies

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The subset of ANA-positive patients with systemic sclerosis (SSc) who lack prototypic SSc-specific autoantibodies (centromere, topoisomerase, RNA polymerase III, “triple negative”) has been poorly characterized. In this study, we aimed to assess clinical associations and prevalence of other autoantibodies in these patients. Patients with ANA+ and triple negative antibodies were identified from two independent SSc cohorts and demographic and clinical data were obtained. Sera were screened for ANA, and autoantibodies were investigated by immunoblots. Patients defined as ANA+ triple negative have equal prevalence of limited and diffuse SSc, and high prevalence of digital ulcers and ILD. These patients have a variety of autoantibodies not commonly clinically assessed, and each has important clinical associations, particularly Ro-52 which is highly prevalent and associated with ILD. ANA+ triple negative patients represent a unique and heterogeneous patient population which should be further characterized.