present study was similar to 1.2-5.5% reported in previous studies [8,9].

Though varying degrees of myalgias are commonly seen; muscle weakness, as seen in this study, is an uncommon manifestation. There are two suggested mechanisms *viz.*, direct invasion of the muscle fibers or release of myotoxic cytokines, particularly tumor necrosis factor (TNF), injuring the affected muscle [10].

Despite a small sample-size and retrospective study design, we found atypical manifestations of dengue fever to be more common than reported, especially encephalopathy. Practitioners need to be aware of the same to ensure prompt recognition and early management.

Contributors: AP: conceived the idea, reviewed the records and analyzed the data; DM: planned the study and helped in manuscript preparation; MJ and JM: important intellectual inputs in the planning and conduct of the study, and manuscript preparation. All the authors were involved in the final approval of manuscript.
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Dengue Associated Hemophagocytic Lymphohistiocytosis: A Case Series

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Hemophagocytic lymphohistiocytosis is a rare complication of dengue. We present 8 cases of dengue associated hemophagocytic lymphohistiocytosis diagnosed in our hospital during the dengue outbreak of 2012. All the cases were treated with a short (4 weeks) course of steroids along with supportive measures, and showed an excellent response.

Keywords: *Dengue fever, Hepatomegaly, Splenomegaly.*

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 clinical features are: fever, hepatosplenomegaly, multiorgan dysfunction and fulminant pancytopenia resembling severe sepsis [1]. Persistence of fever for more than 7 days, with persistent/progressive cytopenias, raised ferritin, and organomegaly suggested the possibility of HLH in our patients with dengue.

Clinical records of children (<15 years) with serologically confirmed dengue infection admitted at the Institute of Child Health, Kolkata during the Dengue outbreak (July 2012 to November 2012) were reviewed. Out of 358 dengue, 8 developed HLH. The diagnosis of HLH was based on the criteria laid down by the Histiocytic Society in 2004. The clinical and laboratory features are listed in ***Table* I**.

The children were given supportive therapy in form of blood component transfusions, as and when required, along with broad spectrum antibiotics. Definitive therapy was administered in the form of parenteral steroids

TABLE I CLINICAL AND LABORATORY FEATURES IN THE STUDY CHILDREN

Clinical Parameters	*No. (%)*	*Laboratory Parameters*	*No. (%)*	*Range*
Fever >7 d	8 (100%)	Anemia (<9 g/dL)	7 (88%)	6.7-9.2
Rash/mucositis	5 (63%)	Thrombocytopenia (<100000/cmm)	8 (100%)	8.8-9.6
Hepatomegaly	8 (100%)	Neutropenia (ANC<1000)	6 (75%)	352-1622
Splenomegaly	7 (88%)	Raised CRP (>6 mg/L)	8 (100%)	82-216
Bleeding manifestations	6 (75%)	Raised SGPT (>50IU/L)	7 (88%)	42-288
Lymphadenopathy	2 (25%)	Raised Ferritin (>500 ng/L)	8 (100%)	1832-64,600
Edema	8 (100%)	Raised LDH (>500U/L)	8 (100%)	872-2680
Ascites	8 (100%)	Raised d-dimer (>1000)	8 (100%)	6800->10,000
Pleural effusion	8 (100%)	Raised triglyceride (>265 mg/dL)	7 (88%)	242-638
ARDS	2 (25%)	Decreased fibrinogen (<1.5 g/dL)	5 (63%)	86-220
Myocarditis	4 (50%)	Altered PT/APTT	8 (100%)	
Hypotension	6 (75%)	Hemophagocytes in bone marrow	6 (75%)	
Encephalopathy	2 (25%)			
Joint pain	2 (25%)			

(dexamethasone 10 mg/m^2 in 3-4 divided doses/day) and continued till child was hemodynamically stable or accepting oral feeds. It was switched to oral dexamethasone in a in a tapering dose for 21 d (5 mg/m^2 for next 7 days and then 2.5 mg/m^2 for further 7 days, and ultimately tapered off over the next 7 days). In seven children, fever subsided within 48-72 h of starting steroids; reversal of cytopenias and regression of hepato-splenomegaly occurred over the next 4-7 days. Serum ferritin started normalizing within a week. Intravenous immunoglobulin was used (1 g/kg) in one patient as a rescue measure as the child did not show any significant improvement after 48 hours of starting steroids.

HLH is a potentially life-threatening condition with protean clinical manifestations [2]. HLH secondary to infections (IAHLH) is the commonest, especially in tropical countries. Any infection (virus, bacteria, fungi, protozoa) can give rise to HLH. In the study by Ramachandran, *et al*. [3], dengue was found to be the leading organism accounting for 5 among 43 cases of HLH. Veerakul, *et al*. [4] also reported a series of 52 pediatric patients with HLH; 15 were infection associated out of which 3 were caused by dengue. Tan, *et al*. [5] also reported a comprehensive report of six cases of dengue associated HLH in adults. In our series, we had 8 cases of dengue associated HLH among 358 admitted dengue patients over a time period of 5 months.

Persistence of fever beyond 7-8 days is unusual in dengue and if it persists, secondary sepsis or dengue associated HLH should be kept in mind. In our series, all 8 children presented with fever of more than 7 days along with persistent or progressive cytopenias, unusual organomegaly with clinical deterioration and sterile cultures.

Early identification and treatment with dexamethasone, even a short course of 4 weeks, can give rise to a good outcome.

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