Hemophagocytic Lymphohistiocytosis: A Life Threatening Rare Entity

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Background

Hemophagocytic Lymphohistiocytosis (HLH) is a rare disorderofthe immune system,affecting macrophages that grow abnormally and accumulate in body organs which include liver, spleen, bone marrow, CNS and skin. There are 2 types of HLH:

Primary: An inherited condition where there is a genetic mutation in:

- Perforin (PRF-1)
- Gene encoding Munc 13-4 protein which alters the NK & Tcell function.

Secondary: Triggered by infections (viral, bacterial, fungal), autoimmune disorders, primary immune deficiencies or cancer.

Materials & Methods

Hereby we are presenting 3 cases of HLH, diagnosed with the established criteria at our institution:

Case 1

4 year old boy, case of Griscelli Syndrome with hypopigmented hair came with recurrent episodes of fever, bilateral ear discharges and abdominal distention since past 2 years, presented with significant pallor with moderate hepato-splenomegaly.

Case 2

3 year old boya case of Malignant Histiocytosis with fever and severe pancytopenia presented with significant hepato-splenomegaly and lymphadenopathy.

Case 3

4 year old boy, with massive hepatosplenomegaly with severe anemia and fever since 1 month, presented with abdominal distention, pallor and seborrhoeic dermatitis.

Results & Conclusions

All our 3 cases were investigated, and satisfied the diagnostic criteria of HLHand were started on treatment as per HLH 2004 protocol.