Image 1. Screenshot of graphical user interface of the scoring program.

Conclusion: In this presentation we outline the successful development of a tool to measure the learning curve in enthesitis. We hypothesize that knowledge about the learning curve and inter-rater reliability in enthesitis US obtained by our tool might contribute to future US curricula, structured reporting and deep learning algorithms.

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Educational cases

POS1452

DE NOVO LUPUS NEPHRITIS FOLLOWING THE INTRODUCTION OF BELIMUMAB

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Background: Belimumab inhibits the activity of the soluble cytokine BlyS (B lymphocyte stimulator) and is recommended for use in moderate refractory systemic lupus erythematosus (SLE). A recent randomised controlled trial reported that Belimumab improves the outcomes for patients with lupus nephritis when used with standard therapy.

Objectives: We present two cases of lupus nephritis that developed in SLE patients without pre-existing renal disease shortly after commencing treatment with Belimumab.

Methods: Both patients are Afro-Caribbean females with similar immunological profiles, including ANA, dsDNA, anti-Sm, anti-Ro and anti-RNP antibodies. The first was 59 years old with a long standing diagnosis of SLE since 1996 that had required previous Cyclophosphamide for neuropsychiatric lupus. She was most recently taking Hydroxychloroquine, Mycophenolate and Prednisolone having previously failed with Azathioprine twice. Belimumab was commenced in February 2020 due to worsening arthritis, mouth ulcers, pleuritis and systemic features associated with a significant rise in her dsDNA and low complement. Her SLE-DAI score was 13. After six months of treatment she developed proteinuria for the first time. Her urine protein creatinine ratio (uPCR) was measured at 205mg/mmol and a subsequent renal biopsy revealed features of active Class IV and V lupus nephritis. Belimumab was changed to Rituximab. Mycophenolate was stopped due to persistent neutropenia and Tacrolimus was introduced instead. After 5 months treatment her uPCR is now 33mg/mmol.

The second patient was 37 years old with a recent diagnosis of SLE in 2018. She presented with inflammatory arthritis, oral and nasal ulcers, and cytopenia. She responded poorly to Azathioprine and was intolerant of Mycophenolate. Her most recent treatment was Hydroxychloroquine and Prednisolone. She was commenced on Belimumab in February 2020 due to active mucocutaneous and musculoskeletal features of lupus with a SLEDAI score of 12. The mucocutaneous

features of lupus responded well but she developed proteinuria seven months later, and by November 2020 her uPCR was 149mg/mmol. Belimumab was switched to Rituximab and initially her uPCR continued to rise but it has now fallen to 43mg/mmol.

Results: The occurrence of new lupus nephritis soon after the initiation of Belimumab monotherapy has been reported previously and our cases raise further concerns. A prospective observational study recently reported significantly increased rates of new lupus nephritis developing in patients receiving Belimumab in addition to standard care compared to those receiving standard treatment. An association between Belimumab and the development of de novo lupus nephritis has not yet been conclusively established but it would create a significant challenge in how Belimumab is used and consented for in SLE, especially if it becomes more widely used to treat lupus nephritis. The mechanism by which Belimumab may increase the risk of, or trigger, lupus nephritis is currently unclear but may result from increased activity in BlyS associated pathways following the blockade of the BlyS pathway.

Conclusion: These two cases raise questions about the role of using Belimumab in patients at risk of developing lupus nephritis. We therefore recommend caution in its use and recommend active monitoring of renal parameters especially in patients with poor clinical and serological response to Belimumab.

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POS1453

OVERLAPPING FEATURES OF RELAPSING POLYCHONDROSIS AND SWEET'S SYNDROME: COULD THIS BE VEXAS (VACUOLES, UBIQUITIN A1E MUTATION, X-LINKED, AUTOINFLAMMATORY, SOMATIC) SYNDROME?

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Background: An 83-year-old male presented to the infectious diseases team with intermittent fevers associated with tension headaches, malaise and fatigue for two years, with recent worsening. He had noted a progressive decline in his mobility over the past year following replacement of his right knee for osteoarthritis. His past medical history included atopic eczema, malaria and bilateral cataract extractions.

On examination, he had a widespread non-scaly annular urticated rash. He had persistently raised inflammatory markers (CRP 40mg/L, ESR 82mm/hour), normocytic anaemia (Hb 102 g/L, MCV 101.9 fL), lymphopenia (0.70 x10⁹/L), fluctuating eosinophilia (0.88-1.25 x10⁹/L) and a mildly elevated lactate dehydrogenase (243IU/L). A thorough work up for pyrexia of unknown origin showed no evidence of infection/vasculitis/immune pathology. The only positive finding was prior exposure to schistosomiasis, treated with a single dose of praziquantel. An echocardiogram and serial PET-CT scans were normal. Given the deterioration in mobility since the right knee replacement, an MRI knee was performed, and cobalt/chromium levels to look for allergic responses to the prosthesis, but these tests did not reveal any findings of significance.

An initial haematology work-up did not identify an abnormality of concern (screening for a myeloproliferative neoplasm including BCR-ABL studies were negative and mast cell tryptase was not elevated). Due to a worsening skin rash, he was reviewed by dermatology where a skin biopsy showed features of superficial neutrophilic dermatosis which can be observed with Sweet's syndrome. However, it was felt that this was an unlikely explanation for the persistent systemic symptoms. He proceeded to a bone marrow biopsy which showed reactive features including vacuolation of myeloid precursors with normal cytogenetics.

During follow up appointments, the patient described new recurrent violaceous patches with episodes of inflammation of the pinna of the ear, suggesting a diagnosis of relapsing polychondritis and so the patient was started on high dose prednisolone (80mg per day [1mg per kg]) and referred for rheumatological assessment. He had an excellent response to prednisolone (fever, ear swelling and rash subsided). The overlapping features of relapsing polychondritis and Sweet's syndrome in an elderly man suggested a diagnosis of VEXAS (vacuoles, E1 enzyme, X-linked, autoinflammatory and somatic) syndrome. The prednisolone dose was rapidly reduced to 10mg per day and the patient was commenced on methotrexate, as a steroid-sparing agent. Further blood tests have been sent for genetic analysis for VEXAS syndrome but results are pending.

Objectives: N/A

Methods: N/A

Results: N/A

Conclusion: VEXAS syndrome is a newly identified genetically defined syndrome, described by Beck et al in October 2020 consisting of somatic mutations in the *UBA1* gene, affecting bone marrow stem cells. In a study of 25 patients with this mutation, diagnostic/ classification criteria for relapsing polychondritis (n=15), Sweet's syndrome (n=8), polyarteritis nodosa (n=3) or giant cell arteritis (n=1) were met and patients often had severe refractory disease with overlapping systemic inflammatory and haematologic features. Features of VEXAS include the presence of vacuoles in myeloid cells, somatic mutations in the *UBA1* (ubiquitin-activating enzyme) gene, X-linkage (therefore only occurring in males), in older people with autoinflammatory syndromes. Although VEXAS syndrome is a relatively rare condition, it was a relevant consideration in this case.

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HPR Measuring health (development and measurement properties of PROs, tests, devices)

POS1454-HPR AESTHETIC IMPAIRMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS: A CASE CONTROL STUDY USING A SEMI-QUANTITATIVE SCALE FOR BODY IMAGE ASSESSMENT

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Background: Systemic sclerosis (SSc) is a fibrotic autoimmune disease characterized by dermatological involvement. Skin involvement can alter appearance, which can have a psychological impact(1). Assessment of body image could be central in optimizing care. Yet, data are scarce(2).

Objectives: The main objective of our study was to assess aesthetic impairment measured on a visual aesthetic scale (AES) (3) in patients with SSc compared to a group of healthy subjects. Secondary objectives were to assess associations between aesthetic impairment and standardized questionnaires for aesthetic impairment as well as clinical, biological, psychological/quality of life, and functional parameters of SSc.

Methods: The "Sclero-esthet" study is a descriptive single-center study of two populations: patients with SSc, included in a referral center for systemic and autoimmune diseases at Lille Hospital, France, and healthy controls.

Results: This study, from January 2017 to October 2017, included 88 patients (69 [78.4%] women) with a median age of 52 years (range: 43 to 59) and 88 controls (49 [55.7%] women) with a median age of 45 years (range: 32 to 55). Perception of physical changes assessed using the AES was greater in cases than in controls (3.7 ± 0.3 vs 2.8 ± 0.3 , $p=0.028$), with statistical correlation with ASWAP. SSc patients with anxiety or depressive symptoms had significantly higher AES scores. Cases presented poorer quality of life than controls.

Conclusion: The AES appears to be a good tool to evaluate aesthetic impairment. Correlations found with psychological and quality of life parameters support the need to improve patient management in this field.

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POS1455-HPR RELATIONSHIP BETWEEN PSYCHOLOGICAL FACTORS AND MEASURES OF RHEUMATOID ARTHRITIS ACTIVITY

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Background: The patient-reported outcomes are important components of quantitative methods of rheumatoid arthritis (RA) activity assessment which are used to choose the appropriate drug therapy. The value of these parameters can be significantly affected not only by the inflammatory process, but also by the psychological characteristics of the patient and, in particular, by hardness [1].

Objectives: To study the relationship between psychological factors and signs of RA activity.

Methods: Patients with RA who met the EULAR/ACR 2010 criteria, and observed at the V.A. Nasonova Research Institute of Rheumatology were included. Clinical examination was performed including patient global assessment (PGA), physician global assessment (PhGA), pain measurement on a visual analog scale, tender joint count (TJC), swollen joint count (SJC). The functional status was determined by HAQ, the quality of life – by SF-36 EQ-5D, the nature of pain – by painDETECT, the presence of anxiety and depression – by HADS. Patients also completed Hardiness Survey questionnaire to assess hardiness (HDS) and 3 components of the HDS – commitment (CMT), control (CT) and challenge (CLN). Disease activity was evaluated with DAS28, CDAI, and RAPID3. All patients signed informed consent to participate in the study. Analysis of the data was performed using Spearman's rank test, Fisher exact test, chi-square and t-tests.

Results: 85 patients with RA were included. There were 69 women and 16 men. Mean age was 56.7 ± 13.1 years, disease duration – 7.6 ± 2.7 years. 72 patients were positive for rheumatoid factor, 75 – for anti-cyclic citrullinated peptide antibody. CDAI showed high activity in 15, moderate – in 37, low – in 30, and remission in 3 patients, DAS 28 – in 10, 55, 12, and 8, and RAPID3 – in 24, 25, 15, and 21, respectively. 24 patients had subclinically or clinically expressed anxiety and 15 – subclinically or clinically expressed depression (≥ 8 according to HADS). In 31 patients, the painDETECT questionnaire revealed possible or probable neuropathic pain. Mean HDS was 84.8 ± 21.7 , CMT – 38.9 ± 9.2 , CT – 29.4 ± 8.6 , CLN – 17.3 ± 7.1 . These values were comparable with the corresponding population data for this age group. There was a significant inverse correlation between HDS and RA activity measures, including SJC, TJC, DAS28 ($p<0.05$), pain, PGA, PhGA, CDAI, RAPID3, and HAQ ($p<0.01$). In addition, HDS and all its components positively correlated with quality of life, assessed by SF-36 and EQ-5D ($p<0.01$). In patients with subclinically and clinically expressed anxiety and depression, HDS, CMT, and CT were significantly lower than in patients without anxiety and depression ($p<0.01$), while the values of CLN in these groups did not differ significantly.

Conclusion: The results of the present study suggest that low HDS may be one of the significant factors determining RA activity level because it does not allow patients to adapt adequately to a stressful situation produced by the disease.

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