Conclusions: Idiopathic NIHF, is a very serious condition with high fetal mortality and limited effective therapy, so prevention is very crucial. APS prophylaxis regimen, based on anti-platelets and anti-coagulant therapy during pregnancy, may have promising effects.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5798

AB1161 FAMILIAL MEDITERRANEAN FEVER AS AN OUTCOME OF UNDIFFERENTIATED ARTHRITIS

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Background: The term «undifferentiated arthritis» (UA) was proposed to emphasise the heterogeneity of unclassifiable arthritides and their potential into a definable form of arthritis. A patient with UA have an early stage of defined arthritis that will meet criteria in time, a forme fruste or partial form of a classifiable disease, an overlap of more than one disease entity, or an arthritis of unknown or in that may (or may not) become differentiated in the future. The heterogeneity associated with the term «UA» emphasises the need for continued follow-up and reassessment of the diagnosis and management of these patients.

Objectives: The aim of this study was revelation of MEFV gene mutations in patients with UA.

Methods: We have examined 80 patients (34 male, 46 female, mean age 36,4 ±3.4 years) with UA. The patients were observed every 6 months in follow-up period of 5 years. The anamnestic and treatment data were obtained. Joint disease activity scores and presence of extra-articular manifestations were determined. The CBC, urinalysis, serum concentrations of creatinine, bilirubin, transaminases, glucose, CRP were determined every 6 months, X-ray examination and ultrasonography of joints were performed once a year. Molecular-genetic analysis of 12 MEFV-mutations, common for Armenians, were carried out in Medical Genetic Centre of Armenia.

Results: From 80 investigated patients with UA 10 had repeated episodes of mono- and oligoarthritis of ankle and/or knee joints with local skin hyperemia and hyperthermia, without subsequent joint deformities, 45 – sacroiliitis (26 bilateral, 19 unilateral), accompanied by enthesopathy, 2 – joint syndrome, resembling rheumatoid arthritis. In 23 patients joint syndrome was accompanied by erythema rash, livedo reticularis, photosensitivity and alopecia. The latter group of patients was diagnosed as SLE-like syndrome. All patients didn't fulfil accepted classification criteria of any autoimmune or autoinflammatory disease. The 80% of investigated patients had no classic febrile attacks of abdominalgia and/or thoracalgia, specific for FMF. In remaining 20% of patients febrile attacks hadn't preceded joint syndrome, but appeared during 5 year follow-up period.

All investigated patients had MEFV gene mutations, which's compositions (homozygous or compound heterozygous) were enough to confirm Familial Mediterranean Fever (FMF). The most common mutations were: M694V–41.2%, V726A–18.8%, M680I–10%. The most common compositions were M694V/M694V, M694V/V726A, M694V/M680I.

Conclusions: As FMF is widely distributed in Mediterranean region, and it had changed its phenotype in last decades, as well as taking into account the increasing rate of migration worldwide, every single case of UA, which doesn't fulfil classification criteria of any disease, should be tested for presence of MEFV-mutations. The diagnosis of FMF changes the approach to follow-up, management, outcome and prognosis of UA.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3682

AB1162 DIAGNOSTIC UTILITY OF LYMPH NODE BIOPSY IN DIFFERENTIAL DIAGNOSIS OF IGG4-RELATED DISEASE, IDIOPATHIC MULTICENTRIC CASTLEMAN'S DISEASE AND PRIMARY DISSEMINATED MALT-LYMPHOMA

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Background: Lymphadenopathy (LA) is a frequent and challenging syndrome in rheumatic patients. It requires comprehensive clinical and expert pathological evaluation. IgG4-related disease (IgG4-RD) combines a group of fibroinflammatory conditions characterised by formation of tumor-like lesions with unique morphological features and hyper-IgG4 secretion in different organs and tissues. LA is frequent in IgG4-RD and doesn't have a unique morphology. Idiopathic multicentric Castleman's disease (iMCD) is a rare lymphoproliferative disorder of a hyper-IL-6 spectrum with obligate lymph nodes (LN) affection and less frequent

extranodal lesions. Pathologic features of the LN in iMCD overlap with IgG4-LA and there can be IgG4 hypersecretion in iMCD patients as well. Primary disseminated MALT-lymphomas (DMALT) are also in the spectrum of differential diagnosis because of the involvement of salivary and lacrimal glands.

Objectives: To evaluate the diagnostic utility of LN biopsy in differential diagnosis of IgG4-RD, iMCD and DMALT.

Methods: Retrospective study. We reviewed medical records from 2009 to 2017 and identified 13 pts. who were examined in our clinic due to some rheumatological diagnosis suspicion and whose leading symptom at the onset of the disease was LA. All patients had prior multiple LN biopsy (25 LN biopsies in total) which was reviewed by an expert pathologist during our examination. 11 pts. had extranodal lesions and underwent extranodal biopsy.

Results: Eight pts. were men, 5 women with average age at the onset 34,7 years (15–71 years). The directional diagnoses established on the LN pathology were as following (in some cases a few diagnosis): iMCD (4 pts), non-Hodgkin lymphoma (3 pts), reactive LN (12 pts). 11 pts. had some extranodal lesions (3,9 per patient, from 1 to 8): orbit – 8, major salivary glands – 8, hepatosplenomegaly – 5, lungs – 5, thyroid – 5, kidneys – 3, sinusitis – 3, skin, cholangitis – 2 each, retroperitoneum, mediastinum, pancreas and soft tissues – 1 each. Due to orbital and major salivary glands involvement some patients had directional diagnosis of Sjogren's syndrome or IgG4-RD. In all 13 pts. the directional diagnosis was changed to some other based on the extranodal biopsy pathology results (in 11 pts) and/or clinical presentation (in 2 pts with isolated LA). LN pathology was not conclusive in all cases. In all cases LN pathology fell into I-IV type of LN morphological picture as reported by J. Ferry et al.¹ Clinical presentation in 13 pts see in table 1. The final diagnosis were: IgG4-RD in 7 pts, iMCD in 2 pts and DMALT in 4 pts.

Abstract AB1162 – Table 1. Clinical-laboratory features of patients

	IgG4-RD (n=7)	iMCD (n=2)	DMALT (n=4)
Median age at the onset, years	26	17	49
Constitutional symptoms, N (%)	1 (14)	2 (100)	1 (25)
Orbit involvement, N (%)	4 (57)	0 (0)	4 (100)
Major salivary glands involvement, N (%)	4 (57)	0 (0)	4 (57)
Atopic, N (%)	4 (57)	0 (0)	0 (0)
Extranodal lesions, N (%)	7 (100)	1 (50)	4 (100)
Blood eosinophilia, N (%)	4 (57)	0 (0)	0 (0)
Blood thrombocytosis, N (%)	0 (0)	2 (100)	0 (0)
Elevation of IL-6 > 2N, N (%)	0 (0)	2 (100)	1 (25)
Elevation of CRP > 2N, N (%)	0 (0)	2 (100)	0 (0)
Serum IgG4 > 200 mg/dl, N (%)	7 (100)	2 (100)	0 (0)
Autoantibodies (ANA, RF, anti-SSA/Ro), N (%)	3 (43)	0 (0)	1 (25)
Hypocomplementemia, N (%)	3 (43)	0 (0)	1 (25)
Serum monoclonal secretion, N (%)	1 (14)	0 (0)	3 (75)
B-cell clonality in the tissue, N (%)	0 (0)	0 (0)	4 (100)
Tissue IgG4+/IgG+ plasma cells > 40%, N (%)	7 (100)	0 (0)	0 (0)

Conclusions: It seems to be very challenging to set a reliable differential diagnosis based on the LN pathology thus extranodal biopsy is preferable. Orbital and major salivary glands involvement is a feature of IgG4-RD or DMALT, but not iMCD. Prominent constitutional symptoms with high laboratory inflammatory markers (CRP, IL-6) are characteristic of iMCD.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7318

AB1163 MY BEHCET'S DISEASE AND MY MENSTRUATION CYCLE: OBSERVATION FROM AN IRISH COHORT

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Background: The menstrual cycle is regulated by the rise and fall of sex hormones in the body. Literature has demonstrated anti-inflammatory properties in both progesterone and oestrogen hormones.¹ There has been recent interest to determine the association between Behçet's Disease (BD), a poorly understood autoinflammatory disorder and menstruation.

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.