

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/mm³, Hb:7.1 g/dL, ANS:920/mm³, Platelets:41.000/mm³, 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