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).

REFERENCES

- 1 Sweat F, et al. *Arch Pathol.* 1964;**78**:69–72
- 2 Constantine VS, Mowry RW. *J Invest Dermatol.* 1968;**50**:419–23

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.

REFERENCES

- 1 Guseva NG. System scleroderma and Nenoaiay neeaiaaiaiey and pseudosclerodermic syndromes. *Moscow: Medicine*, 1993: **269**
- 2 Jimenez SA, Hltraya E, Varga J. Pathogenesis of scleroderma. *Collagen. Rheum Dis Clin North Am.* 1996;**22**(4):647–74

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 spirograph. 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

Objectives To assess the difference in frequency of pulmonary and renal involvement, as well as lungs insufficiency and renal failure between patients (pts) with limited cutaneous Systemic Sclerosis (lcSSc) and diffuse cutaneous Systemic Sclerosis (dcSSc). **Methods** Group of 138 pts with Systemic Sclerosis (SSc), 127 females and 11 males, age 28–75 years (median 53), were followed for 3–26 (median 13) years period. Extent of skin involvement was recorded, and pts were examined for radiographic signs of pulmonary fibrosis, carbon monoxide transfer (TCO) factor (done in 28 pts), and renal involvement and failure (urine sediment, creatinin clearance, arterial tension). Difference of visceral involvement, pulmonary insufficiency and renal failure between pts with lcSSc and dcSSc was correlated.

Results Among 138 pts, 105(76%) had lcSSc, and 33(24%) dcSSc. Pulmonary lesion were found in 40/105 pts with lcSSc, and in 25/33 pts with dcSSc ($p = 0,004\text{-}\chi^2$ test). Renal lesion was detected in 11/105 pts with lcSSc, and in 8/33 pts with dcSSc ($p = 0,04\text{-}\chi^2$ test). Signs of lung insufficiency developed in 30/105 pts with lcSSc, and in 18/33 pts with dcSSc ($p = 0,04\text{-}\chi^2$ test). Lower creatinin clearance and high arterial blood pressure were detected in all patients with recorded renal lesion during the follow up period (in 11/105 pts with lcSSc, and in 8/33 pts with dcSSc). Renal failure was significantly more frequent in dcSSc pts ($p = 0,04\text{-}\chi^2$ test). In 7(21%) pts with dcSSc no criteria for pulmonary and renal involvement were present during the follow up period. On the other hand, in 14(13%) pts with lcSSc pulmonary insufficiency or renal.

Conclusion Diffuse cutaneous lesions were prognostic sign of lungs and kidneys involvement, as well as pulmonary insufficiency and renal failure in most of the patients, but about 20% of the dcSSc patients were without severe visceral lesions during the 13 years of follow up period. On the other hand, 13% of patients with limited cutaneous lesions had bad prognosis and developed pulmonary insufficiency or renal failure in first 5 years of disease duration.

AB0112 SEVERE PULMONARY HYPERTENSION (PH) IN A PATIENT WITH SYSTEMIC SCLEROSIS (SSC) AND ANTIPHOSPHOLIPID SYNDROME (APS) – CASE REPORT

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Background PH is a well-recognised complication of SSc and may be secondary to interstitial lung disease or present as isolated PH due to vascular changes. The coincidence of SSc and APS is not common and only a few reports can be found in the literature.

Objectives We describe here a case of a patient with severe PH in a course of limited SSc and secondary APS.

Methods

Results A 63-year old Caucasian woman with 50-years history of Raynaud phenomenon and 3-years history of mild dyspnea on exertion, previously untreated, suddenly developed fever and chest pain. She was admitted to a general hospital, diagnosed as having right pleuropneumonia and cardiac insufficiency, and treated with antibiotics, ACE inhibitors, diuretics and small doses of methyl-digoxine which led to disappearance of pleural effusion and leg oedema. Since fever, chest pain, and dyspnea persisted she was sent to Department of Rheumatology and Internal Medicine, Medical Academy of Bialystok. Physical examination revealed sclerodactyly, multiple telangiectasias on the face and hands, microstomia, systolic murmur with pulmonart II-nd tone accentuation, and a palpable liver. ECHO-DOPPLER revealed high pulmonary artery pressure (PAP) of 86 mmHg with distention of the right ventricle. There were no signs of interstitial lung disease in high resolution computed tomography. However, examination of bronchoalveolar lavage fluid showed elevated percentage of lymphocytes. Perfusion scintigraphy of the lungs showed pulmonary embolisms/thrombosis. USG-DOPPLER of the lower limbs showed no signs of vein thrombosis. The patient's ESR was 145/hour, serum CRP-253 mg/dl, antinuclear antibodies positive in a titer of 1:1280, speckled, thrombocyte number – 115 000/mikrol. The patient was diagnosed as having SSc complicated with PH. In addition to previous therapy, treatment with cyclophosphamide (1000 mg i.v.), low molecular weight heparin (LMWH) and nifedipine was started. On the third day after cyclophosphamide infusion dyspnea worsened and lower limb oedema appeared, PAP increased to 115 mmHg, and blood platelets fell to 80 000/mikrol. Anticardiolipin antibodies were detected in a titer of IgG-96 ACG and IgM-36 ACG. Secondary APS was diagnosed. LMWH was replaced with intavenous infusion of heparin followed by oral anticoagulants and nifedipine dose was increased to 120 mg/day resulting in PAP fall to 90 mmHg, less dyspnea and disappearance of leg oedema. The patient now feels well with PAP of 60 mmHg.

Conclusion The case described here indicates that APS may participate in the pathogenesis of PH complicating SSc and highlights the need to consider determining ACL antibodies/lupus anticoagulant in SSc patients.

AB0113 ANALYSIS OF CLINICAL MANIFESTATIONS IN PATIENTS WITH CREST VARIANT OF SCLERODERMA

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Background

Abstract AB0113 Table 1

Clinical manifestations	Interstitial lung disease	Renal involvement	Hypertension	ECG changes	Arthritis	Tenosynovitis	Resorption of distal phalanx	Ulcerations	Sicca syndrome
ISSc	50.0%	29.4%	52.9%	50.0%	20.6%	17.6%	58.8%	91.0%	8.8%
dSSc	58.5%	29.4%	70.6%	64.7%	17.6%	23.5%	82.3%	100%	11.8%
Statistical significance	NS	NS	NS	NS	NS	NS	NS	NS	NS