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

Objectives The aim of the study was to ascertain frequency of clinical manifestations in patients with CREST variant related to skin involvement in SSc.

Methods Since 1993 we have treated 184 patients with systemic sclerosis (SSc) at the Institute of Rheumatology in Belgrade. Within this group there were 51 (27.7%) – 49 females and 2 males, with complete clinical manifestations of CREST variant. Mean age of patients was 55.7 years and disease duration 10.9 years. Thirty-four patients (66.7%) had limited cutaneous SSc (lSSc) and 17 (33.3%) diffuse cutaneous SSc (dSSc).

Results Our results are in following Table 1:

Conclusion According our results, visceral involvement in systemic sclerosis does not depend on distribution of skin sclerosis. So we think that CREST variant of SSc can be classified as separate form of scleroderma.

AB0114 RAYNAUD'S PHENOMENON: CORRELATION BETWEEN CLINICAL MANIFESTATIONS AND NAILFOLD CAPILLARY MICROSCOPY

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Background

Objectives To determine the correlations between clinical characteristics and nailfold capillary patterns in patients with Raynaud's phenomenon (RP).

Methods Analysis of clinical and immunological features (RP: phases; time duration in years, symmetry, localization; skin and organ involvement, ANA tests) and nailfold capillary patterns were assessed in 150 patients referred for evaluation of RP during the period 1999–2000. Chi-Square and Fisher's tests were used for statistical analysis.

Results 142/8 female/male; mean age: 42 ± 15.6 years; RP duration: 5.7 years. 47% of RP were referred without diagnosis, 5.5% as primary RP (pRP) and 49% as secondary RP (sRP): connective tissue diseases (CTD) in 73% of cases. Microangiopathic patterns detected were: 38% normal, 25% functional (capillary pallor/hyperemia); 20% CTD pattern without specific capillary abnormalities of scleroderma (SSc), and 17% with SSc-pattern (? slow pattern? 7.6%; ?active pattern? 9.7%). Patients with slow pattern SSc were older than the patients with other patterns: mean average: 53.8 years ($p < 0.05$). RP with uniphasic colour changes were observed principally in patients with normal or functional pattern and RP with biphasic colour changes in patients with SSc-pattern ($p < 0.02$). Significant association between ?slow? pattern SSc and anticentromere antibodies was detected ($p < 0.004$) and between ?active? pattern SSc and positive ANA test with nucleolar pattern. Anticentromere antibodies were detected in 9% of patients showing functional pattern with capillary pallor and ?active? pattern SSc was observed in 42% of positive Scl-70 antibodies patients. Sclerodermatous skin changes was associated with SSc-pattern and functional capillary pallor pattern ($p < 0.001$) and a higher proportion of soft tissue swelling in hands and organ involvement was detected in patients with ?slow? pattern-SSc.

Conclusion Functional pattern with capillary pallor showed a significant association with uniphasic RP, sclerodermatous skin changes and negative ANA test. Only 42% of patients with positive Scl-70 antibodies exhibited ?active? pattern.

AB0115 SCLERODERMA-LIKE CHANGES IN A DIABETIC PATIENT: A CLINICAL CASE

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Background Diabetes mellitus is a metabolic systemic disease that can be associated, particularly when glucose metabolism control is poor, with skin changes and sometimes finger contractures of the hands that clinically resemble scleroderma. Distinction between the two different conditions is important, because they have distinct therapeutic approach and prognostic.

A 55 year-old white woman came to our Unit because of skin changes and articular contractures. She had diabetes mellitus since a gestation eleven years ago and was being followed in the Endocrinology department for this reason. Her disease had been resistant to antidiabetic oral treatment and was complicated with a severe polyneuropathy in glove and stocking pattern. Control of glucose metabolism was poor and she was being taught to start insulin therapy. She had been also diagnosticated of an autoimmune thyroiditis and recent arterial hypertension. We observed a thin woman with very shiny skin over her fingers and unreducible contractures of some of them in both hands. The skin of her distal legs was bilaterally swelled and sclerotic too. She referred a Raynaud phenomenon, which we weren't able to provoke with cold water. There were no other musculoskeletal findings or symptoms suggesting CREST or systemic disease beyond diabetes. Her ANA and anti-DNA were negative, as was anticentromere antibody and Scl70. The hand x-rays showed neither calcifications nor erosions. A capillaroscopy was performed, showing some megacapillaries and structural disorganisation. No avascular areas were found. We concluded, in face of the analytic results, that this was a scleroderma-like manifestation of diabetic cheiroarthropathy and not systemic or limited scleroderma. The changes seen in the capillaroscopy could be due to the cheiroarthropathy itself.

Objectives

Methods

Results

Conclusion

AB0116 TREATMENT OF SYSTEMIC SCLEROSIS WITH ANTI-TNF-ALPHA ANTIBODIES. A CASE REPORT

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Background In february 1999 a 39 year old woman developed progressive swelling of the hands, arthralgias, Raynaud's phenomenon, dyspepsia and reduced physical strength.

For this reasons, positive ANA's (no anti topoisomerase I antibodies, pathologic oesophagus manometry and clinical signs of lung involvement including typical skin lesions (thickening, necrosis, erythema anulare, etc.) systemic sclerosis was diagnosed.

In the course of disease DMARDs such as penicillamin and cyclophosphamide showed no efficacy over a period of 18 months. Relatively high doses of methylprednisolone (up to 250 mg/d; not below 20 mg/d) were needed to reduce the clinical signs of the disease.

In december 2000 anti-TNF-alpha-antibody therapy (infliximad) was started in a dosage of about 5 mg per kg body weight.