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A RARE CASE OF RAPID ONSET FATAL HLH SECONDARY TO LARGE GRANULAR T-CELL LYMPHOCYTIC LEUKEMIA

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INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a life-threatening, hyper-inflammatory condition that can affect all age groups and result in multi-organ failure and death. Primary HLH occurs due to a genetic mutation, while **secondary HLH** is associated with viral/bacterial/fungal infections, autoimmune diseases, **malignancy**, and immunodeficiencies. As such**, prompt treatment is warranted with immunosuppression coupled with cytotoxic chemotherapy.**

AIM

We report a rare case of **rapid onset fatal secondary HLH in** a patient with **underlying large granular T-cell lymphocytic leukemia.**

METHOD

Case report of a patient who presented at Sarasota Memorial Hospital in March 2024.

CASE PRESENTATION

A 66-year-old male with history of chronic neutropenia since 2000, and large granular T-cell lymphocytic leukemia diagnosed 2019, on chronic immunosuppression with cyclosporine 150 mg and prednisone 20 mg daily, who presented due to fevers, rigors, and night sweats for one week in duration.

No other infectious symptomatology was noted at the time. He reported **two similar recent hospitalizations within the same year** with symptomatic improvement on empiric antibiotics.

On presentation, the patient was febrile **101.3°F.** Physical examination was remarkable for non-tender **splenomegaly**.

Infectious workup with procalcitonin, lactic acid, urinalysis, blood cultures, viral panel (including adenovirus, CMV, and EBV) were negative. **Flow cytometry showed excess myeloid with left shift.**

Table 1. Pertinent lab work during the admission.

Labs	Results
White Blood Cells (10^3/µL)	0.3
Absolute Neutrophil Count (10^3/µL)	0.20
Hemoglobin (gm/dL)	8.8
Hematocrit (%)	26.6
Platelet count (10^3/µL)	39
AST (U/L)	79
Ferritin (ng/mL)	29,617
Triglycerides (mg/dL)	249
Fibrinogen (mg/dL)	159
Soluble CD25 (pg/mL)	69,350

The patient fulfilled the Modified 2009 HLH criteria, exhibiting an HScore of 286 points (indicating a >99% probability).

IV steroid was initiated, with plans for continued oral dexamethasone 20 mg daily and initiation of etoposide at 100 mg/m² twice weekly for 2 weeks.

Despite empiric antibiotics, antifungals and dexamethasone, patient continued to have worsening clinical status and multiorgan failure limiting initiation of chemotherapy. Unfortunately, patient passed away within days of his discharge to hospice.

Fig 1. CT abdomen/pelvis; The spleen is enlarged measuring 18 cm craniocaudal by 17.5 cm anterior posterior by 7 cm transverse.



CONCLUSIONS

Due to the malignant inflammation resulting in rapid clinical decline, **a high index of suspicion for secondary HLH is imperative to promptly establish diagnosis and initiate appropriate intervention.**

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