NOVEL DIAGNOSTIC ASPECTS OF ANTI-PM/SCL AUTOANTIBODIES

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Autoantibodies (aab) to the polymyositis/scleroderma (PM/Scl) complex, also known as the human exosome complex, are mainly found in patients with polymyositis/scleroderma overlap syndrome. The PM/Scl autoantigen consisting of nine core proteins and several associated proteins is involved in RNA processing. The majority of the anti-PM/Scl reactivity is directed against the PM/Scl-75c and PM/Scl-100 proteins, but many of the other exosome components also have some contribution to the autoantigenicity of the complex. Historically, the most frequently applied method was the combination of indirect immunofluorescence (IIF) as a screening test followed by confirmation assays. In IIF anti-PM/Scl antibodies have been reported to produce a nucleolar staining pattern on HEp-2 cells. In 2005, it was reported that a significant portion of anti-PM/Scl positive patients contain antibodies that stain cytoplasmic structures, possibly due to anti-GWB antibodies. After the discovery of the major PM/Scl epitope (PM1-Alpha) several studies revealed that an ELISA system based on this peptide antigen is superior for the detection of anti-PM/Scl autoantibodies compared to traditional methods for the detection of those aab. For example, the prevalence of anti-PM1-Alpha in patients with PM/Scl overlap syndrome was 55% compared to 20-30% with conventional techniques. In addition, C1D (18 kDa) a novel PM/Scl associated protein has been described as a major target in sera from PM/Scl overlap syndrome patients. Interestingly, a considerable number of samples of those patients displayed anti-C1D reactivity in the absence of anti-PM/Scl-75c/100 antibodies.