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BOOK OF ABSTRACTS

P 40

ANTIBODIES TO COLLAGEN TYPES I AND IV IN SCLERODERMA
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In the present study, using ELISA, we examined the presence of IgG, IgM, IgA, IgD and IgE antibodies (AB) to collagen I and IV in sera of 20 female patients with scleroderma, who ranged in age from 20-65 years. Based on the extent of cutaneous involvement they were divided into 2 subsets (according to Le Roy, 1988) - 10 patients had limited cutaneous sclerosis (lSSc) WITH ACROSCLEROSIS only, and 10 had diffuse systemic sclerosis (dSSc) with skin changes on the TRUNK. The majority of patients were positive for anticollagen IgA and IgE AB. AutoAB to interstitial collagen I were noted for IgA in 60% of dSSc patients vs. 40% of lSSc and for IgE in 50% of patients in each subset. Most significantly, 70% of dSSc patients vs. 10% of lSSc had IgA autoAB to collagen IV. For IgE to collagen IV were positive 60% of dSSc and 50% of lSSc patients. In conclusion, these preliminary results suggest that IgA antibodies to collagen type IV may play a role in the development of pathological process concerning basement membranes in dSSc. Further longitudinal studies of the same patients are warranted to define its significance as a predictor of this process.

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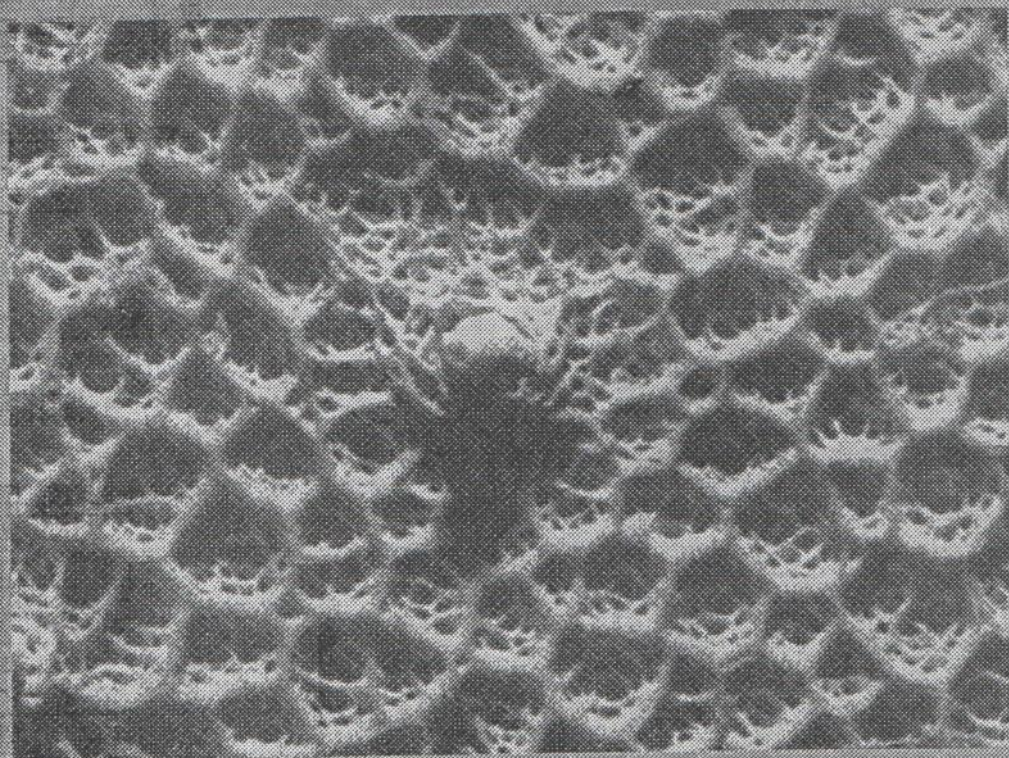
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in the development of vasculopathy, involving the adrenal vessels and leading to bilateral adrenal suppression. We interpret them as a "new reason" for the development of Morbus Addison. We include melanoderma in the list of the dermatological markers of antiphospholipid syndrome.

P-099 SCLEROMYXEDEMA WITH PRONOUNCED SCLERODERMA-LIKE FEATURES

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Scleromyxedema, an entity displaying clinical similarity to systemic sclerosis (SSc) with characteristic mucin deposits and fibroblast proliferation, can present diagnostic difficulty. The differentiation is of practical significance since the internal involvement, if present, differs considerably, with no vascular and immunological abnormalities characteristic of SSc; in about half of the cases is associated paraproteinemia, mostly of IgG lambda, type with no myeloma. We present a female, 42-year-old, in whom the erythematous, oedematous and slightly indurated skin lesions appeared at the age of 29 years, and within 3 years developed diffuse skin sclerosis recognised as SSc. She was seen by us at the age of 37 yrs., and at that time the face was mask-like, there was sclerodactyly, she had moderate muscle involvement and swallowing difficulties. Noticeable were retroauricular nodules and elevated skin-coloured papules on the forehead. Facial features with slowly progressing ectropion, pronounced sclerodactyly without osteolysis and digital pits, presence of papules and nodules and histopathologic picture (proliferation of fibroblasts with mucin deposits) were characteristic of scleromyxedema. There were no visceral changes of SSc type, and esophagus did not show abnormal peristalsis. No paraproteinemia was disclosed within 10-years of her illness. The presented case, in spite of mimicking SSc, has all characteristics of scleromyxedema. Scleromyxedema should be recognised as scleroderma-like entity differing from SSc in the pathogenesis, the course and prognosis.

P-100 ERYTHEMATOUS FACIAL NODULES IN ASSOCIATION WITH ERYTHEMA NODOSUM OF THE LOWER LEGS

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Although erythema nodosum is usually confined to the lower legs, other presentations may occur. However, we know of no published images of this on the face.

A forty-three year old housewife presented with a seven day history of skin lesions appearing over the lower legs and face. Her only symptoms were of a recent upper respiratory tract infection; she was taking no medication. On examination there were symmetrical, tender, red-purple raised plaques from 1 cm to 7 cm diameter on the lower legs and red nodules in the peri-orbital areas of the face. No other abnormalities were detected. Chest X-ray and

serum angiotensin converting enzyme assay were normal. All lesions resolved spontaneously without scarring over the next six weeks with no specific treatment; the patient has remained well since.

Although erythema nodosum typically occurs on the shins, it can develop on the forearms and thighs; it has also been described on the face (Vesey CMR, Wilkinson DS. Erythema Nodosum. A Study of Seventy Cases. *Br J Dermatol.* 1959; 71: 139-55) The synchronised history of the facial and limb lesions strongly suggests association here. The facial lesions in this case appeared more nodular than those on the lower legs. Such variation in clinical appearance has been previously described (Ryan TJ. Cutaneous vasculitis. In: *Textbook of Dermatology* (Rook AJ, Wilkinson DS, Ebling FJG, eds), 6th edn. Oxford: Blackwell Scientific Publications, 1998; 2196-202.) Facial erythema nodosum is otherwise poorly documented. The incidence of facial erythema nodosum is not known.

We wish to highlight the association between these striking facial lesions and the classical appearance of erythema nodosum on the legs.

P-101 ELASTIN PEPTIDES IN THE SERUM WITH PATIENTS WITH SYSTEMIC SCLEROSIS

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The aim of this study was to investigate elastin turnover activity by quantification of circulating elastin-derived peptides (CEDP) in the sera of patients suffering from systemic sclerosis (SS). Using the ELISA method we tested the sera of 26 SS patients aged between 20-65 years and with disease duration from 2 to 23 years during the exacerbation and chronic phase. The sera of 24 healthy matched subjects served as controls. SS patients with severe angiostclerosis and rapidly progressing disease had significantly higher levels of CEDP during exacerbation compared to the control group ($p < 0.001$). In the chronic phase CEDP levels showed a trend towards decreasing. In two patients with highest CEDP levels (4 times more compared to the rest of the groups and 6 times more than the controls) a progressing disease and involvement of several internal organs in various degree was observed 12 months after first assay. It is proposed that elevated CEDP levels may be used as marker of the disease duration and have certain prognostic potential for SS patients.

P-102 CUTANEOUS VASCULITIS - AN UNUSUAL PRESENTATION

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Cutaneous vasculitis comprises of a wide spectrum of clinical syndromes and histopathological findings of vascular inflammation and changes in adjacent tissue. A case of necrotising vasculitis with widespread cutaneous ulceration of the face and limbs and auto-amputation of toes but with no

P-135 LYMPHOCYTOPENIA IN PATIENTS WITH LUPUS ERYTHEMATOSUS IS INVERSE CORRELATED WITH PRESENCE OF ANTINUCLEAR ANTIBODIES

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LE is an autoimmune disease characterised by a variety of immunological deviations. Autoantibody production against nuclear structures (ANA) and dsDNA is one of the best known pathologic features.

To a lesser extent cellular parameters have been investigated in patients with LE. Therefore we analysed in 73 patients with LE (38 women, 35 men; aged from 21 to 86 years, mean 46 ± 15.7 S.E.M.; discoid LE [DLE] n = 38, subacute cutaneous LE [SCLE] n = 22, systemic LE [SLE] n = 13) and 20 healthy control persons absolute lymphocyte counts, peripheral lymphocyte subsets (CD3+, CD3+CD4+, CD3+CD8+) by using FACS-analysis and a variety of autoantibodies in their sera by indirect immunofluorescence, ELISA and Western-blot.

We found lymphocytopenia (<1500 cells/ μ l) in 37% of patients with DLE, in 86% with SCLE and in 77% with SLE. The mean lymphocyte counts were [cells/ μ l]: DLE = 1745, SCLE = 1136, SLE = 1228, healthy controls = 1784. Statistic analyses (student's t-test) revealed significant differences between healthy controls and SCLE (p = 0.00021) and between healthy controls and SLE patients (p = 0.012). Among T-cell-subsets CD3+CD4+ cells were the cell population with the most prominent decrease of absolute cell counts (Mean T-helper-cell-counts [cells/ μ l]: DLE = 708, SCLE = 446, SLE = 469, health control = 676). Moreover we found an inverse correlation between lymphocyte cell-counts and titres of antinuclear antibodies (coefficient of correlation: -0.5).

Taken together our results show that not only patients with SLE, but also patients with cutaneous LE have diminished lymphocyte counts as compared to healthy controls, which are inverse correlated with the titres of ANA.

P-136 FIBRINOLYSIS ABNORMALITIES IN SYSTEMIC SCLEROSIS AND SYSTEMIC LUPUS ERYTHEMATOSUS

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We examined the 45 SLE (Systemic lupus erythematosus) and 45 PSS (Progressive systemic sclerosis) patients' serum fibrinolytic activity. The purpose of the present study was to assess the changes in the activity of plasma plasminogen and proactivator of plasminogen. A significant increase of plasminogen and plasminogen proactivator levels was observed in 47% of PSS and in 49% of SEL patients. 12% of PSS and 12% of SEL patients showed a significant decrease in plasminogen and proactivator of plasminogen activity. Plasminogen and proactivator of plasminogen are stored into endothelial cells and (or) linked at their surface. Endothelial injury resulted in significantly increased of plasminogen and its proactivator. Our findings are in agreement with the

hypothesis of a microvessel injury and release of endothelial related proteins in PSS and SEL patients.

P-137 IMMUNOFLUORESCENT DIAGNOSTICS OF CHILDREN DERMATOSIS

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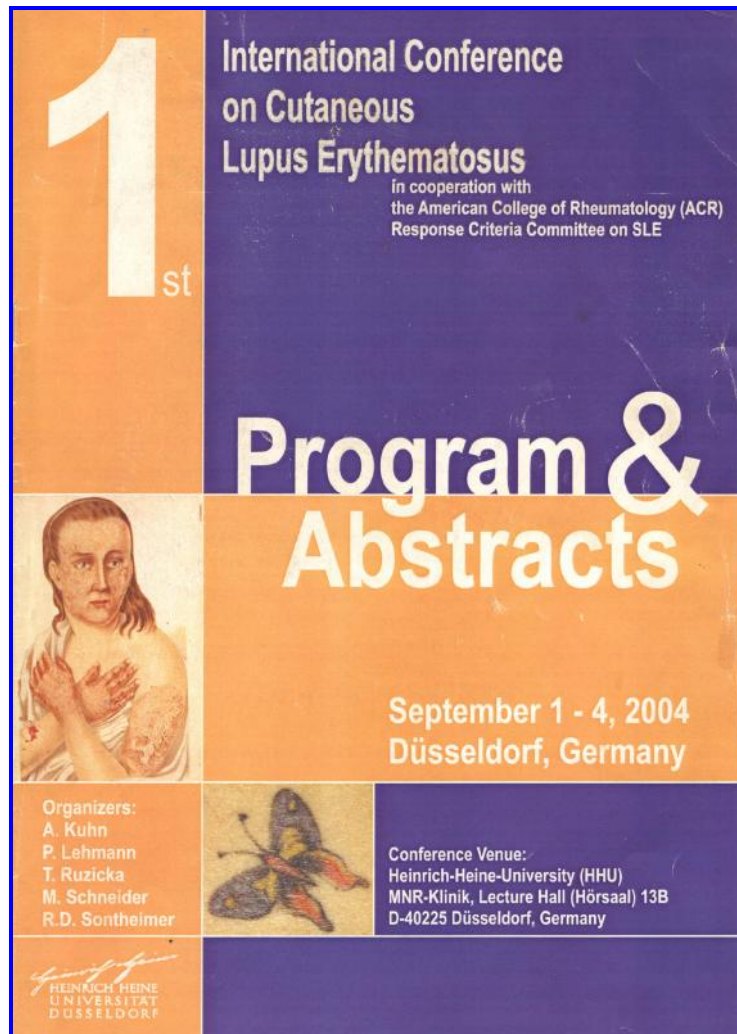
The group of patients consisted of 44 children with dermatosis, 27 of them 16 to 18 years old with alopecia areata of unknown etiology, 16 children 5 to 16 years old with bulbous dermatosis, and one child 12 years old with system affection of connective tissue where diagnosis was not more specified. For detection of antibodies marked animal antiserum was used.

based on direct immunofluorescence in all children with alopecia areata antibodies in hair follicle were detected. In the child with system affection of connective tissue antibodies detect in nuclei of epidermal cells, in area of basal membrane /BM/, upper part of corium and in walls of blood vessels and therefore we were assuming Sharp's syndrome, where diagnosis was complete after indirect immunofluorescent examination /ANA with its specifications/. From bulbous dermatosis, by finding of antibodies fixed in immunocytes beneath epidermis in area of junction zone /JZ/ and in blood vessels, in 2 patients we have diagnosed erythema exsudativum multiforme, by finding of antibodies of type IgA in the upper part of corium we have diagnosed in 13 patients dermatitis herpetiformis, by finding IgA in area of JZ with positive UIF, IgA in area of BM we have diagnosed in one child linear dermatosis.

P-138 ADAMANTIADIS-BEHÇET'S DISEASE: INTERLEUKIN-8 IS INCREASED IN SERUM OF PATIENTS WITH ACTIVE ORAL AND NEUROLOGICAL MANIFESTATIONS AND IS SECRETED BY SMALL VESSEL ENDOTHELIAL CELLS

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The serum levels of several cytokines were assessed in 94 patients with Adamantiades-Beheçet's disease (ABD), aged 11-68 years, during the active (n = 75) and the non-active stage (n = 19) of the disease. Seventy-five healthy individuals served as controls. In a second group of experiments, the involvement of dermal microvascular endothelial cells in interleukin (IL)-8 secretion was investigated. Immortalised human dermal microvascular endothelial cells (HMEC-1) were maintained in vitro with serum samples of 18 patients with ABD or with IL-1 α , tumour necrosis factor (TNF)- α and IL-8. HMEC-1 maintained in serum of healthy individuals served as control. Cytokine levels in serum samples and



(12)

SERUM LEVELS OF ELASTIN AND TYPE IV COLLAGEN DERIVED PEPTIDES IN PATIENTS WITH LUPUS ERYTHEMATOSUS: ASSOCIATION WITH DISEASE ACTIVITY AND PROGRESSION BY 2-YEAR FOLLOW-UP

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The aim of the study was to measure elasin and collagen turnover activity and to establish the connection between to changes in the serum levels of elastin and type IV collagen derived peptides and the clinical manifestations in 34 systemic lupus erythematosus and 10 subacute cutaneous lupus erythematosus patients. Using ELISA method, we tested the sera during the exacerbation and chronic phase. The sera of 25 healthy matched subjects served as controls.

