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Case Report
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HIV Infection in Child Presenting as Hemophagocytic Lymphohistiocytosis

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Abstract

A 9 year boy presented with fever and lymphadenopathy of two year duration. Bone marrow examination revealed haemophagocytic lymphocytes. A diagnostic criterion for Hemophagocytic lymphohistiocytosis was fulfilled. HIV was positive. There was no evidence of any other associated infection. Child improved on therapy.

Keywords: Hemophagocytic lymphohistiocytosis; HIV; Child

Introduction

Hemophagocytic lymphohistiocytosis (HLH) is a rare but potentially fatal disease of overactive histocytes and lymphocytes. It is characterized by fever, lymphadenopathy, hepatosplenomegaly, panocytopenia and severe constitutional symptoms. HLH may be primary or secondary to systemic infections, malignancy, immunodeficiency and / or drug therapy. Infection has an important role in the etiology of HLH [1]. HIV infection alone or in concert with other infection and malignancy has been reported to be cause of HLH in adults [2,3]. Although there has been reports of other infection causing HLH in HIV infective children, HIV alone causing HLH in children has been rarely reported. We report ko a case of HLH caused by HIV in a 9 years old boy.

Case Presentation

A 9 yr boy presented with fever, swellings in cervical and inguinal region and abdominal distension of two year duration. He had history of recurrent infections and paleness of body for last one year. Child had received blood transfusion a year back. Two of his siblings had expired at one and four year of age. Physical examination revealed pallor, generalized lymphadenopathy and hepatosplenomegaly.

Laboratory investigations revealed a hemoglobin 7.8 g/dl, TLC 1800/mm^3($P_{50}L_{45}M_3E_1$) Platelet -75000/mm^3, peripheral smear revelaed dimorphic anemia, ESR 92 mm, urea 30 mg/dl, creatinine 0.7 mg/dl, blood culture and urine culture were sterile. Results of fungal and mycobacterial culture were negative. Bone marrow examination revealed fair number of histiocytes, few showing lymphophagocytosis with fair number of degenerated cells suggestive of HLH. Serum ferritin levels were 159.40 ng/ml (7.00-140.00), Serum Triglyceride-253 mg/dl (<150.0).

Ultrasound abdomen revealed splenomegaly with enlarged mediastinal lymph nodes. HbSAg was negative, Antinuclear antibodies and anti dsDNA did not reveal any abnormality. Results of HIV ELISA test were positive. Subsequently mother was tested for HIV and the result came out to be positive. The patient was started on highly active antiretroviral therapy (HAART). The patient recovered after institution of HAART therapy.

Discussion

The incidence of HLH is estimated to be approximately 1.2 cases per million individuals per year. According to HLH 2004 criteria [4], the diagnosis of HLH is established if five out of eight following diagnostic criteria are met: (i) fever, (ii) splenomegaly, (iii) bicytopenia (with at least two of the following – (i) Hb <9 g/dl, (ii) platelets <100000/µl and neutrophils <1.0 x 10^3/µl) , (iv) hypertriglyceridemia (>265 mg/dl) or hypofibrinogenemia (≤150 mg/dl), (v) hemophagocytosis in bone marrow, spleen or lymph node without evidence of malignancy, (vi) low or absent NK cell activity, (vii) hyperferritinemia (≥500 µg/l) and (viii) increased soluble plasma CD25 levels.

This child had fever, splenomegaly, bicytopenia and hypertriglyceridemia and bone marrow

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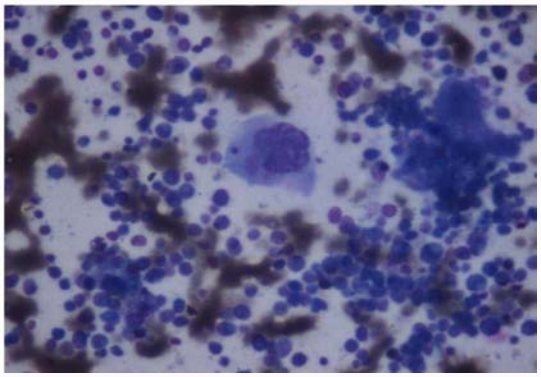

Figure 1: Bone marrow aspirate–fair number of histocytes showing few lymphophagocytosis (lymphocyte ingested by macrophage). Fair number of degenerated cells also seen.

examination showed hemophagocytosis. HLH was attributed to HIV infection alone, because no underlying malignancy, drug reaction or other infectious process could be demonstrated. Hemophagocytic syndrome caused by HIV infection alone has not been widely reported. Most of the reports of HLH revealed that cases occurred in the advanced stages of HIV infection and patients were suffering from infections and malignant disease. In our case, HLH occurred during the acute stage of HIV infection.

HLH is a highly fatal disease if left untreated. Chemotherapy using dexamethasone, cyclosporine and etoposide is used for familial and EBV associated hemophagocytic syndrome. Apart from EBV infection, treating the underlying infection is associated with 60-70% recovery rate in reactive hemophagocytic syndrome [1]. HIV associated HPS has been successfuly treated with HAART [5]. Our patient was treated with HAART received abacavir, lamivudine and nevirapine. CD4 count is 688 and child is doing well.

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