Methods Thirty patients with SSc aged 16 to 68 years (mean age 47.46 ± 2.41 yrs) were included in the study. Nineteen pts had limited cutaneous SSc (lSSc), and eleven – diffuse cutaneous SSc (dSSc). The morphological changes of skin capillaries were estimated by the nailfold capillary microscopy. The skin microcirculation was evaluated by laser Doppler flowmetry (LDF).

Results All studied patients showed an abnormal nailfold capillary pattern: enlargement of the capillary loop (dilated capillaries) and the loss of capillaries either diffusely or in limited areas. Depending on a degree of the nailfold capillaries changes the patients were divided into 3 groups: with mild (17 pts), moderate (14 pts) and severe (9 pts) changes. The level of basal skin blood flow in first group of the patients was 0.624 ± 0.031 v. In second group of the patients basal skin blood flow was 0.361 ± 0.050 v and there was also significantly below, when compared with first group of patients ($p = 0.002$). The lowest level of basal skin blood flow 0.212 ± 0.063 v was observed in the patients of third groups and also was significantly smaller than in the patients of first group ($p < 0.001$). There wasn't a statistically significant difference in basal skin blood flow levels between 2nd and 3rd groups of the patients. The level of basal skin blood flow closely correlated with number of capillaries ($r = 0.73$; $p < 0.001$). In the patients with dSSc the greater decrease of number of capillaries was observed, than in the patients with lSSc. Basal skin blood flow in dSSc was considerably below, than in lSSc (0.438 ± 0.040 v and 0.250 ± 0.063 v respectively; $p = 0.02$).

Conclusion Thus, the microcirculation abnormalities closely correlated with structural changes of capillaries in SSc.

AB0107 MULTIPLE FIBROSIS: TWO CASES OF SUCCESSFUL TREATMENT

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Background

Objectives Multiple fibrosis (MF) is a slowly progressive disease of unknown cause characterised by localised connective tissue accumulation with the formation of multiple dense sclerotic foci leading to the internal organ dysfunction. It's a rare and often fatal disease resulting in the visceral organ insufficiency. The most common localizations of the fibrotic lesions are retroperitoneal, mediastinal and lung fibrosis, pseudotumor of pancreas, sclerosing cholangitis. A disease process is usually difficult to stem and often impossible to reverse. There is no specific treatment for MF.

Methods

Results We report two cases of successful MF treatment with the combination of D-penicillamine (DPA) and steroids (S). A. 36 yr old male presented with 4 yr history of ?cancer like? symptoms: progressive fatigue, weight loss, subfebrile fever, accelerating ESR (40 mm/h), pulmonary (dry cough, dyspnoea, atelectasis of the left lower lobe of the lung by X ray and dense ?tumour? 60 mm. in diameter in the core part of the left lung with a massive ?infiltration? spreading along the lower pulmonary vein by thoracotomy) and eye (loss of sight and right eye pain, unilateral scleral congestion) manifestations. With suspicion on the lung and eye cancers the total left pulmonectomy and right eye enucleation were carried out without any state improvement. Two yr later fasciitis, hepatosplenomegaly and new round foci in the

right lung (15 mm) and pancreas (27×35 mm) were found. MF was confirmed by histopathological finding and the combined therapy with DPA 1000 mg/d and oral steroids 30 mg/d (decreasing to 15 mg daily after 0.5 year and remaining the same for 1.5 year) was prescribed. Over the subsequent 4 months the general condition of the patient improved, the temperature subsided, fasciitis and abdominalgia disappeared completely, the pt gained 7 kg of weight, ESR diminished to normal levels. In a year we revealed obvious decreasing of the liver and spleen sizes and complete disappearance of the foci in the right lung and pancreas. During a 9 yr follow up we did not find any fibrotic foci in tissues, all laboratory tests are within normal limits. The pt receives daily maintenance dose of 125 mg DPA and 7.5 mg S. No side effects of the drugs have been noticed.

The second patient, a 39 yr old female with Ormond's disease (retroperitoneal fibrosis which led to squeeze the ureters and bilateral hydronephrosis) had also been operated on – left nephrectomy – before a correct diagnosis was made. The administration of DPA 500 mg/d and S 20 mg/d check the progression of the disease during two years.

Conclusion Our results support data on successful use of the combined therapy with DPA and S in MF. Such drug treatment allows to get a reverse development of the fibrotic lesions and to maintain a long stable remission of the disease.

AB0108 COMPLICATIONS AND FORMAL PATHOGENESIS OF PROGRESSIVE SYSTEMIC SCLEROSIS. A RETROSPECTIVE STUDY

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Background Progressive systemic sclerosis (SSc) is an autoimmune disorder anatomically characterised mainly by successive fibromuscular intimal proliferation of the blood vessels and by the increase of collagen fibres in various organs. The disease is, in general, characterised by the extent of the process and by the stage of the changes.

Objectives The aim of this study was to determine the main complications of SSc in various organs and histologically characterise the interstitial fibrotic process of SSc patients who died in different stages of their disease.

Methods A randomised autopsy population of 11 in-patients (female 10, average age: 53.6 years; male 1, age of 65 years at death), with clinically confirmed SSc was investigated retrospectively. 30–100 tissue samples from each patient were serially cut and stained with H-E, or picrosirius red F3BA.^{1,2}

Results In 10 of 11 cases, SSc was complicated by a wide spectrum of histological changes of the blood vessels. The lungs showed complex abnormalities such as vascular changes, interstitial pneumonitis, fibrosis or honeycomb-lungs in 6 of 11 patients. In the heart the complex vascular changes were accompanied by multifocal myocardiocytolysis and/or endo-myocardial fibrosis, which led to circulatory failure in 5 of 11 cases. Complex nephropathy in 10 of 11 patients was characterised by interstitial nephritis and/or fibrosis, by mesangioproliferative or membranous glomerulonephritis, and by multifocal cortical (tubular) necrosis. These renal changes led to uremia in 6 of 11 cases. Multifocal acute pancreatitis (of vascular origin) was

observed in 6 of 11 cases. Different tissues and organs were affected to varying degrees, and the anatomic changes were simultaneously present in different stages at various sites. In all cases SSc was the basic disease which led to death.

Conclusion Anatomic changes could be seen in the early, advanced or late stages of the disease, essentially related to the chronology of the progressive sclerotizing process. SSc can be considered to be a progressive multifocal process characterised by simultaneous histological changes in different stages of their progression. In the course of the disease new foci form, which increase and become confluent, ultimately leading to diffuse, systemic interstitial sclerosis. (Supported by ETT Project T-11 226/99 of the Hungarian Ministry of Health).

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AB0109 CARDIAC ABNORMALITIES IN SYSTEM SCLERODERMA

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Background During last decade the morbidity rates in system scleroderma (SSc) have been increased. The bridge-head of aggression in SSc is huge, since there are actually no organs without connective tissue in the human organism. It also determines a high frequency of different visceral complications in SSc. The cardiac lesions in SSc are frequent visceral manifestation of illness, very often resulting in fatal consequences. In this context we attempted to analyse the main forms of cardiac pathology in SSc.

Objectives We examined 52 patients (pts) with SSc. Among them women were predominant (38/52, 73%). The age of patients and duration of disease varied from 29 to 48 and from 2 to 9 years, the average age of patients and average duration of disease constituted 36.2 ± 1.1 and 4.6 ± 0.4 years respectively. The patients with moderate activity of pathological process (degree I and II) were absolute majority (46/52 pts, 88.4%). Visceral implications were diagnosed in 42/52 pts (80.7%), among them cardiac lesions – in 41/52 pts (78.8%).

Methods Electrocardiography (ECG), echocardiography (EchoCG).

Results ECG abnormalities were detected in 41/52 pts (78.8%). Cardiac arrhythmia was determined in 32/52 pts (61.5%). Its most frequent form was extrasystole (18/52 pts, 34.6%): ventricular (9/52 pts, 17.3%), atrial (6/52 pts, 11.5%), atrioventricular (3/52 pts, 5.8%), single (8/52 pts, 15.4%) and group (10/52 pts, 19.2%) extrasystole. Violations of conductivity were also frequent: atrial – in 18/52 pts (34.6%), ventricular – in 16/52 pts (11.5%), right bundle-branch block – in 6/52 pts (11.5%). These abnormalities correlated with duration of disease, but did not depend on activity of SSc. Among 41 patients with ECG abnormalities the decrease of voltage was detected, possibly due to expanded cardiosclerosis.

EchoCG was conducted to determine myocardial contractility. In 27/52 pts (51.9%) was diagnosed LVEF reduction on 5%, in 11/52 pts (21.2%) – on 7–8%, in 3/52 pts (5.8%) – on 10% and more. The mitral valve diseases and pericardial implications were detected in 8/52 pts (15.4%) and 11/52 pts (21.2%) respectively.

Among all these patients the phenomena of myocardial dystrophy and hypoxia, violation of conductivity, excitability and cardiac output were detected that resulted in chronic heart failure. In many cases the heart failure was latent, resulting in late diagnosis and treatment.

Conclusion Carditis was diagnosed among more than three fourth of patients with SSc. The main reason of its development seems to be system sclerosis.

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AB0110 PULMONARY FUNCTIONAL ABNORMALITIES IN ASYMPTOMATIC PATIENTS WITH SYSTEMIC SCLEROSIS

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Background Systemic sclerosis (SSc) is a multisystem disease and pulmonary manifestations are common in SSc patients. Involvement of the lungs in SSc patients is detected with x-ray and functional pulmonary testing. The relationship of the development of the pulmonary abnormalities to duration of the disease and other organ involvement remains unclear.

Objectives The present study was designed to determine the pulmonary function indices in the SSc patients without symptoms of the lung involvement.

Methods Twenty-six women with definite diagnosis of SSc, aged 45.4 ± 11.2 yrs were studied. Control values were obtained from 20 healthy age-matched women. Additionally, the SSc group was compared to the predicted spirometric data based on large population studies. The resting pulmonary function tests were performed with EDS-Sprint spiograph. The measurements were done between 8.00–9.00 AM.

Results Vital capacity and maximal voluntary ventilation were decreased in the SSc patients. Forced vital capacity and one-second forced expiratory volume were also decreased but the later was normal when corrected for reduction of the forced vital capacity. Forced expiratory rate at 50% and 75% of forced vital capacity was impaired in the patients. Forced expiratory flow rate at 25% of forced vital capacity did not differ from the controls. The rate of incidence of abnormal pulmonary functional test in the SSc patients was 30–70 per cent.

Conclusion Pulmonary involvement is common in SSc patients without overt manifestation of the lung disease. The lung function testing is a simple, cheap and safe method for evaluation of the respiratory system, and is recommended as a routine test for all patients with SSc.

AB0111 PROGNOSTIC SIGNIFICANCE OF LIMITED AND DIFUSSE FORM OF SKIN CHANGES IN SYSTEMIC SCLEROSIS

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Background