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CLINICAL MANIFESTATIONS OF PROGRESSIVE SYSTEMIC SCLEROSIS IN KOSOVA

KLINIČKA MANIFESTACIJA SISTEMSKE SKLEROZE NA KOSOVU

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Progressive systemic sclerosis is a connective tissue disease that is characterized by skin thickening and internal organ involvement. Patients with systemic sclerosis are classified as having either limited or diffuse cutaneous changes. Internal organs that are involved in systemic sclerosis are lungs, heart, kidney and digestive system.

Aim of this study is to evaluate clinical manifestations of systemic sclerosis in our country.

From January 2005 to December 2009, 30 patients were referred to the Rheumatology Department of Internal Clinic in Pristina for evaluation of clinical manifestations of systemic sclerosis. All the patients underwent evaluation including history and physical examination, lung and hands X-ray, pulmonary physiology testing, esophagography, echocardiography and rheumatologic serologic testing. The diagnosis of systemic sclerosis was based on clinical features: calcinosis, Raynaud phenomenon, distal esophageal dysmotility, sclerodactyly and scattered telangiectasia, interstitial lung disease, heart and kidney involvement. To all patients were investigated laboratory tests such as erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor. In two patients were investigated anti-Scl-70 and anti-nuclear antibodies (ANAs).

In our study 30 patients were investigated with mean age 48 years, where 25 of them were females and 5 males. Based on the degree of cutaneous involvement, patients with systemic sclerosis were 26 or 86.66% with limited systemic sclerosis and 4 or 13.33% with diffuse cutaneous systemic sclerosis. Calcinosis was present in 7

or 23.33% of patients, Raynaud phenomenon 30 or 100%, distal esophageal dysmotility in 22 or 73.33%, sclerodactyly in 6 or 20%, telangiectasia in 22 or 73.33%. Other clinical manifestations also were present: interstitial lung disease in 17 or 56.66%, cardiac involvement (asymptomatic pericardial effusion) in 6 or 20% and kidney involvement with high blood pressure in 2 or 6.66%. Regarding laboratory tests, all patients had high erythrocyte sedimentation rate (up to 40) and positive C-reactive protein, in two patients anti-Scl-70 and anti-nuclear antibodies (ANAs) were positive. Similar to our study, also other authors presented clinical features of patients with systemic sclerosis, based on clinical signs and symptoms. Lomeo et al. reported the clinical manifestations of patients with systemic sclerosis evaluated over a 9-year period. They concluded that esophagography, lung and hands X-ray and pulmonary physiology testing might facilitate the diagnosis of systemic sclerosis. Poormoghim et al. in their study reported a high incidence of Raynaud phenomenon, telangiectasia, and esophageal disease. In our study, we have identified several clinical manifestations in patients with systemic sclerosis that may help in evaluation of the disease.

Based on our study Raynaud phenomenon, telangiectasia and distal esophageal dysmotility were more common. Our study concludes that limited cutaneous systemic sclerosis is more prevalent than diffuse systemic sclerosis.

Keywords: systemic sclerosis, limited cutaneous changes