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CHRONIC LYMPHOCYTIC LEUKEMIA WITH RICHTER'S TRANSFORMATION PRESENTED WITH IMMINENT TUMOR LYSIS SYNDROME

AUTHORS

Modather Grain MD, 1.
Mutasim Idriss MD, 2.
Mohamed Bouzed MD, 3.
Zhuting Tang MD, 4.
1,2,4. Internal Medicine Residency, Unity Hospital, Rochester, NY, USA
3. University of Tripoli, Tripoli, Libya.

modather.grain@gmail.com
m.bouzed@uot.edu.ly

On behalf of my co-authors, we wish to extend our heartfelt graditude to our supporters and affiliates.

INTRODUCTION

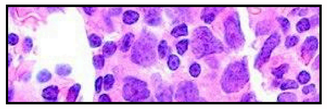
Richter's Transformation (RT) is a serious complication of Chronic Lymphocytic Leukemia (CLL), which is the most common adult leukemia in Western countries (1). RT occurs in 2-10% of CLL patients (2), leading to the development of an aggressive diffuse large B cell lymphoma (3). The median overall survival is only 5-8 months after diagnosis (4).

CASE PRESENTATION

65-year-old male with CLL, not on active treatment, admitted for impaired renal function and high LDH, suggesting disease transformation and possible tumor lysis syndrome. He reported two weeks of decreased appetite and fatigue. On admission, he was stable with hepatosplenomegaly and generalized lymphadenopathy

Initial laboratory workup showed elevated elevated renal function and markedly elevated LDH. CT revealed extensive lymphadenopathy and splenomegaly. Subsequent lymph node and bone marrow biopsies confirmed the diagnosis of diffuse large B-cell lymphoma (DLBCL), stage IV B, indicating Richter's transformation.

IImmunohistochemistry (IHC) and flow cytometry of the biopsy specimens showed that the atypical cells were positive for CD20, CD5, CD23, and BCL2. The Ki-67 stain demonstrated an increased proliferation index of 80%. C-MYC staining was borderline positive. Epstein-Barr virus-encoded small RNA (EBER) in situ hybridization (ISH) was negative.



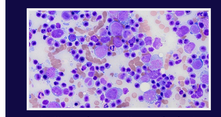
Geimsa stain showing CLL with large lymphoid cells

COURSE OF DISEASE

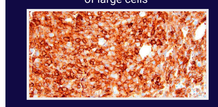
Despite aggressive treatment, the patient was readmitted with recurrent tumor lysis syndrome. After two R-CHOP cycles, response was poor. The patient's condition worsened rapidly, complicated by ongoing tumor lysis syndrome, volume overload, and anuria.

Follow-up scans showed no improvement in spleen size or lymph nodes. Rising LDH levels indicated refractory disease. Hemodialysis was planned for worsening kidney issues.

GDOX salvage chemotherapy was proposed. However, the patient died before starting this treatment or hemodialysis.



H&E Stain showing CLL with cluster of large cells



Large cells positive for CD5



CT abdomen at initial presentation

DISCUSSION

Richter's transformation (RT) is a very rare but severe complication of CLL. It involves the emergence of an aggressive lymphoma, typically DLBCL. This transformation significantly worsens the prognosis and presents diagnostic and management challenges. The underlying mechanisms of RT are complex and not fully understood, but genetic alterations, particularly TP53 mutations, play a crucial role. RT can arise through clonal evolution from the preceding CLL or as an independent secondary neoplasm.(5)

Diagnosing RT requires a high level of suspicion and typically involves imaging studies, bone marrow biopsy, and lymph node biopsy. Histological examination is essential for confirmation. Treatment options are limited and generally less effective than those for de novo DLBCL, with chemotherapy regimens like R-CHOP being commonly used despite poor outcomes. Early detection and prompt treatment initiation are vital for improving patient outcomes.

CONCLUSION

This case highlights the aggressive nature of Richter's transformation, emphasizing the significant challenges in managing this condition, particularly in the context of severe complications such as refractory tumor lysis syndrome and acute renal failure. It underlines the need for early recognition, prompt intervention, and the development of more effective treatment strategies for this aggressive form of lymphoma.

KEYWORDS

Richter transformation, CLL, Diffused large B-Cell lymphoma, Tumor lysis syndrome

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