No. 24

Case series of Hemophagocytic lymphohistiocytosis in Buali Hospital, Ardabil, Iran in July 2018

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Background and Objective

we present an one year-old boy and a 4-month old girl from Iran with hemophagocytic lymphohistiocytosis (HLH) associated with Leishmania. HLH is not an independent disease but rather a life-threatening clinical syndrome that occurs in many underlying conditions and in all age groups[1].It is commonly appears in infancy[2]. In our cases, treatment with was dexamethasone and Etoposide started and second case responded and first case unfortunately died.

Case1

a one-year-old boy was admitted to the Buali Hospital, Ardabil, Iran on July 2018, with a 3-week history of fever, Hepatosplenomegaly and pancytopenia. Serum testing showed elevated transaminase levels (ALT:750,AST:820), hypertriglyceridemia (TG:420), hyperferritinemia (Ferritin:9780) and fibrinogen level was less than 50 unit. Hemophagocytic lymphohistiocytosis was diagnosed on bone marrow examination that showed HLH with Leishmania body (fig-1). The patient was tested for various infectious agent that all of them were negative expect Leishmania, (HBS Ag: negative HCV Ab: negative HIV Ab: negative), DAT for Leishmania was positive. After one week she doesn't have response to antibiotic.Glucantime was added. But unfortunately he didn't response to treatments and died.

Case2

4 month old girl was admitted to the hospital because of fever, pallor and Hepatosplenomegaly for 2 week. Intermittent high grade fever persisted with anorexia and weight loss. Examination revealed fever, pallor and petechiae. Abdominal examination revealed Hepatosplenomegaly. Respiratory, cardiac and neurological were normal. Lab data showed pancytopenia, liver function tests were abnormal (ALT: 672 AST: 1449 ALP: 386 Bill Total: 8 Bill Direct: 3.6). Ferritin was very high (16493). Coagulogram revealed coagulopathy and hyperfibrinogenemia. Renal

function and electrolytes were normal. Abdominal sonography showed Hepatosplenomegaly. DAT WAS 1/1600. Fever persisted and Glucantime was started. Bone marrow examination (BMA) revealed hem phagocytosis with leishman bodies. (Fig-1) Treatment with dexamethasone and Etoposide result in a dramatic resolution of all signs and symptoms within 10 day.

Conclusion

In conclusion, HLH secondary to Leishmania is extremely rare and potentially fatal. The diagnosis is often missed due to overlapping clinical features and negative bone marrow. A high index of clinical suspicion, repeated marrow evaluation with culture and or serology is often required to establish leishmaniasis [6]. Leishmania must be considered and excluded in patients with HLH before immunosuppression is considered.as a result we understand Treatment with dexamethasone and Etoposide provides a dramatic response in these cases.

Keywords: Visceral leishmaniasis, Hemophagocytic lymphohistiocytosis, Hepatosplenomegaly