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HEMATOLOGIC MALIGNANCY-ASSOCIATED HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN ADULTS: A 24-YEAR SINGLE-CENTER RETROSPECTIVE ANALYSIS

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INTRODUCTION

Hemophagocytic Lymphohistiocytosis (HLH) is a systemic inflammation disorder secondary to immune dysregulation. Malignancy Associated HLH is a puzzling diagnosis given both conditions overlapping presentation. There are currently no established treatment guidelines for malignancy-HLH.

AIM

Evaluate the clinical characteristics, treatment outcomes, and challenges in diagnosing Hemophagocytic Lymphohistiocytosis associated with hematological malignancies over a 24-year period.

METHOD

We conducted a retrospective review from patients from Harris Health to identify adult patients with confirmed HLH and the combined diagnosis of malignancy and HLH between 2000 and 2024.

Hemophagocytosis phenomena was demonstrated in all cases based on bone marrow biopsy or IL2 receptor positive.

RESULTS

Sociodemographic characteristics

Among those with malignancy associated HLH, the median age was 46 years (range 22-71), with a male predominance of 79% and 61% of cases were Hispanics.

Clinical characteristics

Out of a total of 76 cases of HLH documented over a 24-year period, 38% were associated with malignancies, 26% with infections, and 21% with autoimmune diseases.

The primary malignancies included Hodgkin lymphoma (43%) and B-cell neoplasms (18%).

Other neoplasms associated with HLH were T-cell/NK-cell neoplasms, Kaposi sarcoma, multiple myeloma and very uncommon malignancies such as peripheral T-cell lymphomas, primary hepatic T-cell lymphoma and EBV associated lymphoproliferative disorder resembling polymorphic PTLD.

Co-infections

EBV co-infection was observed in 25% of patients, with an additional 25% being HIV-positive. One patient presented disseminated histoplasmosis.

Treatment

Treatment approaches encompassed combined HLH and lymphoma therapies in 46% of cases, lymphoma-directed therapy in three patients, and HLH-specific regimens in eight cases.

Prognosis

Clinical response was achieved in 29% of cases, while 32% experienced refractory malignancy-HLH and required hospice care. Among evaluable cases, 71% attained complete remission of lymphoma. Notably, 43% of HIV-positive patients exhibited clinical response.

HLH cohort	N (%)
Total cases of HLH	76
Malignancy associated	28 (38%)
Other diagnosis associated	48 (62%)
Infection associated	20 (26%)
Autoimmune associated	16 (21%)
Primary HLH	2 (3%)
Other	10 (13%)
Acute liver failure	24
Malignancy associated	10 (42%)
Other diagnosis associated	14 (58%)

CONCLUSIONS

Herein, we describe our unique experience of hematological malignancies-HLH on a cohort of 24 years.

The diagnosis is challenging, often delayed, and the prognosis is dismal in refractory cases despite currently available rescue therapies.

Furthermore, we describe the association between HLH and T cell lymphomas and viral infections.

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