Hemophagocytic Syndrome Associated with Dengue Hemorrhagic Fever

The term virus associated hemophagocytic syndrome describes a disorder characterised by a benign generalized histiocytic proliferation with marked hemophagocytosis associated with a systemic virus infection. The mortality for patients with infection associated hemophagocytic syndrome is very high(1). We describe a 12-year-old girl who had fatal hemophagocytic syndrome associated with dengue hemorrhagic fever. A literature search shows fewer than 20 case reports of dengue related hemophagocytosis(2).

A 12-year-old, previously healthy girl presented with fever for 5 days. On examination she was febrile, sick looking with normal vital signs. Her liver was palpable 3 cm below the right costal margin, and spleen was not palpable. She had malena on the second hospital day. She was started on intravenous fluids, and ceftriaxone. She developed tachypnea, epigastric tenderness and hypotension on fourth hospital day. She was resuscitated with IV fluids, fresh frozen plasma and dopamine support. She had high grade spiky fever throughout hospital stay ranging between 101^{o}F-105^{o}F. On 11th hospital day she developed dyspnea and her chest *X*-ray showed bilateral consolidation. She was started on intravenous ceftazidime and within few hours her respiratory distress rapidly progressed and she died inspite of ventilatory support.

Her complete blood count showed a total WBC count of 2.9 x 10^3/µL, Hb of 9.4 g/dL and platelet count of 90,000/µL. Repeat blood counts done six times during hospital stay showed persistent leucopenia, thrombocytopenia and anemia. Blood and urine culture were sterile. Anti dengue IgG was positive (40.20 pu) and IgM was also positive (11.4 pu) suggestive of secondary dengue infection. Bone marrow biopsy study showed features of erythrophagocytosis.

A study by Chih-Jungchen, *et al*.(3) has shown that 61.1% of children with hemophagocytosis had infection associated hemophagocytosis. Normally fever does not persist for more than 7 days in dengue hemorrhagic fever and thrombocytopenia resolves in a few days. But children with dengue fever associated hemophagocytosis have an atypical evolution of dengue hemorrhagic fever with prolonged fever and persistent abdominal pain(4). Our child had prolonged high spiky fevers for 16 days and persistent cytopenias and sucummbed to sepsis inspite of antibiotic therapy. Clinicians should consider the possibility of hemophagocytosis in children having dengue hemorrhagic fever complicated with prolonged spiky fever and persistent pancytopenia.

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