

Response Letter for Case Report Entitled “Pregnancy Induced Haemo-phagocytic Syndrome (HPS)”

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To the Editor,

Thanks for your interest in the case report entitled “Pregnancy Induced Haemo-phagocytic Syndrome (HPS).”

The Histiocyte Society HLH study group revised the diagnostic guidelines of HLH [1]. The diagnosis of HLH is established either by a molecular diagnosis consistent with HLH (primary HLH) or by having five out of eight parameters. They are fever, splenomegaly, cytopenia affecting more than two cell lineages, hypertriglyceridemia and/or hypofibrinogenemia, hemophagocytosis in the bone marrow, spleen, or lymph nodes without evidence of malignancy, low or absent natural killer cell cytotoxicity, hyperferritinemia, and elevated soluble CD25.

The common causes of HPS like infections, autoimmune, or malignant diseases were not present in this patient. Even after the diagnosis of haemo-phagocytic syndrome was established, the pregnancy as the primary cause for HPS was not thought. According to criteria [1],

she was suffering from HLH because patient had fever, splenomegaly, cytopenia more than two cell lineages, hypertriglyceridemia, hyperferritinemia, and hemophagocytosis in bone marrow. The dramatic recovery after abortion led us to think it to be a pregnancy Induced. There was no need of any medications or interventions in this woman.

Liver biopsy was done once platelets counts recovered subsequent to abortion. We also agree that for the diagnosis of HPS, only demonstration of haemo-phagocytosis is not necessary [1].

Reference

1. Henter JI, Aricò M, Elinder G, et al. Familial hemophagocytic lymphohistiocytosis. Primary hemophagocytic lymphohistiocytosis. *Hematol Oncol Clin North Am.* 1998;12:417–33.