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AB0121 CYTOSTATICS EFFECT ON THE DEVELOPMENT OF MALIGNANT AND LYMPHATIC PROLIFERATIVE FORMATIONS DURING SJÖGREN'S DISEASE (SS)

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Background Treatment of rheumatological pts by alkylating cytostatics is undoubtedly of positive effect on the process of the disease but in a long term can stimulate the development of neoplasms and lymphatic proliferative diseases.

Objectives To detailise the degree of risk of above mentioned pathologies in SS pts who for a long time took low doses of chlorobutin or cyclophosphane – from 5 to 15 years.

Methods In the 1st group of 300 pts aged 15–75 years (46.4 (1.7) with disease duration of 5–25 years (8.7(1.9) 200 pts took 5 mg of Prednisolone daily, 1.8 g for a year, 0.9–1.8 g for the subsequent years combined with chlorobutin by 2 mg daily; 720 mg for a year and 360–720 mg for the next years. 100 pts had cyclophosphane by 200 mg twice a month i.v.; 4.8 g for a year and in the next 5 years yearly dosage was 2.4–4.8 g. 2nd group included 100 female pts who took only Prednisolone according the same scheme. 3d group of 100 female pts had no antirheumatic therapy. Age of pts in 2nd and 3d groups was 18–78 years (46.2(2.3) and disease duration of 4–26 years (9.2(2.1). Minimal frequency of blastomas and lymphatic proliferative diseases (1.6%) development was in the 1st group (pseudolymphoma in 1 pt, lymphoma – 2 cases, cancer of stomach in 1 and of skin in 1 pt). In the group treated only by GCS 7 cases of lymphoma were found and 1 case of stomach cancer (8%). Pts who had no antirheumatic therapy had 7 pseudolymphomas 9 lymphomas, 2 cases of Valdenstrem crioglobulinemia, 2 cases of stomach cancer and 1 pulmonary cancer (21%). Out of them pseudolymphomas and lymphomas were diagnosed in 18%.

Results

Conclusion Long term of taking of low doses of cytostatics (chlorobutin, cyclophosphane) combined with low doses of GCS does not promote the development of blastoma process in SS pts and decreases the frequency of transformation of the disease in malignant lymphomas.

FRI0212 CORRELATION BETWEEN HISTOPATHOLOGICAL FINDINGS, SERO-IMMUNOLOGICAL PROFILE AND MARKERS OF DISEASE ACTIVITY IN SICCA SYNDROME

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Background We evaluated sero-immunological parameters and markers of disease activity in ninety-two patients all female who underwent lip biopsy for histological assessment of sicca syndrome, according new preliminary criteria proposed for classification of Sjogren Syndrome (S. S).

Objectives The aim of our study was to compare the histopathological score index according to Chisolm and Mason classification with twenty-two parameters of disease activity.

Methods We have classified thirty-seven patients (group A) with focus score I-II with probable S. S (positivity of 3/6 item) and

fifty-five patients (group B) with focus score III-IV with defined S. S (positivity of 4/6 item). The mean age of two group was: group A 60 ± 11 years, group B 64 ± 9.8 years. The statistical comparison between the two group was performed with T-Student for 20 parameters and chi-square method for ANA and SSA/SSB positivity.

Results The comparison between the two groups show statistically significant differences for sero-immunological parameters such as gamma globulin (p < 0.01), SSA/SSB (p < 0.001). Moreover the group B shows higher levels of ESR (p < 0.005), fibrinogen (p < 0.02) and alpha1-antitrypsin (p < 0.005).

Conclusion This data show the important role of lip biopsy in the diagnosis of S:S and suggest the possible correlation between high histopathologic focus score and immunological markers of disease activity.

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FRI0213 RAYNAUD'S PHENOMENON IN PRIMARY SJÖGREN'S SYNDROME. PREVALENCE AND CLINICAL CHARACTERISTICS IN A SERIES OF 320 PATIENTS

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Background

Objectives To determine the prevalence of Raynaud's phenomenon (RP) in a large series of patients with primary Sjögren's syndrome (SS) and to identify the clinical and immunological features related to its presence.

Methods In a cross-sectional study, we investigated 320 consecutive patients with primary SS (294 women and 26 men; mean age at onset 60 years; range 16–87 years). All patients fulfilled 4 or more of the diagnostic criteria for SS proposed by the European Community Study Group in 1993. Diagnosis of RP was done in SS patients with intermittent attacks of digital pallor and/or cyanosis, in the absence of any other associated disease or anatomical abnormalities.

Results Raynaud's phenomenon (RP) was present in 40 (13%) patients. All were women, with a mean age of 57 years (range, 18 to 78 years). RP precedes onset of sicca symptomatology in 18 (45%) patients. The main triggering factor was cold exposure, which induced RP in all patients, emotional stress in 12 and 2 patients had labour predisposition. Fifteen (38%) patients required pharmacological treatment with calcium channel blockers (12 patients) or angiotensin-converting enzyme inhibitors (2 patients) in the cold seasons. One patient required treatment with intravenous prostacyclin for ischaemic complications. When compared with SS patients without RP, those with RP showed a higher prevalence of articular involvement (50% vs 31%, p = 0.031), cutaneous vasculitis (30% vs 11%, p = 0.003), antinuclear antibodies (95% vs 65%, p < 0.001), anti-Ro/SS-A (59% vs 31%, p < 0.001) and anti-La/SS-B antibodies (44% vs 20%, p = 0.003).

Conclusion We found RP in a 13% of patients with primary SS. Interestingly, RP was the first autoimmune symptomatology in almost 50% of these patients. These patients constituted a subset of SS with a higher frequency of some extraglandular features and positive immunological markers. In contrast, RP have a

milder course in primary SS, with no vascular complications and needing pharmacological treatment in only 40% of patients. The clinical course of RP seems to be milder in patients with primary SS than in other systemic autoimmune diseases such as systemic sclerosis.

FRI0214 INCIDENCE OF PANCREATIC AUTOANTIBODIES IN PRIMARY SJÖGREN SYNDROME

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Background Primary Sjögren (SjS) syndrome is one of the prototypes of autoimmune diseases.

Objectives We aimed to work out the incidence of pancreatic autoantibodies in this disease.

Methods Seventy-six patients (F/M: 71/5, mean age: 49 ± 13 years) diagnosed as primary SjS syndrome according to the San Diego criteria between February 1997 and December 2000, have been included in the study. Twenty two, age matched (52 ± 10 years) female patients with secondary SjS due to rheumatoid arthritis were defined as the control group. Healthy control group included 22 age matched (F/M: 21/1) patients. Immunosuppressive drug usage, liver or renal failure were within the exclusion criteria. Serum samples for anti-insulin (AIA), islet (ICA) and anti-insulin receptor (IRA) antibodies were obtained from all patients after an 18 h fasting.

Results The incidence of anti-insulin antibody was found as 17%, while it was 7% and 17% for islet antibodies and anti-receptor antibodies, respectively (Table 1). These values were not statistically significant when compared with secondary SjS and healthy control groups ($p > 0.05$).

Abstract FRI0214 Table 1 Incidence of pancreatic autoantibodies

	Primary SjS%	Secondary SjS%	Healthy control%	p
AIA	17 (n = 13)	22 (n = 5)	18 (n = 4)	$p > 0.05$
IRA	17 (n = 13)	18 (n = 4)	18 (n = 4)	$p > 0.05$
ICA	7 (n = 5)	13 (n = 3)	13 (n = 3)	$p > 0.05$

Conclusion Incidence of pancreatic auto-antibodies in primary SjS with observed hyperactivity of B cells, was not found statistically different when compared with patient and healthy control groups.

FRI0215 SJÖGREN'S SYNDROME MANIFESTATIONS IN PRIMARY BILIARY CIRRHOSIS

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Background

Objectives Study the Sjögren's Syndrome (SS) manifestations in a sample of patients with Primary Biliary Cirrhosis.

Methods From October 1999 to May 2000 81 patients from the hepatology outpatient department were studied. We used the european criteria for the diagnostic of SS. Extraglandular clinical

manifestations, analitical and immunological study was evaluated using the unity protocol.

Results 90% were women. The mean age was 61 ± 11 y. Clinical evolution was of 6 years. Ocular dryness was present in 60 (74%) and oral dryness in 60(76%). Schirmer and/or bengal's rose were positive in 21(26%). The most common extraglandular manifestations were: Fibromialgy in 42(51%), Raynaud's phenomenon in 21(26%), CREST's syndrome in 13(16%), thyroopathy in 21(26%), antythyroidal antibodies were positive in 19(24%)(Ten of them had hipothyroidism and two of them goitres), arthritis in 7(9%)(Two of them were erosive), inflammatory osteoarthritis in 5(6%), vasculitis in 3(4%), ANA >80 in 47 (58%), Rheumatoid factor in 27(33%), SS-A/Ro antibodies in 14 (17%) and SS-B/La in 8(10%). 9 CREST's patients were positive for anticentromere antibodies. 41(50%) had 4 or more SS European criteria. A minor salival gland biopsy was realised in 53 patients. 39 of them were positive (73%). 41 of the biopsed patients had 3 or more criteria. 31 of this 41 patients has a positive biopsy (75%). The other 12 patients had 2 or less criteria, just 5 of them had a positive biopsy (41%).

Conclusion There is a high prevalence of SS (50%) in patients with CBP. Keratoconjunctivitis sicca's prevalence is 66%. Subjective involvement of oral and ocular dryness are 74% and 76% respectively. Fibromialgy is the rheumatological clinic more frequent, appearing in more than 50% of the patients.

FRI0216 ANTIBODIES TO RECOMBINANT 60-KD RO, 52-KD RO AND 48-KD LA PROTEINS IN PRIMARY SJÖGREN'S SYNDROME. USEFULNESS OF COMBINATION OF ANALYTICAL METHODS FOR DETECTING ANTI-RO AND ANTI LA ANTIBODIES

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Background

Objectives To determine the reactivity and specificity of sera from primary Sjögren's syndrome (SS) patients to recombinant 60-Kd Ro, 52-Kd Ro and 48-Kd La proteins, and to correlate the results with clinical and laboratory findings.

Methods Sera from 46 patients with primary SS diagnosed according to modified 1997 European criteria, were classified in 2 groups based on the presence of anti-Ro and anti-La detected by commercial enzyme linked immunosorbent assay (ELISA). Group A (n = 32) had anti-Ro positive sera, and group B (n = 14) had anti-Ro and anti-La negative sera, but positive antinuclear antibodies and/or rheumatoid factors. Autoantibody reactivity and specificity to recombinant 60-Kd Ro, 52-Kd Ro and 48-Kd La were analysed by ELISA and Western blot.

Results Forty-two out of 46 primary SS sera (91.30%), had reactivity to either natural or recombinant antigens or both. Anti-60-Kd Ro was found in 27 (58.69%) of all sera, of these, 22 (68.75%) sera were of group A, and 5 sera (35.71%) of group B. Anti-52-Kd Ro was highly prevalent since 38 out of 46 sera (62.60%) were positive. Among group A, 31 out of 32 (96.87%) sera were positive for 52-Kd Ro, and 7 out of 14 (50%) in group B. Anti-recombinant 48-Kd La was also highly prevalent, 38 (82.60%) of all sera were positive. In group A, 31 (96.87%) of sera were positive, while 5 (35.71%) of sera of group B, were positive. Ten (71.42%) of 14 patients of group B had antibodies for at least 1 of 3 recombinant antigens: 52-Kd Ro, 60-Kd Ro or