of mantle cell lymphoma who developed autologous GVHD after ASCT, not previously reported. Fifty-four-year-old male patient was diagnosed with Stage 4A Mantle cell lymphoma according to the Ann Arbor staging system based on the result of inguinallymph node excisional biopsy performed in July 2021. Three courses of R-CHOP,DHAP treatment was started for the patient who did not respond after R-CHOP treatment. Complete response was achieved after three cycles of R-DHAP therapy,and mobilization was performed with filgrastim and plerixafor protocol. Then, autologous SCT was performed with the BEAM protocol(Karmustine 300mg/m2, Etoposide 200mg/m2, Cytosine Arabinoside $2 \times 100$mg/m2, Melphalan 140mg/m2). Post-transplant neutrophil engraftment occurredon the+14th day and platelet engraftment on the +32nd day. Piperacillin Tazobactam was started due to fever and sore throat on the 3rd day of ASCT. Teicoplanin was added to the treatment after the fever persisted on the 5th day of ASCT. Piperacillin Tazobactam treatment was discontinued due to acute phase reactant increaseand fever on ASCT +7th day and Meropenem was started. Teicoplanin treatment was stopped on the +16th day after transplantation, and Meropenem was stopped on the +17th day. On the +14th day of autologous SCT, complaints of itching and rash started on the patient's body The biopsy result of the patient who underwent skin biopsy was compatible with grade 1-2 GVHD The patient was started on high-dose corticosteroid therapy. On the+21st day, the complaint of itching and on the +28th day, the skin findings disappeared. On the +28th day of ASCT, corticosteroid therapy was tapered and discontinued. The patient is still being followed without disease. **Conclusion:** Autologous GVHD is an autoimmune syndrome initiated by autoreactive T cells that recognize major histocompatibility complex (MHC) class II antigens.CD8+ T cells recognize MHC class II determinants .There are three types of autologous GVHD: 1. spontaneous autologous GVHD 2.induced autologous GVHD; using cyclosporine, tacrolimus, interferon-$\alpha$, interferon-y and alemtuzumab to induce the effect of GVT; GVHD induced by transfusion of non-irradiated blood products in patients with Hodgkin lymphoma, Non-Hodgkin Lymphoma (NHL), chronic lymphocytic leukemia, and acute myeloid leukemia 3. induced by transfusion of non-irradiated blood products.In the study of Drobyski et al. in 2008, it was reported that autologous GVHD developed in 5 of 386 patients who underwent autologous SCT. Response to steroids was not good. It was the first transplant of 223muliple myeloma patients. Only 2 patients developed autologous GVHD. Autologous GVHD developed in 3 of 27 patients with a second transplant.Autologous GVHD did not develop in Hodgkin lymphoma, non-Hodgkin lymphoma and AML patients. Therefore, it will be important to consider this complication while planning the second autologous stem cell transplant in these patients .Our case presents skin GVHD developing after autologous SCT. However, since our patient has lymphoma and/or did not take a proteasome inhibitor before, it is important by distinguishing it from other cases.When the literature is examined, it is thought that the preparation regimens and post-transplant CsA application prepare the ground for autologous GVHD In addition, it is known that patients with hematological malignancies who are female, given high-dose CD34+ stem cells,bortezomib, lenalidomide, pomalidomide and alemtuzumab used in their pre-transplant treatments are at risk for autologous GVHD.

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## OP 14

## ACUTE GRAFT-VERSUS-HOST DISEASE (AGVHD) PRESENTING AS STEVENS-JOHNSON SYNDROME (SJS)

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**Case report:** This case report describes a 34-year-old male with AML, intermediate risk, initially treated with standard 7 +3 induction chemotherapy followed by high-dose cytarabine consolidation. Despite achieving medullary remission, minimal residual disease(MRD) persisted. The patient underwent allogeneic-HSCT from an HLA-matched sibling donor. At the 33rd-month post-transplant, the patient, in full donor chimerism, developed a pituitary macroadenoma and hypopituitarism, alongside CNS relapse and medullary remission was confirmed in the bone marrow. Management included cranial radiotherapy and pituitary hormone replacement. Subsequent bone marrow relapse was treated with salvage chemotherapy (high-dose cytarabine and mitoxantrone), achieving medullary remission. Persistent CNS disease necessitated intrathecal triple therapy until cerebrospinal fluid clearance. MRI response was also obtained. A second allogeneic HSCT was performed from the same donor using a myeloablative FLU-TBI conditioning regimen. GVHD prophylaxis consisted of Cyclosporine-A and Methotrexate. On post-transplant day +25, the patient presented with severe cutaneous manifestations with some bullous lesions initially suspected as aGVHD or drug eruptions. The patient was initiated on a high-dose corticosteroid (2 mg/kg prednisolone) as a primary treatment. However, due to an inadequate response, ruxolitinib(10 mg twice-daily) was added to the treatment regimen after the first week. Dermatological evaluation raised suspicion of SJS, leading to IVIG administration. The skin biopsy report indicated the possibility of grade 2 GVHD, although the possibility of a drug reaction could not be excluded. Significant clinical improvement was observed within one week of ruxolitinib initiation. Corticosteroids were tapered over six weeks to physiological replacement doses. Ruxolitinib was continued for 56 days before gradual discontinuation. Cyclosporine was maintained with target trough levels of 100-250 ng/mL and

discontinued on day +90 post-transplant. This case highlights the diagnostic challenges in differentiating aGVHD in the post-HSCT setting from SJS/TEN-like presentations. It emphasises the importance of rapid intervention and the potential efficacy of JAK inhibitors in steroid-resistant cutaneous GVHD.

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**Adult Hematology Abstract Categories**

**Other Diseases**
**OP 15**

## RAB27A MUTATION AND EBV INFECTION ASSOCIATED HEMOPHAGOCYTIC SYNDROME: A CASE REPORT

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**Objective:** Hemophagocytic lymphohistiocytosis (HLH) is a nonmalignant immune regulation disorder within the histiocytosis group of diseases. It is a clinical condition in which there is fever, hepatosplenomegaly and cytopenia due to dysfunction of cytotoxic T-lymphocytes and natural killer (NK) cells, activation of macrophages and T-lymphocytes, excessive production of proinflammatory cytokines and hemophagocytosis. It can be primary (familial) and secondary. HLH may also be seen in the course of some immune deficiencies. We aimed to present a case of HLH that developed due to EBV infection and Griscelli syndrome with RAB27A mutation. **Case Report:** An 11-month-old male patient who presented with complaints of fever and swelling in the neck was hospitalized with respiratory distress and poor general condition. On physical examination hepatosplenomegaly, cervical lymphadenopathy and silver-gray hair color was detected. It was learned that the patient had a sibling who died at 3 months old with a similar phenotype. In his tests; wbc:10600/mm3, neu:5000/mm3, lympho:5100/mm3, hb:6.3 gr/dl, plt:29,000/mm3, ALT:167 IU/l, AST:410 IU/L, total bilirubin:2.7 mg/dl, direct bilirubin:2.6 mg/dl, sodium:126 mmol/L, albumin:2.3 g/L, LDH:765 U/L, fibrinogen:69 mg/dl, ferritin: 59334 ng/ml were detected. Hemophagocytosis was observed in the bone marrow. The patient was started on the HLH 2004 chemotherapy protocol, but died within the first 24 hours of treatment. The patient's tests at the time of admission showed EBV VCA IgM: 7.77 (positive), EBV PCR: 72,000 copies. A homozygous c.149delG (p. Arg50Lysfs*35) mutation was detected in the RAB27A gene. **Conclusion:** Griscelli syndrome is a rare autosomal recessive disease that can be accompanied by silver-gray hair, hypopigmentation, recurrent fever and infections, immune deficiency and neurological disorders. Primary HLH is common in our country due to the high prevalence of consanguineous marriages. The patients who cannot be diagnosed early and develop HLH can be fatal, so early diagnosis of these patients is of vital importance.

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**OP 16**

## A CASE OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN A CHILD: A RARE AND CHALLENGING DIAGNOSIS

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**Case report:** Hemophagocytic lymphohistiocytosis (HLH) is an aggressive and life-threatening increased inflammatory syndrome characterized by over-activation of the immune system. Although it is more common in the first years of life, it can be seen in children and adults of all ages. HLH is divided into primary (familial) and secondary (sporadic). Secondary HLH may be associated with malignancy, rheumatologic diseases, chemotherapy or immunosuppressive therapies, but the most common cause is infections. Many viral, bacterial, fungal and protozoa infections can be triggers for both secondary and familial HLH. A 7-year-old male patient was admitted to our clinic with complaints of fever, malaise and anorexia. Blood tests revealed WBC:2260/mm$^3$, Hb:7.1 g/dL, ANS:920/mm$^3$, Platelets:41.000/mm$^3$, Fibrinogen:127 mg/dL, Triglycerides:247 mg/dL, Ferritin:687 ng/mL and splenomegaly on physical examination. Bone marrow aspiration (BMA) examination revealed hemophagocytosis. HLH was diagnosed according to HLH-2004 criteria and HLH-2004 protocol including dexamethasone and cyclosporine treatment was started. Fever, cytopenia and splenomegaly developed again during outpatient follow-up of the patient who responded adequately to the treatment. Ferritin level increased above 3000 ng/mL and another BMA was performed. This time, amastigotes were seen in addition to hemophagocytosis in the BMA evaluation. Leishmania rapid diagnostic test (RK-39) and PCR were positive. Liposomal amphotericin B treatment failed to elicit an adequate response. Meglumine Antimoniate (Glucantime) treatment was then initiated. No mutation was detected in the HLH genetic examination sent at the time of