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Scleral depression saves lives

Drew Scoles, Mina Massaro-Giordano, Albert M. Maguire, Vivian Lee*

Department of Ophthalmology, Scheie Eye Institute, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, USA

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1. Case report

A 44-year-old man with no past medical history presented for retinal evaluation due to new onset floater of the left eye. His slit lamp exam was unremarkable including the lids, lashes and conjunctiva. Bilateral dilated fundus exam was unremarkable. When scleral depression was performed on the left eye, a salmon colored tongue-like conjunctival mass appeared from the superior fornix; prolapsing each time the lid was indented with the depressor, and receding when pressure was removed (Fig. 1). The lesion was biopsied, which disclosed B-cell large cell (DLBCL) intermediate grade non-Hodgkin's lymphoma (Fig. 2). Staging examination including bone marrow biopsy determined that the lymphoma was confined solely to the conjunctiva. Patient was treated with 38 gray total external beam radiation to the orbit as well as chemotherapy with cyclophosphamide, doxorubicin, and vincristine (CHOP) with complete remission of disease.

On routine follow-up, 10 years after his initial radiation treatment, he was noted to have a small area of telangiectasia adjacent to the fovea in the left eye consistent with mild radiation retinopathy. At present, nearly 20 years from initial diagnosis, he has had no recurrence of his lymphoma, his visual acuity remains 20/20, and his radiation retinopathy has not progressed (Fig. 3).

2. Discussion

Conjunctival lymphomas comprise a large proportion (25%) of all periocular lymphoma.[1] However, initial presentation by prolapsing a lesion during scleral depression has not been previously reported. By far the majority of conjunctival lymphomas are B-cell non-Hodgkin lymphomas, of which subtypes include: extra-nodal marginal zone (81%); and rarer types, such as follicular (8%), mantle cell (3%) and DLBCL (3%) as described here. Conjunctival lymphoma is generally found in the elderly in the 6th to 8th decades, often presenting as a visible salmon colored mass with discomfort or pressure sensation. The majority of conjunctival B cell lymphomas are primary (80%). More aggressive forms, such as DLBCL, are present in the ocular adnexa as secondary involvement in approximately 25%.[1,2] Prognosis in DLBCL is predicted by the International Prognostic Index, which includes: patient's age, lactate dehydrogenase, Eastern Cooperative Oncology group performance status, Ann Arbor stage, and presence of extranodal disease.[3] Unlike most conjunctival lymphomas, adjuvant chemotherapy in addition to excision and radiotherapy is recommended for DLBCL because of its high-grade malignant status.[1] The prognosis is less favorable for adnexal DLBCL with 44% experiencing recurrence or progression, 36% overall survival, and 47% disease specific survival at 5 years.[2]

* Corresponding author. Scheie Eye Institute, 51 N 39th Street, Philadelphia, PA, 19104, USA.
E-mail address: Vivian.Lee@pennmedicine.upenn.edu (V. Lee).

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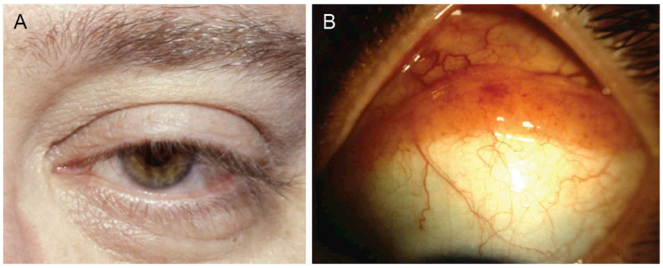

Fig. 1. External photograph of left eye in primary gaze (A), and with eyelid retracted revealing salmon colored patch of conjunctiva (B). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

3. Conclusion

Scleral depression may lead to incidental diagnoses of life-threatening conditions due to prolapse and exposure of forniceal tumors. Early diagnosis of DLBCL in this case led to an atypical outstanding outcome.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

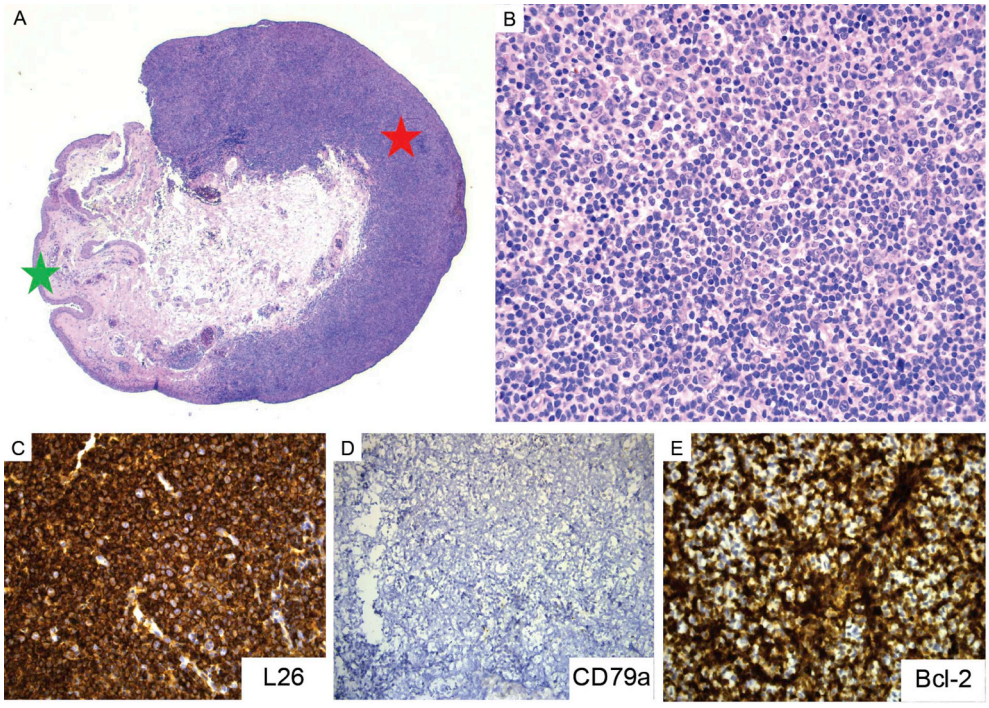

Fig. 2. Photomicrograph demonstrating dense atypical lymphocytic infiltration of conjunctiva (red star) adjacent to unremarkable conjunctiva (green star) at low magnification (A) and high magnification (B). Atypical lymphocytes are L26 and Bcl-2 positive (C, E) while CD79 negative (D). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

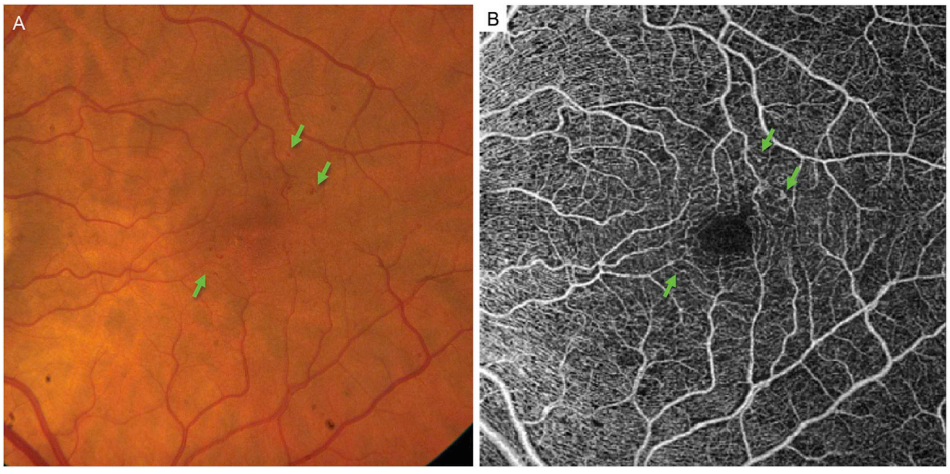

Fig. 3. Color fundus photograph (A) and OCT angiography (B) of the left eye revealing clusters of microaneurysms (green arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

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Verbal informed consent for publication of this report was obtained from the patient.

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