Disease progression in systemic sclerosis-overlap syndrome is significantly different from limited and diffuse cutaneous systemic sclerosis.

Abstract:

Systemic sclerosis (SSc)-overlap syndromes are a very heterogeneous and remarkable subgroup of SSc-patients, who present at least two connective tissue diseases (CTD) at the same time, usually with a specific autoantibody status. To determine whether patients, classified as overlap syndromes, show a disease course different from patients with limited SSc (lcSSc) or diffuse cutaneous SSc (dcSSc). The data of 3240 prospectively included patients, registered in the database of the German Network for Systemic Scleroderma and followed between 2003 and 2013, were analysed. Among 3240 registered patients, 10% were diagnosed as SSc-overlap syndrome. Of these, 82.5% were female. SSc-overlap patients had a mean age of 48±1.2 years and carried significantly more often 'other antibodies' (68.0%; p<0.0001),
including anti-U1RNP, -PmScl, -Ro, -La, as well as anti-Jo-1 and -Ku antibodies. These patients developed musculoskeletal involvement earlier and more frequently (62.5%) than patients diagnosed as lcSSc (32.2%) or dcSSc (43.3%) (p<0.0001). The onset of lung fibrosis and heart involvement in SSc-overlap patients was significantly earlier than in patients with lcSSc and occurred later than in patients with dcSSc. Oesophagus, kidney and PH progression was similar to lcSSc patients, whereas dcSSc patients had a significantly earlier onset. These data support the concept that SSc-overlap syndromes should be regarded as a separate SSC subset, distinct from lcSSc and dcSSc, due to a different progression of the disease, different proportional distribution of specific autoantibodies, and of different organ involvement.