CLINICAL COURSE, PROGNOSIS AND CAUSES OF DEATH IN MIXED CONNECTIVE TISSUE DISEASE

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Objectives: To study the survival rate and prognostic indicators of mixed connective tissue disease (MCTD) in a Hungarian population.

Methods: 280 patients with MCTD diagnosed between 1979 and 2011 were prospectively followed. Clinical features, the autoantibody profile and mortality data were assessed. Prognostic factors for survival were investigated and survival was calculated from the time of the diagnosis by Kaplan-Meier method.

Results: A total of 22 patients died: the causes of death were pulmonary arterial hypertension in 9 patients, thrombotic thrombocytopenic purpura in 3 patients, infections in 3 patients and 7 patients died due to cardiovascular events. The 5, 10, 15 year survival rates were 98%, 96% and 88%, respectively.

Univariate analyses showed that the presence of PAH (p=0.0071, RR: 3.664), serositis (p< 0.001, RR: 4.79), secondary antiphospholipid syndrome (p=0.039, RR: 2.634), cancer (< p0.001, RR: 9.3) and cardiovascular events (p< 0.0001, RR: 3.625) increased the risk of death.

We found a close association between cardiovascular events and the presence of anti-cardiolipin (IgG/IgM: RR: 2.925, 95% CI: 1.50-5.7; IgA: RR: 3.059, 95% CI: 1.15-8.1) and anti-β2-glycoprotein antibodies (IgG/IgM: RR: 2.79, 95% CI: 1.41-5.5; IgA: RR: 6.2, 95 % CI: 2.2-17.7).

Conclusions: During the last decade the number of cardiovascular events increased in MCTD. Beside the anti-U1RNP, the presence of anti-cardiolipin and anti-β2-glycoprotein may play a role in the development of the vascular damage. We believe that monitoring accelerated atherosclerosis and cardiovascular risk factors is an important diagnostic and follow-up task in the modern management of MCTD.