Objectives: The objective of this study was to evaluate the effect of menstruation in triggering exacerbations of Behçet's disease in a Northern European cohort.

Methods: 18 female patients from our rheumatology department satisfying the International Study Group for Behçet's Disease (ISGBD) criteria were recruited. A questionnaire was conducted via telephone to determine whether their exacerbations of BD were correlated to their menstrual cycle.

Results: All 18 patients responded to the questionnaire, with the mean age of 38.8 years and mean age of menarche of 13 years. Four (22.22%) patients were in menopausal state. Half (nine) of the patients reported that their BD flare ups were correlated to their menstrual cycle. Exacerbations experienced include oral aphthosis (88.9%), arthralgia (55.6%), genital ulcerations (44.4%), lethargy (44.4%), skin lesions (11.1%) and headaches (11.1%). Six of the seven patients (86%) who were on contraception were on a progesterone containing contraception. Four out of nine (44%) who did not notice any exacerbations during menstruation stated that they were on progesterone containing contraceptives. It is noteworthy that 10 patients (55.56%) had previous pregnancies while three patients experienced an episode of miscarriage and 1 had a stillbirth.

Conclusions: Our results demonstrated that the disease activity in BD is related to the menstrual cycle, which is contributed by the female sex hormones. The study supports previous hypothesis that the abrupt decline in progesterone during onset of menstruation is associated to disease flare in BD.² Studies comprising larger cohorts should be conducted to further support and strengthen this evidence.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2133

AB1164 IMMUNOGLOBULIN USE IN GRANULOMATOUS MYOSITIS WITH BACKGROUND OF THYMOMA AND SYSTEMIC LUPUS ERYTHEMATOSUS

Y.K. Kwok, M.H. Leung. Division of Rheumatology, Department of Medicine, Queen Elizabeth Hospital, Hong Kong, Hong Kong

Background: Contracting granulomatous myositis is rare in clinical practice.

Objectives: To report a case with systemic lupus erythematosus presented with contracting granulomatous myositis and previous history of thymoma which responded to intravenous immunoglobulin (IVIG).

Methods: A case report from a local hospital in asia

Results: A forty-two year old gentleman presented with polyarthralgia, raynaud's phenomenon, photosensitivity with elevated ANA and anti-dsDNA was diagnosed to have SLE in 2014 in rheumatology clinic. His lupus activity was stable on hydroxychloroquine (HCQ) and low dose steroid for the arthritis control. In May 2015, an anterior mediastinal mass was noted on CT thorax for symptoms of chronic cough and chest pain. Operation was performed in Aug 2015 with incomplete excision with invasion into lung, pericardium and nodal metastases. Pathology showed thymoma. Adjuvant radiotherapy 60 Gys in 30frs was given and completed in Dec 2015. He had stable lupus activity all along until Aug 2017. He was hospitalised with sudden onset of proximal muscle weakness which rendered patient unable to walk. Elevated CK level was up to 5354 IU/L. There was no evidence of concurrent infection. Myositis panel showed Strongly positive anti-PM/SCL, borderline positive Anti Mi2 alpha and PM Scl 100. Dysphagia was documented by speech therapist. Muscle biopsy of right quadriceps was performed by neurologist and showed granulomatous myositis. Electromyography (EMG) showed myopathic changes. Lupus myositis was diagnosed with dysphagia. Patient was started on IVIG (0.4 gm/kg/day) for 5 days and high dose steroid (prednisolone 30 mg daily orally) was initiated concurrently. CK showed good response and normalised with improvement in the lower limb power within one month. Patient was able to walk and discharged home. However he was re-admitted one month later for generalised tightening of 4 limbs and contractures over elbows and knee joints with intense pain and limited range of movement. CK level was normal in this admission. PET-CT showed a small (2.2 × 1.4 cm SUV Max5.5) right anterior mediastinum lesion near to surgical site. It was suspicious of local recurrence. Cardiothoracic surgeon was consulted and suggested not for further surgery since of high operative risk. In view of worsening of contractures and weakness over lower limbs, despite normalisation of CK level and quiescent lupus serology,

further courses of IVIG monthly were commenced. Steroid dosage was able to titrate down to prednisolone 9 mg daily orally gradually with improvement in 4 limbs power and halting of further contractures. No more dysphagia was documented afterwards. With the incomplete excision of thymoma, further discussion with oncologist and neurologist on future treatment plan ensued. Role of IVIG in controlling the underlying autoimmune phenomenon attributed by the underlying malignancy was unknown. IVIG was stopped after 6 courses and oncologist proceeded with palliative chemotherapy.

Conclusions: IVIG use in contracting granulomatous myositis in a lupus patient with underlying thymoma may be useful.

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Acknowledgements: Division of Neurology, Department of Medicine, Queen Elizabeth Hospital

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3843

AB1165 MONOCLONAL GAMMOPATHY IN RHEUMATIC DISEASES AND ITS ROLE IN PREDICTING MALIGNANT TRANSFORMATION

Y. Yang¹, Y. Liu², L. Chen³, Y. Jia³, Z. Li³. ¹Department of Rheumatology and Immunology; ²Department of Hematology; ³Department of Rheumatology, Peking University People's Hospital, Beijing, China

Background: The monoclonal gammopathy (MG) constitutes a group of diseases characterised by the proliferation of a single clone of plasma cells or B-lymphocytes. The spectrum of conditions capable of producing MG mainly includes a premalignant state known as monoclonal gammopathy of undetermined significance (MGUS) and a group of malignant hematologic disorders. In previous studies, significantly increased risk of MG was seen in patients with a history of various rheumatic diseases. We analysed 41 hospitalised patients with underlying rheumatic diseases who were diagnosed with MG at our institute from 2010 to 2017, in order to identify clinical clues for early diagnosis, as well as the risk factors for MG and malignant hematologic neoplasias in patients with rheumatic diseases.

Objectives: To analyse the clinical spectrum, laboratory characteristics and outcomes of monoclonal gammopathy (MG) in patients with rheumatic diseases.

Methods: Screening for the presence of MG was performed in 872 inpatients with rheumatic diseases from January 2010 to July 2017. A total of 41 patients were enrolled. Their clinical and biological features in addition to outcomes were described. For each patient with primary Sjögren syndrome (pSS), 2 age- and sex-matched pSS patients without MG were selected as controls. Risk factors for the presence of MG and malignant haematological neoplasias were assessed.

Results: MG was observed in patients with various rheumatic diseases, with SS the most frequent type. Serum M protein was detected in 37 patients. M components were observed in urine in the other 4 patients. High ESR, albumin/globulin inversion, rheumatoid factor positivity, hypergammaglobulinemia, hypocomplementemia were common features, presented in more than half of the 41 patients. Patients with pSS, when complicated with MG, showed a higher rate of abnormal urine NAG (71.4% vs 15.8%, $p=0.025$), higher levels of ESR [55.0 (53.5) mm/h vs 21.0 (31.8) mm/h, $p=0.001$], ESSDAI [26.0 (25.0) vs 12.0 (9.0), $p=0.006$] and ClinESSDAI scores [24.0 (25.0) vs 10.5 (10.0), $p=0.011$]. Multivariate analysis revealed that the disease activity, assessed by either ESSDAI [adjusted OR 1.127 (95%CI 1.015–1.251), $p=0.025$] or ClinESSDAI [adjusted OR 1.121 (95%CI 1.011–1.242), $p=0.030$], was the only independent risk factor for the presence of MG. During the follow-up, 2 patients had transient serum M protein, 2 had isotype switch, 1 progressed to multiple myeloma (MM) and another 2 experienced renal injuries attributed by monoclonal or polyclonal plasma cell interstitial infiltration. Seven (17.1%) of the 41 MG patients presented haematological neoplasias, 4 with MM, 2 with smouldering multiple myeloma and 1 with B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) type. The presence of light-chain MG was associated with the development of MM [OR 17.5 (95%CI 1.551–197.435), $p=0.041$], but not with an increased risk of lymphoma or SMM.

Conclusions: MG was observed in patients with various rheumatic disorders, with SS being the most common type. The presence of MG might associated with higher disease activity. The development of haematological neoplasias including MM and lymphoma was seen in this setting. Therefore, we recommend the screening for MG and close monitoring for potential malignant transformation in patients with rheumatic diseases as needed.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7575

AB1166

THE ASSOCIATION OF THE EARLY ONSET OF REMITTING SERONEGATIVE SYMMETRICAL SYNOVITIS WITH PITTING OEDEMA (RS3PE) SYNDROME WITH DIPEPTIDYL PEPTIDASE-4 (DPP4) INHIBITOR

Y. Yoshioka, M. Mitsuhashi, A. Kato, D. Kanai, A. Suda, S. Nagaoka.
Rheumatology, Yokohama Minami Kyosai Hospital, Yokohama, Japan

Background: Remitting seronegative symmetrical synovitis with pitting oedema (RS3PE) syndrome is a rare inflammatory arthritis, characterised by absence of rheumatoid factor, symmetrical distal synovitis, pitting oedema of the hands and feet.

In recent years the use of dipeptidyl peptidase-4 (DPP4) inhibitor has increased and some reports have described the association of RS3PE syndrome with DPP4 inhibitor.

Objectives: : We have tried to investigate the association of RS3PE syndrome with DPP4 inhibitor in our hospital.

Methods: : In Japan DPP4 inhibitor was released in December 2009, so we retrospectively analysed background, treatment and clinical course of 25 patients with RS3PE syndrome diagnosed between ^{December 2009} and December 2016 in our hospital. We divided them in two groups according to DPP4 inhibitor prescription and compared two groups.

Results: : Our cases included 18 males and 7 females, and the mean age of RS3PE syndrome onset was 76 years old. The mean follow-up period was 32.5 months. Six patients had diabetes mellitus and DPP4 inhibitor was prescribed in five of six patients (83.3%). (sitagliptin 3 cases, teneligliptin 1 case, alogliptin 1 case). The duration of RS3PE syndrome onset after DPP4 inhibitor prescription was mean 22.9 months, and two cases developed within a half year, two cases after two years.

Compared with non DPP4 inhibitor group, the mean age of RS3PE syndrome onset was significantly low (70 vs 78.5, $p=0.023$), and HbA1c (NGSP) was high (7.3% vs 6.02%, $p=0.00022$) in DPP4 inhibitor group.

The occurrence of flare was four cases in non DPP4 inhibitor group and zero in DPP4 inhibitor group, but was not statistically different ($p=0.275$). Other clinical features were not significantly different.

Abstract AB1166 – Table 1. Clinical characteristics of the patients

Characteristics	DPP-4 inhibitor (N=5)	non DPP-4 inhibitor (N=20)	P value
Mean age – yr	70 (±7.91)	78.5 (±6.61)	0.023
Male sex – no.(%)	5 (100%)	13 (65%)	0.119
CRP, mg/dl	7.97 (±5.08)	7.45 (±5.86)	0.86
HbA1c (NGSP), %	7.3 (±0.68)	6.02 (±0.57)	0.00022
Medications			
MTX			
Patients, no.(%)	2 (40%)	9 (45%)	
Dose, mg/week	8 (±0)	7.33 (±1)	0.081
Oral prednisolone			
Patients, no.(%)	5 (100%)	18 (90%)	
Dose, mg/day	12 (±4.47)	10.8 (±4.62)	0.625
Malignancies – no.(%)	0 (0%)	1 (5%)	0.61
Flare – no.(%)	0 (0%)	4 (20%)	0.275

Conclusions: : DPP4 inhibitor group was significantly younger than non DPP4 inhibitor group, and the possibility that DPP4 inhibitor contributed to the early onset of RS3PE syndrome was suggested.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2829

Diagnostics and imaging procedures

AB1167

METHOD COMPARISON OF AESKUSLIDES ANCA FOR THE DIAGNOSIS OF ANCA-ASSOCIATED VASCULITIS

A. Lerner¹, T. Matthias¹, K. Prager², T. Barth², S. Reuter², J. Bär², S. Bush², M. Walle². ¹RandD, AESKU.KIPP Institute; ²RandD, Aesku. Diagnostics GmbH and Co. KG, Wendelsheim, Germany

Background: AESKUSLIDES ANCA is an indirect immunofluorescence assay used to detect anti-neutrophil cytoplasmic autoantibodies (ANCA) in human serum. This *in vitro* diagnostic assay is used as an aid for the diagnosis of ANCA-associated vasculitis (AAV) in conjunction with other clinical and laboratory findings.

Methods: A method comparison of ethanol and formalin fixed granulocytes was carried out between AESKUSLIDES ANCA (AESKU. Diagnostics) and the NOVA Lite ANCA of INOVA. 507 clinical serum samples (comprising 135 serum samples from patients with AAV and 375 samples from patients with other diseases) were analysed by standard IFA protocols. Results were obtained by manual processing and reading.

Results: In this cohort, AESKUSLIDES ANCA Ethanol slides show higher sensitivities (48.5% vs. 36.4%) and specificities (69.3% vs. 55.2%) compared to INOVA. AESKUSLIDES ANCA Formalin slides show higher sensitivities (50.0% vs. 37.9%) and similar specificities (90.7% vs 91.5%) compared to INOVA.

Conclusions: AESKUSLIDES ANCA Ethanol showed higher diagnostic sensitivity (48.5%) and specificity (69.3%) compared to the predicate assay NOVA Lite provided by INOVA (36.4%, 55.2%). This is due to the fact, that AESKU assay detects more positives in the AAV cohort, and less positives in the other disease groups. AESKUSLIDES ANCA Formalin showed a diagnostic sensitivity (50.0%) compared to the predicate assay NOVA Lite provided by INOVA (37.9%). However, the diagnostic sensitivity was comparable between the two (90.7% vs 91.5%).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6319

AB1168

IS MY CLINICAL EXAMINATION ADEQUATE TO EVALUATE DISEASE ACTIVITY IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA)? A COMPARATIVE ASSESSMENT OF CLINICAL AND ULTRASOUND (US) EXAMINATION OF 40 KNEES AND ANKLES IN 10 CHILDREN WITH JIA

A. Shivpuri, M. Agarwal, S. Sawhney. *PEDIATRIC RHEUMATOLOGY, SIR GANGA RAM HOSPITAL, NEW DELHI, India*

Background: JIA is the commonest rheumatologic disease of childhood with a quoted prevalence of 1:1000. Assessment of children with JIA includes: Clinical, laboratory and more recently US evaluation of joints. Rapid attainment of inactive disease is critical for a good long term outcome. There is a debate in the rheumatology community about added advantage of US examination over clinical assessment of joints. This study was done to compare clinical versus US evaluation of knees and ankles in children with JIA.

Objectives: 1. To clinically examine and scan knees and ankles of 10 children with JIA

2. To determine sensitivity and specificity of clinical examination of knees and ankles vis a vis US scan findings

3. To compare results of knee and ankle examination both clinical and US

Methods: 1. 10 consecutive children with JIA were evaluated for active arthritis of knees and ankles and documented on a pre designed proforma.

2. Children were then examined with US on same day by same observer and joint effusion and or synovitis was captured using an E9/S7 GE machine with a linear transducer 6–15 MHz.

Results: 10 children aged 6–16 years, (6F and 4M) who attended the out-patient paediatric rheumatology clinic at our hospital with JIA were clinically evaluated and US examination performed on same day by same examiner with prior consent. Using the standard ESSR protocols, US knee and ankle joints were examined in longitudinal view and if any effusion or synovitis detected was confirmed on transverse view. The presence of grey scale synovial proliferation or anechoic effusion was taken as a positive US finding.

Knee-Sensitivity of clinical examination-100%, specificity-91.7%.

Ankles-Clinically 8 had swollen ankle, On US only 5 children had swollen ankle (tibio talar TT) joint. 3 who had a swollen ankle clinically had no synovial hypertrophy or effusion in the TT joint but had tenosynovitis(TS) of adjacent tendons: 1 each has a TS of the Extensor Digitorum Longus, Tibialis Anterior, Tibialis Posterior. Sensitivity-60% and specificity-66.7%.