Methods: The charts of 3335 BS pts who were registered between 2000 and 2010 were reviewed retrospectively. Pts who fulfilled ISG criteria for BS after 40 years of age were defined as older onset, while those who fulfilled the criteria before 30 as classic onset. For each older onset chart, 2 consecutively registered early onset charts were selected. Only clinical manifestations at presentation were recorded. A clinical activity index (1) was modified and calculated.

Results: There were only 134(70 M/64 F) pts with older onset BS, which gave a prevalence of 4% in the whole cohort. Age of onset was 40-44 years of age in 54 pts, 45-49 years in 47 and 50+ in the remaining 32. As controls 268(163 M/105 F) classic onset pts were selected. Demographic and clinical characteristics among older and classic onset pts are described for males and females separately, in Table. The frequency of skin manifestations, arthritis and eye disease as well as the mean clinical activity scores were significantly higher among male classic onset pts compared to older onset male pts. Interestingly, the frequency of those with positive pathergy test, vascular involvement and severe eye involvement did not seem to be different among older and classic onset male pts. On the other hand, clinical characteristics and total activity scores were similar between older and classic onset groups among females (Table). The main limitation is that the information was based solely on patient's charts and outcome information was not available.

Conclusion: Compared to classic onset pts, males tend to be less frequent in older cohort. At presentation, older onset male pts had significantly less frequent skin, joint, eye disease, and significantly lower total activity scores compared with classic onset pts. There was no difference between the classic and older onset group, among females.

Table. Clinical characteristics of older age-onset and classical-onset BS pts (Males/Females)

	Older onset males (n=70)	Classical onset males (n=163)	P	Older onset females (n=64)	Classical onset females (n=105)	P
Age at ISG criteria fulfilling, yrs	47.0 ± 6.0	24.2 ± 4.5		46.6 ± 5.7	23.1 ± 4.4	
Oral ulcer, n (%)	69 (99)	163 (100)	-	64 (100)	105 (100)	-
Genital ulcer, n (%)	60 (86)	137 (84)	0.845	51 (80)	90 (86)	0.307
Papulopustular lesions, n (%)	49 (70)	133 (82)	0.050	39 (61)	71 (68)	0.377
Nodular lesions, n (%)	15 (21)	76 (47)	0.0001	29 (45)	60 (57)	0.135
Arthritis, n (%)	10 (14)	45 (28)	0.028	13 (20)	25 (24)	0.597
Eye involvement, n (%)	24 (34)	88 (54)	0.006	26 (41)	41 (39)	0.839
Visual acuity: <0.1 on either or both eye	8 (33)	27 (33)	0.941	4 (15)	9 (23)	0.538
Vascular involvement, n (%)	13 (19)	27 (17)	0.710	2	0	0.142
Pathergy positivity, n (%)	39 (56)	94 (58)	0.876	34 (53)	49 (47)	0.346
Activity score, mean ± SD	5.60 ± 3.36	6.38 ± 3.15	0.020	5.20 ± 2.21	4.79 ± 2.39	0.111

*: Pathergy test was made only in 70 older onset and 163 younger onset male pts, 64 older onset and 105 younger onset female pts.

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Disclosure of Interests: Gul Guzelant Ozkose: None declared, Yilmaz Ozyazgan Speakers bureau: ABBVIE, Cem Mat: None declared, Vedat Hamuryudan Consultant for: Abbvie, Amgen, BMS, Jansen, MSD, Pfizer, UCB, Speakers bureau: Abbvie, Amgen, BMS, Jansen, MSD, Pfizer, UCB, Hasan Yazici: None declared, Emire Seyahi: None declared

DOI: 10.1136/annrheumdis-2019-eular.4386

FRI0593

PATHOLOGICAL FEATURES OF LYMPH NODE IN JAPANESE PATIENTS WITH ADULT-ONSET STILL'S DISEASE

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Background: Adult-onset Still's disease (AOSD) rarely occurs in adults and no specific serological or pathological findings have ever been reported. Although multiple lymphadenopathy often develops in AOSD patients, only few reports have described their pathology so far.^{1,2}

Objectives: To examine pathological features of lymph node (LN) in AOSD patients and their usefulness in diagnosis in Japan.

Methods: This is a single center, retrospective study. The subjects were 115 Japanese patients who visited our department and received LN biopsy from April 2010 to July 2018. Of these, 12 patients, in whom AOSD was suspected by clinicians at the time of biopsy, were analyzed. AOSD patients were diagnosed according to the Yamaguchi's criteria³, except the item "excluding malignant lymphoma".

Results: The median age of the 12 cases (2 males, 10 females) was 60.5 years old (range: 18-81). Paracortical hyperplasia was shown in 7, follicular hyperplasia in 1, histiocytic necrotizing lymphadenitis (HNL) in 3, and diffuse large B cell lymphoma (DLBCL) in 1. Of 9 who fulfilled Yamaguchi's criteria, TAFRO syndrome was diagnosed in one. As for the other 8, diagnosis of AOSD was made and successfully treated as AOSD. Although 1 of the 8 had pathological features of HNL, we treated her as AOSD because her clinical feature and laboratory data were not compatible with HNL. As for the 3 who did not meet the criteria, their clinicopathological diagnosis was HNL in 2 and DLBCL in 1. We could find no associations between the clinical features and lymph node patterns in AOSD as previously reported.

Conclusion: In patient who fulfilled the criteria, most cases showed paracortical hyperplasia in LN as previously reported from outside Japan and no cases had malignant lymphoma. However, in diagnosing AOSD, LN biopsy is needed to exclude AOSD-mimicking malignant lymphoma especially in the case that does not fulfill the criteria.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2770

FRI0594

COMPARISON OF CLINICAL FEATURES BETWEEN HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS COMPLICATED WITH RHEUMATIC DISEASES OR TUMOR

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Background: Hemophagocytic lymphohistiocytosis (HLH) covers a wide spectrum of related life-threatening conditions featuring ineffective immunity characterized by an uncontrolled hyperinflammatory response, which including autosomal recessive familial HLH (FHL), familial erythrophagocytic lymphohistiocytosis, viral-associated hemophagocytic syndrome, and autoimmune-associated macrophage activation syndrome (MAS).

Objectives: Our study was to compare the difference between HLH related with rheumatic diseases or tumor.

Methods: We retrospectively reviewed the patient data with definite bone marrow hemophagocytosis in the inpatient department of the Third Affiliated Hospital of Sun Yat-sen University between 01/11/2011 to 31/05/2018. The clinical manifestations, laboratory exam and prognosis were compared between patients related with tumor and rheumatic diseases.

Results: 7 patients with rheumatic diseases and 5 patients with tumor were enrolled. In patients with rheumatic diseases, 6 with adult-onset Still's disease and 1 with SLE. In patients with tumor, 4 with lymphoma and 1 with undefined tumor. The clinical features were compared between the two types of patients as below.