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## POS0749 ASSOCIATION BETWEEN IMMUNE-SEROLOGICAL PROFILE AND PULMONARY MANIFESTATIONS IN COLOMBIAN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS (LES)

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**Background:** Pulmonary involvement is common in Systemic Lupus Erythematosus (SLE) patients with varying degrees of parenchymal, vascular, and pleural compromise. In GLADEL, pulmonary involvement was reported in 28.4% of the cohort, but its occurrence ranges between 30-90% due to diversity in populations and the methods used to define it.

**Objectives:** To describe the immune-serological profile of a Colombian cohort of SLE patients and to establish its association with pulmonary manifestations.

**Methods:** Retrospective analysis of observational data from the follow-up of a cohort of adult patients with SLE. We included 559 patients that fulfilled the SLICC 2012 classification criteria with at least 6 months of disease history and being treated in a rheumatology specialized medical center between 2015 and 2018. The immuno-serological profile was characterized, and pulmonary involvement was monitored for 1 year. Diagnosis of pulmonary involvement was performed with the rheumatologist report in the clinical chart. Prevalence of pulmonary manifestations and immune-serological profile was determined, and logistic regression was performed afterward adjusted by age, sex, and level of education to establish the association between pulmonary manifestations and a positive auto-antibodies profile.

**Results:** The median age of the cohort was 45 years, 96.5% were female. Pulmonary involvement was documented in 113 patients (20.5%) at the beginning of the study. Their frequency was: pleuritis (14.3%), lupus pneumonitis (3.6%), pulmonary hypertension (3.2%), interstitial lung disease (ILD) (2.3%), pulmonary embolism (2.3%), lung fibrosis (2.14%), alveolar hemorrhage (1.4%), shrinking lung (0.2%). At 1 year of follow up. there were no statistically significant differences in the frequency of pulmonary manifestations. As for the immune-serological profile, there were positive ANA in 92%, anti-dsDNA in 53.1%, anti-B2GP IgM 15.2%, anti- B2GP IgG in 17.2%, and ENA in 97.2%; as for the ENA 41.7% had positive anti-RNP, 40.2% anti-Ro, 36.4% anti-SM and 16.5% anti-La. Low complement levels was characterized as follows: C3 53.1% and C4 29.2%. In the logistic regression adjusted by age, sex and level of education, there was an association between anti-SM and pulmonary manifestations with an adjusted OR of 1.85; 95% CI 1.13-3.01.

**Conclusion:** An association between anti-SM positivity and pleuro-pulmonary manifestations was found. In other cohorts with a greater size, anti-La and anti-RNP have been associated with pulmonary involvement (OR 2.51; 95% CI 1.39-4.57 and OR 1.32; 95% CI 1-1.75 respectively). Anti-RNP positivity has been associated in particular with ILD, pulmonary hypertension and shrinking lung. Although these manifestations prevalence was similar in our cohort, an association with this antibody was not found (OR 1.01, 95% CI: 0.2-4.9). This could be explained by the smaller sample size. As for anti-La positivity, its prevalence in our cohort was less than what was found in the GLADEL cohort (16.5% vs 24.3% respectively). It is possible that this could explain the poor association between anti-La positivity with the presence of pulmonary manifestations in our study compared with those of the GLADEL cohort. There are data that indicates that anti-SM and anti-RNP simultaneous positivity is related mainly to pleuritis OR 1.98 (95% CI: 1.31-3); and this kind of involvement was found to be more frequent in our study. Our results suggest an association between positive anti-SM and pulmonary manifestations in Colombian patients with SLE, pleuritis in particular.

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## POS0750 THE CHARACTERISTICS OF SYSTEMIC LUPUS ERYTHEMATOSUS WHO HAVE CONSTITUTIONAL SYMPTOMS AND LYMPHADENOPATHY

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**Background:** Systemic lupus erythematosus (SLE) is the prototype of systemic autoimmune diseases. It may affect any organ system and may present with a variety of clinical symptoms. Lymphadenopathy (LAP) is a nonspecific finding of SLE and often associated to the active disease state. LAP associated SLE pose a significant diagnostic challenge especially in the presence of constitutional symptoms.

**Objectives:** In this retrospective cohort study, we evaluated the clinical and imaging characteristics of SLE patients who presented with LAP and constitutional symptoms.

**Methods:** SLE Patients with lymphadenopathy who had constitutional symptoms were reviewed retrospectively. The characteristics of the patients such as age, sex, disease duration, clinical features, and laboratory tests including autoantibodies against ANA, dsDNA and ENA were recorded. Imaging findings such as positron emission tomography–computed tomography (PET/CT), thoracal or abdominal CT or diagnostic lymph node biopsy were identified.

**Results:** There were 28 patients in this study. The mean age at the time of LAP was 38.9±15.2 years, 22 (78.6%) were female. All patients were fulfilling SLICC 2012 classification criteria for SLE. LAP was the presenting feature of SLE in 15 (53.6%) patients. The most common clinical features associated with LAP were arthritis (42.8%), fever (35.7%), acute lupus rash (32.1%) and serositis (28.5%). There were 11 (39.2%) who had significant proteinuria and 4 (14.3%) had class III or IV renal histology. Anti-phospholipid antibodies were present in nearly one-third of the patients and 12 of them had venous or arterial thrombosis or pregnancy co-morbidity. Cervical LAP was present in 57.1%, axillary LAP in 82.1% and inguinal in 36%. On CT, the prevalence of thoracal and abdominal LAP and hepatomegaly and splenomegaly were 61.5%, 37,5%, 40% and 16% respectively. The pericardial and pleural effusion and intra-abdominal free fluid were 19.2%, 30.8% and 10.2% respectively. There were 19 (67.8%) patients who underwent PET/CT imaging and all were reported as reactive LAP. In terms of SUV max 7 (36.8%) of them had a threshold ≥2.5. 17 (60.7%) patients required lymph node biopsy; 2 did not include lymphoid tissue, 4 (30.7%) with atypical lymphoid hyperplasia suggestive of viral infection and remaining were reactive lymphoid hyperplasia. Autoantibody testing showed that anti-dsDNA was positive in 66.7% and the most common anti-ENAs were anti-nucleosome (35.7%) and anti-Sm-RNP (35.7%) followed by anti-SSA (25%), anti-ribosomal P (17.9%) and anti-histone (14%). The clinical and laboratory features of the clinical and laboratory features of the group is summarized in Table 1.

**Table 1. Data at the time of LAP**

| | |
|---|---|
| Age, years | 38.9±15.2 |
| Disease duration, months | 3 (2- 300) |
| SLE-DAI, median (min-max) | 9 (2-30) |
| Sex, female n(%) | 22 (78.6) |
| **Clinical features** | **n (%)** |
| Acute cutaneous | 9 (32.1) |
| Arthritis | 12 (42.8) |
| Fever | 10 (35.7) |
| Other constitutional symptoms | 18 (64.2) |
| Serositis | 8 (28.5) |
| Renal involvement | 11 (39.2) |
| Neurologic involvement | 1 (3.6) |
| Leucopenia | 7 (25.0) |
| Trombositopenia | 5 (17.8) |
| Hemolitic anemia | 7 (25.0) |
| ANA (+)>1/320 | 28 (100) |
| Anti-dsDNA (+) | 18 (66.7) |
| Anti-Sm(+) | 5 (17.9) |
| Anti-phospholipid antibody (+) | 9 (32.1) |
| Low complement | 16 (88.5) |

**Conclusion:** We showed that SLE patients who had constitutional findings and LAP could be the presenting feature and may confer a significant diagnostic challenge in which the patients might require invasive procedures and advanced imaging modalities. PET/CT revealed reactive changes in lymph nodes in all patients but lymph node biopsy revealed atypical changes in some patients suggestive of viral infections. On the other hand, LAP patients with constitutional symptoms represents a severe phonotype of the SLE as there were significant renal and thrombotic disease.

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## POS0751 COMORBIDITY AND LONG-TERM OUTCOME IN PATIENTS WITH CONGENITAL HEART BLOCK: PRELIMINARY DATA OF THE ITALIAN REGISTRY ON THE IMMUNE-MEDIATED CONGENITAL HEART BLOCK

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**Background:** Congenital heart block (CHB) is due to placental transfer of maternal anti-Ro/SSA autoantibodies to the fetus. The prevalence of CHB has been estimated as 1-2% in anti-Ro/SSA women while the recurrence rate is 16-19% (1). This condition is associated with a high rate of fetal/neonatal mortality and most of the cases requires pacemaker (PM) pacing. Given the rarity of CHB, limited data are available regarding the long-term follow-up of the offspring other than the cardiovascular complications.

**Objectives:** The results of the Italian Registry of the autoimmune congenital heart block were recently described (2). A peculiarity of this cohort was that most of the mothers had an established diagnosis of systemic autoimmune disease at CHB detection, in contrast with other registries where CHB was mostly incidentally detected in healthy women. Here we report an update, with the preliminary data regarding the long-term outcome of patients with CHB, their unaffected siblings and health controls born from mothers positive for Ro/SSA.

**Methods:** Data regarding demography, treatment, maternal, neonatal outcome, and follow-up were collected through an online electronic datasheet. A dedicated questionnaire was created with the aim to investigate general health, cardiovascular follow-up, and frequency of autoimmune diseases.

**Results:** One-hundred and five cases of CHB in 99 patients were included from 1969 to December 2020. CHB was mostly detected in utero (97 cases, 92.3%) with 8 neonatal cases. Third degree CHB occurred in 71 cases (67.6%). Child mortality was observed in 29 (27.6%) cases: 20 in utero, 7 during neonatal period and 2 during childhood. Overall, a PM was implanted in 54 out of the 85 live births (63.5%). Then, our cohort was divided into 2 subgroups: pregnancy that occurred before (N=61) and after 2010 (N=44) with the aim to evaluate possible differences among the subgroups. Whereas mortality, PM, CHB degree were similar, CHB more frequently occurred in the last 10 years among Ro/SSA asymptomatic carriers than in the group of pregnancies before 2010 (53.6% vs 32.8%, p=0.038). Questionnaires from 14 surviving CHB cases, 8 unaffected siblings 12 controls born from mothers Ro/SSA positive were collected. Among CHB cases, 6 were males and 8 females, median age 12 years (range 6-28). All presented a third degree CHB, 10 required a neonatal PM pacing and one had an implantable ECG recorder. PM was substituted at least once in 9 patients, the oldest patient had to change it four times. No dilated cardiomyopathy occurred and most of the patients maintain an annual follow-up. Two cases of autoimmune diseases were registered among CHB cases, one idiopathic juvenile arthritis and one Cogan's vasculitis, both born from mothers with Sjogren Syndrome. Four cases of neurodevelopmental disorders occurred: three cases of learning disabilities (one in each group) and one case of speech disorder in the sibling group. In addition, a CHB case presented a stress disorder linked to frequent hospitalizations.

**Conclusion:** This registry is an ongoing project aiming at collecting all Italian CHB. Moreover, here we reported the preliminary data concerning the evaluation of long-term follow-up of CHB patients. Our data, even if need to be confirmed in larger cohort, seems reassuring: no differences were reported comparing CHB patients with unaffected siblings or controls.

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## POS0752 INTERSTITIAL LUNG INVOLVEMENT IN SYSTEMIC LUPUS ERYTHEMATOSUS: RELEVANT CLINICAL CHARACTERISTICS. OUR EXPERIENCE IN A THIRD LEVEL HOSPITAL

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**Background:** Lung involvement[1] in Systemic Lupus Erythematosus (SLE) is heterogeneous, with the pleura being more frequently affected. Associated diffuse interstitial lung disease (ILD) is rare (3-13%) with difficult diagnosis and therapeutic management.

**Objectives:** Assess the main clinical-epidemiological characteristics of patients diagnosed with SLE with ILD in a third level hospital and analyze the possible relationship between them.

**Methods:** Descriptive observational study of patients diagnosed with SLE (SLICC criteria) with ILD in our hospital between 1973 and 2020. The following clinical-epidemiological characteristics were evaluated such as sex, mean age at diagnosis, presence of cardiovascular risk factors (CVRF), smoking, baseline cardiac and respiratory comorbidities, laboratory markers (autoimmunity, CRP and vitamin D), baseline SLEDAI, manifestations of SLE, interstitial pattern on HRCT, correlation with conventional radiology (Rx), respiratory function tests, clinical and physical examination at diagnosis.

**Results:** 455 patients diagnosed with SLE were included, of whom 20 had ILD (4.4%). 65% were women with a mean age at diagnosis of 63 ± 16.23 years. 30% presented ILD as the first clinical data for the diagnosis of SLE. 30% had jobs with exposure to chemicals. 25% were ex-smokers and 10% were active smokers. 10% had associated CVRF, highlighting HT (35%). 30% had valve disease on echocardiography such as mitral regurgitation (30%) and tricuspid regurgitation (20%). Only 10% had associated bronchopathy. 35% and 25% associated, respectively, pleural serositis and secondary Sjogren's syndrome. Arthritis and skin involvement were documented in 60% for both clinical domains. 40% had SLEDAI> 6.

At the diagnosis of ILD, 90% had symptoms such as dyspnea (75%) and cough (60%). 80% had crackles on examination. 85% had alterations in the X-ray and 100% in the CT scan with the NINE patterns as the main one (65%) with progression at 2 years despite treatment in 25%. Spirometry differentiated an obstructive pattern (25%), restrictive (20%) and normal (55%) with a tendency to stability at one year (40%) with the prescribed treatment. 90% had a decrease in diffusion, the majority being mild (67%). Analytically, 67% had elevated levels of CRP and hypovitaminosis D; 40% elevated DNA titers, 50% hypocomplementemia and 60% ENAS positive, highlighting Ro (45%); La (15%) and RNP (15%). Leukopenia and thrombopenia (30% and 10% respectively) were also observed.

In our study, we did not find any statistical significance between the variables analyzed, due to the small sample size.

**Conclusion:** 1)ILD is an infrequent manifestation in SLE, it usually associates respiratory symptoms and pulmonary functional repercussions with the diagnosis. The most characteristic radiological pattern is the NINE.