Visceral Leishmaniasis Associated with Hemophagocytic Lymphohistiocytosis

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To the Editor: An 18-mo-old toddler hailing from Nepal presented to us with persistent fever of 4 mo duration, anemia and splenohepatomegaly. Laboratory studies demonstrated pancytopenia (hemoglobin 8.2 g/dl, TLC 3000/mm³, platelets 72,000/mm³), hypertriglyceridemia (742 mg/dl), hyperferritinemia (2075 ng/ml) with features of hemophagocytosis on bone marrow aspiration (BMA) suggesting a diagnosis of hemophagocytic lymphohistiocytosis (HLH), as it met ≥ 5 out of 8 of the HLH diagnostic criteria. Investigations for malaria, Epstein-Barr virus and HIV were negative. The Anti-rK39 ELISA was positive. On this basis, a diagnosis of Visceral leishmaniasis (VL) with secondary HLH was made. The patient was treated with single dose 10 mg/kg liposomal amphotericin B (LamB). Child showed clinical improvement and on follow-up after 8 wk there was no fever, spleen size had significantly decreased and blood counts had normalized.

VL is caused by the protozoa *Leishmania donovani* (LD) and is an uncommon cause of HLH, representing ~2.1% of all such cases [1]. VL-HLH poses as diagnostic dilemma as features of VL and HLH overlap, often delaying the diagnosis.

Only a handful of cases of pediatric VL-HLH have been reported in literature with the median age of diagnosis being 2 y [2]. However, it might be under-recognized rather than being uncommon as a prospective multicentre study demonstrated that up to 41% of children with VL developed HLH [3].

In our child, BMA did not demonstrate LD bodies which are diagnostic of VL. They may not be seen in up to 36% cases of VL because of the paucimicrobial nature of the disease. The

Anti-rK39 ELISA has 97% sensitivity and 98.9% specificity and is a valuable tool in diagnosing VL [4].

Clavijo et al. studied children with VL, with and without HLH and found that there were no differences in outcome [1]. Specific treatment for HLH is generally not needed and most respond to treatment for VL alone as seen in our patient. We used only a single dose of LamB which we believe is sufficient, though others have used longer courses [2, 5].

This case illustrates the importance of recognizing VL as an etiology of HLH, especially in patients from endemic areas and timely administering single-dose LamB to prevent life-threatening complications.

Compliance with Ethical Standards

Conflict of Interest None.

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