ESC criteria was performed. Presence of HFpEF was assumed if patients had NTproBNP >125 pg/mL, and either left ventricular hypertrophy (LVH; averaged septal-posterior wall thickness >11 mm) or left atrial dilation (LAD; LA Diameter >43 mm), regardless of presence of dyspnea. Kaplan-Meier plots were generated, and hazard ratios (HR) with 95% confidence intervals were computed using Cox regression with adjustment for age.

Results: Out of 764 patients (mean age 51 years, 70% female) 46% had rheumatoid arthritis (RA), 34% systemic autoimmune diseases (SAI; connective tissue disease or vasculitis), and 20% spondylo-arthritis (SpA); 248 of these patients (mean age 61±13 years, 62% female, RA 46%; SAI 32%; SpA 22%) had valid echocardiographic data (4 patients with LV ejection fraction <50% were excluded; follow-up data was missing in 3 cases). After a median follow-up time of 5.4 years, 20.6% of patients (group 1–4: n=19/5/8/1 respectively) had died or suffered a CV event (myocardial infarction 4.1%; stroke 1.8%; decompensated HF 1.8%; resuscitation 0.9%).

In univariable analysis NTproBNP >125 pg/ml (HR 3.6; 1.9–6.8, $p=0.0001$), LAD or LVH (HR 2.3; 1.1–4.5, $p=0.02$), and age per 5 years (HR 1.4, 1.2–1.6, $p<0.0001$) were significant predictors for an increased risk for death or CV event. Compared to the referent group consisting of patients with no signs of LVH or LAD in the presence of normal NTproBNP (group 4; n=59, 23.8%), patients with echocardiographic criteria for HFpEF (group 1; n=84, 33.9%) had a 7-fold increased risk for death or CV event: HR 7.2 (1.6–31.9; figure). The event risk for both patients with elevated NTproBNP but absent LVH or LAD (group 2; n=49, 19.8%) as patients with normal NTproBNP but presence of LVH or LAD (group 3; n=56, 22.6%) was also 5- to 6-fold increased: HR 5.5 (1.2–25.8) and 5.5 (0.9–18.2), respectively.

Conclusions: In patients with RD with an increased baseline CV risk, echocardiographic criteria suggestive of HFpEF are highly relevant indicators of worse outcome (7-fold increased risk for death or CVE), in particular in conjunction with an elevated NTproBNP value, irrespective of clinical presentation.

REFERENCE:

- Breunig, M., S. Kleinert, et al. (2017). "Simple screening tools predict death and cardiovascular events in patients with rheumatic disease." *Scand J Rheumatol*:1–8.

Acknowledgements: This study was supported by the Competence Network Heart Failure Germany (BMBF grant 01 GI0205/01GI1202A) and the Comprehensive Heart Failure Centre Würzburg (BMBF grant 01EO1004).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4875

AB1345

EVALUATION OF MACROPHAGE ACTIVATION SYNDROME IN HOSPITALISED FEBRILE PATIENTS WITH KIKUCHI-FUJIMOTO DISEASE BASED ON THE 2016 EULAR/ACR/PRINTO CLASSIFICATION CRITERIA

S.S. Ahn, H.J. Kim, H.W. Kim, S.M. Jung, S.-W. Lee, M.-C. Park, Y.-B. Park, J. J. Song. *Division of Rheumatology, Department of Internal Medicine, Yonsei University College of Medicine, Seoul, Korea, Republic of Ireland*

Background: The incidence and the risk of MAS in Kikuchi-Fujimoto disease (KFD) have been poorly understood, probably because of the rarity of the disease.

Objectives: To evaluate the 2016 classification criteria for MAS in hospitalised febrile patients with KFD and compare the clinical features of MAS in KFD with those of systemic lupus erythematosus (SLE) and adult-onset Still's disease (AOSD).

Methods: The records of febrile patients with KFD, hospitalised between November 2005 and April 2017, were reviewed. Patients were evaluated for MAS using the 2016 classification criteria. Clinical and laboratory features between patients with KFD with and without MAS, as well as those of patients with MAS in KFD, AOSD, and SLE were compared.

Results: Among 78 patients hospitalised with KFD, 24 had MAS during admission. Patients with MAS had longer hospital stays than patients without MAS and only patients with MAS required intensive unit care and experienced in-hospital mortality. Patients with MAS had higher C-reactive protein, aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase, and ferritin levels and lower lymphocyte count, platelet count, and total protein and albumin levels. MAS patients with KFD, AOSD, and SLE exhibited different clinical characteristics

according to the underlying disease; however, MAS patients with KFD required less intensive immunosuppressive treatment than those with AOSD and SLE.

Abstract AB1345 – Table 2. Comparison of baseline laboratory findings of patients with MAS and without MAS

Variable	Without MAS (n=54)	With MAS (n=24)	p-value
General laboratory variables			
WBC count (/μL)	3475.0 (2350.0–5000.0)	3285.0 (2335.0–5905.0)	0.996
Haemoglobin (g/dL)	12.5 (11.7–13.4)	11.7 (10.3–13.2)	0.102
Neutrophil count (/μL)	1925.0 (1310.0–3050.0)	2206.5 (1375.0–4833.5)	0.378
Lymphocyte count (/μL)	1045.0 (860.0–1370.0)	600.0 (490.0–940.0)	<0.001
ESR (mm/h)	48.5 (27.0–64.0)	52.5 (39.5–72.5)	0.242
CRP (mg/L)	15.0 (7.5–37.1)	52.6 (27.7–97.1)	<0.001
Creatinine (mg/dL)	0.7 (0.6–0.8)	0.7 (0.5–1.1)	0.349
ALT (IU/L)	21.5 (14.0–38.0)	44.5 (23.0–153.0)	0.003
Albumin (mg/dL)	4.0 (3.7–4.3)	3.4 (3.1–3.7)	<0.001
LDH (IU/L)	439.5 (291.0–611.0)	771.5 (460.0–1213.5)	0.003
Laboratory variables selected for MAS classification			
Platelet count (× 1000/μL)	195.5 (155.0–260.0)	139.0 (94.5–184.0)	<0.001
AST (IU/L)	29.0 (22.0–39.0)	93.5 (62.5–191.0)	<0.001
Ferritin (ng/mL)	242.8 (112.0–405.2)	1449.7 (812.9–4463.1)	<0.001
Triglyceride (mg/dL) [†]	89.0 (60.0–106.5)	133.0 (90.3–161.3)	0.076
Fibrinogen (mg/dL) [§]	342.0 (338.0–346.0)	317.5 (227.5–401.0)	0.695
Bone marrow biopsy findings*			
Presence of hemophagocytosis	0/7 (0.0)	2/12 (16.7)	0.509

Data are expressed as median (interquartile range) or n (%).

ALT, alanine aminotransferase; AST, aspartate aminotransferase; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; LDH, lactate dehydrogenase; MAS, macrophage activation syndrome; WBC, white blood cell

Abstract AB1345 – Figure 1. Comparison of HScore and the probability of having hemophagocytic syndrome in patients with KFD with and without MAS

Comparison of HScore (A) and the probability of having hemophagocytic syndrome (B) in KFD patients with MAS and without MAS

HScore, hemophagocytic syndrome score; KFD, Kikuchi-Fujimoto disease; MAS, macrophage activation syndrome.

Conclusions: The 2016 classification criteria were useful in identifying KFD patients with poor clinical outcome. Although treatment for patients with MAS in KFD, AOSD, and SLE differed according to the underlying disease, the 2016 classification criteria for MAS might aid in stratifying high-risk patients and provide guidance for treatment.

REFERENCES:

- International journal of dermatology. 2016;55(10):1069–75.
- Ann Rheum Dis. 2016;75(3):481–9.

Acknowledgements: None.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5007