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CASE REPORT PEER REVIEWED | OPEN ACCESS

A fatal case of hemophagocytic lymphohistiocytosis in an elderly male

Maysoon T Hussain, Saurabh Dubey, Mohamad Akil, Nasir Gondal

ABSTRACT

We present a case of an 87-year-old male with hemophagocytic lymphohistiocytosis (HLH). Hemophagocytic lymphohistiocytosis is a life-threatening disorder characterized by dysregulated immune activity leading to end-organ damage. Hematologic malignancies appear to be the main cause of secondary HLH. Diagnostic criteria include fever, cytopenia, splenomegaly, hypertriglyceridemia, hypofibrinogenemia, a ferritin level greater than 500 ng/mL, low NK-cell activity, and elevated sIL2Ra levels greater than or equal to 2400 U/mL. Five out of eight of the criteria are necessary to establish a diagnosis of HLH. Our patient met five out of eight diagnostic criteria for HLH (fever, cytopenia, splenomegaly, hypertriglyceridemia, and ferritin level greater than 500 ng/mL). Bone marrow aspirate revealed hemophagocytosis. Bone marrow biopsy revealed marginal zone B cell lymphoma, which is presumed to be the underlying cause of his condition. Due to poor functional status, the patient was a poor candidate for curative treatment, and the family chose not to pursue this route. The patient was treated with Decadron 20 mg intravenous (IV) push daily until he ultimately died, likely due to respiratory failure due to heart failure (HF).

Keywords: B cell lymphoma, Cytopenias, Hemophagocytic lymphohistiocytosis, Unexplained fever

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INTRODUCTION

Hemophagocytic lymphohistiocytosis is a life-threatening disorder characterized by dysregulated immune activity leading to severe inflammation and end-organ damage. Primary HLH is genetic and emerges in early childhood, whereas secondary HLH presents in adulthood, with a mean age of 50, in response to triggers such as malignancy, infection, and autoimmune disorders. Hematologic malignancies, particularly non-Hodgkin lymphomas and CLL, appear to be the main cause of secondary HLH, accounting for 56% of cases in one study [1]. Diagnostic criteria include fever, cytopenia, splenomegaly, hypertriglyceridemia, hypofibrinogenemia, a ferritin level greater than 500 ng/mL, low NK-cell activity, and elevated sIL2Ra levels greater than or equal to 2400 U/mL [2]. Five out of eight of the criteria are necessary to establish a diagnosis of HLH.

Objective

We present a case of an 87-year-old male with HLH. Few case reports of HLH in people older than 80 years old have been reported in literature [3, 4].

CASE REPORT

An 87-year-old male presented to the emergency department due to worsening confusion and

functional decline. He had a history of hypertension, hypercholesterolemia, asthma, atrial fibrillation, diabetes mellitus, and prostatectomy. His family reported that he had recurrent falls, weight loss, loss of appetite, lethargy, chills, and fever of around 100–101°F. On examination, the patient had facial puffiness, pale skin, abdominal distension, and ecchymoses, and was oriented to person only. He had a recent admission during which he was treated with IV Zosyn for suspected pneumonia. However, his white blood cell (WBC) count remained elevated, blood cultures were negative, and erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) had increased despite treatment with antibiotics. Computed tomography (CT) scan of the abdomen and pelvis to evaluate further for sources of infection revealed splenomegaly. Ultimately, fever of non-infectious etiology was suspected, and the patient underwent a rheumatologic workup, iron studies, and tumor marker evaluation. These studies revealed elevated IgG subclass 4 (142.3), elevated haptoglobin (296), elevated ferritin (2,200). CEA (carcinoembryonic antigen), free PSA (prostate-specific antigen), and free PSA ratio were within normal limits.

The hematology/oncology service was consulted for evaluation for possible leukemia or lymphoma given the patient's splenomegaly, monocytosis, and unexplained fever. Workup revealed elevated ferritin, triglycerides, fibrinogen, LDH, and IgG4. He therefore met five out of eight diagnostic criteria for HLH (fever, cytopenia, splenomegaly, hypertriglyceridemia, and ferritin level greater than 500 ng/mL). Serum protein electrophoresis was within normal limits. Bone marrow aspirate revealed left shift, increased erythroid series, and hemophagocytosis (Figure 1). Flow cytometry showed no evidence of clonal expansion, possibly due to the paucicellular nature of the specimen. Bone marrow biopsy revealed marginal zone B cell lymphoma (nodular pattern of involvement, CD5 and CD10 negative, and CD20 positive). On biopsy, the overall marrow cellularity was 60%, dyserythropoiesis with <2% blasts was visualized, as well as rare hemophagocytosis. The patient was treated with Decadron 20 mg IV push daily with marginal improvement. Due to poor functional status, following discussion with the family, they decided against curative treatment and ultimately made the decision to change the code status of the patient to do not resuscitate (DNR)/do not intubate (DNI). The patient died shortly afterward, likely as a result of respiratory failure attributable to heart failure (HF) (Figure 1).

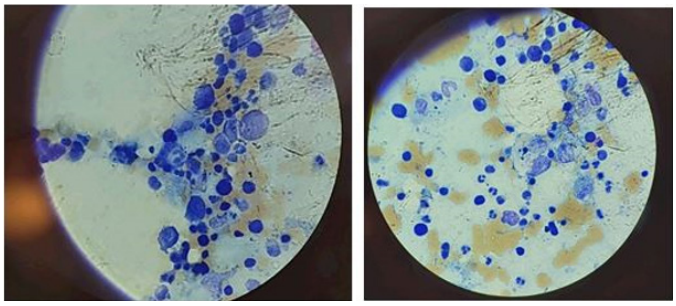

Figure 1: Bone marrow aspirate sample under microscope showing left shift, increased erythroid series, and hemophagocytosis of bone marrow elements.

DISCUSSION

Hemophagocytic lymphohistiocytosis can be diagnosed when five out of eight of the diagnostic criteria are present. Interestingly, our patient had elevated fibrinogen whereas 50–80% of patients with HLH have low fibrinogen, and increased fibrinolytic activity is associated with HLH [5, 6]. Hemophagocytosis on bone marrow biopsy is a hallmark of the disease. Our patient's bone marrow biopsy revealed marginal zone B cell lymphoma, which is likely the underlying cause of his secondary HLH. The prevalence of secondary HLH can be hard to estimate, as it often goes unrecognized until hemophagocytosis is identified by autopsy [7]. Mortality is high, (ranging from around 2–19.5% in rheumatologic causes, to 72% in some lymphomas), even with treatment [1, 8]. For this reason, early diagnosis and aggressive treatment is imperative.

CONCLUSION

Hemophagocytic lymphohistiocytosis is a life-threatening disorder and should be considered in cases that present as refractory fever in the context of cytopenias and/or splenomegaly when infectious or other causes are ruled out.

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Author Contributions

Maysoon T Hussain – Design of the work, Acquisition of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Saurabh Dubey – Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mohamad Akil – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Nasir Gondal – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.