SAT0465

CLINICAL MANIFESTATIONS AND PROGNOSIS OF SLE WITH CLINICALLY SIGNIFICANT ANTIPHOSPHOLIPID ANTIBODIES

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Background: Antiphospholipid antibodies (aPLs) have been described in 20% >40% of SLE patients, with 50%>70% of patients with SLE and aPL showing the clinical features of APS after 20 years of follow-up¹. It assumes that aPLs-positive SLE patients would have a more severe clinical phenotype and worse prognosis than those without aPLs.

Objectives: We decided to investigate the clinical manifestations and prognosis of SLE with clinically significant aPLs in a multiple centre SLE cohort.

Methods: A follow-up study to investigate the prognosis of SLE, has been conducted in 26 centres across Jiangsu province as described before². SLE patients fulfilled ACR/SLICC criteria. Patients who had ever recorded first admissions and detected aPLs during the 1999±2009 decade were followed and checked for their survival status in 2015. Clinically significant aPL were defined as: positive LA test, aCL IgG/IgM antibodies>99 th percentile and/or aβ2GPI>99 th percentile on two or more occasions at least 12 weeks apart.

Results: 1) Among 1372 SLE patients, 495 patients was reported the aPLs minutely, and 146 cases was with clinically significant aPLs. Compared with aPLs negative SLE patients, the proportion of men, and the rates of oral ulcer, neuropsychiatric involvement, dsDNA, antinuclear antibody and C3 were significantly higher in aPLs positive SLE. (table 1)

	aPLs positive n=146	aPL negative n=349	P
Gender (male)	5	32	0.017
Oral ulcer	29	44	0.047
Thromboembolism	16	18	0.031
Neuropsychiatric	14	20	0.000
C3 decrease	119	248	0.015
Anaemia	109	214	0.005
ANA positive	138	317	0.048
Anti dsDNA positive	83	169	0.009

2) There were 20 deaths in aPLs positive SLE group and 52 deaths in aPLs negative SLE group during the average follow up of 7.38±0.56 years and 7.54±0.47 years respectively. There was no significant difference in survival curves by Kaplan Meier survival analysis (p=0.776). (Picture 1)

3) Multivariate Cox regression analysis revealed that long time of diagnosis (HR 4.205, p<0.05), SDI>1 in admission (HR 11.982, p<0.01), neuropsychiatric involvement (HR 2.826, p<0.05), and increased serum creatinine (HR 8.403, p<0.01) were independent predictors of mortality. (table 2)

factors	Univariate			Multivariate		
	HR	95% CI	P	HR	95% CI	P
Time of diagnosis>1 year	5.26	2.24–12.32	0	4.20	1.06–16.56	0.04
SDI≥1 on admission	4.37	1.67–11.40	0.003	11.98	2.85–50.25	0.001
Neuropsychiatric	4.52	1.89–10.84	0.001	2.82	1.01–7.85	0.048
Increased serum creatinine	4.39	1.87–10.30	0.001	8.40	2.20–32.04	0.002

Conclusions: In this study, we observed that around one-third of patients had clinically significant aPLs, and such autoantibody positivity was associated with a different clinical and serological profile. However, the mortality between aPLs

positive and negative SLE patients had no significant difference. SLE patients presented with vital organ damages rather than active disease at initial hospitalisation are likely to have a poor outcome, especially neuropsychiatric involvements and renal insufficiency.

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SAT0466

SERUM VITAMIN D DEFICIENCY IS ASSOCIATED WITH ACTIVE RENAL DISEASE IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Objectives: 25-hydroxyvitamin D (25(OH)D) deficiency is common in systemic lupus erythematosus (SLE) as well as chronic kidney disease. In this study, we investigated the association of 25(OH)D deficiency and renal involvement in SLE patients.

Methods: Two hundred seventy-two SLE patients and 138 control subjects were enrolled; 102 patients with active nephritis, 42 patients with inactive nephritis, and 128 patients with non-renal disease. Serum 25(OH)D levels were measured, and clinical and laboratory data were obtained from medical records.

Results: Mean serum 25(OH)D levels were significantly lower in SLE patient than control subjects (19.6 ng/ml versus 21.7 ng/ml, p=0.006). Out of 272 patients, 61.8% were vitamin D deficient (defined as <20 ng/ml). Patients with active nephritis had lower serum 25(OH)D levels (16.9 ng/mL) than patients with inactive nephritis (20.2 ng/mL) and without nephritis (21.5 ng/mL) (p=0.030, p<0.001), but there was no difference between the inactive nephritis and non-renal disease. Moreover, serum 25(OH)D levels were positively correlated with complement C3 (γ=0.135, p=0.026) and C4 (γ=0.159, p=0.009), but inversely with anti-dsDNA antibody level (γ=-0.156, p=0.010). Analysis of receiver operating characteristic curve for differentiating active nephritis and non-renal disease revealed an area under the curve (AUC) of 0.676, which is better than those of anti-dsDNA antibody (AUC=0.585, p=0.038) and complement C3 (AUC=0.509, p=0.001), C4 (AUC=0.538, p=0.008).

Conclusions: Vitamin D deficiency is more common in SLE patients with active nephritis, and its level could be a potential marker for active renal disease in SLE. A prospective cohort study is needed to further elucidate the causal relationships over time.

Disclosure of Interest: None declared

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SAT0467

CEREBRAL VENOUS THROMBOSIS OCCURRENCE IN SYSTEMIC LUPUS ERYTHEMATOSUS WITHOUT ANTIPHOSPHOLIPID ANTIBODY SYNDROME: A MONOCENTRIC SERIE OF 10 CASES

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Background: Cerebral venous thrombosis (CVT), which includes cerebral vein and dural sinus thrombosis, is a rare disorder that can lead to significant morbidity and mortality. Its occurrence in SLE in the absence of APS has been rarely reported. In this study we aimed to describe a cohort of SLE patients suffering from CVT without APS.

Methods: We collected retrospectively clinical and biological data of patients with confirmed CVT in the Pitié-Salpêtrière cohort of SLE (n=1352 patients). Patients fulfilled ACR SLE criteria. The diagnosis of CVT was confirmed by brain imaging studies. Exclusion criteria were patient with a lupus anticoagulant or IgG/IgM anti-cardiolipin antibodies or anti-β2 glycoprotein-1 abs. We searched on PUBMED database for case report of this association published in English until 31 August 2017. Lupus flares were defined according the SELENA Flare instrument.

Results: We included 10 patients (8 women and 2 men). The median (range) age at diagnosis of CVT was 28 years.⁹⁻⁵³ The CVT occurred with a median delay of 4

years (0–11) after the diagnosis of SLE. At the time of the CVT diagnosis: no patients had a past medical history of thrombotic event or miscarriage or foetal loss; 7 patients had a lupus flare (5 lupus nephritis [1 class I, 1 class V, 2 class IV and 1 class III+V], 4 immune thrombocytopenia, 2 autoimmune hemolytic anaemia, 3 cutaneous lupus, 1 serositis and 1 arthritis); 7 patients were treated with corticosteroids, 4 with hydroxychloroquine and 4 with immunosuppressive drugs. Other potential precipitating factors of CVT were: 2 nephrotic syndromes, 2 anaemia and 1 hyperhomocysteinemia. CVT was symptomatic for 9 patients: 8 headaches, 3 epilepsy and 1 sensitive-motor deficit. The diagnosis of CVT was confirmed by magnetic resonance imaging (MRI) for 9 patients and cerebral angiography for 1 patient. The median delay between the onset of clinical symptoms and the diagnosis of thrombosis was 10 days.^{3–37} Nine patients presented a single localisation of CVT (superior longitudinal or lateral or cavernous sinus, or cortical cerebral vein). Only 1 patient had thrombosis of both lateral and sigmoid sinus. Cerebral infarction or haemorrhage was seen for 2 patients. Corticosteroids and immunosuppressant treatment were increased or introduced because of a concomitant lupus flare for 2 patients. All patients were treated with heparin followed by vitamin K antagonists for 7 patients or apixaban for 2 patients. One patient received long-term heparin. After a median survey of 19.5 (1–120) months: anti-coagulant drugs were stopped for 6 patients; recanalisation were complete in 7 of the 7 patients assessed on brain MRI; no patient had a residual neurological damage; only 1 patient had a new vascular event in the form of a brain haemorrhage. In addition to these 10 cases we found 17 cases from a literature review.

Conclusions: CVT is a rare event in SLE in absence of APS. In our cohort SLE was often active at the time of the CVT occurrence. Most of the CVT were limited in extension and severity. The outcome on anticoagulant treatment was favourable without residual neurological damage. The occurrence of CVT in SLE is not an indication by itself to increase or to introduce corticosteroids or immunosuppressive drug.

Disclosure of Interest: None declared

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SATURDAY, 16 JUNE 2018

Scleroderma, myositis and related syndromes

SAT0468 SCLERODERMA -INTERSTITIAL LUNG DISEASE: DOES MAINTENANCE THERAPY MAKE A DIFFERENCE

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Background: Interstitial lung disease (ILD) is the commonest cause of death among Scleroderma (SSc) patients. The evidence regarding efficacy of cyclophosphamide (CYC) in SSc-ILD results mainly from two high-quality RCTs.^{1,2} In the scleroderma lung study 1, after 12 months of therapy with oral CYC, a significant difference in mean forced vital capacity (FVC) was noted favouring CYC group. However, the effect waned off by the end of 24 months of follow up, indicating that maintenance therapy might help in preserving the initial benefits.³

Objectives: To study the long-term effect of maintenance therapy in SSc related interstitial lung disease (SSc-ILD)

Methods: This is a retrospective data analysis of forty-three patients with SSc-ILD divided into two groups, those who received maintenance therapy with either mycophenolate (MMF) or azathioprine (AZA) and the other group without any maintenance therapy following CYC induction. Response to treatment was classified as improved ($\geq 10\%$ increase in FVC from baseline), stable (change in FVC of 0%–10% from baseline) and worsened ($\geq 10\%$ decrease in FVC or fall of FVC to $<40\%$ of predicted). Results were expressed as percentages for categorical variables and mean with standard deviation (SD) for continuous variables. Chi square test was used for comparison of responses between two groups of patients. Following induction patients were divided into groups: those They were assessed at three years.

Results: Baseline characteristics of the study population are shown in table 1. Maintenance therapy was given in 29 (67.4%) patients and among them favourable outcome (improve +stable) was noted in 23 (79.3%) patients and unfavourable outcome was noted in 6 (20.7%) patients. Fourteen (32.6%) patients did not receive maintenance therapy and among them, favourable outcome was noted in 4 (28.6%) patients and unfavourable outcome in 10 (71.4%) patients. This difference in the rates of favourable outcome between both the groups was statistically significant ($p=0.001$). Figure 1 summarises the response rates of the patients of these two groups.

Abstract SAT0468 – Table 1. Baseline characteristics of the patients (n=43)

Characteristic	Maintenance group (n=29)	No maintenance group (n=14)
Females (%)	93	85.7
Mean age in years(SD)	40.9 (± 10.9)	42 (± 10.5)
Type of Scleroderma		
Limited cutaneous (%)	44.8	57.1
Diffuse cutaneous (%)	55.1	42.9
ANA positivity (%)	75.9	78.6
PAH (%)	34.5	28.6
Mean FVC (SD)	57.6 (± 13)	55.6 (± 15.2)

Conclusions: Maintenance therapy up to 3 years (either with AZA or MMF) following IV-CYC induction, resulted in a favourable outcome and helps in preserving the initial beneficial effects achieved with IV CYC.

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SAT0469 “INTERSTITIAL LUNG DISEASE IN IDIOPATHIC INFLAMMATORY MYOPATHIES: ULTRASOUND ASSESSMENT OF PLEURAL IRREGULARITIES AND COMPARISON WITH HIGH RESOLUTION COMPUTED TOMOGRAPHY”

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Background: Interstitial Lung Disease (ILD) is one of the most frequent and significant extra-muscular manifestations in Idiopathic Inflammatory Myopathies (IIM). Although high resolution computed tomography (HRCT) remains the gold standard technique for the evaluation of ILD, during the last years several authors proposed ultrasound (US) quantification of pleural irregularities (PIs) as a possible method to detect lung involvement in IIM patients.

Objectives: The objective of our study was to evaluate the prevalence of PIs and to compare the results obtained to the data provided by HRCT.

Methods: Thirtyseven patients (24 females, 13 males, mean duration of the disease 5.21 $\pm$ 7.6 and median age 62.24 $\pm$ 1.8 years) with a diagnosis of IIM according to Bohan and Peter criteria (17PM, 16DM, 1MCI, 3overlap syndromes) who required HRCT evaluation were enrolled. All patients underwent rheumatological clinical evaluation, including dyspnoea measurement by means MRC scale and disease activity assessment according to IMACS criteria (muscle enzymes, MMT8, VAS, HAQ, MYOACT). Patients were also asked to complete Patient Reported Outcome (PRO) questionnaires as Leicester Cough Questionnaire (LCQ). All the patients underwent a thoracic US evaluation in 53 anterior and posterior thoracic areas by Esaote MyLab Gold ultrasound device with 8–18 MHz