JMSCR Vol||3||Issue||12||Page 8635-8637||December 2015

www.jmscr.igmpublication.org

Impact Factor 3.79
Index Copernicus Value: 5.88
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: http://dx.doi.org/10.18535/jmscr/v3i12.27

IGM Publication Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Dengue with Hemophagocytic Lymphohistiocytosis-Rare Presentation

CASE REPORT

Authors

Dr Shweta Nair[1], Dr Abir Lal Nath[2], Dr Rajdeep Pal[3]

[1]MBBS, PG Student MD Pediatrics Department of Pediatrics DY Patil Medical College
[2]MBBS, PG Student MD Pediatrics Department of Pediatrics DY Patil Medical College
[3]MBBS, PG Student MD Pediatrics DY Patil Medical College
DY Patil Medical College Navi Mumbai India

ABSTRACT

A 1 and a half-year-old female child presented with fever, cough and cold since 2 weeks. Investigations revealed Dengue IgG, IgM Positive with pancytopenia and diagnosed to have HLH on bone marrow examination in view of unusual course. She was managed symptomatically along with antibiotics for associated urinary tract infection. It has not been reported in literature earlier.

Hemophagocytic Lymphohistiocytosis (HLH) is a rare macrophage related hyper inflammatory disorder that presents as prolonged fever and a sepsis like syndrome. It is a rare and life threatening complication of dengue.

KEYWORDS: *Dengue, Hemophagocytic lymphohistiocytosis, pancytopenia*

INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a heterogeneous group of clinical syndromes characterised by activation and subsequent uncontrolled non-malignant proliferation of T-lymphocytes and macrophages, leading to a cytokine storm. The presenting features are fever, hepatosplenomegaly, multiorgan dysfunction and fulminant pancytopenia resembling severe sepsis [1].HLH is a potentially life-threatening condition with protean clinical manifestations [2].

We reporta 1 and a half-year-old female child who came with fever of two weeks duration along with a maculopapularrash. Investigations revealed pancytopenia, hyperferritinemia, hypertriglyceri-demia, and hypofibrinogenemia. In our child, unusual persistence of dengue fever pattern for 2 weeks, with pancytopenias, hyperferritinemia, hypertriglyceridemia, and hypofibrinogenemia and organomegaly suggested the possibility of HLH after which the bone marrow examination confirmed the diagnosis.

CASE REPORT

A 1 and a half-year-old female child presented with fever, cough and cold for two weeks. Physical examination revealed a maculopapular rash over the trunk and extremities with generalized flushing of the face. Pallor was present. Investigations revealed pancytopenia

(Hb-8.3g/dl, Platelets-98000/uL, Neutrophils-38.9). Dengue IgG, IgM were positive. Serum iron studies showed hypertriglyceridemia (371mg/dl), hypofibrinogenemia (145mg/dl), hyperferritine-mia (1741ng/ml), hypertriglyceridemia (371mg/dl).

BUN/creatinine was normal. LFT was within normal range.USG abdomen showed hepatomegaly.Blood culture was sterile. Urine culture which showed E. Coli growth. Since the child was having an unusual course so the child was worked up for HLH. The child was started on antibiotic therapy as per culture report. Bone marrow biopsy revealed hemophagocytosis.

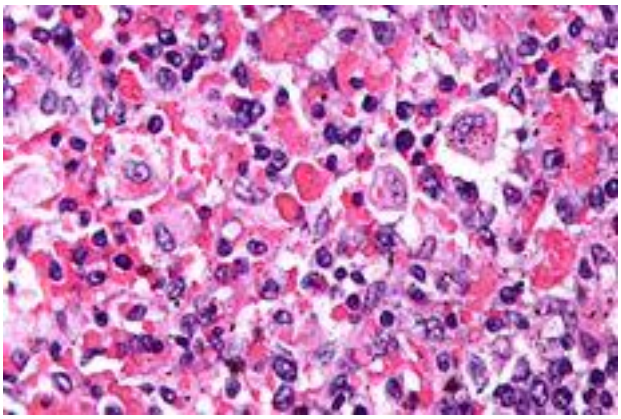

FIG.1: Bone marrow picture showing Hemophagocytosis.

Child was not started on steroids as the child recovered spontaneously which has not been reported in earlier literature.

DISCUSSION

Hemophagocytic syndrome (HLH) is a nonmalignant disease characterized by an expansion of the monocyte - macrophage population and intense hemophagocytosis[(3)]. HLH should be considered in the differential diagnosis of children with sepsis or presumed sepsis that do not respond to the conventional treatment. Persistence of fever beyond 7-8 days is unusual in dengue and if it persists, secondary sepsis or dengue associated HLH should be considered as described in our child.

Primary hemophagocytic lymphohistiocytosis (ie, familial erythrophagocytic lymphohistiocytosis [FEL]), an inherited form of hemophagocytic lymphohistiocytosis syndrome, is a heterogeneous autosomal recessive disorder found to be more prevalent with parental consanguinity. Secondary hemophagocytic lymphohistiocytosis (ie, acquired hemophagocytic lymphohistiocytosis) occurs after strong immunologic activation, such as that which can occur with systemic infection, immunodeficiency, or underlying malignancy. Both forms are characterized by the overwhelming activation of normal T lymphocytes and macrophages, invariably leading to clinical and hematologic alterations and death in the absence of treatment.[(4)]

The importance of the association between HLH and infection lies in the fact that both forms of HLH may be preceded by infection. HLH may also mimic infectious diseases such as overwhelming bacterial sepsis.Typical laboratory findings include bicytopenia or pancytopenia, hepatic impairment with coagulopathy, hypofibrinogenemia, elevation of serum LDH and triglyceride levels, and ferritinemia.

Park et al. in their cohort of 23 patients with secondary HLH found that high fibrinogen at the time of diagnosis, not the rate of decline in ferritin, was associated with prolonged survival, but in our child there was hypofibrinogenemia and hyperferritiniemia.In the study by Ramachandran, *et al*. [5], dengue was found to be the leading organism accounting for 5 among 43 cases of HLH.

Diagnosis is confirmed by cytological or pathological examination of bone marrow or tissue specimens.

HLH should be considered in the differential diagnosis of children and adolescents with prolonged fever, hepatosplenomegaly, and cytopenia. For children with primary HLH, the first step is to suppress the overactive immune system, commonly by treatment with a combination of steroids and chemotherapy. The patients who have no suitable donor for stem cell

transplantation are treated with HLH-2004 protocol, which includes etoposide, dexamethasone, cyclosporine A.

In our case the child had urinary tract infection and recovered spontaneously without the use of steroids.The association of urinary tract infection and HLH is not reported earlier in literature. Prompt recognition and early institution of appropriate therapy may result in good outcome, particularly in infection-associated HLH similar to our child.

REFERENCES

1. Loy T, Alberto AD, Perry MC. Familial erythrophagocytic lymphohistiocytosis. Semin Oncol. 1991; 18:34-9.
2. Janka G, Elinder G, Imashuku S, Schneider M, Henter J. Infection- and malignancy-associated hemophagocytic syndromes: secondary hemophagocyticl-ymphohistiocytosis. Hematol OncolClin North Am. 1998; 12:435-44.
3. Fisman DN. Hemophagocytic syndromes and infection. Emerg Infect Dis 2000. Nov-Dec;6(6):601-608 10.3201/eid0606.000608
4. Feldmann J, Le Deist F, Ouachee-Chardin M, et al. Functional consequences of perforin gene mutations in 22 patients with familial haemophagocytic lymphohistioc-ytosis. *Br J Haematol*. Jun 2002;117(4): 965-72.[Medline].
5. Ramachandran B, Balasubramanian S, Abhishek N, Ravikumar KG, Ramanan AV. Profile of hemophagocytic-lymphohistiocytosis in children in a tertiary care hospital in India. Indian Pediatr. 2011; 48:31-5.