Concurrent presentation of AML/MDS and marginal zone lymphoma : an unusual case of bilineage hematologic malignancy

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Introduction and objective:

Marginal zone lymphomas (MZLs) are indolent B-cell lymphomas managed from observation to systemic therapy depending on disease burden.
Their **co-occurrence with MDS or AML** is **exceptionally rare**, with no clear treatment guidelines.
We report a **rare case** of **disseminated MZL** evolving into **high-risk MDS/AML**, highlighting **diagnostic complexity** and **therapeutic challenges** in overlapping myeloid and lymphoid neoplasms.

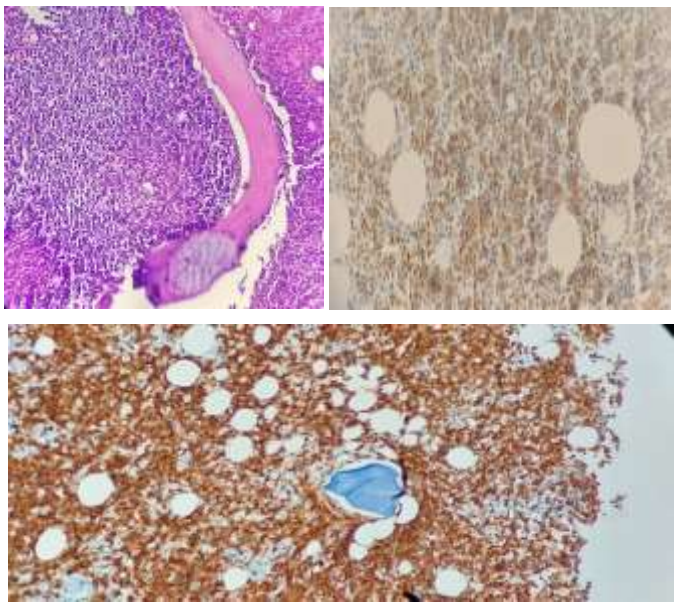

Figure 1 : Photomicrographs of the bone marrow biopsy, showing histology and immunohistochemistry findings consistent with marginal zone lymphoma (MZL), a low-grade B-cell non-Hodgkin lymphoma. Immunostaining reveals positive labeling of lymphocytes with B-cell markers CD20 and PAX5.

Patient and methods:

A 70-year-old man with chronic smoking and prior thyroidectomy presented with **symptomatic anemia** (Hb 4 g/dL) and **bicytopenia** (platelets 80 G/L).
Bone marrow showed **multilineage dysplasia** (1% blasts, normal karyotype); imaging revealed **perirenal infiltration** and **multiple lymphadenopathies**.
Initially diagnosed with **low-risk MDS**, treated with **erythropoietin** without response.
After one year, progression to **MDS-EB2 (17% blasts)** with **MZL infiltration** confirmed on biopsy.
NGS identified high-risk mutations (**ASXL1, RUNX1, TET2, EZH2**).
Treatment started: **Azacitidine + Venetoclax (AZA-VEN)**.

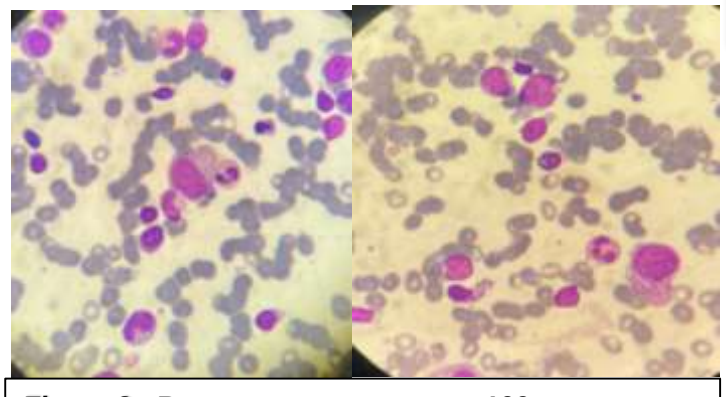

Figure 2 : Bone marrow smear at ×100 magnification showing 17% blasts of medium to large size, with a high nuclear-to-cytoplasmic ratio, regular nuclear contours, fine chromatin, and basophilic cytoplasm, occasionally with sparse granules.

Résultats et discussions:

During AZA-VEN therapy, the patient developed severe thrombocytopenia, leading to a fatal hemorrhagic stroke.
Co-occurrence of **MDS-EB2 and disseminated MZL** is **extremely rare**, posing major diagnostic and therapeutic challenges.
According to the **2022 ICC**, the case fits **MDS/AML with myelodysplasia-related mutations**, associated with **poor prognosis**.
Although **AZA-VEN** shows promising response rates in MDS/AML, **profound cytopenias** remain a major limitation.
This case highlights the need for **molecular-guided management** and **close hematologic monitoring** during treatment.

Conclusion

This case highlights the **high hematologic toxicity** of AZA-VEN, particularly in elderly patients or those with **concurrent myeloid and lymphoid malignancies**.
It underscores the need for **further research** to develop **individualized treatment strategies** that optimize efficacy while minimizing complications and improving outcomes in patients with dual hematologic neoplasms.