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HEMOPHAGOCYTIC SYNDROME: A REPORT OF 6 PATIENTS IN A 15-YEAR RETROSPECTIVE STUDY

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Hemophagocytic syndrome is a class II non-Langerhans cell lymphohystiocytosis (HLH) that is distinguished in familial type (FHL) and secondary group of diseases, which are infection (IAHS), malignancy (MAHS) and rheumatic disease (RAHS) associated syndromes. We report six children with HLH during a 15-year retrospective study. Age of onset was 27.83±31.24 months. All patients were hospitalized with FUO. Initial clinical findings were hepatosplenomegaly, rash, serositis. Laboratory investigation revealed anemia, neutropenia, trombopenia. Liver function tests, ferritin, triglycerides were elevated. Fibrinogen was low. Myelogram showed hemophagocytosis. Blood cultures were negative. Serology tests were indicative for EBV in 2 patients and CMV in 1. Treatment included gancyclovir for 2 patients with EBV and 1 with CMV infection. Gamma globulin and corticosteroides were administered to all and etoposide to 3 of them. Disease duration was 1.91±0.49 months. Follow up period is 5 months to 7 years. Three children, 2 with EBV infection and 2 without documented cause of the disease are considered to have VAHS and remain asymptomatic. One child died with central nervous system hemophagocytosis and it is speculated to have FHS. The last child continues with fever and rheumatic rash, but hepatoslenomegaly resolved and hemoglobin, triglycerides and fibrinogen are normalized, while leucocytes and platelets are increased. Possible systemic juvenile idiopathic arthritis is regarded as underline condition. Conclusion: HLH is a condition of consideration in FUO. The etiology is diverse as well as outcome. Prognosis is poor especially for FHS for which recent genetic analysis achievement helps to diagnosis, close follow up and treatment.