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Autor(en) des Beitrags:
Yang, JM; Hildebrandt, B;
Luderschmidt, C; Pollard, KM

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Human scleroderma sera contain autoantibodies to protein components specific to the U3 small nucleolar RNP complex.

Abstract:
OBJECTIVE: To determine whether antifibrillarin autoantibodies in scleroderma patients are associated with autoantibodies to protein components specific for U3 small nucleolar RNP (U3 snoRNP).

METHODS: Sera from 220 scleroderma patients were examined for antinucleolar autoantibodies (ANoA) and for antibodies to fibrillarin and the U3 snoRNP-specific proteins Mpp10 and hU3-55K. Clinical correlates were determined for the different autoantibody specificities.

RESULTS: Fifty-nine of the 220 patients were positive for ANoA, and 31 of these patients were antifibrillarin positive. Anti-hU3-55K was found in 10 patients, all of whom were antifibrillarin positive. Twenty-nine patients had anti-Mpp10 antibodies; 23 of these were antifibrillarin positive and 6 were antifibrillarin negative. ANoA, including antifibrillarin, anti-hU3-55K, and anti-Mpp10, were associated with diffuse, rather than limited, systemic or localized scleroderma. Esophageal and lung involvement were more common in patients with antifibrillarin and anti-Mpp10 antibodies, and the highest frequency was in patients with anti-Mpp10 alone.

CONCLUSION: Antifibrillarin autoantibodies are associated with autoantibodies to protein components specific to U3 snoRNP, particularly Mpp10. The prevalence of anti-Mpp10 antibodies in antifibrillarin-positive patients suggests that the U3 snoRNP particle
is a source of immunogenic/antigenic material for the anti-snoRNP response in scleroderma. Autoantibodies to snoRNP components were more frequent in patients with diffuse scleroderma than in those with either the limited systemic or localized forms. The increased expression of these antibodies in patients with the more severe form of scleroderma, coupled with the observations that fibrillarin expression is positively linked to collagen expression in fibroblasts and that fibrillarin is overexpressed in scleroderma fibroblasts, suggests a source of snoRNP to initiate and maintain these autoantibody responses.