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T-111 BENEFICIAL EFFECT OF PLASMAPHERESIS IN SYSTEMIC SCLEROSIS WITH SEVERE RAYNAUD'S PHENOMENON

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Systemic sclerosis (SSc) is a generalized, multisystem disease in which characteristic vascular alterations are seen together with severe fibrosis of skin, synovium, and certain internal organs chiefly the intestinal tract, lung, heart, and kidney. The presence of Raynaud's phenomenon in 95% of SSc patients called attention to a possible vascular pathogenesis of the disease. Raynaud's phenomenon in SSc is probably due in part to intimal thickening of the digital arteries and is partially reversible with plasmapheresis.

Four SSc patients with severe Raynaud's phenomenon were treated with seven plasmapheresis exchanges, twice a week. Raynaud's phenomenon, especially the reaction upon cold provocation was disappeared and objective improvement in loosening of skin was occurred in all patients. Digital skin ulcers were healed with plasmapheresis in one patient. Pulmonary involvement, presented in three patients, was improved in one patient, based on computed tomography and CO diffusing capacity. The levels of circulating immune complexes and IgG were decreased, and hemoglobin levels were increased after plasmapheresis in all patients. In two patients, elevated levels of creatine kinase were normalized.

Our results suggest that plasmapheresis is beneficial in SSc patients, especially with severe Raynaud's phenomenon. It can also be considered to treat severe ischemia of the digits.

T-112 ANA AND ANTI-ADRENAL ANTIBODIES IN PATIENTS WITH SYSTEMIC CONNECTIVITE TISSUE DISORDERS

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To study a correlation of serum level between antinuclear autoantibodies (ANA) and autoantibodies to adrenocorticocytes (au:AC) we investigated sera from 76 patients with systemic connective tissue disorder (SCTD) as well as from 10 healthy donors in control group. Of 76 50 patients had systemic lupus erythematosus (SLE), 6 – scleroderma, 6 – Sjogren's syndrom (SS) and 14 – mixed connective tissue disease (mCTD).

We used indirect immunofluorescence assay ("The Binding Site Ltd", UK) with different substrates namely with monkey adrenal cortex sections for the detection of au:AC and with Hep2-cells for ANA, their titers and subtypes such as autoantibodies (au:) to Sm ag, SS-A/Ro and SS-B/La ag, hnRNP ag, whereas for detection of specific autoantibodies against double-stranded DNA (au:dsDNA) Critidia Luciliae cells were used.

Neither ANA nor au:AC were found in control group whereas all patients with SCTD had ANA.

Among SLE patients 75% had au:dsDNA, 20% had au:Sm ag, 20% had au:SS-A/Ro and au:SS-B/La ag, 4% had au:hnRNP ag, 26% had au: to cytoplasmic components SCLERODERMA patients had au: against Scl-70, centromeres and nucleolin in 50, 25 and 25%, respectively. All mCTD patients had au: UI-RNP ag but 15% had au:dsDNA. Both au:SS-A/Ro ag and au:SS-B/La ag were found in SS patients. Most SCTD patients (95%) had ANA titers being considered medium (range from 1: 160 to 1:640) and high (>1:640).

Au:AC were detected in 20% of SCLERODERMA, in 50% of SS, in 60% of mCTD and in 85% of SLE patients. High titers of ANA in SLE SS and mCTD patients correlated with au:AC to nuclei visualized in all three adrenal cortex zones and two zones (glomerular and steroid-producing) in SCLERODERMA cases, whereas medium titers of ANA were related to au:AC specific to steroid-producing zone only.

Patterns on Hep-2 and monkey adrenal cortex cells were the same in 90% of cases that points to similar tendency of autoimmune process. Data obtained show that adrenal cortex dysfunction in SCTD patients can partially be due to a failure caused by au: to nuclei and cytoplasm components. Anti-adrenal au: occurrence is the most frequent at SLE compared to other types of SCTD and as higher serum ANA titer as higher damage risk is at SCTD.

Sjoegren's Syndrome

T-113 PRIMITIF GOUGEROT-SJÖGREN SYNDROME (ABOUT 24 CASES)

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We report a retrospective study about 24 cases of the primary Gougerot-Sjögren syndrome (PGSS) collected at the rheuymatologic department in UHC Ibn Rochd in Casablanca during 4 years (1991.1994).

This group contains 23 women and 1 man, their mean age is 53.75 years old.

The predominant revealing symptoms were the extraglandular manifestations in 75% of the cases, particularly the articular ones (62.5%). In addition to the ocular manifestations (87.5%) and the salivary ones (83.3%), other manifestations have been noticed: articular, pulmonary, hepatic, cutaneomucous and neuropsychiatric manifestations.

The diangnosis of the PGSS is based particularly on the biopsy of the accessory salivary glands that was evocative of the Gougerot-Sjögren syndrome in 65.2% of the cases, the existence of a kiratoconjunctivites sicca in 84.6% of the cases and the electrophoresis shows a hypergammaglobulinemia in 52.9 of the cases.

An inflammatory anaemia has been noticed in 25% of the cases, a sedimentations rate > 50 mm at the first hour in 33.4% and a positive rheumatoid serology in 31.8% of the cases. The antinuclear antibodies were present in 2 cases among 18 cases that were a matter of search.

The treatment of 18 among our patients was based on the corticotherapy per Os (prednisone), the posology was the most often from 1/4 to 1/2 mg/kg/day.

The evolution was, on the whole, benign; we don't demonstrate any symptom that suggests a malignant transformation. However, the recession is insufficient, and doesn't permit a long term evolutive appreciation.

T-114 SJÖGREN'S SYNDROME: COMPARISON OF SUBJECTIVE AND OBJECTIVE FINDINGS

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Sjögren's Syndrome (SS) is an autoimmune systemic disorder characterized by decreased salivary and lachrymal functions. Dry mouth and eyes are the most common and prominent symptoms. The decline in salivary secretion leads to difficulties in speaking and swallowing, resulting in both an increased risk for infections in the mucous membranes of the mouth, and rapidly progressing caries. These symptoms are often very severe and consequently lead to a reduction in the quality of life. Due to the noticeable oral changes in these patients, the dentist may be the first to suspect SS. In this study, the clinical findings and subjective complaints of 31 SS patients and their salivary flow rates are compared in order to determine the presence of any correlation among these three parameters. The results revealed that in 27 out of 31 patients (87%), dry mouth was the most common complaint, and difficulty in speaking, chewing and swallowing were the other prominent subjective findings. On the other hand, the objective clinical examination had supported the subjective findings and showed a decrease of whole salivary flow rate below 0.1 ml/min in 51% of SS patients.

Spondylarthropathies

T-115 CASE REPORT: ANKYLOSING SPONDYLITIS COEXISTING WITH UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE

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Ankylosing Spondylitis rarely coexist with a connective tissue disorder and few cases are described in medical literature. We describe a case in which Ankylosing Spodylitis is associated with undifferentiated connective tissue disease. A female, 24 years old, Caucasian, came to our Service for the onset by far more than a year of fever, migrating arthralgia, muscle pain and weakness. She referred low back pain since 4 months, a malar butterfly rash during in the summer '98. Personal remote history was negative for other diseases. Physical examination showed a positivity for Schober test and tests for sacroiliac pain; no other pathological signs were found. Laboratory features were: microcitic anemia, increased ESR and acute phase proteins. Chest XR and abdominal ultrasonography were normal. LE test, Schirmer test, stool guaiac, bacterial cultures (feces, urine, troath and cervical sample), viral and bacterial serology, were negative. Auto-antibody findings were: ANA speckled and ENA at low title (anti-RNP, anti-SS-B, anti-Sm, anti-centromere) and anti-BPI. Negative findings were RA-test, LAC, anticardiolipin antibodies, anti-ds-DNA and the other auto non-organ specific antibodies. Circulating immune complexes were moderately increased. B27 allele was found positive with HLA tipizzation. Nailfold capillary abnormalities were not specific. AP XR view of sacroiliac joints showed grade II bilateral sacroiilitis, confirmed successively by computed tomography.

The patient was treated with corticosteroids, NSAIDs and sulfasalazine, with the resolution of the symptoms and the normalisation of inflammatory laboratory tests.

This case satisfies ESSG and New York criteria for AS with associated symptoms and laboratory tests suggestive for undifferentiated connective tissue disease.

It seemed useful to us to describe this case because in our clinical experience we did found, sometimes, AS coexisting with laboratory features and/or clinical signs of a connective tissue disorder. Since few case are described in medical