

**Results** Although age has been demonstrated to be a risk factor for severe bleeding episodes in patients placed on long-term anticoagulation, we decided to start hydroxychloroquine and combination therapy with warfarin (INR range between 2.5 and 3.5) and low-dose aspirin according to EULAR recommendations, considering the patient's good clinical condition and absence of comorbidities. The patient was informed about the nature of the disease and the need for treatment with a high risk of bleeding. In December 2021, despite vaccination with two doses, he contracted COVID-19 infection and warfarin treatment was replaced with low molecular weight heparin at prophylactic dosage, but after some days he presented dyspnoea with desaturation. A lung CT scan showed bilateral pulmonary embolism, attributable to intercurrent viral infection and to APS (not completely controlled after warfarin withdrawal), and so clinicians decided to replace treatment with warfarin. After one month, patient had been discharged from hospital in good clinical health.

**Conclusions** Accurate diagnosis and treatment of late-onset APS represent a challenge due to the lack of knowledge of this disease in the elderly.

**PO.3.70 CASE REPORT OF A SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT WITH AN ULCERATIVE NECROTIC VASCULITIS: FEATURES OF THERAPEUTIC APPROACHES**

<sup>1</sup>A Shumilova\*, <sup>1</sup>E Naryshkin, <sup>2</sup>E Stolyarevich, <sup>1</sup>T Reshetnyak. <sup>1</sup>V.A.Nasonova Research Institute of Rheumatology ~ Moscow ~ Russian Federation; <sup>2</sup>City Clinical Hospital No. 52 of the Moscow Department of Health ~ Moscow ~ Russian Federation

10.1136/lupus-2022-elm2022.99

**Background** Systemic lupus erythematosus (SLE) is a heterogeneous chronic rheumatic autoimmune disease with a range of clinical manifestations, characterized by impaired activation of cellular and humoral links of immunity, uncontrolled hyperproduction of autoantibodies of a wide spectrum to nuclear antigens and the formation of immune complexes that cause immuno-inflammatory damage to tissues and organs. Vasculitis in connective tissue disease (CTD) is quite rare, it is reported in approximately 10% of patients with CTD, SLE shows the highest association rate.

**Purpose** To report a case of a patient with SLE with ulcerative necrotic vasculitis of the upper and lower extremities with the formation of dry necrosis in combination with lupus nephritis (LN).

**Methods** A 26-year-old female patient with SLE for 1 year was diagnosed based on ulcerative necrotic vasculitis of the upper and lower extremities with the formation of dry necrosis, Class V LN (proteinuria, erythrocyturia, cylindruria), adhesive pericarditis, immunological disorders (highly positive antibodies to Sm+, anti-Ro/SS-A+), ANF hep-2 positivity.

**Results** An anamnesis of the disease, physical, laboratory and instrumental examination and treatment tactics are presented in table 1.

**Conclusions** Necrotizing vasculitis is always secondary to SLE and it is necessary to exclude antiphospholipid syndrome. Vessels of any size may be involved, but mainly medium vessels vasculitis in SLE is reported. The most common clinical manifestations are ischemic lesion, ulcers, gangrene etc. It may be an early warning sign for systemic involvement. The combination of glucocorticoids, hydroxychloroquine, cytostatic (azathioprine), anti-B-cell therapy (rituximab) is highly effective.

**Abstract PO.3.70 Table 1** Clinical and laboratory manifestations of the disease and treatment tactics

Age	February 2020 (25 years old)	March 2020 (25 years old)	June-July- August 2021 (26 years old)	September 2021 (33-34 years old)
<b>Manifestations</b>				
Raynaud's syndrome	■	■	■	■
Dry necrosis		■ of the fingers of the hands	■ of the fingers of the hands and the skin of and lower extremities	■ Post-ulcerated hyper-pigmented defects on the skin of the shins and thighs, forearms on both sides, ulcerative defect without signs of infection, not covered with a crust, up to 2 cm in diameter with a corolla of hyperemia around the circumference. The stumps of the fingers of the right hand: dry necrosis of the nail phalanx of the I finger, proximal phalanges of the II and V fingers, II and III finger stumps with signs of ulcerative defects covered with crusts.
Swelling of the hands			■	
Pain and burning in the area of the stump of the hands			■	■
Impaired nitrogen excretion function of the kidneys			■ Creatinine up to 170 mmol/l, Urea up to 17.35 mmol/l	
Lupus nephritis			■ Proteinuria 0.26g/l, Leukocyturia 15-20p/s, Erythrocyturia 5-8p/s, Hyaline cylinders 7-8p/s	■ Proteinuria 1,324 g/day (<0.5), Erythrocyturia (<5)
Hematological disorders			■ Anemia up to 63 g/l, leukocytosis 12.7 10 <sup>9</sup> /l, ESR-59->75mm/h, CRP++	■ Leukocytosis 10.4 10 <sup>9</sup> /l (4.0-9.0), ESR-35mm/h (<30), anemia up to 107 g/l (120-140)
Antiphospholipid antibodies and ANCA			■ LA-60.50 (positive), the rest was negative	Negative
Antinuclear antibodies			■ RNP+, Sm/RNP+, SSA/Ro 60kD, ribosomes+	■ anti-Ro/SS-A-45.6U/ml (0-25), Antibody to Sm-29.5U/ml (0-25)
ANF hep-2 positivity				■ 1/320 sp, cytopl
Duplex scanning of the arteries of the upper extremities				■ Reduction of blood flow in the radial and ulnar arteries in the right and left upper extremities.
X-ray of the hands				■ Arthritis of the wrist joints and metacarpophalangeal joints, absence of most distal and middle phalanges of both hands, stumps of proximal phalanges of both hands.
Echocardiography:				■ The pericardium is slightly thickened and compacted, unidirectional movement of the pericardium leaves
Nephrobiopsy				■ Membranous nephropathy. Considering the immunofluorescence data (C1q glow) corresponds to the Class V lupus nephritis.
Diagnosis	Raynaud's syndrome	Raynaud's syndrome	Burger's Disease. Raynaud's syndrome	SLE, acute at the beginning of the process, high activity (SLEDAI-2k=22): ulcerative necrotic vasculitis of the upper and lower extremities with the formation of dry necrosis, Class V lupus nephritis (proteinuria, erythrocyturia, cylindruria), adhesive pericarditis, immunological disorders (Sm+, anti-Ro/SS-A+), ANF+
Treatment tactics	-	Thoracic sympathectomy was performed without effect, then amputation of the phalanges of the II-V fingers of both hands. Patient did not receive any therapy.	Hemotransfusions of the Erythrocyte mass. Methylprednisolone 16 mg/day.  Since August 2021: Rivaroxaban 10 mg/day, Acetylsalicylic acid 100 mg/day.	Methylprednisolone 16 mg/day, Hydroxychloroquine 200 mg/day, Nadropanin calcium 0.6->0.9/day, Acetylsalicylic acid 100 mg/day; Azathioprine 100 mg/day; Intravenous Rituximab 1000 mg №1; Intravenous Alprostadil 20 mcg №10.

No recurrence of vasculitis was detected within 7 months after correction of therapy.

**PO.3.71 A UNIQUE CASE OF A DELAYED DIAGNOSIS OF STRONGLYLOIDIASIS HYPERINFECTION IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS, LYMPHOPLASMATIC LYMPHOMA, AND KIDNEY INJURY**

<sup>1</sup>D Bosnic\*, <sup>2</sup>I Aurer, <sup>3</sup>S Bulimbašić. <sup>1</sup>University Hospital Center Zagreb, Division of Clinical Immunology and Rheumatology, Department of Internal Medicine, ~ Zagreb ~ Croatia; <sup>2</sup>University Hospital Center Zagreb, Division of Haematology, Department of Internal Medicine, ~ Zagreb ~ Croatia; <sup>3</sup>University Hospital Center Zagreb, Department of Pathology and Cytology ~ Zagreb ~ Croatia

10.1136/lupus-2022-elm2022.100

**Purpose** To report a unique case of a delayed diagnosis of stronglyloidiasis hyperinfection in a patient with systemic lupus erythematosus (SLE), lymphoplasmatic lymphoma and proteinuria with microhaematuria. Stronglyloidiasis stercoralis is a rare nematode endemic to the tropical and subtropic regions and immunocompromised patients are at risk of infection. Stronglyloidiasis hyperinfection has been described in SLE cases previously, yet not also co-occurring with lymphoplasmatic lymphoma.

**Methods** A 67-year-old female with a fifteen year history of SLE was referred to the immunology outpatient clinic due to malaise and ataxia progressing over several months. Before the referral, neurological, radiological, and hematological workup was performed, which objectified the clinical symptoms, showed CNS affection, and confirmed remission of a lymphoplasmatic lymphoma treated using an B-R protocol over the previous year. Physical examination findings were consistent with previous chronic SLE sequelae, yet a decrease in body weight was noted, as well as a mild anemia, proteinuria (175 mg/dU) with microhaematuria, and increased eosinophile count. Workup was broadened, including kidney biopsy, bone marrow biopsy, parasite serology, and stool ova and parasite test.

**Results** Follow-up laboratory showed a further increase in eosinophile count (25%), positive Stronglyoides stercoralis serology (immunoglobulin G ELISA), and stronglyoides larvae in stool samples. Bone marrow biopsy showed no abnormalities and kidney biopsy analysis ruled out lupus nephritis as a cause of proteinuria and haematuria. Single oral dose of ivermectin (200 µg/kg) was administered. One month after treatment the eosinophile count normalized and follow-up stool sample was free of larvae. Proteinuria and haematuria had resolved and other chronic morbidity remains under remission.

**Conclusion** Stronglyoidiasis stercoralis hyperinfection should be considered when differentiating between possible causes of general deterioration in multi-morbid SLE cases, presenting even in areas not considered endemic. Paraneoplastic syndrome remains a possible etiology of the observed kidney injury.

**PO.3.72 EXTRACELLULAR VESICLES OPSONIZED BY MONOMERIC C-REACTIVE PROTEIN ARE ACCESSIBLE AS AUTOANTIGENS IN SYSTEMIC LUPUS ERYTHEMATOSUS**

<sup>1</sup>J Karlsson\*, <sup>1</sup>J Wetterö, <sup>2</sup>L Potempa, <sup>3</sup>F Mobarrez, <sup>1</sup>C Sjöwall. <sup>1</sup>Linköping University ~ Linköping ~ Sweden; <sup>2</sup>Roosevelt University College of Pharmacy ~ Schaumburg, Illinois ~ USA; <sup>3</sup>Uppsala University ~ Uppsala ~ Sweden

10.1136/lupus-2022-elm2022.101

**Purpose** Under certain conditions, the pentameric (p) structure of C-reactive protein (CRP) can dissociate into the more proinflammatory isoform monomeric (m)CRP. Autoantibodies against mCRP have been reported by us and others in systemic lupus erythematosus (SLE), and their levels appear to associate with renal involvement and increased disease activity. The origin of this autoreactivity thus calls for further investigation. Extracellular vesicles (EVs) have been proposed as important mediators of lupus pathogenesis by exposing nuclear antigens on their surface, thereby serving as adjuvant for autoantibody generation and subsequent immune complex-formation. Herein, we investigated pCRP and mCRP on EVs in plasma from patients with SLE and potential associations with manifestations, disease activity and organ damage.

**Methods** A flow cytometry protocol was established to detect pCRP and mCRP on EVs in 67 well-characterized patients from a regional Swedish lupus registry, and 60 age- and sex-matched controls. Other proteins, such as surface-bound complement protein (C)3, 4, and 4d, were also measured. Clinical data was available for the patients and plasma levels of C3 and C4 were measured by nephelometry. SLE disease activity was assessed by SLEDAI-2K and organ damage evaluated by SLICC/ACR damage index (SDI).

**Results** The levels of mCRP and pCRP on EVs were higher in patients than in controls (median with (interquartile range):  $6.6 \times 10^{-4}$  ( $3.4 \times 10^{-4}$  –  $1.2 \times 10^{-3}$ ) vs  $9.3 \times 10^{-5}$  ( $6.8 \times 10^{-5}$  –  $4.7 \times 10^{-4}$ ) for mCRP, and  $5.7 \times 10^{-4}$  ( $3.5 \times 10^{-4}$  –  $1.1 \times 10^{-3}$ ) vs  $1.1 \times 10^{-4}$  ( $6.0 \times 10^{-5}$  –  $4.1 \times 10^{-4}$ ) for pCRP, respectively;  $p < 0.001$ ). mCRP was more abundant on EVs in patients with active SLE (SLEDAI-2K  $\geq 5$ ) compared to those with quiescent disease ( $p < 0.01$ ). Furthermore, mCRP surface levels correlated weakly with SLEDAI-2K ( $\rho = 0.25$ ,  $p < 0.05$ ), and even stronger with modified SLEDAI-2K ( $\rho = 0.41$ ,  $p < 0.001$ ), but levels were significantly lower in patients with established organ damage. EV-bound mCRP correlated with SDI ( $\rho = -0.30$ ,  $p < 0.05$ ), a correlation which was stronger among patients with lupus nephritis ( $\rho = -0.61$ ,  $p < 0.01$ ). Furthermore, soluble plasma complement protein 4 (C4) correlated significantly with C4d on microparticles ( $\rho = 0.27$ ,  $p < 0.05$ ), particularly in those with active SLE ( $\rho = 0.73$ ,  $p < 0.05$ ).

**Conclusions** Our results suggest that EV-bound CRP, especially mCRP, could indeed be of relevance for disease progression, and that opsonized EVs might provide a potential autoantigen source.