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.