

## **RECURRENT HEMOPHAGOCYTTIC LYMPHOHISTIOCYTOSIS SYNDROME (HLH) IN A PATIENT WITH COMMON VARIABLE IMMUNE DEFICIENCY**

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Autoimmune features have been described in common variable immune deficiency. We describe a 13 year old male with three episodes of symptoms consistent with hemophagocytic lymphohistiocytosis (HLH). The patient presented at ten months of age with idiopathic fulminant hepatitis and splenomegaly. Extensive workup was negative for autoimmune or infectious etiologies. At age 7 he developed a febrile illness with sequestration (Hgb 6.8, WBC 1.7, ANC 250, platelets 14,000). Hypogammaglobulinemia and impaired specific antibodies were noted. Recurrent febrile illnesses resulted in splenomegaly and pancytopenia. In 1998, the patient developed hepatitis, atypical lymphocytosis, and hemophagocytosis. Liver biopsy revealed periportal lymphocyte expansion with nodule formation. He responded to corticosteroids. Recurrent hemophagocytosis episode occurred in 2002 after an EBV viral illness requiring high dose IV steroids. Liver biopsy in 2003 revealed loss of bile ducts and periportal fibrosis. In 2004, the patient developed fever, lymphocytosis, anemia, thrombocytopenia, hypofibrinogenemia, hypertriglyceridemia and hepatomegaly with coagulopathy. HLH-2004 therapy including etoposide pulses, dexamethasone, cyclosporine and intrathecal hydrocortisone was initiated. He went into remission and therapy was discontinued. There was no mutation in Perforin 1 gene (PRF1) or SH2D1A. The patient relapsed in November 2004 after a febrile illness. Reinduction was commenced and the patient is now in his second remission. Unrelated stem cell transplant is being considered. We conclude that HLH can be a life threatening complication in CVID.