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623.MANTLE CELL, FOLLICULAR, AND OTHER INDOLENT B CELL LYMPHOMAS: CLINICAL AND EPIDEMIOLOGICAL

Leukostasis Twist in Mantle Cell Lymphoma: Systematic Review and Case Report

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Introduction:
Mantle cell lymphoma (MCL) is an aggressive B-cell non-Hodgkin lymphoma. Patients often present with lymphadenopathy, early satiety, and B-symptoms. Despite up to 92% of MCL patients having peripheral blood involvement by flow cytometry, presentation with leukemic involvement and hyperleukocytosis are uncommon.
Hyperleukocytosis, marked by a white blood count (WBC) >100 x 10^9/L, often leads to symptomatic leukostasis due to tissue hypoxia, generally found in acute leukemias. We conducted a systematic review of leukostasis in MCL, and present a case we encountered in our clinical practice of a non-nodal MCL variant showing leukostasis, spontaneous tumour lysis syndrome (TLS), and severe anemia to identify treatment strategies and outcomes.
Methods:
We searched PubMed for peer-reviewed articles and abstracts on "mantle cell lymphoma," "hyperleukocytosis," and "leukostasis." All articles and abstracts that discuss hyperleukocytosis leading to leukostasis in MCL were included.
Results:
The search yielded eight results. After exclusions, five case reports and abstracts were included. These studies featured different MCL variants, presenting WBCs ranging from 121 to 1227 x 10^9/L, and various leukostasis symptoms. Leukapheresis was used in all five published cases, leading to lower WBCs and symptom improvement. Results are summarized in *Table 1.*
Our patient was found to have the small lymphocyte variant of MCL, with WBC of 543.3 x 10^9/L on presentation. Our facility does not have leukapheresis capability. As a result, our initial treatment consisted of only supportive care and escalating doses of corticosteroids, with close monitoring for TLS. To our knowledge, this report is the first demonstration that IV methylprednisolone, without leukapheresis, is effective for acute management of leukostasis. Our patient's WBC reduced from 543.3 x 10^9/L to 230 x 10^9/L, leading to improved neurological function and oxygenation. Steroids were particularly useful in our patient with secondary, MCL-related warm autoimmune hemolytic anemia.
Conclusion:
This review highlights the importance of recognizing non-specific leukostasis symptoms, even in patients with mature B-cell lymphoproliferative disorders. Leukapheresis is effective in treating leukostasis in MCL patients. In hospitals without leukapheresis, escalating doses of IV methylprednisolone can be considered as a safe and effective strategy.

Disclosures Crowther: *Bayer:* Honoraria; *Eversana:* Consultancy; *Syneos Health:* Consultancy; *Hemostasis Reference Laboratory:* Consultancy; *Precision Biologics:* Consultancy; *Astra-Zeneca:* Consultancy; *Treasurer, American Society of Hematology:* Membership on an entity's Board of Directors or advisory committees; *CSL Behring:* Honoraria; *Pfizer:* Honoraria.

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Selected Articles	Underlying Subtype of MCL	Clinical Presentation, and Relevant Investigations	Management	Outcome
Smith et al., 2004	Pro-lymphocytoid	76-year-old male. Oculodynia, blurred vision, visual field defects. WBC 1227 x 10^9/L.	Leukapheresis daily for 7 days. Chemotherapy. Hydroxyurea.	WBC remained >100 x 10^9/L. Patient passed away 6 months after diagnosis.
Kwan et al., 2015	Small lymphocyte	88-year-old female. Respiratory compromise. No interstitial or alveolar infiltration on chest X-ray or CT thorax. On 6L nasal cannula. WBC 465 x 10^9/L.	Two whole blood volume leukapheresis.	WBC down to 221 x 10^9/L in 150 minutes. Respiratory rate from 28 to 18. Arterial oxygen saturation from 91% to 97% on 6L nasal cannula.
Salloum et al., 1998	Uncertain	49-year-old male. Post-initiation of chemotherapy with Filgrastim. WBC 121 x 10^9/L. Paracentral visual blurred spots, dizziness, and weakness. Retinal hemorrhages.	Leukapheresis. Filgrastim held.	Dizziness, generalized fatigue resolved. Vision improved gradually.
Rovnan et al., 2020	Small lymphocyte	64-year-old male. Cough, shortness of breath, with subsequent acute respiratory failure requiring intubation. WBC 336 x 10^9/L. Infectious workup negative.	IV methyl-prednisolone 500 mg daily followed by leukapheresis	Unknown duration of methylprednisolone, or clinical response. Leukapheresis decreased WBC to 38 x 10^9/L. Improved respiratory symptoms. Extubated to room air.
Nguyen at al., 2011	Blastoid	73-year-old male. Fatigue and thoracic pain. WBC 630 x 10^9/L. ECG showed ST elevations. Elevated troponin and CK.	Leukapheresis daily x3 days. No angiogram. After second leukapheresis, patient initiated on chemotherapy.	WBC decreased to 174 x 10^9/L after third leukapheresis. Full recovery.
Case Presentation	Small lymphocyte	75-year-old female. Shortness of breath, gait instability, weakness. Hypoxemic, chest X-ray showed bilateral infiltrates. Ischemic changes on ECG, elevated troponin. WBC 543.3 x 10^9/L. Hb 32 g/L.	IV methylprednisolone at escalating doses from 25 mg once daily to 150 mg once daily, 4 days total. Concurrent allopurinol for TLS prophylaxis.	WBC decreased to 230 x 10^9/L on Day 5, and 149.7 x 10^9/L on Day 9 (with no further steroid or chemotherapy administration between Day 4 and 9). Leukostasis signs and symptoms resolved.

Table 1: Summarizes the clinical presentation, relevant bloodwork and diagnosis, management, and outcomes of the articles yielded from our literature search. MCL, mantle cell lymphoma; WBC, white blood count; TLS, tumour lysis syndrome; Hb, hemoglobin.

Figure 1