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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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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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.

Table 1. Comparing of clinical features between patients with HLH complicated with rheumatic diseases or tumor

Clinical features	Patients with rheumatic diseases (n=7)	Patients with tumor (n=5)
Age (mean)	17-66 yrs (38.7)	29-59 (47.8)
Male/Female	1/6	3/2
Fever	100%	100%
Cytopenias (≥ 2 blood cell lines)	85.7%	100%
Abnormal liver function tests	71.4%	100%
Hypofibrinogenemia	28.6%	100%
Hypertriglyceridemia	85.7%	80%
Hyperferritinemia	100% (4/4)	100%
Increased lactate dehydrogenase	83.3% (5/6)	80%
Lung involvement	57.1%	100%
Splenomegaly/Hepatomegaly	85.7%	75% (3/4)
Fulfill the 2004 HLH criteria	71.4%	100%
Treatment Corticosteroid	100%	100%
Cyclosporine	42.9%	0
Etoposide (VP-16)	14.3%	40%
Cyclophosphamide	14.3%	20%
Prognosis	71.4%	40%
Control during inpatient period	28.6%	60%
Dead or give up		

Conclusion: Clinical features of HLH patients complicated with tumor or rheumatic diseases were comparable in many aspects. Yet patients with tumor have more hypofibrinogenemia and lung involvement, and tend to be treated with VP-16 and Chemical therapy, and have much worse prognosis.

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FRI0595

MAY CANCER RISK BE DECREASED IN A LARGE COHORT OF CHILD AND ADULT FAMILIAL MEDITERRANEAN FEVER PATIENTS?

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Background: Recent studies suggested that the cancer incidence may be lower among FMF patients compared with healthy controls.

Objectives: We have assessed cancer risk among FMF patients with or without amyloidosis to confirm this hypothesis.

Methods: We studied FMF patients, diagnosed and followed-up in Hacettepe University hospitals. Patients were identified from the hospital registry by using the ICD-10 code for FMF. Presence of amyloidosis was also noted. Data related to cancer were collected from patient files and oncology hospital registries. We calculated age- and gender-specific standardized incidence rates according to the Turkish National Cancer Registry data at 2014.

Results: Total of 3899 FMF patients (120 patients had also amyloidosis) were included. Median age was 22 and 56% were females. Thirty-eight (0.1%) patients had cancer during the study period. Type and distribution of cancers were as follow; females: 7 breast, 5 lymphoma, 4 ovarian, 2 renal, 2 central nervous system, 2 connective tissue sarcoma, 1 lung, 1 colorectal, 1 gastric, 1 endometrium, 1 leukemia and 1 yolk sac cancer; males: 2 leukemia, 1 prostate, 1 colorectal, 1 renal, 1 pancreas, 1 lymphoma, 1 Kaposi sarcoma and 2 unclassified adenocarcinoma. The overall cancer risk among patients with FMF was significantly lower in both males (SIR 0.42 (95% CI 0.21-0.75), p=0.002) and females (SIR 0.65 (95% CI 0.44-0.93), p=0.019). The overall cancer risk among patients with FMF and amyloidosis was (SIR 1.21 (95% CI 0.49-2.52), p = 0.73) without gender difference. Although the overall cancer risk seems decreased, an increased risk was found among female patients under 20 years-old (SIR 3.44 (95% CI 1.26-7.64), p = 0.003), however, there was

no predominant type of cancer in this group. Detailed SIR values according to sex and age groups are given in **Table 1**.

Conclusion: Cancer incidence was significantly lower in FMF patients, confirming the results of a previous study. However, we found an increased cancer risk among female patients under 20 years-old. Further prospective studies especially in this age group are needed to confirm this association. If so, early cancer screening may be considered in this particular patient group. Additionally, we found no increased cancer risk in FMF patients having amyloidosis. Possible underlying mechanisms need to be explained.

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. SIR values for different age cut-offs in both sexes

Gender	Age	Observed/Expected Cancer Cases	SIR	95% Confidence interval	P value
Female	<20- years	5/1.45	3.44	1.26-7.64	0.003
	≥50- years	17/24.8	0.68	0.41-1.075	0.10
	Overall	28/43.08	0.65	0.44-0.93	0.019
Male	<20- years	3/1.83	1.64	0.42-4.46	0.55
	≥50- years	6/15.17	0.39	0.16-0.82	0.009
	Overall	10/23.9	0.42	0.21-0.75	0.002
Amyloidosis	Overall	6/4.9	1.21	0.49-2.52	0.73

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FRI0596

AZATHIOPRINE AND GLUCOCORTICOID COMBINATION MIGHT BE A GOOD TREATMENT OPTION TO ACHIEVE REMISSION IN PATIENTS WITH IGG4-RELATED DISEASE

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Background: IgG4-related disease is a recently recognised inflammatory disease of unknown etiology, often seen in men over the age of 50 and may affect many organs and systems with elevated serum IgG4 levels and typical histopathological features.

Objectives: The aim of this study is to determine the demographic and clinical characteristics of patients with IgG4-related disease.

Methods: Patients diagnosed as having IgG4-related disease by their typical histopathological findings and imaging features and/or increased serum IgG4 concentrations (> 135 mg/dl) from two university hospital in İzmir were included in the study.

Results: There were 53 patients with a mean age of 51.49 yrs (69.8% male). The most common involvement was retroperitoneal fibrosis (54.7%), followed by the cardiovascular system (CVS) involvement (45.3%) (Table 1). While 22 patients had at least two organ involvement, the most common association was retroperitoneal fibrosis and CVS involvement (15 patients). Serum IgG4 levels were studied in 36 patients (67.9%) and found to be higher levels in 20 patients. (55.5%) (Table 2). In 44 patients (83%), acute phase reactants (APRs) were increased at the time of the diagnosis. There was no correlation between the extent of the disease and serum IgG4 levels and initial erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) values. 28 patients (52.8%) were diagnosed by imaging, 9 (17%) by imaging and IgG4 elevation, 5 (9.4%) by imaging and histopathology, 10 (18.9%) by imaging, histopathology and IgG4 elevation and 1 patient (1.9%) by only histopathology.