a common pathogenesis. New prospective studies involving large patients series are needed in this regard.

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AB1159

VIRUS-NEGATIVE LYMPHOCYTIC MYOCARDITIS: CLINICAL AND DIAGNOSTIC FEATURES FROM A MONOCENTRIC ITALIAN COHORT

S. Sartorelli, C. Campochiaro, G. De Luca, C. Candela, G. Cavalli, L. Dagna. *Unit of Rheumatology, Immunology, Allergy and Rare Diseases (UnIRAR), IRCCS San Raffaele Hospital, Milan, Italy*

Background: Virus-negative lymphocytic myocarditis (VNLM) is defined by endomyocardial biopsy (EMB) established histological, immunological and immune-histochemical criteria; it may occur as a distinct disease or in the context of systemic autoimmune or inflammatory disorders.

Objectives: To describe the demographic, clinical, histological and immune-histochemical features of VNLM from a monocentric Italian cohort.

Methods: 42 patients (mean age 45.57±14.9 years; male to female ratio 1:1) were diagnosed with EMB-proven VNLM at our Centre from January 2015 to December 2017. In all patients, comprehensive demographic, clinical and histological data were collected.

Results: The most common initial clinical feature was chest pain (40.5%), followed by palpitations (26.2%) and syncope (23.8%). Aborted sudden cardiac death (SCD) was the first manifestation in 3 cases, while arrhythmias were overall present in 47.6% of patients, being life-threatening in 10 of them. Interestingly, 4 patients had only few constitutional symptoms and 2 patients were completely asymptomatic. The distribution of traditional cardiovascular risk factor reflected that of the general population, apart for a more common familiarity for SCD (31.7%) and for autoimmunity (31.7%). Serum levels of troponin T and NT-proBNP were increased in 40.5% and 30.9%, respectively. Both echocardiography and standard ECG were unremarkable in half of the patients, while nearly all patients (92.5%) had at least one Lake-Louise criterion at cardiac magnetic resonance (CMR) evaluation. The most common CMR finding was delayed enhancement in 90% of cases, while T2-oedema was found in 21 patients (50%). Left ventricular ejection fraction was reduced in 50% of patients; a concomitant pericardial effusion was detected in 22.5% of cases. Abnormalities on 24h-ECG-Holter tape were overall detectable in 20 patients (47.6%), with ventricular ectopic beats and non-sustained ventricular tachycardia being the most common findings.

Despite positivity for ANA in 42.8% patients, only 4 patients could be diagnosed with a systemic autoimmune disease. Anti-heart antibodies (AHA) and anti-intercalated disks antibodies (AIDA) were positive in 21 patients (50%) and 12 (28.6%) patients, respectively.

On EMB, myocarditis was classified as active in 23 cases (54.8%) and as chronic in 18 (42.3%), while 7 patients (16.7%) had evidence of both features. CD3 +T-lymphocytes>7/mm² were detectable in 27 patients (64.3%), necrosis in 20 patients (47.6%), oedema in 28 patients (66.7%), while only 4 patients showed signs of vasculitis or thrombotic microangiopathy. At time of diagnosis, myocardial fibrosis was evident in 73.8% of EMBs and dilated cardiomyopathy in 6 patients (14.3%). All patients were treated with steroids and azathioprine as first line therapy, and 17 patients (40.5%) were initially referred for device implantation.

Conclusions: VNLM is an overlooked disease characterised by a broad spectrum of clinical features and peculiar immune-mediated hallmarks. The early recognition of myocarditis, allowing a prompt therapeutic intervention, should be a major goal for rheumatologists.

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AB1160

A SYNDROME OF RECURRENT IDIOPATHIC HYDROPS FETALIS, RESPONDING TO ANTI-PLATELETS/ANTI-COAGULANT PROPHYLAXIS. IS IT A NEW ENTITY OR A PART OF MATERNAL HYPERCOAGULATION STATE; THROMBOPHILIA OR ANTI-PHOSPHOLIPID SYNDROME (APS)?

S. Aamar. *Hadassah Mount Scopus, Hebrew University Medical School, Jerusalem, Israel*

Background: Anti-phospholipid (aPL) syndrome (APS) is defined in the presence of anticardiolipin (aCL), anti-β2 glycoprotein-I antibodies, or lupus anticoagulant (LAC) with hypercoagulability, including arterial/venous thrombosis episode (s), or pregnancy morbidity; early spontaneous abortions, stillbirth or prematurity with (pre)eclampsia. Beside these classification criteria the increased awareness for seronegative APS and non-criteria clinical manifestations, necessitate additional laboratory diagnostics for aPL's. Use of preventive treatment protocol; low-dose (100 mg/d) aspirin (ASA) and/or daily subcutaneous (SC) heparin, is highly effective and improves fetal vitality and pregnancy outcome. Beside APS, stillbirth and fetal loss may result from thrombophilia and Hydrops fetalis (HF). HF is described as fetal pathological fluid accumulation in serous cavities and soft tissues. It is accompanied with placental thickening and hydramnios. Most of the cases refer to non-immune (NIHF), not caused by red cell alloimmunization. Almost third of NIHF are idiopathic (iNIHF). The pathogenesis of tissue hypoxia with capillary leakage remains unclear. With advanced in-utero therapy mortality is still high (50%–95%).

Objectives: To describe the entity of recurrent idiopathic NIHF, resulting in habitual miscarriages. To document the use of APS prophylactic regimen for fetal loss, and its effect on iNIHF occurrence.

Methods: Data from medical files of women with previous iNIHF, who were treated in the rheumatology clinic, Hadassah Mount Scopus Hospital in Jerusalem, between years 2002–2017 were summarised (table 1).

Results: The present series illustrates the impact of the prophylactic regimen of APS in preventing obstetrical morbidities, including miscarriages and fetal death due to Hydrops fetalis (table 1). Thrombophilia and aPL profiles were normal. Five women who had multiple early abortions and 8 pregnancies with iNIHF, following treatment had a total of 12 successful pregnancies with uneventful delivery to healthy babies.

ANA:anti nuclear antibodies, IUFD:intra uterine fetal death, IVF:in vitro fertilisation, CLX:Clexan=Enoxaparin (SC 40 mg/d), HCQ:Hydroxy-Chloroquine (200 mgx 2/d), PRD:Prednisone (10–20 mg/d)

Abstract AB1160 – table 1. summary of cases

patient	Age (yrs)	Pregnancies before prophylaxis	Outcome (mother age, outcome, gestation age wks)	Treatment (Prophylaxis during pregnancy)	Pregnancies on prophylaxis (maternal age)	Outcome	Positive findings
1	38	10(x7 IVF)	x1(21, preterm, 30), x2(22, 23, iNIHF/IUFD, 31), x7 abortions,<10)	CLX, ASA, HCQ	x3(25, 27, 29 Spontaneous (no IVF), on HCQ, PRD 5 mg/d	Uneventful	ANA+, aCL+, LAC+, Raynaud's, Livedo Reticularis
2	43	2	x2(20, 21 IUFD/iNIHF, 27,23)	ASA, PRD	x3(23, 26, 29	1-mild HF, live birth 34 wks. 2+3 uneventful	Migraine, low platelets
3	31	2	x1(22, abortion, 8), x1(23 IUFD/ iNIHF, 24)	ASA, CLX	x2(25, 27	Uneventful	None
4	27	3	x1(20, IUFD/iNIHF, 25), x1(21, abortion,<10), x1(22 IUFD/iNIHF, 26)	ASA, CLX, PRD	x2(23, 25	Uneventful	Raynaud's, Alopecia
5	48	4	x1(25 normal, term), x2(26, 27, abortions,<10), x1(29, IUFD/iNIHF, 30)	CLX 60 mg/d	x2(30, 33	Uneventful	None

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

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AB1161 FAMILIAL MEDITERRANEAN FEVER AS AN OUTCOME OF UNDIFFERENTIATED ARTHRITIS

V. Vardanyan¹, K. Ginosyan¹, A. Simonyan¹, S. Bakhshyan². ¹Internal Medicine, Yerevan State Medical University; ²"Cito" Medical Centre, Yerevan, Armenia

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.

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

V. Vasilyev¹, E. Sokol¹, S. Palshina¹, B. Chaltsev¹, N. Probatova², A. Pavlovskaya², N. Kokosadze², A. Kovrigina³, T. Safonova¹, I. Gayduk⁴. ¹Nasonova Research Institute of Rheumatology, Moscow, Russia; ²N.N. Blokhin Russian Cancer Research Center; ³National Hematology Research Center; ⁴Moscow State University of Medicine and Dentistry, Moscow, Russian Federation

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

W.L. Ng^{1,2}, F. Adeeb^{1,2}, A. Sebastian^{1,2}, A. Anjum^{1,2}, M. Brady^{1,2}, M. Gillespie¹, F. Irwin², B. McCarthy², J. Devlin^{1,2}, A. Fraser^{1,2}. ¹Rheumatology, University Hospital Limerick; ²Rheumatology, Croom Orthopaedic Hospital, Limerick, Ireland

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.