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Zainab Fatima
East Tennessee State University

Haroon Rahman
East Tennessee State University

Sonia Kumari Oad
East Tennessee State University

Elnora Spradling
East Tennessee State University

Devapiran Jaishankar
East Tennessee State University

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Fatima, Zainab; Rahman, Haroon; Oad, Sonia Kumari; Spradling, Elnora; and Jaishankar, Devapiran, "Lymphoma at First Sight: A Rare Case of Mantle Cell Lymphoma Presenting as Isolated Periorbital Swelling" (2020). *Appalachian Student Research Forum*. 26.
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Lymphoma at First Sight: A Rare Case of Mantle Cell Lymphoma Presenting as Isolated Periorbital Swelling

Authors: Fatima,Zainab[1], Rehman, Haroon[2], Oad,Sonia K [2], Spradling, Elnora[2], Jaishankar, Devapiran[2]

Author affiliations: 1. Internal Medicine , East Tennessee State University, Johnson City, TN, 2. East Tennessee State University, Johnson City, TN

INTRODUCTION

- Mantle cell lymphoma (MCL) represents a heterogenous subtype of non-Hodgkin lymphoma (NHL), which can present in three distinct clinicopathologic variants: indolent type MCL, classic type MCL and blastoid type MCL.
- Despite the different variations, MCL, in general, is almost always associated with advanced-stage disease at diagnosis, with a strong predilection for significant extra-nodal involvement, usually to the bone marrow, CNS, peripheral blood and the gastrointestinal tract.
- However, the literature review reveals ocular involvement is a more rarely described extra-nodal site of involvement by MCL. Among the reported cases, the orbit was most commonly involved, followed by the eyelid and the lacrimal gland.

CASE DESCRIPTION

- We report a 63-year-old male who presented with a nine-month history of progressive symptoms of periorbital swelling and eyelid apraxia, causing bilateral visual disturbances. The patient was initially treated for presumed blepharospasm by his ophthalmologist with botulinum toxin injections; however, his periorbital edema continued to worsen, and he developed a discrete nodule in his right lower eyelid.
- Biopsy of the right eyelid nodule revealed classic type MCL with immunohistochemical testing positive for CD20, CD5, cyclin D1, SOX11 and Ki67 proliferative index of 40%. Fluorescence in situ hybridization (FISH) analysis detected (11;14) translocation.

CASE DESCRIPTION (continued)

- Mantle Cell Lymphoma International Prognostic Index Combined Biologic Index (MIPIb) score was calculated to be 6.5 points based on his age, ECOG performance status of 0-1, normal serum LDH, normal white blood cell count and elevated Ki67 proliferative index, stratifying patient into the high-risk group, with an estimated median overall survival of 37 months.
- Due to the bulky MCL involvement in the palpebral conjunctiva affecting his vision and eyelid function, he was immediately treated with radiation therapy to the bilateral orbits. PET-CT revealed adenopathy above and below the diaphragm. Bone marrow biopsy revealed focal involvement (5-10%) by MCL.
- Brain MRI revealed MCL infiltration in the bilateral orbits and lacrimal glands. Upper and lower endoscopy revealed multiple polyps positive for MCL. Given the advanced stage of the disease and his high-risk stratification, he was started on intensive induction chemotherapy with rituximab, dexamethasone, cytarabine, and carboplatin and received prophylactic intrathecal methotrexate. Systemic imaging after completion of four cycles of treatment revealed near resolution of the majority of the lymphadenopathy and all of the lymph nodes no longer demonstrated any significant metabolic activity.
- He completed two additional cycles of systemic chemotherapy and is currently being evaluated for autologous hematopoietic stem cell transplantation in complete remission-1 given his excellent response to treatment, his young age, high-risk disease at diagnosis, and good performance status.

CONCLUSION

Despite the diffuse and extensive systemic disease, interestingly, our patient did not exhibit any constitutional or metastasis-associated symptoms and only presented with isolated periorbital swelling.
Our case emphasizes the rare extra-nodal site of involvement by MCL and encourages all medical providers to remain cognizant of the varying ways in which MCL can present clinically.

PATHOLOGY

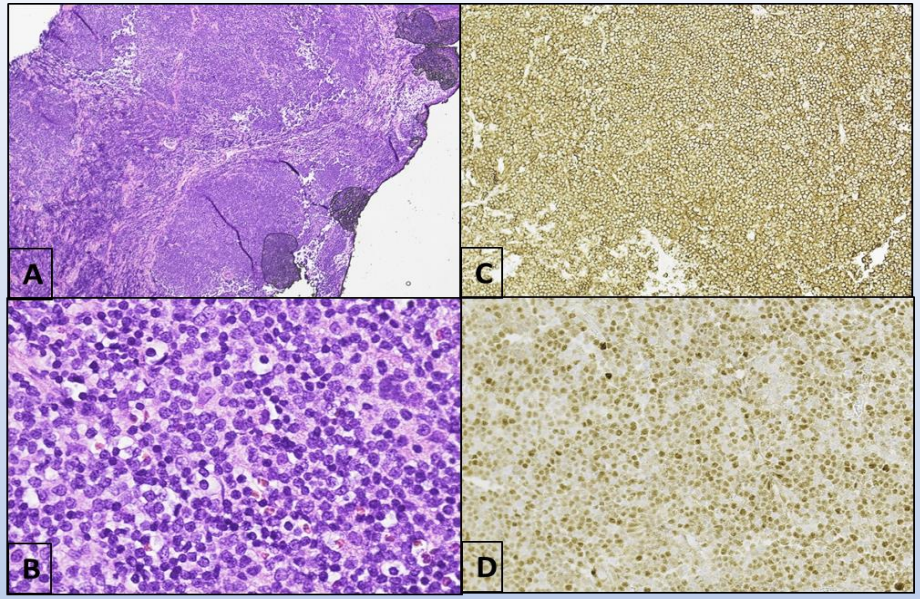

Figure A, The lid biopsy shows diffuse to vaguely nodular lymphoid infiltrate replacing the biopsy. **Figure B**, The tumor cells are small to medium sized round lymphoid cells. They show round nuclei with variable nuclear membrane irregularity, inconspicuous nucleoli and scant cytoplasm. On immunohistochemistry the tumor cells are positive for CD20 (membranous stain) **Figure C,** and Cyclin D1 (nuclear stain), **Figure D** consistent with mantle cell lymphoma staining pattern.

REFERENCES

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