the last follow-up visit was unrelated to age or the duration of SLE, but it correlated with the initial SLEDAI score (Spearman $p < 0.05$); the higher the initial SLEDAI score, the higher the SLICC score at the end of follow-up. The SLICC score was significantly lower (Mann Whitney $p < 0.05$) in patients with anti-ENA antibodies, particularly those who had anti-Ro/SS-A (mean 1.11 vs 1.79) or anti-La/SS-B antibodies (mean 0.52 vs 1.7). SLICC score showed no relation with anticardiolipin antibodies. The 8 patients who died in the course of the study had significantly higher SLICC scores (mean 2.87 vs 1.44).

Conclusion High SLEDAI score at the onset of SLE and the absence of anti-ENA antibodies, particularly anti-Ro/SS-A or anti-La/SS-B antibodies were associated with significantly higher SLICC scores, suggesting that these factors may indicate poor prognosis in SLE.

FRI0123 CUMULATIVE TISSUE DAMAGE IN PATIENTS WITH JUVENILE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS

I Monteagudo, FJ López-Longo, CM González, AI Turrión, CA Montilla, M Moreno-Zazo, N Caro, R Del Castillo, C González-Montagut, L Cebrián, E Carmona, L Carreño. *Rheumatology, Hospital General Universitario Gregorio Marañón, Madrid, Spain*

10.1136/annrheumdis-2001.158

Background In earlier studies, we found that the juvenile onset of systemic lupus erythematosus (SLE) did not significantly modify the clinical expression of the disease.¹ Although juvenile-onset SLE may have a more sombre prognosis than adult-onset SLE, there have been few comparative studies of these types of SLE.

Objectives To compare the Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index (SLICC/ACR) of a cohort of patients with juvenile-onset SLE and patients with adult-onset SLE.

Methods We calculated the SLICC/ACR scores of 236 patients diagnosed as SLE between 1988 and 2000. The SLE disease activity index (SLEDAI) of the first hospital study was calculated retrospectively.

Results In 61 patients (25.8%), SLE had begun before the age of 19 years. This group included 52 women (85.2%). The mean age at onset of SLE was 13.4 years (SD 3.2) and mean duration of the disease was 9.9 years (SD 6.5). The comparison of patients with juvenile-onset SLE and a group of 175 patients with adult-onset SLE did not reveal significant differences in sex, duration of the disease, and SLEDAI score at the onset of the disease (mean 16.31; SD 8.26 vs 14.08; SD 8.08). There were no differences in the SLICC/ACR scores obtained at the last follow-up visit (mean 1.6; SD 1.7 vs 1.45; SD 1.94) ($p > 0.05$), although kidney disease (proteinuria over 3.5 g/day, treatment by dialysis or transplantation) (24 of 25; 39.3% vs 42 of 211; 24%) ($p: 0.03$; OR: 2; 95% CI: 1.1–3.8) and avascular necrosis (4 of 25; 6.6% vs 2 of 211; 1.1%) ($p: 0.04$; OR: 6; 95% CI: 1.08–3.4) were more frequent in patients with juvenile-onset disease. Both groups had a similar mortality (1 of 61; 1.6% vs 7 of 175; 4%) ($p > 0.05$).

Conclusion Although kidney disease and avascular necrosis were more frequent in patients with juvenile-onset SLE, the SLICC tissue damage score was similar in patients with juvenile-onset SLE and adult-onset SLE.

REFERENCE

- 1 Lupus 1999;8:287–92

FRI0124 DOES MIXED CONNECTIVE TISSUE DISEASE HAVE A LESS FAVOURABLE PROGNOSIS THAN SYSTEMIC LUPUS ERYTHEMATOSUS?

FJ López-Longo, CM González, AI Turrión, M Moreno-Zazo, R Del Castillo, N Caro, L Cebrián, C González-Montagut, E Carmona, S Gómez, I Monteagudo, L Carreño. *Rheumatology, Hospital General Universitario Gregorio Marañón, Madrid, Spain*

10.1136/annrheumdis-2001.159

Background Mixed connective tissue disease (MCTD) has been considered to have a good prognosis, although some patients present severe clinical manifestations like pulmonary hypertension.

Objectives To compare the cumulative tissue damage occurring in patients with MCTD and systemic lupus erythematosus (SLE).

Methods We compared the cumulative tissue damage index scores of patients with MCTD and SLE studied between 1988 and 2000. The consequences of MCTD and SLE and their treatment were evaluated using the Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index (SLICC/ACR) obtained at the last follow-up visit; scleroderma lesions were not taken into account in patients with MCTD.

Results We studied 37 patients with MCTD and 236 patients with SLE. There was no difference in the relative proportion of women in with MCTD and SLE (32 of 37; 86.5% vs 211 of 236; 89.4%). Mean age (40 years; SD 17.6 vs 39; SD 15.8), mean age at onset of disease (29 years; SD 17.6 vs 29; SD 14.7), and mean duration of disease (11 years; SD 7.9 vs 9.7; SD 6.9) were similar in both groups. There were no differences in mortality (1 of 37; 2.9% vs 8 of 236; 3.4%) or SLICC score (1.43; SD 1.11 vs 1.49; SD 1.88). Patients with MCTD had more musculoskeletal sequelae (20 of 37; 54.1% vs 36 of 236; 15.3%) (OR: 6.5) ($p < 0.005$) and pulmonary fibrosis (5 of 37; 13.5% vs 9 of 236; 3.8%) (OR: 3.9) ($p < 0.05$) than patients with SLE. Patients with SLE had more kidney disease as defined by the SLICC (66 of 236; 28% vs 3 of 37; 8.1%) ($p < 0.01$).

Conclusion In spite of the differences observed, the cumulative tissue damage and mortality were similar in MCTD and SLE, after excluding from consideration scleroderma lesions in patients with MCTD.

FRI0125 CEREBRAL SINGLE-PHOTON EMISSION COMPUTED TOMOGRAPHY (SPECT) IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

¹CM González, ¹FJ López-Longo, ²A Ortega, ²M Suárez, ¹N Caro, ¹R Del Castillo, ¹C González-Montagut, ¹L Cebrián, ²MI Almoguera, ¹I Monteagudo, ¹L Carreño. ¹Rheumatology; ²Nuclear Medicine, Hospital General Universitario Gregorio Marañón, Madrid, Spain

10.1136/annrheumdis-2001.160

Background Hypoperfusion observed in cerebral SPECT studies of patients with SLE has been found to be related with clinical activity in 10 patients with SLE and no neuropsychiatric manifestations.¹

Objectives To study the clinical significance of cerebral SPECT in unselected patients with SLE.

Methods We performed cerebral SPECT in 57 women with SLE seen consecutively in our clinic. In the same week that SPECT was performed, SLEDAI, ESR, the concentrations of anti-DNA antibody and the C3 and C4 complement fractions, and the SLICC/ACR tissue damage index were determined.

Results The mean age of patients was 37 years (SD 13.1) and the mean duration of the disease was 9.7 years (SD 6.3). Cerebral SPECT revealed hypoperfusion in 49 patients (86%), which was diffuse in 33 (57.9%) and patchy in 16 (28.1%). The most frequent sites of hypoperfusion were the frontal, temporal, and parietal cortical areas. Normal perfusion or mild hypoperfusion (unappreciable) was observed in 22 patients (38.6%) and moderate or intense hypoperfusion (appreciable) was observed in 27 (47.4%). Appreciable hypoperfusion was unrelated to age, duration of SLE, or the concentrations of anti-DNA antibodies and the C3 and C4 fractions. Patients with appreciable cerebral hypoperfusion had a more active clinical disease (mean SLEDAI 13.92; SD 8.44 vs 4.56; SD 4.15) (Mann-Whitney $p < 0.005$), more cumulative tissue damage (mean SLICC 2.66; SD 2.84 vs 1.03; SD 1.51) ($p = 0.035$), and higher ESR (mean 28.7; SD 22.5 vs 17.7; SD 13.3) ($p = 0.023$) than patients with a normal SPECT. Cerebral hypoperfusion was related with neuropsychiatric manifestations present at the time of study (17 of 27; 63% vs 3 of 30; 10%) (OR: 15.3) or cumulative (19 of 27; 70.4% vs 8 of 30; 26.7%) (OR: 6.5), whether mild (OR: 5.5) or severe (OR: 8.2).

Conclusion Cerebral hypoperfusion detected by SPECT was related with concomitant and previous neuropsychiatric manifestations, the clinical activity of SLE, and cumulative tissue damage.

REFERENCE

1 Rev Esp Reumatol. 2000;27:202

FRI0126 CORTICOSTEROID THERAPY-INDUCED INJURY IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

I Koni, M Kawano, S Kitajima, H Mutoh, K Katano, R Inoue, H Mabuchi. *Second Department of Internal Medicine, School of Medicine, Kanazawa University, Kanazawa, Japan*

10.1136/annrheumdis-2001.161

Background Corticosteroid therapy is dramatically effective in patients with systemic lupus erythematosus (SLE). Long-term steroid therapy, however, is associated with numerous well-known adverse effects such as vascular disease, osteoporosis and avascular bone necrosis.

Objectives To evaluate the association between steroid therapy and its side effects in patients with SLE.

Methods The medical records of 40 patients with SLE, who have been receiving long term follow up in our clinic from 1970 to 2000, were checked with regard to events possibly related to steroid therapy. The risk of complications associated with the cumulative prednisone dose (CPD) and maximal prednisone dose (MPD: high >60 mg/day, 60 mg/day $\geq$ moderate >30 mg/day, low ≤ 30 mg/day) was estimated using multivariate logistic analysis, adjusted for age and sex. In CPD 10 mg of prednisone daily for 10 years (36.5 g) was assessed as a reference unit.

Results Five patients had coronary artery disease and 2 of them died. Hypertension was found in 16 patients, stroke in 5, diabetes mellitus in 13, osteoporosis in 20, avascular bone necrosis in 7 and herpes zoster in 11. There were statistically significant relationships between CPD and osteoporosis (RR 8.5, $p < 0.01$), and coronary artery disease (RR 3.7, $p < 0.05$). The development of stroke also has a tendency related to CPD, whereas herpes zoster and avascular bone necrosis were associated with MPD (RR 5.1, $p < 0.05$ and RR 5.2, $p = 0.1$). Moreover

hypertension and diabetes mellitus were more related to age and body mass index, respectively.

Conclusion In SLE patients long-term corticosteroid therapy frequently induced some adverse effects, which could be identified as two distinct types: one is cumulative dose-related injury like osteoporosis and another is maximal dose-related injury like herpes zoster. Steroid-sparing strategy could be needed in order to minimise cumulative and high-dose corticosteroid exposure.

FRI0127 ATHEROSCLEROSIS AND ARTERIAL DISEASE IN SYSTEMIC LUPUS ERYTHEMATOSUS

¹J Frostegard, ¹E Svenungsson, ²K Jensen-Ustad, ³M Heimburger, ¹L Klareskog, ⁴A Hamsten, ⁵U De Faire, ⁶J Witztum. ¹Department of Rheumatology, ²Department of Clinical Physiology, ³Department of Rheumatology, Huddinge University Hospital, Stockholm, Sweden; ⁴Department of Medicine, ⁵Division of Cardiovascular-Epidemiology, Karolinska Hospital; ⁶Department of Medicine, University of California San Diego, San Diego, USA

10.1136/annrheumdis-2001.162

Background Systemic lupus erythematosus (SLE) patients have increased morbidity and mortality in cardiovascular diseases. Whether this is caused by aggravated atherosclerosis has not been totally clarified. Since atherosclerosis recently has been shown to be an inflammatory disease it is important to study the inflammatory nature of SLE in this context. Furthermore the role of phospholipid antibodies and enhanced thrombogenesis as well as the importance of other cardiovascular risk factors like LDL oxidation need to be studied in SLE patients.

Objectives To determine the cause of the enhanced risk of cardiovascular disease present in systemic lupus erythematosus.

Methods Twenty-six women (mean age 52 yrs) with SLE and a history of arterial disease (SLE cases) were compared with 26 age matched women with SLE but without manifest arterial disease (SLE controls) and 26 age matched population control women (population controls). Common carotid intima-media thickness (IMT) was measured by B-mode ultrasound as a surrogate measure of atherosclerosis. All were subject to clinical and laboratory evaluations with respect to known cardiovascular risk factors. A monoclonal antibody to oxLDL, EO6, was used to determine oxidation epitopes in LDL.

Results SLE cases had increased IMT as compared to population controls (IMT $0.66 \text{ mm} \pm 0.15$ vs 0.59 ± 0.12 , $p = 0.037$) while the IMT of SLE controls ($0.60 \text{ mm} \pm 0.14$) did not differ from that of population controls. Raised plasma concentrations of oxLDL- ($p = 0.033$); dyslipidemia with raised triglycerides ($p < 0.001$) and lipoprotein (a) ($p = 0.002$), and decreased HDL-cholesterol concentrations ($p = 0.03$); raised alpha-1-antitrypsin ($p = 0.002$), antibodies to oxLDL ($p < 0.001$), lupus anticoagulant ($p = 0.007$) and homocysteine-levels ($p = 0.03$) discriminated between SLE cases and SLE controls. SLE cases had been subject to significantly higher cumulative prednisone dose than SLE controls ($p = 0.04$). Disease duration, smoking, blood pressure, BMI or diabetes mellitus did not differ significantly between the groups.

Conclusion We found that arterial disease in SLE is linked to aggravated atherosclerosis. However, unlike the situation in diabetes and hypertension, aggravated atherosclerosis was not present in SLE patients without signs of arterial disease. Dyslipidemia, systemic inflammation and raised concentrations of OxLDL, anti OxLDL antibodies, lupus anticoagulant and homocysteine distinguished women with SLE and arterial disease from both SLE controls and population controls.