SATURDAY, 06 JUNE 2020

SLE, Sjögren's and APS - clinical aspects (other than treatment)

SAT0197

NON MYOCARDIAL CARDIAC INVOLVEMENT IN ANTIPHOSPHOLIPID SYNDROME IN A SPANISH REFERENCE CENTER

Á. Robles Marhuenda¹, J. Álvarez Troncoso¹, A. De Gea Grela¹, G. Daroca Bengoa¹, L. Ramos Ruperto¹, A. Díez Vidal¹, J. J. Ríos¹, C. Soto Abánades¹, E. Martínez Robles¹, A. Noblejas Mozo¹, F. Arnalich Fernández¹. ¹Hospital Universitario La Paz, Department of Internal Medicine, Madrid, Spain

Background: Antiphospholipid syndrome (APS) is a systemic autoimmune disease, associated with a hypercoagulable state and fetal loss and with other clinical manifestations including cardiac involvement. APS occurs as a primary disorder (PAPS) or secondary to another autoimmune disease (SAPS). Due to its vascular nature, various organs and tissues may be affected, including the cardiac system. Cardiac manifestations of APS are valve abnormalities (valve thickening and vegetations), occlusive arterial disease (atherosclerosis and myocardial infarction) and pulmonary hypertension (PH).

Objectives: To assess the prevalence of non-myocardial involvement (valvulopathy and pulmonary hypertension) in a cohort of patients with antiphospholipid antibodies (aPLs).

Methods: Retrospective observational study in a Spanish reference center for systemic autoimmune diseases. All patients with aPLs and performed transthoracic echocardiogram (TTE) were included in the study. Patients were divided between PAPS, SAPS and aPLs carriers. A cohort of 50 patients with systemic lupus erythematosus (SLE) without aPLs was used as a control. Anti-cardiolipin, anti-B2GP1 and lupus anticoagulant antibodies were determined by standard techniques.

Results: A total of 220 patients were reviewed. 145 (65.9%) were female. The mean age was 42 years. Among all patients with aPLs, 102 were PAPS, 73 SAPS, and 45 asymptomatic carriers (silent APS). Patients with aPLs, unlike patients with SLE without aPLs, presented more often pathological TTE (114 patients, 52%) ($p = 0.02$), with more valvular involvement (87, 39%) ($p = 0.005$) and pulmonary hypertension (21, 9.5%, $p = \text{ns}$). Valve involvement was identified in 99 patients: 45 in PAPS, 27 in SAPS, 14 in aPLs carriers and 13 in the SLE without aPLs, these differences being statistically significant ($p = 0.002$). Valvulopathy was asymptomatic in the majority of patients but required valve replacement in two patients. Mitral valve was the most affected, especially in the form of insufficiency (57%), followed by aortic valve, combined mitral and aortic valve, and less frequently the pulmonary valve alone (3 cases).

	aPLs global	PAPS	SAPS	aPLs carriers	SLE w/o aPLs
Total	220 (100%)	102 (46.4%)	73 (33.2%)	45 (20.4%)	50 (100%)
Men	75 (34.1%)	41 (40.2%)	23 (31.5%)	11 (24.4%)	6 (12%)
Women	145 (65.9%)	61 (59.8%)	50 (68.5%)	34 (75.6%)	44 (88%)

Conclusion: Subclinical valve involvement was very common in patients with APS. There was no correlation with other clinical manifestations of APS nor were other risk factors identified. PH was less frequent than valvular involvement in patients with APS. However, despite not being statistically significant, close to 10% of patients with APS had PH compared to 6% of patients without APS.

	aPLs global	PAPS	SAPS	aPLs carriers	SLE w/o aPLs
TTE performed	220 (100%)	102 (46.4%)	73 (33.2%)	45 (20.4%)	50 (100%)
Pathological TTE	114 (52%)				16 (32%)
Valvular involvement	87 (39%)	45 (20.4%)	27 (12.3%)	14 (6.3%)	13 (26%)
Pulmonary hypertension	21 (9.5%)				3 (6%)

Every patient with APS should have an echocardiogram in the initial study protocol in order to rule out both valvulopathy and pulmonary hypertension. This could modify the patient's management both in the short and long term, as well as the prognosis.

References:

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- [2] Tenedios F, Erkan D, Lockshin MD. Cardiac involvement in the antiphospholipid syndrome. *Lupus.* 2005;14(9):691–696.
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SAT0198

PROBABILITY OF PULMONARY HYPERTENSION IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS ACCORDING TO ESC GUIDELINE 2015

H. Baharuddin¹, N. F. Abdul Manaf², Z. Ismail³, K. S. Ibrahim⁴, M. Mohd Zain⁵, S. S. Ch'ng⁵. ¹Universiti Teknologi MARA, Department of Internal Medicine, Rheumatology Unit, Selangor, Malaysia; ²Universiti Teknologi MARA, Department of Internal Medicine, Selangor, Malaysia; ³Universiti Teknologi MARA, Department of Public Health & Preventive Medicine, Selangor, Malaysia; ⁴Universiti Teknologi MARA, Department of Internal Medicine, Cardiology Unit, Selangor, Malaysia; ⁵Hospital Selayang, Rheumatology Unit, Selangor, Malaysia

Background: The prevalence of pulmonary hypertension (PH) in systemic lupus erythematosus (SLE) varied according to the definitions and investigations used. Echocardiography should always be performed when PH is suspected, based on symptoms and signs,¹ which may be difficult to be assessed in patients with SLE because early symptoms of PH, including shortness of breath, fatigue, weakness, angina and syncope can be part of disease manifestation.

Objectives: To investigate the probability of PH in patients with SLE based on 2015 ESC Guideline, regardless of symptoms.

Methods: A cross-sectional study was conducted in a rheumatology centre in Malaysia which included patients aged 18 years and above, who fulfilled SLICC2012 criteria. Exclusion criteria were diagnosis of overlap syndrome and pregnancy. Demographic data and immunology profile were obtained from electronic medical records. TTE was performed by one technician who was blinded to other clinical details. Low, intermediate or high probability of PH was determined based on 2015 European Society of Cardiology (ESC) echocardiography criteria of PH.

Results: A total of 60 patients with SLE were recruited. The mean age was 41.6±10.9 years and SLE disease duration was 11.5±9.4 years. The cardiovascular co-morbidities were hypertension (38.3%), dyslipidaemia (25%), diabetes mellitus (5.0%) and ischemic heart disease (1.6%). Based on 2015 ESC echocardiography criteria for PH, 50 (83.3%) patients had low probability of PH, 8 (13.3%) had intermediate probability of PH and 2 (3.3%) with high probability of PH. (Table 1). Further analysis revealed that two patients with high PH probability were asymptomatic at the time of study. They were treated for active SLE after PH was diagnosed from TTE performed within a year of study period and subsequent RHC confirmed pulmonary artery hypertension (PAH). Among

Table 1. The details of echocardiographic signs of PH in patients with SLE.

Echocardiographic 'signs' of PH		PH probability (no of patients)		
		Low, n=50	Intermediate, n=8	High, n=2
Peak tricuspid regurgitant velocity, m/s	• ≤2.8 • 2.9-3.4 • >3.4	50 0 0	1 7 0	0 0 2
Ventricles	• Right/ left ventricle basal diameter ratio >1.0 • Flattening of interventricular septum	0 0	1 0	2 2
Pulmonary artery	• Right ventricular outflow Doppler acceleration time <105ms and/or mid-diastolic notching • Early diastolic pulmonary regurgitation velocity >2.2m/s • PA diameter >25mm	28 0 4	5 0 2	2 2 2
Inferior vena cava and right atrium	• Inferior vena diameter >21mm with decreased inspiratory collapse • Right atrial area >18cm ²	0 0	0 0	0 2

patients with intermediate probability of PH, one patient had intermittent palpitation and chest pain, while others were asymptomatic including one patient with PAH based on RHC. The most prevalent auto-antibodies among patients with intermediate and high probability of PH were anti-Ro (8 patients), anti-nuclear antibodies (7 patients) and anti-dsDNA (5 patients).

Conclusion: We found 16.6% patients with SLE who had intermediate and high probability of PH, based on 2015 ESC echocardiography criteria for PH. All except one patient had symptoms suggestive of PH at the time of study. RHC performed subsequently on two patients with high PH probability confirmed PAH.

References:

[1] Galie N, Humbert M, Vachiery JL, et al. *European Heart Journal*, 2016; 37(1):67-119

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SAT0199 PARTICULARITIES OF SJÖGREN SYNDROME IN ELDERLY PATIENTS

I. Naceur¹, R. Abida¹, T. Ben Salem¹, F. Said¹, M. Khanfir¹, I. Ben Ghorbel¹, M. Lamoum¹, M. H. Houman¹. ¹Faculty of Medicine, Tunis-El Manar University, Internal Medicine, Rabta university Hospital, Tunis, Tunisia

Background: Sjögren syndrome (SS) is a systemic autoimmune disease mainly described in females at a peak incidence age of 50. It was suggested that elderly onset of disease has particular clinical and biological phenotype.

Objectives: The aim of our study is to determine the particularities of SS in elderly patients.

Methods: Data of 332 patient fulfilling the American European Consensus Group criteria for Sjögren's syndrome over a period time of 18 years were studied. Clinical and biological features of elderly patients (G1) were described and compared to those of patients aged below 65 years old (G2) using the X2 and Fisher test.

Results: A total of 35 elderly were retained: 33 females and 2 males. The mean age of disease onset was 68.8 ± 4.4 years. The average delay (from first sign of the disease to diagnosis) was 1.27 years. The mean age at diagnosis was 70.3 ± 4.7 years. Xerostomia was described by 33 patients (94.3%). Focus score in the minor labial salivary glands pathology was ≥ 1 in 32 patients (91.4%). Two patients had abnormalities in parotid scintigraphy. Xerophthalmia was described by 32 elderly patients (91.4%). Schirmer test was abnormal in 20 cases (57.1%) and Break Up Time test was altered in 20 cases (57.1%). Arthralgia was the most frequent extra-glandular manifestation reported in 74.3% of patients. Fatigue was noted in 12 patients. The other systemic manifestations were: interstitial lung disease (n=13), peripheral neuropathy (n=7), central nervous system involvement (n=5), Raynaud's phenomenon (n=4), myositis (n=2) and renal tubulopathy (n=2). Laboratory findings showed hyperglobulinemia (n=19) and lymphopenia (n=15). Antinuclear antibodies were positive in 25 patients (71.4%) with positive anti-SSA antibodies (n=14) and positive anti-SSB antibodies (n=7). SS was primary in 25 patients. Ten patients had one or more associated autoimmune diseases: cryoglobulinemia (n=4), systemic scleroderma (n=3), systemic lupus erythematosus (n=2), rheumatoid arthritis (n=2) and autoimmune hepatopathy (n=2).

Systemic treatments involved corticosteroids for 15 patients, antimalarial agents for 12 patients, nonsteroidal anti-inflammatory drugs for three patients and immunosuppressant agents for nine patients. One patient developed lymphoma.

Comparative analysis showed that SS diagnosis was made earlier in elderly with an average delay of 1.27 years in G1 vs 3 years in G2 (p=0.02). Fatigue was more frequent in elderly (63.2% vs 23.8%) p<0.01. Positivity of anti-SSA was also more frequent in elderly (64.9% vs 46.7%; p=0.04). Anti-malarial agents were less prescribed in elderly (36.4% vs 55.3%; p=0.03). There was no significant differences between the two groups concerning the other clinical features, laboratory findings, treatment and outcomes.

Conclusion: Regardless of the statistical findings in our study and in the literature, treatment and follow-up of elderly patients with SS must obey to closer attention considering their vulnerability and the complexity of their management.

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SAT0200

RISK FACTORS FOR ADVERSE PREGNANCY OUTCOMES IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS

C. Cetin¹, T. Saraç-Sivriköz², M. Ateş-Tıkız², E. S. Torun¹, A. Ersoy¹, Y. Yalçinkaya¹, A. Gül¹, M. Inanc¹, M. L. Ocal¹, İ. Kalelioğlu², B. Artım-Esen¹. ¹Istanbul Faculty of Medicine, Rheumatology, Istanbul, Turkey; ²Istanbul Faculty of Medicine, Obstetrics and Gynecology, Istanbul, Turkey

Background: Pregnancies of patients with systemic lupus erythematosus (SLE) can be risky both for the mother and the fetus because of disease activity and pregnancy complications.¹

Objectives: In this study, we evaluated the risk factors related to adverse pregnancy outcomes (APO) in our pregnant SLE cohort who were followed up by both Rheumatology and Obstetrics and Gynecology departments at our university.

Methods: 168 pregnancy data were analyzed from 136 patients who fulfilled ACR classification criteria for SLE. The course of pregnancies were monitored and fetal/neonatal outcomes were recorded. Unexplained fetal death after 12 weeks of gestation, neonatal death, preterm birth due to preeclampsia, eclampsia or HELLP and birth of small for gestational age (SGA) infant were defined as APO. Cumulative clinical, laboratory and serological findings, disease activity (SLEDAI-2K) and damage (SLICC/ACR), and conventional risk factors were compared between APO(+) and APO(-) groups.

Results: The comparison of demographics, conventional risk factors and disease characteristics in APO(+) and APO(-) groups are summarized in Table-1. In APO(+) pregnancies, the duration of disease was longer (p <0.05) and the frequency of chronic hypertension was higher (p <0.05) compared to APO(-) pregnancies. Renal and neuropsychiatric (NP) involvement, thrombocytopenia, antiphospholipid syndrome (APS), lupus anticoagulant and anti-cardiolipin IgM positivity were significantly higher in APO(+) group. Mean SLEDAI-2K scores of three trimesters and postpartum 6 months were higher in APO(+) patients compared to APO(-) patients (2.2 ± 3.6 vs 1.2 ± 2.04, p <0.05; 4.9 ± 6.03 vs 2.7 ± 5.01, p = 0.02, respectively). Percentage of patients with damage at the beginning of pregnancy and the mean SLICC damage score were significantly higher in APO(+) group compared to APO(-) group (1.8 ± 2.1 vs 0.8 ± 1.3, p <0.05). In APO(+) group, damage was significantly higher in neuropsychiatric, renal and cardiovascular and locomotor systems (p <0.05).

Conclusion: Although an important proportion of SLE pregnancies result in live birth, active disease, especially renal and NP involvement, and presence of damage at the beginning of pregnancy increase the risk of maternal and fetal complications. Furthermore, the presence of APS or antiphospholipid antibody positivity are important risk factors for obstetric complications. In conclusion, pregnancy should be allowed after controlling the disease activity and patients should be closely monitored in coordination with Obstetrics and Gynecology clinics. In case of presence of damage, both the patient and the physician should be aware of a possible adverse pregnancy outcome.

References:

[1] *Ann Intern Med*. 2015 August 4; 163(3): 153–163. doi:10.7326/M14-2235.

Table 1. Demographic data of APO (+) and APO (-) groups, comparison of conventional risk factors, cumulative clinical, serological and laboratory features

	APO (-) (n=111)	APO (+) (n=57)	p
Age	35.1±6.7	34.9±5.9	NS
Age at conception	30.6±5.6	28.9±4.2	NS
Disease duration (months)	141.6±70	166.9±87.9	<0.05
Chronic hypertension, n (%)	6 (7)	11 (19.6)	<0.05
Photosensitivity, n (%)	86 (77.5)	43 (75.4)	NS
Malar rash, n (%)	66 (59.5)	38 (66.7)	NS
Oral ulcer, n (%)	11 (9.9)	6 (10.5)	NS
Arthritis, n (%)	77 (59.4)	42 (73.7)	NS
Serositis, n (%)	17 (15.3)	13 (22.8)	NS
Renal, n (%)	39 (35.1)	30 (52.6)	<0.05
Hematologic, n (%)	78 (70.3)	40 (70.2)	NS
Thrombocytopenia, n (%)	37 (33.3)	30 (52.6)	<0.05
AIHA, n (%)	16 (14.4)	14 (24.6)	NS
Neurologic, n (%)	7 (6.3)	9 (15.8)	<0.05
Anti-cardiolipin IgG, n(%)	28 (25.2)	18 (32.1)	NS
Anti-cardiolipin IgM, n (%)	18 (16.2)	18 (32.1)	<0.05
Lupus anticoagulant, n (%)	26 (23.4)	28 (49.1)	<0.001
Antiphospholipid syndrome, n (%)	28 (25.2)	30 (52.6)	<0.001

(NS=not significant, APO=adverse pregnancy outcome, AIHA=autoimmune hemolytic anemia)

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