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.

REFERENCES:

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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 adenocancer. 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 065 (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 comfirm 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.

REFERENCES:

. 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 unkown 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 Ig-G4-related disease by their typical histopathological findings and imaging features and/or increased serum IgG4 concentrations (> 135 mg/dl) from two university hospital in Izmir 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.