Mantle Cell Lymphoma Presenting as an Eyelid Mass: A Rare Extranodal Manifestation of Aggressive B-Cell Lymphoma

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Introduction

Mantle cell lymphoma (MCL) is an uncommon subtype of non-Hodgkin lymphoma, representing only about 5–7% of cases. It originates from B lymphocytes in the mantle zone of lymphoid follicles and is usually driven by the t(11;14)(q13;q32) translocation, which leads to cyclin D1 overexpression and uncontrolled cell growth.

Case Presentation

A 57-year-old male presented with drenching night sweats and a right lower eyelid lesion. Bone marrow biopsy revealed CD20+, CD5+, cyclin D1+ mantle cell lymphoma (MCL) with high Ki-67, indicating an aggressive phenotype. Imaging revealed diffuse osseous lesions, lymphadenopathy and a large right gluteal soft tissue malignant mass. Bone marrow biopsy confirmed involvement with CD5+ B-cells and MF 1–2 reticulin fibrosis. Serum labs showed mild normocytic anemia and thrombocytopenia. PET-CT and further molecular testing are pending, with plans to start treatment with alternating R-CHOP and R-DHAP.

Results

Serum labs

HGB	9.8
WBC	3.4
PLT	137K

Immunophenotypic markers and interpretation of bone marrow biopsy

CD5+	Aberrant T-cell marker on B-cells
CD19+	Expressed on early B-cell precursors through mature B cells (but not on plasma cells)
CD20+	Pan–B-cell marker; a target of Anti-CD20 therapies
FMC7+	Supports MCL over CLL
CD23-	Normally an IgE receptor on mature B cells; absent in MCL, present in CLL
Lambda restricted	Confirms clonal B-cell population in the bone marrow, supporting MCL as opposed to multiple myeloma
KI-67 (45-50%)	Ki-67 proliferation index >30% suggests an aggressive tumor

MRI right lower extremity w/ and w/o contrast
Innumerable widespread patchy bone marrow signal abnormalities throughout the axial and proximal appendicular skeleton consistent with metastatic disease, lymphoma, or multiple myeloma.
Nonspecific enhancing soft tissue malignant neoplasm in the subcutaneous soft tissues of the right buttock measuring approximately 9 x 8 cm x 4 cm.
A separate malignant soft tissue mass in the subcutaneous soft tissues in the right buttock near the midline, measuring 4.2 cm in surface area by 1.5 cm in thickness.
Significant bilateral inguinal and iliac chain lymphadenopathy consistent with metastatic disease or lymphoma.

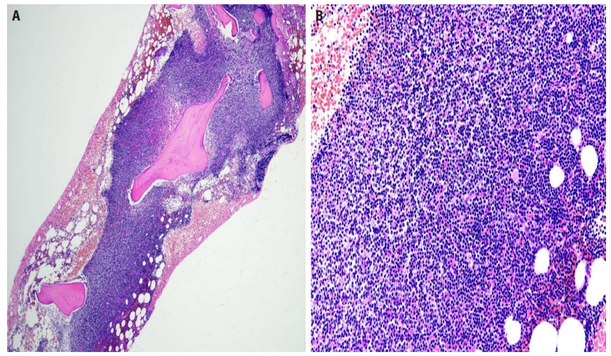
Literature Review

Cutaneous involvement in mantle cell lymphoma is rare, only present in about 1-2% of cases[Akay, Panse]. The median age is 68 years old [Zhou].
There are no population-level percentages of gluteal MCL metastases, however there are isolated case reports of soft tissue MCL involvement.
Extranodal involvement of skin and soft tissues are both unusual for MCL but were both discovered as part of the initial workup for this patient.
Notably, another case reported a patient with previously diagnosed and treated MCL who developed an upper left eyelid lesion misdiagnosed as a benign chalazion, later found to be squamous cell carcinoma[Malsavi]--highlighting the importance of early biopsy.

Results

CT chest without contrast

Bilateral axillary lymphadenopathy with an index left axillary node measuring 1.5 x 2.6 cm



Bone marrow biopsy from a similar case, demonstrating extensive lymphoma involvement[Demianets]

Discussion

This developing case presents an aggressive MCL wherein the skin of the eyelid served as the initial diagnostic site, followed by confirmation of soft tissue, lymph node, marrow and osseous involvement. MCL typically presents at stage IV. Some common extranodal sites include the GI tract, bone marrow, spleen and Waldeyer's Ring. Some of our patient's extranodal manifestations were much more uncommon, as MCL cutaneous involvement makes up ~1-2% of all MCL cases, and only isolated reports describe MCL manifesting in gluteal soft tissues.

This case highlights the importance of obtaining ophthalmology evaluation for atypical appearing eyelid lesions, as this can be a manifestation of aggressive hematologic malignancy. Biopsy of the lesion with immunohistochemistry and molecular profiling was important for diagnosis and prognostication. This case will be followed as the patient completes further treatment and begins chemotherapy.

Resources/References

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