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

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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

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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.