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 intravenous 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