months for central nervous system (CNS). NI revealed the disease in 14 cases (34%). CNS and PNS involvements were respectively found in 27 and 19 cases (six patients had both PNS and CNS). In patients with CNS disorders, headache was the most frequent symptom ($n=11$). Pyramidal syndrome was found in 9 cases and cerebellar syndrome in one patient. Aseptic meningitis was noted in two cases. Brain MRI was performed in 22 cases and showed abnormalities in 82% of cases. T2 and flair weighted hyperintensities were periventricular ($n=11$), subcortical ($n=5$), frontal ($n=3$) and parietal ($n=2$). Myelitis was found in one patient.

Patients with PNS showed paresthesia (92%), motor deficit (22%) and decreased tendon reflex (18%). Sensory-motor neuropathy was described in 4 cases, pure sensory neuropathy in 6 cases (including one case of small fiber neuropathy) and motor neuropathy in one patient. Mononeuropathy multiplex was found in 4 cases and polyradiculoneuritis in one patient. Neuromuscular biopsy was performed in five patients and showed vasculitis in two cases. Trigeminal nerve and cochlear nerve were respectively involved in four and one patients. Antinuclear antibodies, anti-SSA and anti-SSB antibodies were respectively positive in 71%, 50% and 28% of cases. NI were treated with corticosteroids and immunosuppressive therapy in respectively 56% and 41% of cases. Outcome was good in 48% of the patients. Comparison of clinical and biological features in patients with and without NI showed no significant differences, only fatigue was significantly associated to NI (45.5% vs 12.9%; $p=0.005$).

Conclusions: In our series, NI prevalence was similar to other groups. CNS involvements were more frequent than PNS manifestations whereas in other ethnic groups they are less frequent.

Disclosure of Interest: None declared

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FRI0301 CHARACTERIZATION OF FEVER IN HOSPITALIZED PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS: LESSONS FROM THE JIANGSU COHORT

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Background: Fever is one of the main symptoms leading to the admission of patients with systemic lupus erythematosus (SLE) and its causes are usually complicated.

Objectives: To explore the prevalence and clinical characteristics of fever in Chinese SLE patients at the time of their first admission and to find out whether fever is related to a poor prognosis.

Methods: A follow-up study aimed to delineate SLE prognosis had been conducted by our Lupus Collaborative Group under the supervisor of Jiangsu Rheumatology Association to collect the data of patients who had ever recorded first admissions during the 1999–2009 decade (1). Among which, those with admission temperature documented were extracted for the assessment of potential factors associated with fever. The independency of various clinical features linked to fever was established by binary logistic regression analysis, and the relation between fever and patient's survival was determined by Cox regression analysis.

Results: Totally 1,348 patients were enrolled, in which 1,049 (77.8%) had normal temperature, 221 (16.4%) had low and moderate fever and 78 (5.8%) had high fever at the time of their first hospitalization. Compared with those having normal temperature, fever patients were more likely to have short disease duration (RR 0.74), concurrent infection (RR 3.29) and gastrointestinal (RR 1.57) as well as hematological involvements (RR 1.56). High C reactive protein level (RR 2.08) and positive anti-Sm antibody (RR1.55) were the two laboratory factors related to fever at admission. However, age, gender, SLEDAI score or erythrocyte sedimentation rate was not independently associated with fever in this cohort. Cox regression analysis showed that there was no correlation between fever and overall deaths (Figure 1) as well as infection induced deaths.

Figure 1 Cumulative survival rates for patients with or without fever at first admission

Conclusions: Both infection and lupus itself (especially gastrointestinal and hematological involvements) contribute to fever in SLE patients. Although the presence of fever increases the complexity of disease treatment, it does not constitute a risk factor for patient's long-term outcome.

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FRI0302 ELEVATED CYSTEINE-RICH PROTEIN 61 IN SYSTEMIC LUPUS ERYTHEMATOSUS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary arterial hypertension (PAH) is a complex and devastating complication of connective tissue diseases that leads to severe morbidity and mortality. Unlike in Caucasians, it is systemic lupus erythematosus that recognized as the major underlying cause of CTD associated PAH in Asian countries, especially in China [1]. Early diagnosis and intervention is vital for better long-term outcome in SLE-PAH patients. Previous study has demonstrated Cysteine-rich protein 61 (Cyr61) was highly expressed in SLE patients [2]. However, the role of Cyr61 in pulmonary arterial hypertension (PAH) remains unknown.

Objectives: To explore the value of Cyr61 for PAH in SLE patients by comparing the plasma Cyr61 levels in SLE patients with/without PAH.

Methods: Plasma samples from two tertiary medical centers were obtained from 54 patients with definite SLE-PAH, 52 age, gender and SLEDAI matched SLE patients without PAH, and 54 age and gender matched healthy controls. Plasma Cyr61 concentration was measured by enzyme-linked immunosorbent assay.

Results: Plasma Cyr61 concentration in SLE-PAH patients was significantly higher than the matched SLE patients and healthy controls (median (IQR): 172.5 (143.8, 218.2), 124.9 (104.1, 154.7), 58.17 (28.9, 80.4) respectively, $P<0.001$) (Figure1). The sensitivity and specificity of Cyr61 in predicting the presence of PAH in entire SLE patients were 79.6% and 67.3%. Receiver operating characteristic curve analysis showed the area under the curve was 0.757 (95% CI: 0.662–0.852), with 140.6 pg/ml as the cut off concentration (Figure2). Further multivariate logistic regression analyses revealed high Cyr61 level (>140.6) is an independent risk factor for SLE patients to develop PAH (OR:7.822 (95% CI: 2.224–41.138)) (Table 1). Additionally, weak to moderate positive correlations were observed between Cyr61 concentration and serositis, hematological involvement, red blood cell distribution width, right ventricular systolic pressure and right ventricular diameter measured by echocardiography in entire SLE population.

Table 1. Multivariate logistic regression analysis

Variables	OR	95% CI	P value
High Cyr61 level	7.822	2.224 41.138	0.002
Serositis	8.244	1.854 36.663	0.006
Anti-RNP positivity	3.902	1.067 14.267	0.040
Renal disorders	0.174	0.044 0.689	0.013
Anti-dsDNA positivity	0.171	0.046 0.636	0.008

Conclusions: Plasma Cyr61 level was significantly higher in SLE-PAH patients than SLE patients without PAH. Cyr61 may be used as a biomarker for PAH complication in SLE patients.

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[1] Hao, Y.J., et al., Connective tissue disease-associated pulmonary arterial hypertension in Chinese patients. *European Respiratory Journal*, 2014. 44(4): p. 963–972.

[2] Lin, J., et al., Serum Cyr61 is associated with clinical disease activity and inflammation in patients with systemic lupus erythematosus. *Medicine (Baltimore)*, 2015. 94(19): p. e834.

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Vasculitis

FRI0303 THE IGG4:IGG RNA RATIO IS A NEW AND PROMISING DISEASE ACTIVITY MARKER IN GRANULOMATOSIS WITH POLYANGIITIS

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Background: Granulomatosis with Polyangiitis (GPA) is a form of vasculitis characterized by inflammation of blood vessels in lungs, kidneys and the ear, nose and throat region. Regular monitoring and treatment adjustments are common, as the disease activity tends to fluctuate over time. Unfortunately, good markers for disease activity are lacking. This leads to both over- and undertreatment. Immunoglobulin G4 positive (IgG4+) B-cells and plasma cells are implicated in the pathogenesis of GPA, but the level of serum IgG4 does not seem to be a good disease activity marker. Recently we developed a test that indirectly measures the presence of IgG4+ B-cells/plasma cells by measuring the IgG4:IgG RNA ratio¹. We hypothesized that this test could be used as disease activity marker.

Objectives: To test the IgG4:IgG RNA ratio in peripheral blood as a disease activity marker in GPA.

Methods: 27 PR3+ ANCA+ positive GPA patients were included in this cross-sectional study. Mean age was 52 years, 56% were female, and 39% had active disease. For each patient the ESR, CRP, BVAS, and ANCA titre were measured and peripheral blood samples were obtained. Patients were defined as having active disease if the BVAS was ≥ 3 . In addition we included 10 healthy controls, and 63 patients with other immune mediated inflammatory diseases (systemic lupus erythematosus (SLE) (n=24), rheumatoid arthritis (RA) (n=19), primary sclerosing cholangitis (PSC) (n=20)). A validated qPCR test was performed in all groups to measure the IgG4:IgG RNA ratio in peripheral blood samples¹.

Results: The median IgG4:IgG RNA ratio was significantly higher in the GPA cohort (5.7, IQR 2.6 – 19.7) compared to all control groups: 1.2 in SLE (0.7 - 3.3; $p < 0.01$), 2.5 in RA (0.7 - 4.1; $p < 0.05$), 1.6 in PSC (1.0 - 2.8; $p < 0.001$) and 1.3 in HC (0.6 - 1.8; $p < 0.01$). In addition, the median IgG4:IgG RNA ratio was significantly higher in patients with active disease (23.8; IQR 12.1 – 29.1) compared to patients in remission (3.5; IQR 2.0 – 5.5) ($p < 0.0001$). The height of the IgG4:IgG RNA ratio significantly correlated with height of the BVAS ($r^2 = 0.76$, $p < 0.0001$), while the ESR, CRP and ANCA titre did not. Interestingly, IgG4/IgG RNA ratios among patients with active disease were consistently above 9.3, and among patients in remission they were below this threshold.

IgG4:IgG RNA ratio (%) in healthy controls (HC), primary sclerosing cholangitis (PSC), systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and granulomatosis with polyangiitis (GPA). Red dots represent patients with active disease (resp. SLEDAI, DAS28 and BVAS). * < 0.05 , ** < 0.01 , *** < 0.001

Conclusions: The IgG4:IgG RNA ratio distinguishes active GPA from GPA in remission with excellent specificity and sensitivity. Moreover the ratio shows a significant correlation with disease severity, in contrast to ESR, CRP and ANCA titre. Retesting in another, prospective study is indicated to validate the IgG4:IgG RNA ratio as a novel, highly sensitive and specific marker of disease activity in GPA.

References:

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FRI0304 THE UTILITY OF 18F FDG-PET/CT IN DISTINGUISHING BENIGN FROM MALIGNANT RETROPERITONEAL FIBROSIS

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Background: Retroperitoneal fibrosis (RPF) could be idiopathic or secondary to various factors including malignancy. Pathology remains the most reliable diagnostic approach. However, the pathological features have yet been well defined and the feasibility of biopsy procedure depends on several factors such as the proximity of the lesions to abdominal aorta. ¹⁸F FDG-PET/CT may help exclude malignancy by assessing FDG-uptake and mapping the whole body, especially when biopsy procedure is inaccessible.

Objectives: To evaluate the utility of PET/CT in distinguishing benign from malignant RPF.

Methods: Patients with benign or malignant RPF diagnosed between September 2011 and June 2016 were included. Morphologic features, FDG-uptake of retroperitoneal lesions and lymph nodes (LNs) were compared. FDG-uptake of retroperitoneal lesions was assessed visually with the reference of liver and assessed semiquantitatively with SUVmax. LNs were regarded as specific LNs if frequencies of LNs with high FDG-uptake were observed differently between two forms of RPF. ROC analyses were performed to evaluate the diagnostic accuracy.

Results: Seventy-one cases with benign RPF and 21 cases with malignant RPF secondary to lymphoma (15) or metastatic carcinoma (6) with primary sites of pancreas (2), colon (2), stomach (1) and kidney (1) were included. The craniocaudal length, axial width, and distances between anterior or posterior

Table 1. Morphologic features, FDG-uptake of retroperitoneal lesions and lymph nodes (LNs)

Parameters	Benign RPF (71)	Malignant RPF (21)	p value
Morphologic features			
Craniocaudal length, median (IQR), mm	107.0 (80.5,136.4)	164.9 (104.7,229.2)	0.001
Axial width, median (IQR), mm	41.0 (32.3,52.0)	85.0 (62.3,98.5)	<0.001
Aorta lumen to anterior limit, median (IQR), mm	8.9 (6.0,12.0)	20.7 (13.9,45.3)	<0.001
Aorta lumen to posterior limit, median (IQR), mm	2.8 (2.2,4.4)	7.4 (4.5,14.5)	<0.001
FDG-uptake of retroperitoneal lesions			
High FDG-uptake, n (%)	55 (77.5)	21 (100.0)	0.017
SUV max, mean (S.D.)	4.8 (1.7)	12.2 (7.1)	<0.001
LNs with high FDG-uptake, n (%)			
Hilar/Mediastinal	27 (38.0)	13 (61.9)	0.054
Cervical	6 (8.5)	5 (23.8)	0.058
Axillary	4 (5.6)	6 (28.6)	0.003
Retroperitoneal	3 (4.2)	16 (76.2)	<0.001
Supraclavicular	2 (2.8)	8 (38.1)	<0.001
Inguinal	1 (1.4)	5 (23.8)	<0.001
Peritoneal	0 (0.0)	10 (47.6)	<0.001
Number of specific LNs, n (%)	0 (0.0)	2 (1.3)	<0.001

Figure 1. Representative ¹⁸F fluorodeoxyglucose positron emission tomograph images for benign retroperitoneal fibrosis (A) and malignant retroperitoneal fibrosis secondary to lymphoma (B, C) and metastatic carcinoma (D). The retroperitoneal process are circled by circles in the same size with different level of FDG-uptake. D-F are images from one patient with histological diagnosis of colon cancer. FDG-uptake is accumulated in left supraclavicular lymph node pointed by the arrow (E) and in sigmoid colon pointed by the arrow head (F).