AB0674  RETROSPECTIVE STUDY OF A COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS IN A TERTIARY CARE HOSPITAL

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Background: Systemic sclerosis (SS) is an autoimmune disease of unknown etiology, characterized by the presence of fibrosis and vasculopathy in skin and multiple internal organs such as the lungs, the kidneys and the digestive tract. The course of the disease is unpredictable and could remain relatively stable or have a rapid evolution. Multiple studies have been carried out to determine the clinical characteristics and survival prognosis on SS patients.

Objectives: To analyze the demographic characteristics, clinical features, treatment and prognosis in a SS disease cohort.

Methods: We performed an observational and retrospective study of patients with SS. The patients had been attended by the Rheumatology department of a tertiary care hospital. We collected demographic, clinical and analytical variables, as well as treatment and prognosis. We classified the disease employing the LeRoy and Medsger, VEDDOS criteria and 2013 ACR/EULAR criteria.

Results: Of our 43 patients, 36 (83.7%) were female and only 7 (16.3%) were male. The average age was 60.4 years (SD 15.6), the average age at diagnosis was 53.3 years (SD 17.6) and the mean time of evolution of the disease was 7.9 years (SD 6.3). Of all the patients, 3 patients (6.9%) died, the mean age at death being 53.6 years (SD 23.7) and the mean time from diagnosis to death of 19 years (SD 10). The most frequently occurring presentation was limited SS (with 18 patients 41.9%), followed by pre scleroderma with 14 patients (32.6%), diffuse SS with 6 patients (14%) and esclerodermia sine esclerodermia with 2 patients (4.7%). Three patients (7%) were labeled with MCTD. SS was associated with other autoimmune diseases in 20% of patients. Five (11.6%) patients developed neoplasms throughout the course of the study. The rest of the clinical characteristics are listed in Table 1 and 2, as well as the strength of association of these with the type of SS, calculated using chi square and Fischer test.

Conclusion: Worthy of noting in our cohort is the absence of scleroderma in more than 40% of our patients, probably because the new criteria have allowed us to diagnose the disease at an earlier stage and also due to the scarce frequency of puffy fingers with respect to other larger series. Digestive involvement was the most frequent visceral manifestation, followed by pulmonary manifestations, specifically interstitial lung disease (ILD). Despite the small sample size, lung disease was significantly associated with the two forms of systemic sclerosis, but not with the diffuse pattern. Both the ILD and the pulmonary arterial hypertension (PAH) were more frequent in patients with SSD. Mortality in all cases was due to interstitial lung involvement. As is frequently described, SS is associated with other systemic autoimmune diseases, constituting an overlap syndrome.

REFERENCES

AB0675  DECREASED CAPILLARY DENSITY IS ASSOCIATED WITH DECREASED HAND MOBILITY IN SYSTEMIC SCLEROSIS

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Background: Nailfold videocapillaroscopy (NVC) is a simple method to evaluate capillary density and morphology. Systemic sclerosis (SSc) is characterized with decreased hand mobility and joint contractures evolving in hands in early disease stage. Movement ability (MA) of hands can be measured with different methods including Hand Mobility Index in Scleroderma (HAMIS), hand anatomic index (HAI), delta finger-to-palm index (ΔFTP), and number of joints with decreased range of motion (joint contractures-JC).

Objectives: To examine correlation between capillary density and MA of hands in patients with SSc.

Methods: 138 patients with SSc underwent detailed examination of MA and capillary density measured with videocapillaroscopy. 70 patients had limited cutaneous SSc (lcSSc) and 66 had diffuse cutaneous SSc (dcSSc). 48 patients had ≤5 years of disease duration (early SSc) and 88 patients had disease duration>5 years (late SSc) calculated from the first non-Raynauds symptoms.

Results: dcSSc patients had significantly higher HAMIS score than lcSSc patients (4.51/5.65/5. vs. 2.05/4.5; p<0.01). HAMIS score showed negative correlation with capillary density in the entire cohort (rho: -0.269, p<0.01) and in dcSSc patients (rho: -0.487; p<0.0001), but not in lcSSc ones.

HAI showed a significantly higher value in lcSSc patients than in dcSSc patients (3.79/3.19/3.76/ vs. 2.64/3.76; p<0.01). Significant positive correlation between HAI and capillary density was found in dcSSc patients (rho: 0.377, p<0.01).

ΔFTP value than lcSSc patients (73.86/65.63/87.00/ vs. 82.81/73.63/90.5/; p<0.05). Positive correlation was found between capillary density and ΔFTP in the entire cohort, in dcSSc patients even if dcSSc was examined separately as early dcSSc and late dcSSc. No significant correlation was found between capillary density and ΔFTP in lcSSc patients.

The number of JC was significantly higher in dcSSc patients than in lcSSc patients (16/0/28/ vs. 0/11/; p<0.01), and no statistically significant difference was found between early SSc patients and late SSc patients. Strong negative correlation was found between JC count and capillary density in the entire cohort (rho -0.376; p<0.0001), in dcSSc patients (rho -0.501; p<0.0001), in early SSc patients (rho -0.583; p<0.0001) and late SSc patients (rho -0.262 p<0.05), but not in lcSSc patients.

Conclusion: Decreased capillary density correlated with decreased hand mobility and higher number of JC. Tissue ischemia may play role in the development of joint contractures and it might be a contributing factor to decreased hand mobility impairment in SSc.

REFERENCES