Atypical Presentation of Hemophagocytic Lymphohistiocytosis in Severe Dengue Fever

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Abstract

In recent years, there was a surge of severe dengue cases in Malaysia with serious implications on morbidity and mortality. Such patients complicated with hemophagocytic lymphohistiocytosis (HLH) are poorly recognized and mostly fatal. A middle-aged lady, with diabetes and hypertension, complained of feeling unwell for 4 days with lethargy and poor oral intake, but no warning signs or bleeding. Clinically she was dehydrated, hypotensive with non-tender hepatomegaly, but not in leakage. Initial investigations showed positive NS1 antigen, high haematocrit, severe thrombocytopenia, transaminitis, acute kidney injury with lactic acidosis. She was diagnosed as severe dengue and admitted to intensive care. She later deteriorated with drop in Glasgow Coma Scale to 12/15. She was afebrile and jaundiced. Leukocytosis was present, with bicytopenia, coagulopathy and worsening transaminitis. HLH was suspected in the presence of hyperferritinemia and high LDH; this was confirmed by bone marrow biopsy. Treatment was initiated with intravenous dexamethasone and immunoglobulin; subsequently requiring continuous renal replacement therapy. Clinical and biochemical improvement was evident after 48 hours, and further resolution in 5 days, but no neurological recovery. She succumbed due to recurrent nosocomial infections. Here, we report a case of atypical presentation of HLH in dengue, with encephalopathy, in the absence of fever and splenomegaly. Association with neurological involvement, leukocytosis, kidney and liver failure, as illustrated in this case, has been reported to have poor prognosis and high mortality risk. Such presentation is rare, thus its recognition is critical in altering the prognosis of this otherwise deadly disease.

Keywords: Severe dengue fever, encephalopathy, hemophagocytic lymphohistiocytosis (HLH), intravenous dexamethasone, immunoglobulin

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