SOHO ANNUAL MEETING 2024

Diagnostic Challenges and Therapeutic Dilemmas: A Case of Hemophagocytic Lymphohistiocytosis Secondary to Lymphoma

Authors : Asmaa Mohsen[1], Noha M. Badawy[1] , Malaka Awad[1], Walaa Ahmed Yusuf [2], Emad Azmy[1]
1. Clinical haematology unit , internal medicine department , Faculty of medicine , Mansoura University, Egypt
2. Pathology department, Mansoura University Hospitals, Mansoura, Egypt

INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a rare and potentially fatal hyperinflammatory syndrome characterized by excessive activation of the immune system. Diagnosing HLH is particularly challenging due to its overlapping clinical features with other severe inflammatory and infectious diseases

AIM

This abstract discusses the critical importance and difficulty of diagnosing HLH as well as lymphoma, underscoring the need for heightened awareness and comprehensive diagnostic evaluation in affected patients.

METHODS

A case report which includes diagnostic history taking, physical examination, full blood count , Bone marrow aspiration and biopsy , Lymph node biopsy and imaging studies .

.

RESULTS

- We report a 63 year old lady who presented in November 2022 with fatigue, fever, anemia and progressing leukopenia. Initial investigations revealed high lactate dehydrogenase at 1363 U/L, and markedly elevated ferritin (2700 ng/mL) and Soluble CD 25 at 13855 IU/ml. Bone marrow biopsy indicated with normal haematopoietic series.
- CT imaging showed reactive lymphadenopathy, hepatosplenomegaly (Spleen 25 cm). Lymph node biopsy showed a reactive pattern. Revision of Bone marrow biopsy showed some macrophage activity with CD86 positive in 15% of cells. PET CT scan showed a reactive LN and non-active splenomegaly.
- She was started on HLH 2004 protocol after which she improved clinically and on laboratory basis. After 4 months of treatment our lady started steroid withdrawal to represent with sever pancytopenia, fever and progressively increased soluble CD25 to reach 18000 IU/ml. Repeat PET CT showed reactive lymphadenopathy with moderately active splenomegaly (SUV 7.8 Vs 3). Splenectomy was operated and splenic pathology revealed fibro congestive morphology with no atypia. The patient was restarted on higher doses of steroid and continued HLH protocol for another 2 months until she represented with leucocytosis, lymphocytosis and elevated liver function tests. Virology including EBV PCR, immune tests, toxoplasma serology was negative. Repeat Bone marrow biopsy with flow cytometry and axillary lymph node biopsy showed infiltration with diffuse large B-cell lymphoma with Ki 67 at 80%. Our patient started R-CHOP therapy for 3 cycles. Her CBC and lymphadenopathy improved but unfortunately she died of pneumonia later on .

CONCLUSIONS

- HLH is a very rare presentation of haematological malignancy, both lymphoma and HLH are difficult to diagnose and treat which needs frequent testing and different management options for better response

REFERENCES

1- Knauft, J., Schenk, T., Ernst, T., Schnetzke, U., Hochhaus, A., La Rosée, P., & Birndt, S. (2024). Lymphoma-associated hemophagocytic lymphohistiocytosis (LA-HLH): a scoping review unveils clinical and diagnostic patterns of a lymphoma subgroup with poor prognosis. *Leukemia*, *38*(2), 235-249.

2- Zoref-Lorenz, A., Iyer, S. P., Murakami, J., Hofstetter, L., Prakash, R., Weinstein, S., ... & Goyal, G. (2023). Treatments and Outcomes of Lymphoma-Associated Hyperinflammation-a Report from the HLH in Adults Working Group for the Histiocyte Society. *Blood*, *142*, 2537.

CONTACT INFORMATION

Email : asmaamohsen@mans.edu.eg