

Case report: Cytokine hemoadsorption in a case of hemophagocytic lymphohistiocytosis secondary to extranodal NK/T-cell lymphoma

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This case reports on a 50-year-old African male patient, who was admitted to the hospital with a 2-month history of mild epistaxis and diffuse abdominal pain.

Case presentation

- He presented with fever and hepatosplenomegaly on physical exam; laboratory values showed cytotoxicity with undissociated cholestasis, non-oliguric renal failure, pancytopenia with hemolytic anemia, hyperferritinemia, and hypertriglyceridemia
- He required rapid admission to the Intensive Care Unit (ICU) for hemodynamic and respiratory support
- A full microbiological panel was carried out, including screening for imported disease, standard serologies and cultures for bacterial and fungal infection; however, results came back positive for Epstein-Barr virus with a 7.7 Log viremia
- The patient fulfilled the hemophagocytic lymphohistiocytosis (HLH)-2004 diagnostic criteria, and according to the 2018 Consensus Statements by the HLH Steering Committee of the Histiocyte Society, dexamethasone and etoposide were started, together with empiric antibiotic treatment including meropenem and amikacin
- Simultaneously, liver biopsy and bone marrow aspirate were performed, confirming the diagnosis of NK-type non-Hodgkin's lymphoma with secondary hemophagocytosis
- Although the patient received broad-spectrum antibiotics and the recommended HLH treatment, he rapidly deteriorated over the next 48 h with distributive shock and multiorgan dysfunction (renal, neurological, hemodynamic, respiratory, and hepatic) requiring high doses of norepinephrine support (up to 1.5 µg/kg/min), continuous veno-venous hemodiafiltration and invasive mechanical ventilation

- He exhibited significantly increased plasma levels of inflammatory mediators (interleukin (IL)-10 5643 pg/ml, IL-6 233 pg/ml, C-reactive protein CRP 7.7 mg/dl) as well as pronounced hyperlactatemia (8.5 mmol/L) and hyperbilirubinemia (22 mg/dl)
- Due to highly elevated levels of cytokines and refractory distributive shock, rescue therapy with CytoSorb was started within 24 hours of ICU admission

Treatment

- One treatment session with CytoSorb was carried out for a duration of 24 hours
- Cytokine adsorption was performed in parallel to the veno-venous hemodiafiltration circuit

Measurements

- Hemodynamics and requirements for vasoactive substances
- Inflammatory parameters
- Lactate
- Bilirubin

Results

- After starting this procedure, rapid hemodynamic control was achieved with a significant reduction in vasopressor support requirements after only a few hours. Vasopressor support could be completely stopped 37 h after the initiation of CytoSorb therapy
- Cytokine hemoadsorption with CytoSorb also led to a rapid decrease in several cytokines, such as IL-10 (from 5643 to 490 pg/ml) and IL-6 (from 233 to 91.2 pg/ml)
- After a transient increase to 10.3 mmol/l, lactate levels decreased to a level of 5 mmol/l after cessation of hemoadsorption treatment with decreasing values thereafter
- Treatment also resulted in a significant decrease in bilirubin plasma concentrations from 22 to 7.8 mg/dl

Patient Follow-up

- Despite the successful management of the acute phase, 6 days after completion of hemoadsorption and 9 days after ICU admission, the patient died due to thrombotic complications related to the underlying lymphoma with thrombosis of the inferior vena cava, hepatic system, right atrium and partial thrombosis of the thoracic aorta

Conclusions

- In this patient, cytokine hemoabsorption was associated with a rapid decrease in IL-10 levels and significant hemodynamic improvement
- This case report highlights that cytokine hemoabsorption can be an effective and safe rescue therapy in patients with HLH and multiorgan dysfunction, complementary to standard protocol treatments
- The authors suggest real-time monitoring of plasma cytokine concentrations as a tool to monitor the biological effect of cytokine hemoabsorption, optimizing the duration of this procedure