Case Report

A Rare Cause of Hemophagocytic Lymphohistiocytosis: *Fusobacterium* Infection—A Case Report and Review of the Literature

Ghulam Rehman Mohyuddin and Heather J. Male

1 Kansas University Medical Center, 3901 Rainbow Boulevard, Kansas City, KS 66160, USA
2 Division of Hematologic Malignancies and Cellular Therapeutics (HMCT), Department of Internal Medicine, University of Kansas Cancer Center, 2330 Shawnee Mission Parkway, Mailstop 5003, Westwood, KS 66205, USA

Correspondence should be addressed to Ghulam Rehman Mohyuddin; gmohy-ud-din@kumc.edu

Received 23 May 2016; Accepted 20 July 2016

Academic Editor: Tatsuharu Ohno

Copyright © 2016 G. R. Mohyuddin and H. J. Male. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Hemophagocytic lymphohistiocytosis (HLH) is a rare syndrome characterized by excessive activation of the immune system. Viral infections are a common trigger, but bacterial infections are rare precipitants [1]. We present a case of HLH associated with *Fusobacterium* infection, which has only been reported once previously [2].

1. Introduction

Hemophagocytic lymphohistiocytosis (HLH) is a rare syndrome characterized by excessive activation of the immune system. Viral infections are a common trigger, but bacterial infections are rare precipitants [1]. We present a case of HLH associated with *Fusobacterium* infection, which has only been reported once previously [2].

2. Case Presentation

A 19-year-old previously healthy male acutely developed headache, photophobia, diffuse myalgia, fatigue, nausea, and vomiting. Upon presentation he was found to be hypotensive, tachycardic, and febrile. He was given intravenous fluids and started on vancomycin and ceftriaxone. A lumbar puncture was not suggestive of meningitis. Patient progressively developed shock that required use of vasopressors, as well as renal and respiratory failure. Blood cultures grew *Fusobacterium necrophorum*. Given continued fevers despite appropriate antimicrobials, a bone marrow biopsy was performed revealing increased histiocytes with hemophagocytosis. Dexamethasone was added with dramatic clinical improvement. Our case highlights *Fusobacterium* as a rare precipitant of HLH and proves that a high index of clinical suspicion is crucial for early diagnosis of HLH, allowing for prompt initiation of HLH-specific immunosuppressive therapy that can be life-saving.

Hindawi Publishing Corporation
Case Reports in Hematology
Volume 2016, Article ID 4839146, 2 pages
http://dx.doi.org/10.1155/2016/4839146
Due to persistent high oxygen requirements, fevers, and renal failure despite therapy, dexamethasone dosed according to the pediatric HLH-94 protocol at 10 mg/m² daily was initiated on day 6.

A significant improvement in inflammatory markers and renal and hepatic function was noted within 24 hours of starting dexamethasone. The patient was quickly weaned from mechanical ventilation. The hepatic lesion suspicious for hemangiomia was drained and found to be purulent, although cultures remained negative. Ferritin peaked at 966 ng/mL and triglycerides peaked at 725 mg/dL, both normalizing within 2 weeks. Subsequent evaluation showed resolution of the cardiac dysfunction, septic emboli, and hepatosplenomegaly.

The patient was discharged home with a dexamethasone taper and an extended course of ertapenem.

3. Discussion

Diagnosing HLH requires a high index of clinical suspicion, as the presentation is often nonspecific. The criteria for diagnosis are well published, but a clinical diagnosis remains [3].

Although severe infection alone could have explained the multisystem organ failure and hemophagocytosis, our patient did meet 5/8 criteria for HLH. The possibility of HLH was entertained early in our patient’s course because the immune response and the multisystem organ failure despite antimicrobial therapy exceeded what would be expected in a healthy 19-year-old.

In the adult population, an elevated ferritin is not specific for HLH, as renal failure, hepatocellular injury, malignancies, and infections can cause a drastically elevated ferritin level [4, 5]. In our case, we feel we diagnosed HLH early enough to prevent progressive macrophage activation, and thus the ferritin only peaked at 966, a value far lower than that seen in most cases [1, 6, 7].

The premise for treatment of HLH is to stop the trigger and control the overactive immune system. Any underlying cause such as infection or malignancy should be aggressively treated, with consideration of additional immunosuppressive therapy only if there is insufficient improvement. Treatment in adults is largely based on the HLH-94 study in which pediatric patients were treated with an 8-week induction of dexamethasone and etoposide [8]. Etoposide-based regimens are now considered the standard of care for HLH. The ongoing HLH-2004 trial is evaluating the addition of Cyclosporine to that regimen [9].

In certain cases of infection-associated HLH, glucocorticoid monotherapy has been used with success, with the caveat that there will be a low threshold to proceed to full immunosuppressive therapy if a satisfactory response is not achieved rapidly [10, 11].

4. Conclusion

This case report highlights that with early identification of HLH in infection, it can be stopped with steroids alone. It describes the rare association between Fusobacterium infection and secondary HLH. We conclude by emphasizing that a high index of clinical suspicion is crucial for early diagnosis of secondary HLH, and prompt initiation of HLH-specific immunosuppressive and anti-inflammatory therapy in addition to treatment directed against the infectious source can be life-saving in infection-associated HLH.

Competing Interests

Ghulam Rehman Mohyuddin and Heather J. Male have no conflict of interests to declare.

Authors’ Contributions

Both authors took care of the patient, with Dr. Ghulam Rehman Mohyuddin being the resident physician and Dr. Heather J. Male the attending physician.

References
