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**ABSTRACT BOOK**

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## FREQUENCY OF GENETIC MUTATIONS IN CHRONIC LYMPHOCYTIC LEUKAEMIA PATIENTS AND THEIR EFFECT ON SURVIVAL

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Presentation Type Poster

Abstract Category Adult Hematology Abstract Categories-> Chronic Lymphocytic Leukemia

**Introduction and Objective:** Chronic lymphocytic leukaemia (CLL) is the most common type of leukaemia in western world countries. In addition to clinical variables such as gender, age and Rai/Binet stage, cytogenetic abnormalities are among the prognostic factors. In this study, we aimed to investigate whether the frequency of cytogenetic abnormalities in CLL patients followed up in our clinic and their effects on prognosis are compatible with the literature.

**Materials and Methods:** Data of 100 patients diagnosed with CLL who were older than 18 years and younger than 80 years of age and followed up in Erciyes University Faculty of Medicine, Hematology-Oncology Hospital between January 2010 and January 2022 were included retrospectively. The frequency of cytogenetic abnormalities and their effects on prognosis were evaluated by statistical analyses.

**Results:** Of the 100 patients included in the study, 30 (30%) were female and 70 (70%) were male. The mean age of all patients was 67.11±9.47 years and the mean age at diagnosis was 61.32±10.13 years. It was observed that 56% of the patients were positive for any cytogenetic abnormality. The most common cytogenetic abnormality was 13q14 deletion. 13q14 deletion was found to be positive in 40.3% (n:31) of the patients. 17p deletion was found positive in 17% (n:17), 11q deletion in 15.4% (n:10), and trisomy 12 in 12.5% (n:7) of the patients. Some of the characteristics of patients with 17p and 11q deletions, which were evaluated to be associated with poor prognosis, are given in the table (Table 1,2).

**Conclusion:** According to our findings, the presence of cytogenetic abnormalities has an important role in disease risk determination in accordance with the literature. Therefore, cytogenetic anomaly screening should be performed in every patient before treatment. This screening is important in terms of determining the prognosis, stage and treatment options.

Feature		17p deletion Positive (n:100)		17p deletion Negative (n:100)		p
		n	%	n	%	
Survival	Living	10	13.5	64	86.5	0,136**
	Not Living	7	26.9	19	73.1	
Disease process	Progressed	10	21.7	36	78.3	0,244*
	Not progressing	7	13.0	47	87.0	
Stage	Stage 0-1-2	13	14.4	77	85.6	0,064**
	Stage 3-4	4	40.0	6	60.0	

Feature		11q deletion Positive (n:65)		11q deletion Negative (n:65)		p
		n	%	n	%	
Survival	Living	7	13.5	45	86.5	0,405*
	Not Living	3	23.1	10	76.9	
Disease process	Progressed	8	27.6	21	72.4	0,019*
	Not progressing	2	5.6	34	94.4	
Stage	Stage 0-1-2	8	13.8	50	86.2	0,292*
	Stage 3-4	2	28.6	5	71.4	

Table 1: 11q Deletion and Some Characteristics of Patients

Table 2: 17p Deletion and Some Characteristics of Patients

## GRANULOCYTE TRANSFUSION ACCELERATES RECOVERY FROM NEUTROPENIA IN PATIENTS WITH HEMATOLOGIC MALIGNANCIES

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**Introduction:** Neutropenia is the most common and serious consequence of myelosuppressive chemotherapy in patients with hematologic malignancies. The complication of febrile neutropenia, which usually requires hospitalization and empirical wide-spectrum antibiotics, may develop. It often leads to the need for dose reduction or treatment delays, potentially jeopardizing the course of treatment (1).

Granulocyte transfusions can restore granulocyte counts (2) and thus theoretically reduce the risk of infection in such patients. However, it remains unclear whether granulocyte transfusions can improve outcomes in neutropenic patients with chronic infections. We aimed to contribute to this uncertainty in the literature with our study.

In our study, we aimed to demonstrate the efficacy of granulocyte transfusion in neutropenic patients with hematologic malignancy despite recombinant myeloid growth factor therapy.

**Methods:** In this retrospective study, 72 patients who were treated in our hematology clinic between 2016 and 2022 and who met the criteria of our study were included.

Demographic data, hematologic malignancy subtypes, chemotherapy regimens, types of antimicrobial therapy, number of neutropenic days, clinical outcome before and after granulocyte transfusion, and neutrophil count changes in blood parameters were analyzed.

Those with missing information obtained from the patient information management system, those who refused treatment, those who could not find a donor for granulocyte transfusion, and those who died before receiving granulocyte treatment were excluded from the study.

In the study, p-values less than 0.05 were considered significant. The analyses were analyzed with the SPSS 25.0 program.

**Results:** In our study, 56.9% of the patients were male, the most common diagnosis was AML with 65.3%, and the most common CT protocol was high-intensity CT protocol with 61.1%. Regarding the types of antimicrobial treatment received by the patients, 91.7% Gram-/+ was the most common type of treatment. It was observed that 62.5% of the patients recovered from neutropenia after granulocyte transfusion and 37.5% did not recover or exited (Table 1).

It was observed that patients who were neutropenic before chemotherapy were more likely to recover from neutropenia after granulocyte transfusion (p=0.01) and had lower rates of recovery from neutropenia (p=0.04).

It was observed that the time-dependent changes in neutrophil counts of the group emerging from neutropenia after granulocyte transfusion were different. In the study, it was observed that neutrophil counts on day 3, day 2; day 2, and day 1 levels increased significantly compared to the baseline (p=0.01) (Table 2).

Patients with AML had a lower rate of recovery from neutropenia after granulocyte transfusion (p=0.03).

**Conclusions:** Considering the present results, granulocyte transfusion seems to accelerate the recovery from neutropenia in the sample we analyzed. In addition, the diagnosis of the patient, the type of chemotherapy received, and the time of granulocyte transfusion were evaluated as factors affecting the results. However, in light of the data obtained, we believe that prospective studies with a larger number of patients should be conducted to evaluate the consistency of our results.

**Keywords:** chemotherapy; granulocytes; leukemia; malignancy; neutropenia

## CORRELATION BETWEEN 18F-FDG PET/CT SEMIQUANTITATIVE PARAMETERS AND Ki-67 EXPRESSION IN DIFFUSE LARGE CELL LYMPHOMA

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**Presentation Type** Oral

**Abstract Category** Adult Hematology Abstract Categories → Aggressive B-Cell Lymphoma

### Objective

Positron emission tomography (PET) is widely used in staging and evaluating response to treatment in lymphomas. In FDG-PET imaging, the glucose utilization rate in tissues is determined. In FDG-PET images, malignant tissues that use excess glucose appear as areas of high contrast compared to normal tissues. Diffuse large B-cell lymphoma (DLBCL) is the most common type of lymphoma in our country as well as all over the world. In the presented study, it was aimed to investigate whether there is a relationship between SUV (standardized uptake value) values, which are the semiquantitative evaluation of FDG accumulation, and LDH, Ki-67 proliferation index, Beta-2 microglobulin and average age in cases diagnosed with DLBCL.

### Methodology

PET imaging of 40 patients diagnosed with diffuse large B-cell lymphoma included in this retrospective study was performed with FDG PET/CT in the Department of Nuclear Medicine at SBU Sultan Abdulhamit II Han Training and Research Hospital, and routine PET imaging protocol was applied to all patients. SUVmax values obtained from PET imaging of lymphoma patients and clinical and demographic data were statistically analyzed.

### Results

The correlation between SUVmax values in the FDG PET-CT study of diffuse large B-cell lymphoma patients and serum LDH, Ki-67, Beta-2 microglobulin level and average age was examined. According to the Pearson correlation test results, a significant positive correlation was observed between SUVmax values, Ki-67 ( $r= 0.2840$ ;  $p= 0.0379$ ) and LDH ( $r= 0.4184$ ;  $p= 0.0036$ ). However, no statistical significance was detected between SUVmax values, Beta-2 microglobulin ( $r=0.1071$ ;  $p= 0.2554$ ) and age ( $r= 0.08783$ ;  $p= 0.2950$ ) values ( $p>0.05$ ).

### Conclusion

Considering the results of the study, a moderate positive correlation is observed between SUVmax values and clinically LDH, and a weak positive correlation with Ki-67 proliferation index. FDG PET-CT imaging, with its semiquantitative FDG PET parameters, appears to be sensitive in detecting the response of the disease to treatment, during or at the end of treatment, and in estimating the biological aggressiveness and prognosis of the tumor. However, SUVmax values plays a potential role in diffuse large B-cell lymphomas with higher proliferation and more aggressive behaviour in clinical practice.

## IS IT NECESSARY TO EVALUATE ROUTINE HEMOSTASIS TESTS IN MYELOFIBROSIS?

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**Presentation Type** Oral

**Abstract Category** Adult Hematology Abstract Categories -> Myeloproliferative Neoplasms

Hematological malignancies are one of the underlying diseases that frequently cause disseminated intravascular coagulation (DIC). Although the frequency of DIC in myeloproliferative neoplasms has been studied, the frequency and course of DIC in Myelofibrosis (MF) are unknown. Moreover, the effect of early diagnosis and immediate treatment of DIC on the course of the disease is a matter of curiosity. In this study, we aimed to investigate whether ordering DIC tests routinely would be meaningful in detecting subclinical DIC in stable MF outpatients.

### Methodology

MF (primary, post-essential thrombocytosis and post-polycythemia vera) patients who were followed up in the hematology outpatient clinics of Ankara Bilkent City Hospital and who had routine hemostasis tests (INR, aPTT, PT, fibrinogen and D-dimer) resulted within the last one year (31.01.2023-31.01.2024) were evaluated retrospectively.

### Results

Among 137 MF patients, 37 patients who admitted to outpatient follow-up with routine hemostasis tests were evaluated. The median age was 63 (25-83) years. Male/female ratio was 19/18. The patients were clinically stable without clinical findings of disseminated intravascular coagulation such as fever, hypotension, and bleeding. Ten patients had thrombocytopenia ( $<100 \times 10^9$ ), but thrombocytopenia was thought to be related to bone marrow failure. When patients were evaluated with International Society of Thrombosis DIC criteria without including platelet count, overt DIC criteria were not met in any patient. In 3 patients, non-overt DIC was observed. However, all of these 3 patients had a history of chronic liver disease and thrombosis (all 3 were anticoagulated). Therefore, the impairment in hemostasis tests was probably associated with the use of anticoagulant therapy.

### Conclusion

This study showed that routinely ordering hemostasis tests to detect overt or non-overt DIC in stable patients without clinical symptoms may not contribute to patient management. However, this hypothesis can be supported in larger cohorts. We have planned to evaluate the frequency of DIC and accompanying causes in symptomatic myelofibrosis patients with new scoring systems developed specifically for hematological neoplasias with disease-related thrombocytopenia.

## RETROPECTIVE ANALYSIS OF WEEKLY KARFILZOMIB DEXAMETHASONE REGIMEN IN RELAPSED REFRACTORY MULTIPLE MYELOMA : SINGLE CENTER EXPERIENCE

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**Presentation Type** Oral

**Abstract Category** Adult Hematology Abstract Categories -> Multiple Myeloma

**Objective:** Treatment protocols requiring less hospital admissions has become preferable since Covid19 pandemics. Carfilzomib is a second generation proteasome inhibitor .There are strong evidences that weekly carfilzomib at a dose of 70 mg/ m<sup>2</sup> plus dexamethasone ( Kd70) increases progression free survival (PFS). Real life outcomes of relapsed refractory Multiple Myeloma (RRMM) patients managed with Kd70 between 2018-2023 in a tertiary center are analysed in this study.

**Methodology:** Patients with RRMM aged older than 18 years old between 2018 and 2023 in our center were retrospectively analysed. At least 1 cycle of Kd70 ( carfilzomib 70 mg/m<sup>2</sup> on days 1, 8 and 15 plus weekly dexamethasone 40 mg on 21 days cycle ) received 25 patients were enrolled. Patients received carfilzomib other than Kd70 schedule were excluded.

**Results:** Ten female and 15 male patients with a median age of 56 (43-68) years old were enrolled. At the data cut-off time 10 patients were still on Kd70 regimen ,15 patients discontinued Kd70 , 3 of whom due to toxicity . International staging system (ISS) 3 patients were 36 % of total cohort. Three patients had high risk cytogenetics and data of 9 patients were unavailable. Kd70 was commenced on the 4th or later line of therapy in 64% of patients. Ten patients discontinued Kd70 in the first 6 months of therapy due to progression or toxicities.. Overall response rate was 70.5 % in 15 response evaluable patients. Nine patients had VGPR , 3 had PR and 1 Patient has stabile disease as best response.

There was no  $\geq$  grade 3 hematologic toxicity ;14 patients (56 %) had grade 1 thrombocytopenia and 9 (36 %) had grade 1 anemia. The incidence of non-hematologic toxicities was 92 % but mostly low grade. Pneumonia ,nausea and emesis were the most frequent adverse events. Median PFS was 9.5 months and 3 years OS was % 68 at median 36 months follow up.

**Conclusion :** Kd70 shows efficacy and acceptable toxicity profile in RRMM even after multiple lines. The real life data of efficacy of the regimen in the earlier lines should be evaluated.

## THE IMPACT OF DIABETES MELLITUS ON HEMATOPOIETIC STEM CELL MOBILIZATION

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Presentation Type Oral

Abstract Category Adult Hematology Abstract Categories -> Cellular Therapy

### Objective

Factors such as age, underlying hematological disease, chemotherapy and radiotherapy used, and BM infiltration may cause mobilization failure. Several preclinical observed that diabetes mellitus (DM) leads to profound remodeling of the hematopoietic stem cell (HSC) niche, resulting in the impaired release of HSCs.

We aim to examine the effect of DM on HSC mobilization and to investigate whether there is a relationship between complications developing in the DM process and drugs used to treat DM and mobilization failure.

In Erciyes University Bone Marrow Transplantation Unit, 218 patients who underwent apheresis for stem cell mobilization between 2011-2021 were evaluated retrospectively. One hundred and nine patients had a diagnosis of DM, and 109 did not.

Mobilization failure developed in 17 (15.6%) of the patients in the DM group, while it developed in 7 (6.4%) patients in the non-DM group ( $p=0.03$ ). CD34+ stem cell count was 8.05 (1.3-30.2)  $\times 10^6/\text{kg}$  in the DM group, while it was 8.2 (1.7-37.3)  $\times 10^6/\text{kg}$  in the other group ( $p=0.55$ ). There was no statistically significant relationship between glucose and hemoglobin A1c levels and the amount of CD34+ cells ( $p=0.83$  and  $p=0.14$ , respectively).

Using sulfonylurea was the only independent predictor of mobilization failure (OR 5.75, 95% CI: 1.38-24.05,  $p=0.02$ )

## Conclusion

DM should be considered a risk factor for mobilization failure. Further research is needed fully to understand the mechanisms underlying the mobilization failure effects of sulfonylureas and to develop strategies to improve stem cell mobilization in diabetic patients.

## CLINICAL SIGNIFICANCE OF CALR MUTATION IN PATIENTS WITH ESSENTIAL THROMBOCYTHEMIA

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Presentation Type Poster

Abstract Category Myeloproliferative Neoplasms

**Aim:** In essential thrombocythemia (ET), CALR mutation is associated with younger age, higher platelet count and lower rate of thrombosis. We examined the demographic, laboratory and clinical features of the CALR mutation in the Turkish population and its impact on prognosis.

**Materials and Methods:** A cohort of 394 ET patients diagnosed from 1995 and 2022 were included in this retrospective, multicenter study.

**Results:** Clinical and laboratory features of 394 ET patients are summarized in Table-1. In CALR+ patients, the frequency of female gender, mean leukocyte count, Hb and Hct at time of diagnosis are lower and the mean serum LDH level, platelet count and frequency of platelet count  $\geq 1.10^9/\mu\text{L}$  are higher. The frequency of overall arterial and venous thrombosis and only arterial thrombosis before diagnosis ET are significantly lower for CALR+ patients than JAK2V617F+ patients. In subanalysis of patients aged  $\leq 60$  years at diagnosis, frequency of overall arterial and venous thrombosis and only venous thrombosis are lower in CALR+ and triple-negative patients than in JAK2V617F+ patients. In multivariate analysis for thrombosis, CALR mutation was not protective against thrombosis and arterial thrombosis while female gender was protective. CALR+ patients were more frequently exposed to multipl-line therapies and interferon-based therapy. Frequency of malignancy was higher in CALR+ and JAK2V617F+ patients than in triple-negative patients. There was no significant difference for myelofibrotic transformation and bleeding complications between JAK2V617F+, CALR+ and triple negative groups. In survival analysis, overall survival (OS) and thrombosis-free survival (TFS) were similar between mutation groups. The presence of CALR+ mutation was associated with lower leukemia-free survival (LFS).

Conclusion: In our study, CALR+ patients were associated with lower frequency of female gender, and lower leukocyte counts, Hb and Hct levels and higher platelet count and LDH levels. In multivariate analysis, CALR mutation lost its protective effect against thrombosis. In a large cohort of Turkish patients with long follow-up period, our study demonstrated the demographic characteristics of ET patients and impact of CALR gene mutation on age, gender, blood count parameters, spleen size, complications including thrombosis/bleeding/malignancy and myelofibrotic transformation in the Turkish population.

**Table I: : Clinical and laboratory features of 394 ET patients**

	<b>ET Patients (394) (mean ± SD) (n %)</b>	<b>JAK2V61 7F+ (278) (mean ± SD) (n %)</b>	<b>CALR+ (72) (mean ± SD) (n %)</b>	<b>MPL+ (3) (mean ± SD) (n %)</b>	<b>Triple Negative (41) (mean ± SD) (n %)</b>	<b>p*</b>
<b>Gender</b>						
<b>Female</b>	249 (63,2)	183 (65.8) <sup>1</sup>	33 (45.8) <sup>b</sup>	1 (33.3)	32 (78) <sup>a</sup>	0.001 <sup>1</sup>
<b>Male</b>	145 (36,8)	95 (34.2) <sup>1</sup>	39 (54.2) <sup>b</sup>	2 (66.7)	9 (22) <sup>a</sup>	
<b>Age at MPN diagnosis, (mean ± SD)</b>	59,3 ± 16,4	52.5 ± 16.6	49.9 ± 16.5	49.3 ± 24	47.6 ± 16.1	0.142 <sup>2</sup>
<b>≤60</b>	262 (66,5)	178 (64)	50 (69.4)	2 (66.7)	32 (78)	0.174 <sup>1</sup>
<b>&gt;60</b>	132 (33,5)	100 (36)	22 (30.6)	1 (33.3)	9 (22)	
<b>WBC at MPN diagnosis, (mean ± SD) (.10<sup>3</sup> / μL)</b>	10,5 ± 4,1	10.9 ± 4.5 <sup>a</sup>	9.6 ± 3.1 <sup>b</sup>	7.5 ± 2.2	9.9 ± 2.9 <sup>a, b</sup>	0.029 <sup>2</sup>
<b>&lt;11 000 / μL</b>	250 (63,5)	173 (62.2)	49 (68.1)	3 (100)	25 (61)	0.628 <sup>1</sup>
<b>≥ 11 000 / μL</b>	144 (36,5)	105 (37.8)	23 (31.9)	-	16 (39)	
<b>HB at MPN diagnosis, (mean ± SD)</b>	13,4 ± 1,7	13.7 ± 1.6 <sup>a</sup>	12.7 ± 1.8 <sup>b</sup>	13.6 ± 1.1	13.1 ± 1.6 <sup>b</sup>	<0.001 <sup>2</sup>
<b>HCT at MPN diagnosis, (mean ± SD)</b>	40,8 ± 5,1	41.7 ± 4.9 <sup>a</sup>	38.1 ± 5.1 <sup>b</sup>	40.5 ± 3.4	39.7 ± 4.8 <sup>b</sup>	<0.001 <sup>2</sup>
<b>Plt at MPN diagnosis, (mean ± SD) (.10<sup>3</sup> / μL)</b>	904,7 ± 357,6	858.1 ± 306.4 <sup>a</sup>	1078.5 ± 468.3 <sup>b</sup>	796.3 ± 145	922.9 ± 381.8 <sup>a, b</sup>	<0.001 <sup>2</sup>
<b>&lt;1 000 000 / μL</b>	278 (70,6)	207 (74.5) <sup>1</sup>	38 (52.8) <sup>b</sup>	3 (100)	30 (73.2)	0.001 <sup>1</sup>
<b>≥ 1 000 000 / μL</b>	116 (29,4)	71 (25.5) <sup>a</sup>	34 (47.2) <sup>b</sup>	-	11 (26.8) <sup>a, b</sup>	
<b>LDH at MPN diagnosis, (mean ± SD) (U/L)</b>	307,5 ± 169,6	314.6 ± 185.7 <sup>a</sup>	321.5 ± 128.9 <sup>a</sup>	344 ± 281.2	232.1 ± 61.2 <sup>b</sup>	0.010 <sup>2</sup>
<b>Spleen size at MPN diagnosis, (mean ± SD) (mm)</b>	130,1 ± 26,9	132.6 ± 29.8 <sup>a</sup>	125.7 ± 18.2 <sup>a, b</sup>	120	121.8 ± 14.5 <sup>b</sup>	0.016 <sup>2</sup>
<b>CV risk factors</b>		193 (69.4) <sup>1</sup>	41 (56.9) <sup>a, b</sup>	3 (100)	20 (48.8) <sup>b</sup>	0.010 <sup>1</sup>

<b>Revised IPSET risk score</b>		-				
<b>Very low</b>	75 (19)	136 (48.9)	46 (63.9)	1 (33.3)	28 (68.3)	0.090
<b>Low</b>	140 (35,5)	-	2 (2.8)	1 (33.3)	1 (2.4)	
<b>Intermediate</b>	23 (5,8)	142 (51.1)	16 (22.2)	1 (33.3)	6 (14.6)	
<b>High</b>	156 (39,6)		8 (11.1)	-	6 (14.6)	
<b>Thrombosis</b>	128 (32,5)	105 (37.8) <sup>1</sup>	15 (20.8) <sup>b</sup>	0 (0)	8 (19.5) <sup>a,b</sup>	0.004 <sup>1</sup>
<b>Arterial thrombosis before diagnosis</b>	68 (14,7)	59 (21.2) <sup>a</sup>	4 (5,6) <sup>b</sup>	-	5 (12,2) <sup>a,b</sup>	0.005 <sup>1</sup>
<b>Venous thrombosis before diagnosis</b>	35 (6,3)	31 (11.2)	3 (4.2)	-	1 (2.4)	0.055 <sup>1</sup>
<b>Arterial thrombosis after MPN diagnosis</b>	31 (7,1)	21 (7.6)	8 (11.1)	-	2 (4.9)	0.455 <sup>1</sup>
<b>Venous thrombosis after MPN diagnosis</b>	15 (3)	14 (5.1)	1 (1.4)	-	-	0.229 <sup>2</sup>

## BIOCHEMICAL PROPERTIES OF RED BLOOD CELLS IN POLYCYTHEMIA VERA

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Presentation Type    Oral

Abstract Category    Adult Hematology Abstract Categories -> Myeloproliferative Neoplasms

### Objective

Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by an increase in red blood cell mass. Thrombotic complications are the main cause of morbidity and mortality in PV. Elevated hematocrit and increased blood viscosity are crucial risk factors for thrombus formation. The aim of our analysis is to evaluate the biochemical alterations in red blood cells (RBCs) and the hemoglobin structure in patients with PV that may be associated with thrombotic complications.

### Methodology

Blood samples were taken from 20 PV patients and 16 healthy individuals. The isolated RBCs were examined using Raman spectroscopy.

### Results

We found a larger contribution of ferrous heme iron, which is a molecular state typical for deoxyhemoglobin, in PV samples compared to the control samples. Furthermore, a significant increase in the Fe II/Fe III ratio in PV samples was correlated with a higher hematocrit (Hct) to hemoglobin (Hgb) ratio. A positive trend between a higher Fe II/ Fe III ratio and a higher RDW-SD and RDW-CV was observed in PV samples. In RBCs collected from PV patients we observed a less stable hemoglobin structure.

### Conclusion

Higher values of RDW-SD and RDW-CV may reflect a higher Fe II/ Fe III ratio and be a simple indicator of biochemical alterations in RBCs. A higher Hct/ Hgb ratio could indicate higher clonal myeloproliferative potential and be associated with shorter time to thrombosis in patients with PV. Our future analysis will focus on correlating the above observations with the prothrombotic activity to demonstrate a possible link between the biochemical alterations of RBCs and the thrombotic complications in

## RARE ANTI-M ALLOANTIBODY POSITIVITY IN A PREGNANT WOMAN AND THE EFFECTIVENESS OF PLASMAPHERESIS IN TREATMENT: A CASE REPORT

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Anti-M-related alloantibodies are typically IgG antibodies that can lead to hemolysis. These antibodies can cause hemolytic disease of the newborn or hydrops fetalis by crossing the placenta.

In this context, we will discuss a case in which a pregnant patient with recurrent intrauterine ex fetus and anti-M alloantibody positive was treated effectively with plasmapheresis.

During the exams performed on a 24-year-old pregnant woman, it was discovered that she had a positive Indirect Coombs test and her anti-M allo-antibody was also positive. The anti-M antibody titer was greater than 1/4096. In November 2019, during the 10th week of her sixth pregnancy, the patient underwent plasmapheresis due to the presence of ascites and subcutaneous edema in the fetus, which may have been indicative of hydrops fetalis. After the plasmapheresis, the patient was monitored with antibody titer and fetal ultrasound. She continued to undergo plasmapheresis three times a week from the 13th week of pregnancy until the birth of the baby. During the follow-up, the antibody titer decreased to 1/8. In May 2020, a healthy baby girl was born via cesarean section due to transverse presentation at the 35th week of pregnancy.

The patient became pregnant again in September 2021 and was found to be Anti-M alloantibody positive. She underwent plasmapheresis twice a week from the 7th week of pregnancy until the birth of the baby. The patient delivered a healthy male baby via cesarean section due to fetal distress in the 37th week of pregnancy.

Treatment options for hemolysis or hydrops fetalis caused by alloimmunization during pregnancy include intrauterine red blood cell transfusion, immunoglobulin injection, and plasmapheresis. Plasmapheresis was successful in achieving live births in the sixth and seventh pregnancies of a patient who had experienced intrauterine ex fetus in her previous five pregnancies.

## CHRONIC LYMPHOCYTIC LEUKEMIA: IMMUNE DYSREGULATION AND MANAGEMENT - A CASE REPORT

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### Introduction

Hypogammaglobulinemia is the most common immune disorder in chronic lymphocytic leukemia (CLL). In CLL patients, both humoral and cellular immune dysfunction and treatment-related immune dysregulation may occur.

The most common cause of death in CLL patients is infections. Apart from anti-infective treatments given for infection, immunoglobulin replacement can be performed.

We aimed to present three CLL patients with hypogammaglobulinemia who were followed up in our clinic.

### Case report 1

A 69-year-old male patient was diagnosed with CLL(RAI stage IV) in 2018. The patient was treated with Rituximab plus Bendamustine. Before the treatment, the patient's IgG level was 516 gr/dl. During and after the treatment, the patient's IgG level was 355 mg/dl and 285 mg/dl respectively. Subsequently, the patient was monitored without any treatment or immunoglobulin replacement therapy (IGRT).

### Case report 2

An 84-year-old male patient was diagnosed with CLL(RAI IV) in September 2017. At the time of diagnosis, his IgG level was 1720 mg/dl. The patient received Rituximab plus Bendamustine treatment, and after the first cure, he was hospitalized due to neutropenic fever and lung fungal infection. The patient was treated with voriconazole, meropenem, and teicoplanin. His IgG level was 1120 mg/dl at that time. IgG levels measured during follow-up were 525 mg/dl, 496 mg/dl, and 639 mg/dl, respectively. During this period, the patient did not receive IGRT.

### Case report 3

A 56-year-old female patient, who has asthma and Sjögren's disease, was diagnosed with RAI I SLL/CLL in 2018. At the time of diagnosis, her IgG level was 642 mg/dl. During the follow-up period, the IgG levels decreased to 377 mg/dl. In 2019 Rituximab plus Venetoclax treatment was started. Intravenous immunoglobulin (IVIG) was given the patient, who had pneumonia in the 6th month of treatment and did not respond to antibiotic treatment. Post-treatment IgG levels were below 500 mg/dl and frequent infections. Due to the patient's history, IVIG was given regularly.

### Discussion&Conclusion

As a result, hypogammaglobulinemia may be observed in CLL patients throughout the course of the disease, and immunoglobulin replacement may be required in patients who do not respond to anti-infective treatments and whose IgG levels are  $<5$  g/dl.

## THE INTERSECTION OF THALASSEMIA MINOR AND CHRONIC MYELOPROLIFERATIVE DISORDERS: CLINICAL AND LABORATORY IMPLICATIONS

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This study aims to explore the rare co-occurrence of thalassemia minor and Chronic Myeloproliferative Disorders (CMPD), focusing on how thalassemia minor can obscure the diagnostic markers of CMPD, including hemoglobin levels, and the subsequent effects on clinical and laboratory outcomes. The investigation involved 32 patients diagnosed with thalassemia minor alongside CMPD at Cukurova University Faculty of Medicine's Department of Hematology between January 2003 and January 2024. The diagnostic criteria utilized were the "2020 ELN Criteria" for CML and the "WHO Classification of Myeloid Neoplasms and Acute Leukemia, 2016 Revision Criteria" for BCR-ABL negative CMPDs. Our findings highlight a spectrum of CMPD manifestations among the patients, with a significant portion displaying symptoms and signs such as hepatomegaly and splenomegaly. A pivotal aspect of this study was the identification of the JAK2 V617F mutation in the majority of patients, underscoring the prevalence of CMPD despite the potential masking effect of thalassemia minor on certain hematologic markers. The research illuminates the intricate relationship between thalassemia minor and CMPD, emphasizing the need for careful diagnostic consideration to ensure accurate identification and management of these patients. This study contributes valuable insights into the complexities of diagnosing and treating individuals with concurrent thalassemia minor and CMPD, urging a nuanced approach to hematologic evaluation.

Keywords:

1. Thalassemia Minor
2. Chronic Myeloproliferative Disorders
3. Hemoglobin Levels
4. JAK2 V617F Mutation
5. Diagnostic Challenges

## VITALITY AND ENGRAFTMENT TIMES IN ALLOGENIC HEMATOPOETIC STEM CELL TRANSPLANTATIONS MADE WITH FROZEN AND FRESH PRODUCT

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### Background:

Allogeneic stem cell transplants can be collected on the day of transplantation or frozen and stored after collection due to medical or donor-related reasons.

### Aims:

In our study, we compared the product viability, neutrophil and platelet engraftment times between allogeneic peripheral stem cell transplants that were frozen and unfrozen in our center between 2020 and 2023.

### Methods:

Allogeneic transplantation procedures performed between 2020 and 2023 were evaluated retrospectively. Mobilization of allogeneic donors was performed with 10µc/kg GCS-F. Collection operations were carried out with the OptiaSpectra device on the 5th day of GCS-F. After collection, the products were frozen with cryoprotectant prepared with 10% DMSO (dimethyl sulfoxide) and 20% plasma and stored at -80 °C. On the day of transplantation, allogeneic stem cells, which were thawed in a blood melting device at 37°C, were transplanted to the patient.

### Results:

Of the 93 allogeneic transplants performed at our Erciyes University Faculty of Medicine stem cell transplantation center, 68 (73.1%) were performed with fresh products, while 25 (36.9%) were performed with frozen products. Frozen cells were transplanted to patients after an average of 15±44.8 (6-215) days. While the number of CD34+ cells in frozen products was 7x10<sup>6</sup>/kg, this value was 7.06x10<sup>6</sup>/kg in fresh products. While the neutrophil engraftment period was 16 days in allogeneic transplants Performed without freezing, it was 17 days in transplantations performed with frozen products, and the platelet engraftment period was 13 days in allogeneic transplants performed without freezing and 14.5 days in frozen transplants. One of the patients who underwent cryopreservation and two of the patients who underwent noncryopreservation died before neutrophil and platelet engraftment developed. While the viability rate during transportation in frozen products was 96.1%, the viability rate in fresh products was 99.7% . It was observed that the viability rate in two frozen allogeneic stem cell products was 88.8% and 83.44%.

Summary/Conclusion: Considering the values of both viability rates and engraftment times, we observed that, in terms of patient and donor safety, freezing donor stem cells before transplantation does not negatively affect our transplantation process and can be used as an alternative method.

## VENETOCLAX PLUS İBRUTİNİB AS FIRST-LINE TREATMENT IN PATIENTS WITH CHRONIC LYMPHOCYTIC LEUKEMİA- META-ANALYSIS OF RANDOMISED CONTROLLED TRIALS

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Presentation Type Oral

Abstract Category Adult Hematology Abstract Categories -> Chronic Lymphocytic Leukemia

### Objective

Venetoclax-ibrutinib combination has been reported to improve outcomes in the treatment of chronic lymphocytic leukemia (CLL). This study aims to evaluate the efficacy and safety of venetoclax-ibrutinib combination therapy compared to alternative treatments in CLL patients.

### Methodology

A comprehensive search of Cochrane CENTRAL, Ovid Medline, PubMed, and Web of Science up to February 20, 2024, was conducted. Randomized controlled trials (RCTs) directly comparing venetoclax-ibrutinib with alternative treatments were included. Data were extracted from three distinct RCTs, and analysis was performed using an inverse variance-weighted fixed-effects model. The primary efficacy endpoint was progression-free survival (PFS) at 36 months, with secondary endpoints including minimal residual disease (MRD) detection in peripheral blood (PB) and bone marrow (BM). Adverse events (AEs) were assessed for safety outcomes. Subgroup analyses were conducted based on the presence of the IGHV mutation.

### Results

The analysis included three RCTs with a total of 1046 patients, two of which explored venetoclax+ibrutinib, while one remains determined to explore venetoclax+ibrutinib+obinutuzumab. Venetoclax-ibrutinib demonstrated superiority over control regimens in terms of PFS at months 12 (RR 1.10, 95% CI 1.06 to 1.14), 24 (RR 1.21, 95% CI 1.15 to 1.28), 36 (RR 1.27, 95% CI 1.14 to 1.42), and 48 (RR 1.47, 95% CI 1.18 to 1.84), but not at month 60 (RR 1.74, 95% CI 0.88 to 3.43). Additionally, it significantly reduced MRD in PB (RR 1.54, 95% CI 1.42 to 1.68) and BM (RR 1.79, 95% CI 1.57 to 2.03). Overall survival at month 36 was comparable between venetoclax-ibrutinib and control regimens (RR 1.01, 95% CI 0.97 to 1.05). Safety analysis showed lower rates of death (RR 0.49, 95% CI 0.28 to 0.88), and serious AEs (RR 0.86, 95% CI 0.80 to 0.92) with venetoclax-ibrutinib but not for pneumonia (RR 1.36, 95% CI 0.78 to 2.35). However, diarrhea and hypertension were more common. Subgroup analysis suggests potential differences in PFS only at month 24 (p<0.01).

## Conclusion

Venetoclax-ibrutinib combination therapy demonstrates superior efficacy in terms of progression-free survival and minimal residual disease compared to alternative treatments in CLL patients. Although associated with certain adverse events, its safety profile is generally favorable.

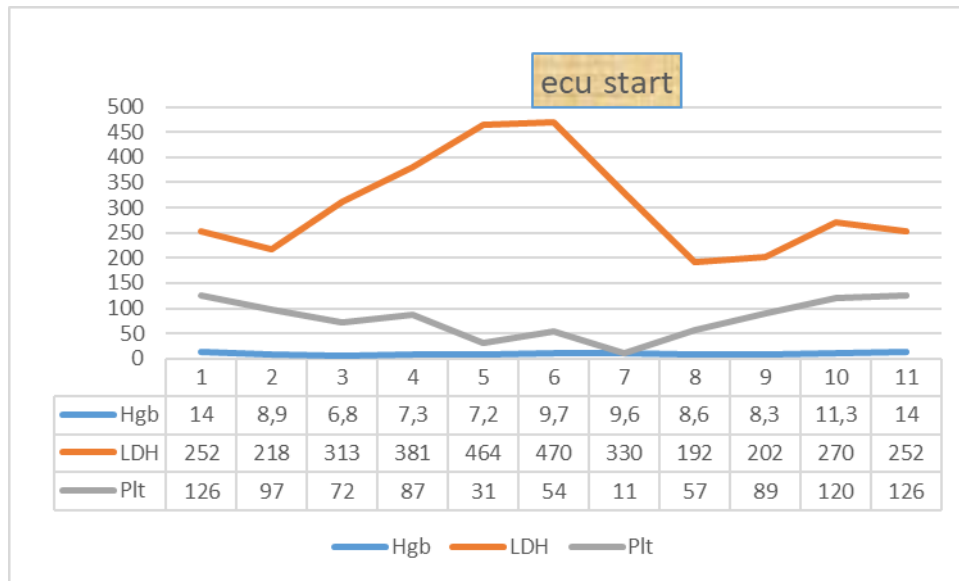
Keywords: Venetoclax, ibrutinib, chronic lymphocytic leukemia, progression-free survival, meta-analysis, hypertension

## TREATMENT OF HEMATOPOIETIC STEM CELL TRANSPLANT-ASSOCIATED MICROANGIOPATHY WITH ECULİZUMAB

Gülşah Akyol, Şerife Emre Ünsal, Neslihan Şanlı Mandacı, Muzaffer Keklik, Ali Ünal

Plasma exchange is the essential treatment for TTP, but it has no known benefit for patients with cancer-induced or drug-induced TMA. The etiology and treatment of microangiopathy that develops after transplantation are not clearly established.

A 62-year-old female patient who underwent surgery and chemoradiotherapy for breast cancer 6 years ago was diagnosed with AML. Upon failure to respond to 7/3 induction therapy, FLAG-IDA-Venetoclax chemotherapy was administered. Achieving remission, the patient underwent Allogeneic HCT from a fully matched sibling. Following transplantation, CMV positivity was treated initially with Sidofovir followed by Valganciclovir. Two months post-transplantation, the patient developed anemia and elevated LDH levels, with schistocytes observed in her blood smear. Prednisolone was initiated at 1mg/kg and reevaluation was scheduled after 3 weeks. 1 session of plasmapheresis was performed on the patient who did not respond to steroids. Concurrently developing hyponatremia and hypokalemia led to consideration of Fanconi syndrome, and treatment with aldactazide was added. At the 3-month post-transplantation follow-up, an increase in schistocytes was noted. Progression of microangiopathy was suspected, prompting initiation of Eculizumab. Weekly dosing of 600 mg for 3 weeks resulted in disappearance of schistocytes upon control examination, and treatment was successfully completed.



Conclusion:

Post-transplantation TMA can be either primary-possibly related to the transplant itself, as in our case, or it can occur following CMV viremia, or it may also be triggered by medications used. Additionally, the treatment of this condition is also challenging

## MARGINAL ZONE LYMPHOMA IN THE LANDSCAPE OF AUTOIMMUNE COMPLEXITY: A CASE REPORT WITH SJÖGREN'S SYNDROME AND HASHIMOTO THYROIDITIS

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### Abstract:

In a compelling case, a 49-year-old woman grappling with autoimmune thyroiditis and ongoing otitis presented with a persistent neck swelling. Diagnostic tests revealed multiple enlarged cervical lymph nodes. A targeted biopsy from these nodes exposed a high concentration of lymphocytes, particularly around the salivary glands, with antibodies confirming Sjögren's Syndrome. Treatment began with hydroxychloroquine and prednisolone.

Further PET-CT scans showed active lymph nodes in the neck, and a deeper biopsy identified these as infiltrated by abnormal lymphocytes, diagnosing the condition as extranodal marginal zone lymphoma (MALToma) of the mucosa-associated lymphoid tissue. This diagnosis prompted a combined treatment strategy, engaging both rheumatology, considering rituximab, and hematology departments. The plan included targeted radiation therapy for the salivary glands and cervical lymph nodes, involving radiation oncology consultations.

This case illuminates the complex interplay between autoimmune diseases and lymphoproliferative disorders, stressing the importance of a team-based approach in diagnosis and treatment. It showcases the necessity for meticulous diagnostic work to identify and treat potential malignancies within autoimmune diseases, highlighting the crucial role of a multidisciplinary team in managing such intricate medical conditions.

### Keywords

- 1)Autoimmune diseases
- 2)Sjögren's Syndrome
- 3)Marginal zone lymphoma
- 4)Hashimoto thyroiditis
- 5)Lymphoproliferative disorders

## MULTICENTRIC CASTLEMAN'S DISEASE IN THE SETTING OF POEMS SYNDROME

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Presentation Type: Poster

Abstract Category: Multiple Myeloma

Case Report: A 59-year-old male with a history of type 2 diabetes mellitus, chronic kidney disease, and protein S deficiency presents with 3 years of worsening lymphadenopathy, renal function, weight loss (36 kg), skin thickening, lower extremity weakness, and gastroparesis. Initial PET/CT imaging shows multiple lymph nodes with SUV max ranging from 1.1 to 5.7, L1 sclerotic lesion, edema, and hepatosplenomegaly. The labs show the following: Hgb: 11.1 g/dL, Creatinine: 2.2 mg/dL, Total Calcium: 8.2 mg/dL, SPEP with M spike of .93 g/dL, Immunofixation shows a monoclonal IgG lambda. Serum-free light chains of kappa: 212, lambda 285, and K:L ratio: 0.74. An L1 bone biopsy reveals a lambda-restricted plasma cell neoplasm. The right axillary lymph node was subsequently sampled, which reveals follicular hyperplasia, vascular hyalinization, atrophic germinal centers, prominent mantle zones, and plasmacytosis. These morphological findings without evidence of clonal proliferation in the involved lymph nodes are consistent with multicentric Castleman disease as part of POEMS syndrome. The patient was initially treated with radiation therapy to L1 and siltuximab, a chimeric monoclonal antibody that binds to interleukin-6 (IL-6). Additionally, the patient received plasma cell-directed therapy, which included daratumumab 1800 mg subcutaneously weekly x 8 followed by every other week x 8 followed by every 28 days; along with lenalidomide 10 mg days 1-21 every 28-day basis with warfarin support and dexamethasone 12 mg weekly.

Discussion:

POEMS syndrome (peripheral neuropathy, organomegaly, endocrine abnormalities, monoclonal gammopathy, and skin changes) is a paraneoplastic process often associated with Multicentric Castleman's Disease (MCD). It shares features with and can be mistaken for plasma cell myeloma (PCM). However, patients with POEMS syndrome present more with sclerotic bone lesions than lytic lesions in PCM. Additionally, serum electrophoresis M-spike is only mildly elevated <2mg/dL in POEMS. In terms of the symptoms, polyneuropathy and endocrinopathies in PCM are rare. While first treated similarly with lenalidomide and stem cell transplant, emerging research on the role of cytokines in driving MCD has allowed for targeted treatments such as IL-6 inhibitors. Continued study is necessary to better diagnose and treat this rare lymphoproliferative process.

## CASE REPORT OF REFRACTORY LARGE B-CELL LYMPHOMA WITH IRF4 REARRANGEMENT

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Presentation Type Poster

Abstract Category Aggressive B-Cell Lymphoma

### Introduction

Large B-cell lymphoma with IRF4 rearrangement (IRF4+ LBCL) presents in young adulthood with involvement of the head and neck region. There is insufficient data on the association of concomitant cytogenetic abnormalities with prognosis. We aimed to present a case of refractory IRF4+ LBCL.

A 20-year-old male patient with no known history of chronic disease or medication, with right cervical swelling which had been growing slowly for 4 months. Physical examination revealed bilateral hypertrophic tonsils with prominent right side. Laboratory analysis revealed no significance in complete blood count, biochemical analysis and peripheral blood smear. The biopsy of the palatine tonsil was performed.

Microscopic examination demonstrated follicular infiltration of neoplastic lymphoid cells with medium-sized blastic morphology. Immunohistochemical examination revealed strong expression of pan-B markers (CD20 CD79a, PAX-5) as well as CD10, bcl-2, bcl-6 and MUM-1. Ki-67 proliferation index was 75% and myc expression was negative with immunohistochemistry. NGS analysis resulted as IRF-4 mutation, as well as BABAM2::JAK2 fusion and bcl-2 mutation that were supportive for IRF4+LBCL.

Positron emission tomography-computed tomography (PET/CT) performed for staging revealed a 3.9x2.8 cm asymmetric hypermetabolic soft tissue (SUVmax:19.2) at the level of the right palatine tonsil, and hypermetabolic activity (SUV max 14.2) at the level of the left palatine tonsil. R-CHOP chemotherapy protocol was initiated. At the end of 4 cycles, a partial response was observed; and the treatment was completed to 6 cycles. However, at the end of treatment, PET/CT showed an increase in the size and metabolic activity of the primary lesion. Repeat biopsy was resulted the same morphology. Salvage chemotherapy followed by autologous stem cell transplantation was planned.

## Discussion

In our knowledge, age has been defined as prognostic parameter for IRF-4+LBCL, Thus, more complex molecular abnormalities have been reported in the adult age group. JAK2 fusion abnormalities are associated with treatment resistance in myeloid and lymphoblastic leukemias, but there is no data on their contribution to the clinical course in IRF4+ LBCL. In our case with primary refractory course, it was considered that additional JAK fusion abnormality may have a negative effect on the clinical course.

## FIRST 1-YEAR RESULTS OF A NEW HEMATOPOIETIC STEM CELL TRANSPLANT CENTER

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Abstract Category Adult Hematology Abstract Categories – Cellular Therapy

### Objective

Hematopoietic stem cell transplantation (HSCT) is used as a treatment option for many malignant or benign hematologic diseases. In our country, the number of centers performing HSCT has been increasing recently. We aimed to share the 1-year transplant data of a newly opened HSCT center.

### Methodology

Our study is a retrospective observational study and the files of bone marrow transplant patients who underwent bone marrow transplantation at Sanko University Hospital between March 2023 and March 2024 were retrospectively evaluated.

### Results

A total of 29 patients (18 F, 11 M) were included in the study. The median age was 56 years (42-70) in 26 patients who underwent autologous transplantation and 56 years (42-70) in 3 patients who underwent allogeneic transplantation. Of the 26 patients who underwent autologous transplantation, 19 were diagnosed with Multiple Myeloma, 2 with DLBCL, 1 with Burkitt Lymphoma, 1 with Mantle Cell Lymphoma, 2 with Hodgkin's Lymphoma and 1 Adult T cell lymphoblastic lymphoma. Of the 3 patients who underwent allogeneic transplantation, 2 were diagnosed with AML and 1 with ALL. (Table 1) In autologous transplantation, melfelan (140-200) was used as a preparative regimen in 19 patients, Mito/mel in 4 patients, and Be-EAM in 3 patient. In allogeneic transplantation, BuCyC was used in 2 patients and TreFLu in 1 patient. The mean stem cell count for autologous transplantation was 6.22x 10<sup>6</sup> CD 34/kg. The number of stem cells given for allogeneic transplantation was 6.5x10<sup>6</sup> CD 34/kg. Neutrophil and thromocyte engraftments in autologous transplants were on day 11 (10-14) and day 12 (10-14), respectively. In allogeneic transplants, neutrophil and thromocyte engraftments were 15 (12-16) days and 15 (12-16) days, respectively. Stem cell infusion was performed on a median of 1 day (21 patients 1 day, 5 patients 2 days) in the autologous group. The duration of hospitalization was 19 days (15-40) in the autologous group and 31 (28-33) days in the allogeneic group. Febrile neutropenia was observed in 15 patients in the autologous group and 3 patients in the allogeneic group. Fungal infection was not observed in both groups. CMV infection was observed in 1 patient in the autologous group and 1 patient in the allogeneic group. Acute and chronic GVHD was not observed. Five patients (5 MM) with a pre-transplant PPD of 10 mm received INH prophylaxis.

Covid 19 infection was observed in 2 autologous and 1 allogeneic transplant patients during pre-transplant preparation. TRM was not observed in the first 100 days (0/29 patients). 1 patient with r/r DLBCL exited during the CART treatment process after progressive disease at oxide +60. Mortality in the first 100 days was 3%. (Table 1)

### Conclusion

Despite the small number of patients in the study and the limited follow-up period, the low TRM and mortality in the first 100 days and the absence of infectious deaths such as febrile neutropenia and fungal infections give hope for the future.

Table 1: Patient Characteristics

	Otolog	Allojenik
n	26	3
Age (median, range)	56 (42-70)	42 (31-51)
Gender (F/M)	16/10	2/1
Diagnosis (MM, NHL, HL, AML, ALL)	19/5/2/0/0	0/0/0/2/1
Indication for transplantation : Relapsed Disease/Primary Consolidation	5/21	0/3
Priming regimen (MEL, Mtx/mel, Be-EAM, BuCyC, TreFlu)	19/4/3/0/0	0/0/0/2/1
Stem cell count (x106 CD 34/kg)	6,22 (3-10)	6,5 (6,5)
Neutrophil engraftment time (median/range) (days)	11 (10-14)	15 (12-16)
Trombosit Engrafman Zamanı (medyan/aralık) (gün) Platelet Engrafting Time (median/range) (days)	12 (10-14)	15 (12-16)
Number of stem cell infusion days 1/2 (days)	21/5	3/0
Length of hospitalization (median/range) (days)	19 (15-40)	31 (28-33)
Febrile Neutropenia	15/26	3/3
Fungal Infection	0/26	0/3
CMV Infection	1/26	1/3
Acute GVHD	0/26	0/3

<b>Chronic GVHD</b>	0/26	0/3
<b>PPD positive (INH prophylaxis)</b>	5/26	0/3
<b>Covid 19 Infection</b>	2/26	1/3
<b>+100 days TRM</b>	0/26	0/3
<b>+100 days mortality</b>	1/26	0/3

## CAN LYMPHOMA MIMIC CROHN'S DISEASE?

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### Introduction

Marginal zone lymphoma originates from B lymphocytes derived from the marginal zone of the germinal follicle. It is one of the low-grade lymphomas and is classified based on splenic, nodal, and mucosal involvement. Crohn's disease, an inflammatory bowel disease, affects the gastrointestinal system transmurally, with the most commonly affected areas being the ileum and proximal colon. A case of Marginal Zone Lymphoma mimicking Crohn's disease is quite rare in the literature. Here, we present a case of Marginal Zone Lymphoma mimicking Crohn's Disease.

### Case

A 65-year-old male with hypertension and hypothyroidism presented with complaints of abdominal pain and diarrhea for the past 2 years. He was initially diagnosed with Crohn's disease based on clinical and histopathological evaluation upon referral to the gastroenterology clinic. He presented three months later with worsening fatigue and weight loss. Bilateral axillary and inguinal lymphadenopathy were noted on physical examination. Laboratory investigations revealed white blood cell count of  $2.37 \times 10000/\mu\text{l}$  (with neutrophils  $370/\mu\text{l}$ , lymphocytes  $1980/\mu\text{l}$ ), hemoglobin 5.3 g/dl, platelet count  $18000/\mu\text{l}$ . Ultrasonography showed bilateral axillary lymphadenopathies measuring up to  $2.5 \times 1$  cm and inguinal lymphadenopathies measuring up to  $3 \times 1$  cm. Further evaluation included bone marrow aspiration and biopsy. Flow cytometry evaluation of bone marrow revealed a suspicious B-cell population with 75% of CD5-/CD19+, FMC7- phenotype showing kappa monoclonality, suggestive of marginal zone lymphoma involvement of bone marrow. Excisional biopsy of axillary lymph node confirmed the diagnosis of marginal zone lymphoma on histopathological examination. The patient received 6 cycles of R-CHOP chemotherapy for widespread nodal and extra nodal involvement of lymphoma. Improvement in blood counts and resolution of symptoms such as diarrhea and abdominal pain were noted following lymphoma treatment.

### Discussion

The initial presentation of lymphoma with involvement of the small intestine may have led to the misdiagnosis of Crohn's disease. Stundiene et al. also reported a case of Marginal Zone Lymphoma mimicking Crohn's disease in their case report. Diseases can present with similar clinical pictures. Besides histopathological evaluation, diagnoses should be reconsidered in cases of clinical suspicion. Differential diagnosis is crucial in diseases that can lead to similar symptoms.

## WHAT HAPPENED SUDDENLY OR ACUTE ABDOMEN? DIFFICULT CASE OF ATRA RELATED PNEUMATOSİS CYSTOIDES INTESTINALIS

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### Introduction

Acute promyelocytic leukemia (APL) is characterized by the presence of atypical promyelocytes in the bone marrow and peripheral blood, is associated with disseminated intravascular coagulation (DIC), therefore urgent treatment is required and All-trans retinoic acid (ATRA) is used to control this clinic. (1). The use of ATRA may cause the development of a fatal syndrome accompanied by peripheral blood leukocytosis in 15% of patients diagnosed with acute promyelocytic leukemia. There are suggestions that early recognition of fever, dyspnea, alveolar edema symptoms and early administration of corticosteroid treatment may reduce the morbidity and mortality associated with this syndrome (2,3). Gastrointestinal tract-related adverse events in patients using ATRA are listed on the product label as abdominal bloating (11%), abdominal pain (31%), anorexia (17%), constipation (17%), diarrhea (23%), dry mucous membranes ( $\leq 77\%$ ). It has been reported as indigestion (14%), gastrointestinal bleeding (34%), nausea ( $\leq 57\%$ ), stomatitis (26%), vomiting ( $\leq 57\%$ ). Pneumatosis cystoides intestinalis is a rare disease characterized by the presence of numerous gas-filled cysts in the intestinal submucosa or subserosa (4). Cysts are most often located in the colon. The etiology of the disease includes high intraluminal pressure, lung diseases, bacterial gas production, malnutrition, chemotherapy and connective tissue diseases. Diagnosis is often made by endoscopy or abdominal computed tomography (5, 6). We present the first case in the literature of ATRA-associated pneumatosis cystoides intestinalis and its management.

### Case Report

A 49-year-old male patient with no known systemic disease and no regular medication use was admitted to our hospital with complaints of fatigue and paleness of the skin. No pathological findings were found in the system query. There were blastic cells containing “fagote “ in the patient's peripheral blood smear. Since the flow cytometry test taken from the patient's peripheral blood was compatible with acute promyelocytic leukemia, the patient underwent bone marrow aspiration and biopsy. ATRA and daunorubicin, cytarabine treatment was started immediately. The patient needed intubation on the 16th day of ATRA treatment due to differentiation syndrome. After fist abdominal tomography was planned for the patient who had abdominal pain on the 9th day of treatment. Diffuse air densities were observed in the wall of the transverse colon (Figure 1). The images were compatible with pneumatosis cystoides intestinalis. Afterwards, the treatment was continued for 4 more days, and when the patient's symptoms included daily weight gain and oral ulcer development, the treatment was discontinued, considering that it might be related to ATRA. Dexamethasone treatment was started.

The patient's abdominal pain subsided within 1 week after discontinuation of ATRA use, and his oral regimen was opened and gradually increased. Dexamethasone treatment was gradually reduced and discontinued. Control abdominal tomography was taken. It was observed that the air values observed in the vicinity of the transverse colon were resorbed.

#### DISCUSSION

ATRA is also associated with many gastrointestinal adverse events such as dry mucous membranes, gastrointestinal bleeding, nausea, vomiting. Pneumatosis cystoides intestinalis is a rare disease and occurs secondary to many conditions such as high intraluminal pressure, lung diseases, and chemotherapy (4-6). There are publications in the literature showing that ATRA use causes scrotal and oral ulcers (7, 8). In our case, we thought that the use of ATRA caused ulcerations in the colon, and the gas formed in the lumen might have passed from these ulcers to the colon wall, causing the formation of gas-filled cyst. In this case, the use of antibiotics and closed oral monitoring at the beginning of the development of pneumatosis cystoides intestinalis did not help in reducing the patient's symptoms. The patient's symptoms improved rapidly after discontinuation of ATRA use. This situation suggests that pneumatosis cystoides intestinalis is related to drug use. Pneumatosis cystoides intestinalis has been reported for many different etiological reasons in the literature. However, in our scans, we did not find any case of this disease developing in association with ATRA use. It is the first case that we think will contribute to literature.



Figure 1: Pneumatosis cystoides intestinalis in transvers colon, gases in transvers colon

## MONOCYTOSIS DEVELOPING IN THE COURSE OF CHRONIC LYMPHOCYTIC LEUKEMIA: CASE REPORT

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Presentation Type : Poster

Abstract Category Adult Hematology Abstract Categories -> Chronic Lymphocytic Leukemia

**Introduction:** Chronic lymphocytic leukemia (CLL) is the most common leukemia in adults. 95 percent are of B cell origin, and it is a malignant disease characterized by the accumulation of mature small lymphocytes in the bone marrow, liver, spleen, lymph nodes and blood. It constitutes 25-30% of all leukemia cases. Monocyte-derived cells, components of the cancer microenvironment, promote chronic lymphocytic leukemia (CLL) cell survival in vitro through direct cell-cell interaction and secreted factors. It is known that circulating absolute monocyte count (AMC) reflects monocyte-derived cells in the microenvironment and that higher monocyte count is associated with increased CLL cell survival in vivo and therefore, it is known to be associated with worse outcomes. Here; We present the case of monocytosis in a 54-year-old female patient with CLL because it suggests recurrence of the disease and is rare.

**Case Report:** A 54-year-old female patient, who had been followed up with the diagnosis of CLL for six years and had no follow-up for 1 year, applied due to mouth sores, weakness and neck swelling that had been present for 10 days. On physical examination of the patient, there were aphthous lesions in the mouth and around the lips, and cervical lymphadenopathies. In simultaneous blood tests, her white blood cell count was 142,000/mL and 70% consisted of monocytes. She also had anemia and thrombocytopenia. Bone marrow biopsy was performed considering there might be leukemic transformation. The biopsy result was compatible with CLL. From the patient's history, it was learned that he had previously received 6 cycles of R-FC, and since it was refractory, he was switched to Ibrutinib treatment. It was learned that he stopped using it due to side effects. Thereupon, R Venetoclax treatment was planned for the patient. In the second week of treatment, the patient's blood count began to improve. At the end of the first month, a decrease in lymphadenopathies was observed. In the fourth month of his treatment, the patient's lymphadenopathies completely regressed and his complete blood count returned to normal. The patient still continues his treatment and follow-up.

**Discussion:** High AMC at diagnosis is associated with poor CLL patient outcomes and accelerated disease progression. Monocyte-derived cells in the CLL microenvironment support CLL cell survival and proliferation. It should be kept in mind that the absolute monocyte count may be increased in chronic lymphocytic leukemia patients compared to the number in normal controls, and this is an indicator of poor prognosis.

## DIAGNOSIS AND TREATMENT OF PRIMARY HEPATIC LYMPHOMA; CASE REPORT

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**Introduction:** Primary hepatic lymphoma accounts for less than 1% of all extranodal lymphomas. It is twice as common in middle-aged males than females. Primary hepatic lymphoma has non-specific symptoms including abdominal pain, fatigue, weight loss and a mass with or without jaundice. There are publications showing an association with hepatitis C. The pathological diagnosis is usually diffuse large B-cell lymphoma or immunoblastic lymphoma. It is a rare disease presenting diagnostic and therapeutic difficulties (1).

Our aim in presenting this case is to draw attention to the diagnosis and treatment of primary hepatic diffuse large B-cell lymphoma.

**Key words:** primary hepatic lymphoma, chemotherapy

**Case report:** A 51-year-old male patient. He was admitted to an external centre about 1.5 years ago with complaints of weight loss and excessive sweating. Due to elevated liver functional tests (KCFT), ultrasonography (USG) and computerised tomography (CT) examinations were performed and a mass was found in the liver. Viral serological tests were negative and the patient was diagnosed as diffuse large B-cell lymphoma NOS (not otherwise specified) with germinal centre phenotype after biopsy. Positron emission tomography (PET) showed a 51\*44 mm geographically circumscribed hypodense lesion at the level of segments 2 and 6-7 of the liver, which was more prominent in late images and showed heterogeneous hypermetabolic activity minimally more than the liver parenchyma (Figure 1) and BCL-2 mutation result T (14:18) (Q32;Q 21) - (IGH/BCL-2): negative. R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, Prednisol) treatment was administered for 6 cycles between May and September 2021. Bone marrow transplantation (BMT) was planned for the patient whose body involvement decreased but did not disappear completely. Then R-ESHAP (Ruximab, Etoposide, Prednisolone, Prednisolone, Cisplatin) CT was administered with mobilisation. After chemotherapy, the patient was evaluated as disease reactivation in the PET council, since the nodular lesions in the liver segments in the PET taken after chemotherapy showed a minimal increase in metabolic activity in late images in the current study. R-GDP (Rituximab, Gemcitabine, Dexamethasone, Sisplatin) was started. Autologous stem cell transplantation (OSCT) preparation was started with CT.

## SYNCHRONOUS CD5 (-) MANTLE CELL LYMPHOMA AND GASTRIC ADENOCARCINOMA: A CASE REPORT

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### Background:

Mantle Cell Lymphoma (MCL) is an aggressive CD5 (+) B-cell malignancy. Up to 10% of the disease lacks CD5 expression. However, limited studies are showing its clinical and biological effects, it has been shown that CD5 negativity improves survival in MCL.

### Aim:

Here, we aimed to discuss a very rare case of synchronous CD 5 (-) MCL and Gastric Adenocarcinoma.

### Case Report:

A 66-year-old male was examined due to anemia and fatigue. In the endoscopy two malignant ulcerated lesions with a sunken center were observed; the first 2 cm in size starting from the proximal corpus, extending towards the greater curvature, and the second 2.5 cm in size starting from the angular incisura and extending towards the lesser curvature (Figure 1A). The biopsy result taken from the lesions was interpreted as an epithelial malignant tumor (a tubular adenocarcinoma) and Ki67 (MIB1) was found to be 80%, P53 (-), MSH 2 and 6 (-), PMS 2 nuclear (+), HER2-neu / CEP17 ratio >2 (Figure 1C). As a result of positron emission tomography performed on the patient for staging purposes, a diffuse increase in metabolic activity in the skeletal system, vertebral, sacroiliac sclerotic, and lytic areas was observed. The widespread involvement in the bone was found to be atypical and the patient underwent bone marrow aspiration biopsy (BMAB). As a result, reticular fiber grade was III, collagen focal (+), immunohistochemistry showed Cyclin-D1 (+), CD5 (-), SOX11 (-), Ki-67 proliferation index was found to be around 5%, and a diagnosis of CD5 (-) MCL, myelofibrosis was made (Figure 1B). A biopsy was taken from the mass lytic lesion in the left sacroiliac region and the result was found to be compatible with CD5 (-) MCL. The diagnosis of early-stage gastric cancer without bone metastasis was confirmed. By searching the literature data on the approach to MCL, it was decided to perform close monitoring without treatment.



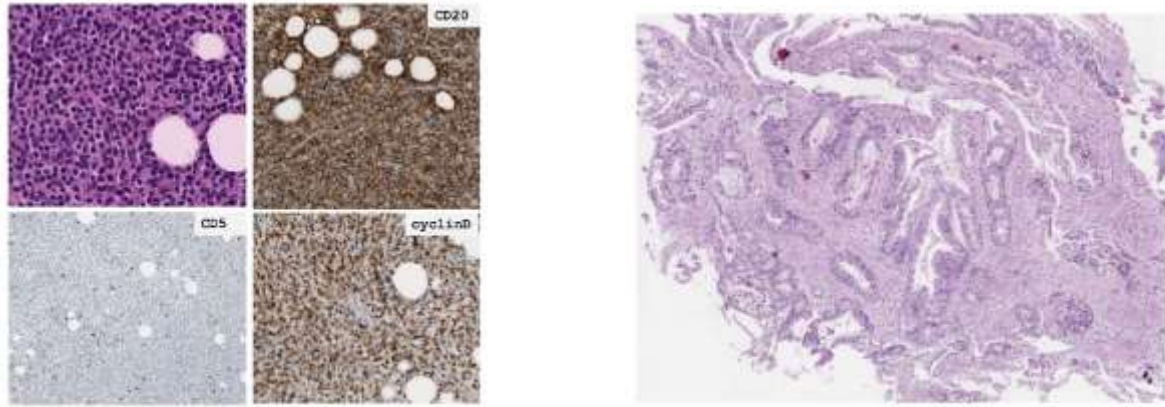


Figure 1.

A: The endoscopy images of the lesions, B: Immunohistochemical analysis of lymphoid cells: CD5 (-); CD20 (+); Cyclin D1 (+); SOX-11 (+),  
C: Microscopic appearance of the lesion.

#### Conclusion:

When the literature was explored, no case report of synchronous gastric adenocarcinoma and CD5 (-) MCL was found. In this case, we emphasized the importance of performing (BMAB) or, if necessary, bone sampling in solid organ malignancies, which are early stages with atypical bone involvement.

## LARGE CELL TRANSFORMATION OF MYCOSIS FUNGOIDES: NOT UNCOMMON, SLIGHTLY BEHIND, SOMETIMES OVERLOOKED

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Mycosis fungoides (MF) is the most commonly encountered primary cutaneous lymphoma. Clinical behavior is usually progressing slowly over many years. Prognosis is based on the extent of body surface area involvement, type of skin lesions, or presence of extracutaneous involvement, as survival with tumor stage (T3) disease is shortened to a median of 35 months.

Large cell transformation (LCT) of MF is a histopathologic evolution of neoplastic small lymphocytes to a clonally identical large cell phenotype, which may occur often at advanced stage but rarely at the patch or plaque stage. The survival is variable among studies which implies a subset of LCT patients has poor outcomes with some prognostic factors.

**Case:** A 65-year-old female patient was assessed for left submandibular lymphadenomegaly with a size of 1.5cm. She had been diagnosed as having MF 12 years ago but had the suspected clinical picture longer than that. Multiple lines therapy including topical steroid, UVB, interferon, and acitretin was used for the disease activity as hyperkeratotic erythematous, squamatic pruritic, plaques, and occasionally with the ulcerations. The left submandibular lymphadenomegaly was the only pathologic lesion with a significant high metabolic activity (SUDmax:24.6) on PET-CT. Lymph node excision was decided. The histology revealed CD3, CD5, CD4, CD30, p63 positive, IRF4/DUSP22 (with FISH) and ALK-1 negative large cell infiltration. LCT of MF was diagnosed. Bone marrow was not involved. Systemic therapy was chosen. According to the reimbursement criteria in the country CHOEP protocol (cyclophosphamide, doxorubicin, vincristine, etoposide, prednisolone) was started. After four courses, she achieved a complete response. She denied autologous stem cell transplantation as consolidation and more two courses of CHOEP were given based on disease aggressiveness. The patient is still in remission and alive for one year.

**Conclusion:** In LCT of MF, there are some defined prognostic factors such as advanced stage at the diagnosis, extracutaneous involvement, high serum LDH level, CD30 positivity, and folliculotropism. Our patient had at least three poor prognostic factors.

Although there are studies indicating poorer outcomes with LCT of MF, we think there is an emerging need to collect the data in the new drugs area.

## A CASE OF CHRONIC MYELOMONOCYTIC LEUKEMIA WITH RECURRENT SKIN INVOLVEMENT

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Presentation Type Oral

Abstract Category Adult Hematology Abstract Categories -> Myeloproliferative neoplasms

### Objective

Chronic myelomonocytic leukemia (CMML) is a clonal disease of hematopoietic stem cells, included in the myelodysplastic/myeloproliferative neoplasia group because it has both myelodysplastic and myeloproliferative features [1]. CMML with skin involvement is rare. Although the skin involvement of CMML is classified differently in different studies, it has been reported as leukemia cutis (LC), blastic plasmacytoid dendritic cell neoplasia (BPDCH), multicentric reticulohistiocytosis and atypical histiocyte/monocyte eruption on the skin. [2,3,4].

### Case Report

While a 71-year-old male patient was being examined for monocytosis, anemia, and hepatosplenomegaly, his tests revealed leukocyte 41000/mm<sup>3</sup>, neutrophil 29000/mm<sup>3</sup>, monocyte 7840/mm<sup>3</sup>, hemoglobin 9.5 g/dL, platelet 75000/mm<sup>3</sup>, and lactate dehydrogenase 712 U/L. The bone marrow blast percentage was 6% at the time of diagnosis. The disease was classified as myeloproliferative type, CMML-1. [5]. NRAS, SF3B1, and TET2 mutations were detected positive. It was evaluated as a high-risk CMML according to the CPSS-mol classification model [6].

Just before azacitidine treatment was started, pink-purple nodular lesions, the largest of which was up to 5 cm, developed on the legs and trunk. The lesion on the left side of the thorax turned into an abscess. A skin biopsy performed with the preliminary diagnosis of LC was not diagnostic due to technical inadequacies. The lesions completely regressed with azacitidine treatment under appropriate antibiotic therapy. Before the fourth course of azacitidine treatment, the same lesions recurred in similar locations. Meanwhile, the monocyte level increased from 1500 to 5600/mm<sup>3</sup>, without worsening of anemia and thrombocytopenia, and without an increase in hepatosplenomegaly. When the skin lesions appeared for the second time, the bone marrow biopsy performed with the preliminary diagnosis of transformation into acute leukemia was again compatible with CMML-1. The second skin biopsy, including deeper layers, showed cells with large cytoplasm, thin chromatin, and a partially large nucleus in the epidermis. These cells were CD68+ and showed suspicious staining with CD14, but did not show staining with MPO, c-kit, CD34, TdT, CD20, CD30, and CD15. It was evaluated as skin infiltration of CMML. The patient, to whom 1000 mg/day hydroxyurea was added to azacitidine

treatment, completed 7 courses of azacitidine treatment with a 'clinical benefit' [7]. Skin lesions disappeared with the combination of hydroxyurea and azacitidine. The patient died from respiratory failure due to influenza pneumonia.

### Conclusion

Skin involvement with various histopathological features may be observed in the course of CMML. It is important not to miss LC and BPDCN due to their impact on prognosis and warning about switching to more intensive treatments.

The case we present is important because CMML has skin involvement without progression criteria such as an increase in blast percentage or organomegaly. The regression of the lesions upon initiation of treatment, their recurrence while receiving active treatment, and their regression again when hydroxyurea is added may be an indication that treatment choices should be re-evaluated when skin infiltration occurs.

## A RARE CASE OF PLASMABLASTIC LYMPHOMA PRESENTING WITH A BULKY RENAL MASS IN AN IMMUNOCOMPETENT PATIENT

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Plasmablastic lymphoma (PBL) is a rare and aggressive subtype of large B-cell lymphomas. PBL patients are predominantly immunocompromised, or HIV infected. Activated B cells undergoing somatic hypermutation and class-switching recombination, are believed to be the tumor cell origin. Histologically, the tumor exhibits a diffuse growth pattern leading to impairment of structure with large, atypical cells resembling immunoblasts. The specificity of PBL cells is lacking B-cell markers (CD19, CD20, and PAX-5) expression and having an immunophenotype similar to plasma cells such as showing CD38, and CD138 positivity. EBV-encoded RNA (EBER) has been seen in around 80% of cases and over 66% of cases present MYC rearrangements.

More than half of PBL arises in the oral cavity but may develop in various tissues or organs and it has been shown regardless of the location and presence of HIV and EBV, PBLs display comparable clinicopathologic features.

Intensive treatment protocols such as DA-EPOCH with the addition of new drugs like bortezomib are recommended treatment for fit patients.

A 59-year-old HIV-negative male presented with decreased urine output, high creatinine (5,85 mg/dl), high serum LDH level (588 IU/L) levels, and anemia (Hb: 6,8 g/dL, MCV:88fL) with bilateral grade 2 hydronephrosis on USG. PET-CT showed a bulky mass in the pelvis sized 18\*15\*25cm which covered nearly the right half of the pelvis and infiltrated the renal parenchyma (SUDmax:7,5), and a mass sized 30\*24 mm in the liver (SUDmax:4,5). A biopsy of the kidney was decided. The histology revealed diffuse large-cell infiltration. The infiltrating cells were CD20, CD3, and PAX5 negative but CD138, and MUM1 positive with a high Ki67 labeling of 90%. EBER expression by in situ hybridization was strongly positive (70%) and clonality by kappa light chain restriction was documented.

He had a monoclonal gammopathy of IgG kappa type. Bone marrow did not show any evidence of lymphoproliferative disease, or plasma cell neoplasia. The karyotyping result was normal. DA-EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin) with bortezomib was chosen as treatment. The response was rapid.

After two courses of chemotherapy serum creatinine decreased to normal levels (0,6 mg/dl) and hemoglobin increased to 8,4 g/dL.

**CONCLUSION:** In our case, PBL arises with a rare form as developing in immunocompetent biology, and in extraoral locations as huge masses.

Intensive chemotherapy combined with antimyeloma drugs such as proteasome inhibitors, immunomodulators, and monoclonal antibodies provides significant improvement in PBL but the management of PBL remains challenging.

The aggressive nature and propensity for relapse dictate there is still a need for clinical studies with large case series especially patients with relapsed and resistant PBL.

## SYSTEMIC AA AMYLOIDOSIS; AS A COMPLICATION OF IDIOPATHIC MULTICENTRIC CASTLEMAN'S DISEASE

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Castleman disease (CD) is a chronic inflammatory process with polyclonal lymphoproliferation resulting in enlarged lymph nodes that can be unicentric or multicentric. A subset of multicentric CD (MCD) is caused by human herpesvirus-8 (HHV-8). HHV-8-negative MCD cases are named idiopathic (iMCD). While the exact etiology of iMCD is not clear yet, dysregulated interleukin-6 (IL-6) expression with high circulating IL-6 concentration seems to be a major contributor, especially for systemic inflammation-related symptoms and generalized lymphadenopathy.

Persistent inflammatory response may be complicated by systemic AA amyloidosis. In AA amyloidosis amyloid fibrils are derived from an acute phase reactant protein serum amyloid A (SAA) protein. IL-6 is one of the major stimulants of SAA. In general, CD disease-associated AA amyloidosis regarding reports are scant in the literature.

Case: A 48-year-old male patient was referred as having treatment-refractory iMCD with multiple mediastinal lymph nodes, emerging monoclonal gammopathy, and massive proteinuria.

In the past, he had been evaluated for infertility and had been found carrying the uracil-DNA glycosylase coding UNG gene mutation as with two siblings. The latter can be associated with a heterogeneous group of congenital and acquired conditions classified as hyperimmunoglobulin M syndromes, stratified in primary immune deficiencies (PID). The patient's story was not consistent with PID.

He had received three cycles of corticosteroids and rituximab for iMCD, which did not provide a significant improvement.

He was exhausted, dyspneic, and striking skinny. Based on the laboratory results as anemia (Hgb: 8,2 gr/dL), high serum CRP:166 mg/L, serum creatinine (3,2 mg/dL) level, hypoalbuminemia (1,6 gr/dL), and massive proteinuria (12 gr/ day), a kidney biopsy was decided. He had monoclonal gammopathy as IgM lambda and IgG kappa type and 0,46 g/dL M-spike on serum protein electrophoresis. Serum IgG and IgA levels were within normal but serum IgM was increased (365mg/dl). Abdominal fat aspiration was found to be amyloid negative. Kidney histology was consistent with AA amyloidosis. Severe iMCD-associated AA amyloidosis was diagnosed. Based on blocking the downstream effects of IL-6, an anti-IL-6 receptor monoclonal antibody tocilizumab which was available in the country, was chosen for treatment. After three courses of tocilizumab, the laboratory findings showed significant improvement (Hgb:8,5 gr/dL. serum CRP <0,6 mg /L, creatinine:2,51 mg/dL, albumine:3.15 gr/dL, M-spike: 0,2 g/dL, and massive proteinuria:9,1 g/day) and regressed mediastinal lymph nodes.

Conclusion: Experience with systemic AA amyloidosis complicated iMCD is scarce. As in our case, anti-IL-6 receptor monoclonal antibody treatment led to improvement in constitutional symptoms and a decrease in acute phase response which may result in regression of amyloid deposits and organ dysfunction.

## PRIMARY HEMOPHAGOCYtic LYMPHOHISTIOCYTOSIS IN AN ADULT FEMALE; GRISCELLI SYNDROME TYPE 2 (GS2) WITHOUT HYPOPIGMENTATION

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Hemophagocytic lymphohistiocytosis (HLH) is an immune dysregulation as inappropriate activation resulting in increased pro-inflammatory cytokines production which leads to systemic activation of macrophages and the subsequent cellular destruction.

In primary HLH, there are genetic mutations that impair the interaction between NK cells, CD8+ cytotoxic T-cells, and antigen-presenting cells. Primary HLH heritability is variable as X-linked, recessive, or dominant. It can be associated with clinical syndromes. Forms without an associated genetic syndrome are referred to as familial HLH.

Griscelli syndrome (GS) type 2 is a rare autosomal recessive disorder, due to a mutation in RAB27A gene. It associates with partial albinism, and immune deficiency which leads to recurrent infections and HLH. Occasionally, GS-2 patients with RAB27A mutation display normal pigmentation.

**Case:** A 22-year-old female patient was referred with unresolved fever for 1 month. She describes two other clinical pictures resembling this, which were without any definite diagnosis and improved with supportive treatment.

She was exhausted and pail. Mild splenomegaly was palpated. Laboratory tests revealed pancytopenia (WBC:200x10<sup>6</sup>/L neutrophil:10x10<sup>6</sup>/L Hgb:8 g/dL PLT:79000x10<sup>6</sup>/L), elevated transaminase (ALT:237 U/L, AST:171 U/L), C-reactive protein (105mg/L), ferritin (4035 ng/mL), triglyceride (320 mg/dL) levels. On abdominal ultrasound, the spleen was 155 mm, and the peripheral smear showed phagocytosed red blood cells. HLH was diagnosed. Since she was an adult first step was screening for secondary HLH causes. All possible causes as infections, autoimmune diseases, or malignant disorders were ruled out. PET-CT showed widespread, increased *bone marrow and spleen* activity with generalised lymphadenomegalies which seems to be reactive.

With suspicion of primary HLH genetic analysis was performed and dexamethasone was started. However, a homozygous mutation in the RAB27A gene was detected which is known as causing a form of the syndrome designated type 2 GS. The treatment was converted according to the HLH-2004 protocol. The clinical picture recovered totally. An allogeneic stem cell transplantation (SCT) was decided.

**Conclusion:** The treatment of HLH is designed to halt any underlying trigger and control the overactive immune system. There are no prospective trials guiding treatment of HLH in adults. It is believed that adults should undergo allogeneic SCT if they have refractory or relapsed disease. On the other hand, the prognosis of GS2 is poor, and SCT is the only curative treatment. Thus, we conclude RAB27A mutations should be investigated in patients with suspected HLH disease which are mostly the case in point for pediatric patients.

## PRIMARY CUTANEOUS FOLLICLE CENTER CELL LYMPHOMA FOLLICULAR TYPE WITH HIGH KI-67 INDICES ARISING ON THE FOREHAD

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While primary cutaneous lymphomas (PCL) are the most frequent extra-nodal lymphomas, primary cutaneous B-cell lymphomas (PCBCL) are relatively rare. Chronic antigenic stimulation is one of the reasons thought to be involved in the pathogenesis. According to the WHO and EORTC classification, three subtypes were defined as primary cutaneous marginal zone lymphomas, primary cutaneous follicle center cell lymphoma (PCFCL), and primary cutaneous large B-cell lymphomas, from which PCFCLs are the most encountered.

Painless, violaceous, solitary or grouped nodules, plaques, and papules are the typical lesions that are not ulcerated. They may occur in all body areas but mostly in the head and neck region. They are rarely multifocal.

Differential diagnosis involves especially systemic nodal follicular lymphomas with secondary cutaneous involvement. The clinical findings and strong CD10 and bcl-2 expressions are in favor of follicular lymphomas.

Case: A 60-year-old male patient referred rapidly growing nodular grouped lesions on the forehead with a size of 10 cm. The lesion was painless and not itchy. He had no constitutional symptoms. He revealed a history of cutaneous B-cell non-Hodgkin lymphoma on the right shoulder and to be treated with combined chemotherapy which provided complete remission after six courses 20 years ago.

Laboratory test results were within normal limits including blood cell counts, serum LDH, CRP level, and kidney and liver function analysis. Histological examination revealed CD20, CD23, CD10 positive and CD3, CD5 negative non-epidermotropic centrocyte infiltration with some centroblasts. Ki 67 proliferation index was 70%. There was no involvement other than the lesion in the PET-CT imaging. PCFCL was diagnosed. Radiotherapy was not appropriate due to the size and depth of the lesion. We started treatment with Rituximab plus Bendamustin regimen. The relapse of the lymphoma diagnosed in the past was not considered, but it was thought that both may have the same origin.

Conclusion: PCFCL is considered an indolent lymphoma with a good prognosis. 5-year disease-specific survival is  $\geq 95\%$ . Some cases may need as in our case, debulking treatment which may also be curative.

## LONG-TERM REMISSION AFTER NELARABINE AND DONOR LYMPHOCYTE INFUSION IN A T-ALL PATIENT RELAPSED AFTER ALLOGENEIC HEMATOPOIETIC STEM CELL TRANSPLANTATION

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An 19-year-old female presented with a bulky mediastinal mass and diagnosed with T-cell acute lymphoblastic leukemia (T-ALL). After Hyper-CVAD chemotherapy, she underwent allogeneic hematopoietic stem cell transplantation (AHSCT) from matched unrelated donor in CR1.

Conditioning regimen was Bu/Cy (myeloablative) and GVHD prophylaxis was short-term methotrexate, cyclosporine and ATG-F. One year after APSCT, 35x32 mm solid mass was observed on the left kidney, concurrent with >95% chimerism. Histopathological examination revealed TDT+ acute lymphoblasts. Bone marrow biopsy was normocellular. No response was obtained with FLAG regimen. Then, nelarabine (1500 mg/m<sup>2</sup> 1.3.5.days every 21 days) with escalated dose donor lymphocyte infusion (DLI) was performed (CD3:5x10<sup>6</sup>/kg, 10x10<sup>6</sup>/kg, 20x10<sup>6</sup>/kg, 50x10<sup>6</sup>/kg). Complete remission was obtained with this therapy and complicated with reversible peripheral neuropathy. The patient is in CR2 for about 4.5 years.

The graft-versus-leukemia (GVL) effect associated with DLI for relapsed hematologic disease after AHSCT is a clinically well-established phenomenon; yet, response rates for most diseases remain low. Therapeutic options for relapsed T-ALL are very limited, especially when disease recurs in the first year after AHSCT. We conclude that the combination of nelarabine and DLI offers a promising approach in relapsed T-ALL post-HSCT, where treatment options are very limited.

## SECONDARY SOLID TUMOR AFTER ALLOGENEIC HEMATOPOIETIC STEM CELL TRANSPLANTATION: A CASE REPORT

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**Background:** We present a case of 56-year-old male diagnosed with Acute Myeloid Leukemia (AML) 21 years ago. He underwent allogeneic hematopoietic stem cell transplantation (AHSCT) from HLA matched sibling donor in CR2 following the conditioning regimen BU/CY plus short-term methotrexate and cyclosporin as Graft Versus Host disease (GVHD) prophylaxis 19 years ago. He has not been complicated with acute or chronic GVHD.

**Case Presentation:** In Jan 2024, he developed massive ascites. Non-portal hypertension type ascites was detected. The analysis of ascitic fluid was consistent with adenocarcinoma. Serum CA 19-9 and CA 125 levels were elevated suggesting gastrointestinal, biliary tract or pancreatic tumor. PET scan was inconclusive to detect primary origin of the tumor. Further investigations including endoscopy, endoscopic ultrasound and colonoscopy were planned.

**Conclusion:** Secondary solid tumors (SST) are a rare but well-defined late complications of AHSCT. SST occur in up to 15% of patients within 15 years of AHSCT with myeloablative conditioning, without reaching plateau. The most common SSTs include squamous cell carcinoma of skin, genitourinary tract and oral cavity; lung and breast cancers. Risk factors of SSTs were reported as smoking, alcohol intake and use of myeloablative conditioning regimens. SSTs are common causes of nonrelapse mortality in long-term survivors and may account for up to 10% of late deaths. Cancer prevention guidelines are largely consensus-driven and follow the recommendations for general population. Herein we report a case diagnosed with SST 19 years after AHSCT bearing 30 pack-year smoking and use of myeloablative conditioning regimen as risk factors.