Publication of the Month

EliA™ Excellence in Autoimmunity

May 05/10:

Avouac J, Airo P, Dieude P, Caramaschi P, Tiev K, Diot E, Sibila J, Cappelli S, Granel B, Vacca A, Wipff J, Meyer O, Kahan A, Matucci-Cerinic M, Allanore Y

Associated Autoimmune Diseases in Systemic Sclerosis Define a Subset of Patients with Milder Disease: Results from 2 Large Cohorts of European Caucasian Patients

The Journal of Rheumatology 2010; 37:3; doi:10.3899/jrheum.090815

Introduction:

Systemic sclerosis (SSc) is a severe connective tissue disorder characterized by alterations of the microvasculature, disturbances of the immune system, and the massive deposition of collagen in connective tissue. Preliminary results suggest that SSc could be associated with other autoimmune diseases (AID).

The aim of the study was to assess the prevalence and potential associations with the systemic sclerosis (SSc) phenotype of additional autoimmune diseases.

Content:

The present study shows that specific autoimmune diseases were found in 114/585 (19%) French and 179/547 (33%) Italians with SSc. For the diagnosis of autoimmune diseases international standard criteria were used. The frequency of AID in the cohorts and the combined population is shown in the following table:

Disease [n (%)]	SSc Patients from France, n = 585	SSc Patients from Italy, n = 547	p	Combined Population, n = 1132
Sjögren's syndrome	44 (7.5)	93 (17)	< 0.0001	137 (12)
Autoimmune thyroiditis	23 (4)	47 (8.5)	0.025	70 (6)
Myositis	19 (3)	16 (3)	0.86	35 (3)
Primary biliary cirrhosis	14 (2)	17 (3)	0.37	31 (3)
Rheumatoid arthritis	8 (1)	3 (0.5)	0.53	11 (1)
Systemic lupus erythematosus	6 (1)	3 (0.5)	0.53	9 (1)
≥ 1 AI disease associated with SSc	87 (15)	152 (28)	< 0.0001	239 (21)
≥ 2 AI diseases associated with SSc	16 (3)	14 (2.5)	0.74	30 (3)
≥ 3 AI diseases associated with SSc	2 (0.3)	2 (0.3)	0.59	4 (0.3)

Differences between both cohorts: Italian patients were significantly older, their disease duration was significantly longer, a higher percentage of patients with the limited cutaneous subtype were found and positive anticardiolipin antibodies were detected more frequently.

21% of this large series of European Caucasian patients with SSc has developed one or more autoimmune diseases known to occur with CTD. Sjögren's syndrome and thyroiditis were predominant in both cohorts. The study only considers a restricted panel of autoimmune diseases known to occur with CTD.

The presence of at least one autoimmune disease was associated with a milder SSc disease which is attended by the presence of ANA, the limited cutaneous subtype, and lower frequency of digital ulceration. This strongly suggests that genetic background would drive predominant autoimmunity.

Comment:

This publication shows that European Caucasian SSc patients often develop at least one other autoimmune disease.

The association of SSc with another autoimmune disease leads to milder SSc-manifestations. If such manifestations appear, other probably hidden autoimmune diseases should also be considered and tested.