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Case Report

Successful Hematopoietic Stem Cell Transplant in the Medical Intensive Care Unit- a Case of Overwhelming Hemophagocytic Syndrome

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Introduction

Hemophagocytic syndrome is a life threatening disorder characterized by severe deregulated activation of the immune system leading to Systemic Inflammatory Response Syndrome (SIRS) and multi organ failure. Its acquired form in adults occur secondary to infections, autoimmune disorders and malignancies [1]. Although usually treated with corticosteroids and immune suppressants, allergenic bone marrow transplantation in the only therapy with long term disease free survival. Bone marrow transplant is generally performed when patients are in clinical remission.

We present a unique case of patient with over whelming Hemophagocytic syndrome presenting with fulminate sepsis that successfully underwent hematopoietic stem cell transplant while he was critically ill in the intensive care unit.

Case Presentation

A 26 year old man initially presented with confusion, fever and chills. He was also found to have thrombocytopenia, anemia. Coagulopathy, acute kidney injury, hyponatremia, elevated LDH, increased ferreting and elevated liver enzymes. Bone marrow biopsy revealed hemophagocytosis with normal cytogenetic. He was started on high dose steroids and topside with clinical remission of his symptoms and hematologic abnormalities. A decision was made to perform matched unrelated bone marrow transplant.

However, he presented to the emergency department on the day of transplant with fever, dispend, fatigue and confusion. He subsequently went into a systole while in ED and after few cycles of CPR regained spontaneous circulation. He was then admitted to the medical intensive care unit. Treatment for presumed sepsis with broad spectrum antibiotics, intravenous fluids and vasopressors was initiated. Over the next few hours, he continued to remain extremely hypertensive requiring four vasopressors.

Laboratory abnormalities include pancytopenia, hyponatremia and extremely high ferreting levels. He was also found to have shock liver and acute kidney injury as well. Cultures remained negative. He then underwent sternal bone marrow biopsy that suggested active hemophagocytosis. A diagnosis of fulminate overwhelming Hemophagocytic syndrome was made.

He was treated with high dose corticosteroids with remarkable improvement in hemodynamics and liberation from mechanical ventilation over 48 hours after admission to medical ICU. He underwent allergenic bone marrow transplant without any complications within 24 hours of admission to medical ICU. His post-transplant course was complicated by moderate graft versus host disease (GVHD) that resolved with systemic steroids. He was seen in the clinic one year post transplant with no history of opportunistic infections, relapse of GVHD or drug toxicity.

Discussion

The international histiocytes society classifies the histiocytic [2] disorders into three categories from class I to class III. The majority of Hemophagocytic syndromes belong to class II with rare malignant histiocytosis falling in class III and is now considered consisting of primary (familial) and secondary (infection- or lymphoma-associated) Hemophagocytic Lympho Histiocytosis (HLH).

Clinical features in patients with these Hemophagocytic syndromes are similar and include idiopathic fever, cytopenias, liver dysfunction, hepatosplenomegaly, coagulopathy, as in our patient [3,4]. Hemophagocytosis can be found on biopsy specimen but is not required.

Hemophagocytic syndrome is characterized by mutual stimulation of the innate and adaptive arms of the immune system leading to persistent hypercytokinemia and SIRS. Early treatment is critical to prevent progressive multi-organ failure [4].

The basis of treatment for these disorders is corticosteroids and topside. Allogeneic BMT has mostly been described for patients with HLH who have an aggressive course, since without bone marrow transplant their prognosis is poor [3,4]. Even when stem cell transplants (SCTs) were successful, some patients still relapsed following SCT and died with a median follow-up of 23 months [5,6].

Our patient represents a unique case of this disorder who was successfully treated with Allogeneic hematopoietic stem cell transplant in medical ICU when he was hemodynamically unstable requiring multiple vasopressors. Intensivists should have a low threshold to suspect this disorder in otherwise healthy individuals presenting with fulminate sepsis like picture without any identifiable source who do not respond to conventional treatment and have profound hyperferritinemia. Bone marrow biopsy should be considered in those patients.

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