Fatal case of EBV induced hemophagocytic lymphohistiocytosis (HLH), presenting with acute abdomen

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Introduction:

Hemophagocytic lymphohistiocytosis (HLH) is a rare hyperinflammatory syndrome with excessive immune activation. Primary or secondary HLH, have a high mortality rate approaching 50%, despite multiple management regiments. In adults, infections, especially EBV, and hematological malignancies are the most common causes. HLH recognition is often delayed, due to unfamiliarity of physicians of this diagnosis. Scores are available to aid physicians recognize HLH, but can be applied if the diagnosis is suspected.

Case description:

40 years old male, from Sierra Leon, presented to the emergency with acute abdomen. Patient had high grade fever, generalized abdominal pain and vomiting. Initial investigations showed massive hepatosplenomegaly, monocytosis, thrombocytopenia and deranged LFT. Patient was admitted as a case of acute cholecystitis. Patient was started on broad spectrum antibiotics but failed to improve and continued to rapidly deteriorate requiring intubation and inotropic support in ICU. Patient was managed as a case of septic shock and underwent open cholecystectomy, however the excised gall bladder couldn't explain the patient's clinical picture. HLH was considered at that phase as supported by additional findings of hypertriglyceridemia, hypofibrinogenemia, hyperferritinemia, thrombocytopenia. Pulse steroids therapy was initiated, with an initial response followed by relapse of condition. Patient was transferred to a facility with hematology service for continuity of care and bone marrow biopsy. Serology showed Positive EBV IgM with EBV DNA found to be 1260000000 copies/mL. Patient continued to deteriorate with multiple organ failure. Patient had an asystolic cardiac arrest and was declared dead on day 23 of hospital admission.

Conclusion:

HLH is an underdiagnosed entity in the critically ill, often misdiagnosed as sepsis or DIC. Physicians needs to be aware of its causes and pathogenesis for early recognition and appropriate treatment as multiple organ dysfunction rapidly develop. If suspected, patient should be followed and managed by a hematology service, as bone marrow biopsy and flow cytometry are needed for the diagnosis. Steroids therapy and etoposide are usually initiated once diagnosis is established. Cyclosporine is also used but high risk of developing PRES if CNS is involved. Despite multiple proposed drug regimen, mortality remains very high, and increases as the diagnosis is delayed.