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SCIENTIFIC LETTER TO THE EDITOR

# Enteric Fever Presenting as Secondary Hemophagocytic Lymphohistiocytosis

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Sir,

Hemophagocytosis is the pathologic finding of aggressive, but non-malignant proliferation of macrophages and histiocytes [1]. Hemophagocytic Lymphohistiocytosis (HLH) has been associated with a variety of infections and malignancies [2, 3]. However, its association with enteric fever is uncommon. We describe a case of enteric fever with an uncommon complication of secondary HLH (sHLH), responding completely with antibiotic treatment alone.

A 10-y-old boy presented with complaints of fever for 5 d and altered consciousness for one day. His younger sibling was admitted to a hospital for the past 10 d as a case of enteric fever.

Examination revealed severe pallor, petechie over body, hepatomegaly (5 cm below right costal margin), splenomegaly (3 cm in splenic axis), altered sensorium ($E_3M_1V_2$) and meningeal signs. Initial diagnosis of meningoencephalitis was kept.

Investigations revealed pancytopenia (Hb 6.9 g%, TLC 800/μL, ANC 384/μL, MCV 86.3 fL, platelets=$10\times10^3$/μL, DLC = $N_{48}L_{48}E_0M_4$, and normocytic normochromic RBCs). Rest of the investigations, including lumbar puncture were normal. In view of pancytopenia, bone marrow examination was performed which showed features of hemophagocytosis (Fig. 1).

His serum triglycerides (265 mg/dl) and serum ferritin (1,791 ng/ml) were high. Thus the diagnosis was established as infection associated hemophagocytosis (IAHS). A widal test done on the seventh day of fever showed positive titres ($T_O$ >1:320, $T_H$ >1:320) rising from baseline tires ($T_O$ <1:40, $T_H$ <1:40) done at day two, done from outside. Repeat blood culture and bone marrow cultures were sterile.

The patient was managed as a case of enteric fever with encephalopathy and secondary HLH. He received multiple transfusion supports and ceftriaxone for fourteen days. He became asymptomatic by fifth day of antibacterial therapy, his blood counts normalised and he was discharged after completion of antibiotic therapy. Association of sHLH with enteric fever has only been reported in a few cases till date in the English literature [4, 5].

Fatality rate of 50% is present in children with sHLH. The treatment includes etoposide, dexamethasone, and cyclosporine. However our case report stresses the fact that management of underlying infection in cases presenting with severe forms of HLH with enteric fever may obviate the need of intensive treatment. Cytopenias are known in enteric fever due to marrow suppression. However, hemophagocytosis is described as an uncommon extra-intestinal complication of enteric fever [4].

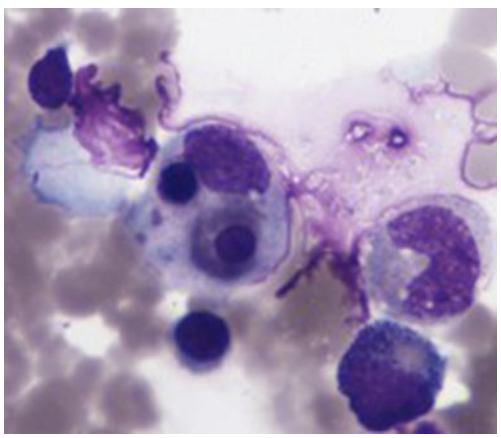

**Fig. 1** Bone marrow examination showing a histiocyte with engulfed lymphocyte and nucleated erythrocyte

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We thus conclude that physicians must possess a high index of suspicion for diagnosing enteric fever amongst patients with HLH as antibacterial treatment alone can be life saving.

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