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Primary cutaneous extranodal marginal zone B-cell lymphoma of the eyelid skin: Diagnostic clues and distinction from other ocular adnexal diseases

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ABSTRACT

A 60-year-old man developed a rubbery thickening and erythema of his left lateral upper and lower eyelids and lateral canthus over several months. He was treated for an extended period of time for blepharitis and chalazia. Incisional biopsy eventually disclosed microscopically a hypercellular lymphoid population sparing the epidermis that surrounded adnexal structures and infiltrated between orbicularis muscle fibers. Immunohistochemically, the lesion was found to be composed of neoplastic, kappa-restricted B cells with an equal number of reactive T cells and small reactive follicles. The diagnosis was a primary cutaneous extranodal marginal zone B-cell lymphoma of the eyelid skin (EMZL). We review the distinguishing clinical, histopathologic, and immunohistochemical features of cutaneous EMZL and contrast those with EMZL of other ocular adnexal sites. Also offered is a differential diagnosis of cutaneous lymphomas of the eyelid skin, which are predominately T-cell lesions.

1. Introduction

Extranodal marginal zone B-cell lymphoma (EMZL), also called mucosa-associated lymphoid tissue lymphoma (MALT), is the most common lymphoma of the ocular adnexa (conjunctiva, orbital soft tissues, and lacrimal gland).[5] Primary and secondary cutaneous lymphomas of the eyelid skin are quite uncommon and are usually of T-cell origin, as are cutaneous lymphomas of other sites.[23] We report a case of primary cutaneous extranodal marginal zone B-cell lymphoma of the upper and lower eyelid skin and lateral canthus and discuss the morphologic distinctions between cutaneous

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EMZL of the eyelid skin and the much more commonly encountered EMZL of other ocular adnexal sites.

2. Case report

2.1. *Clinical history*

A 60-year-old generally healthy man presented to his ophthalmologist with a 1-month history of sore and red left eyelids. He was diagnosed as having bilateral meibomian gland disease with eyelid telangiectasias and thickening and erythema of the left lateral canthus (Fig. 1A), as well as left upper and lower eyelid chalazia. Over the following months, he was treated with warm compresses, tobramycin/dexamethasone ophthalmic drops, oral doxycycline and eventually an intralesional steroid injection of the "chalazia," with temporary relief. Six months after his initial presentation, he was referred to an ophthalmic plastic surgeon because of persistence of the disease and progressive eyelid swelling. There was a suggestion of eyelash thinning, and the lesion was rubbery on palpation. A biopsy of the left upper and lower eyelids was performed. A subsequent systemic evaluation by an oncologist revealed no hematologic laboratory

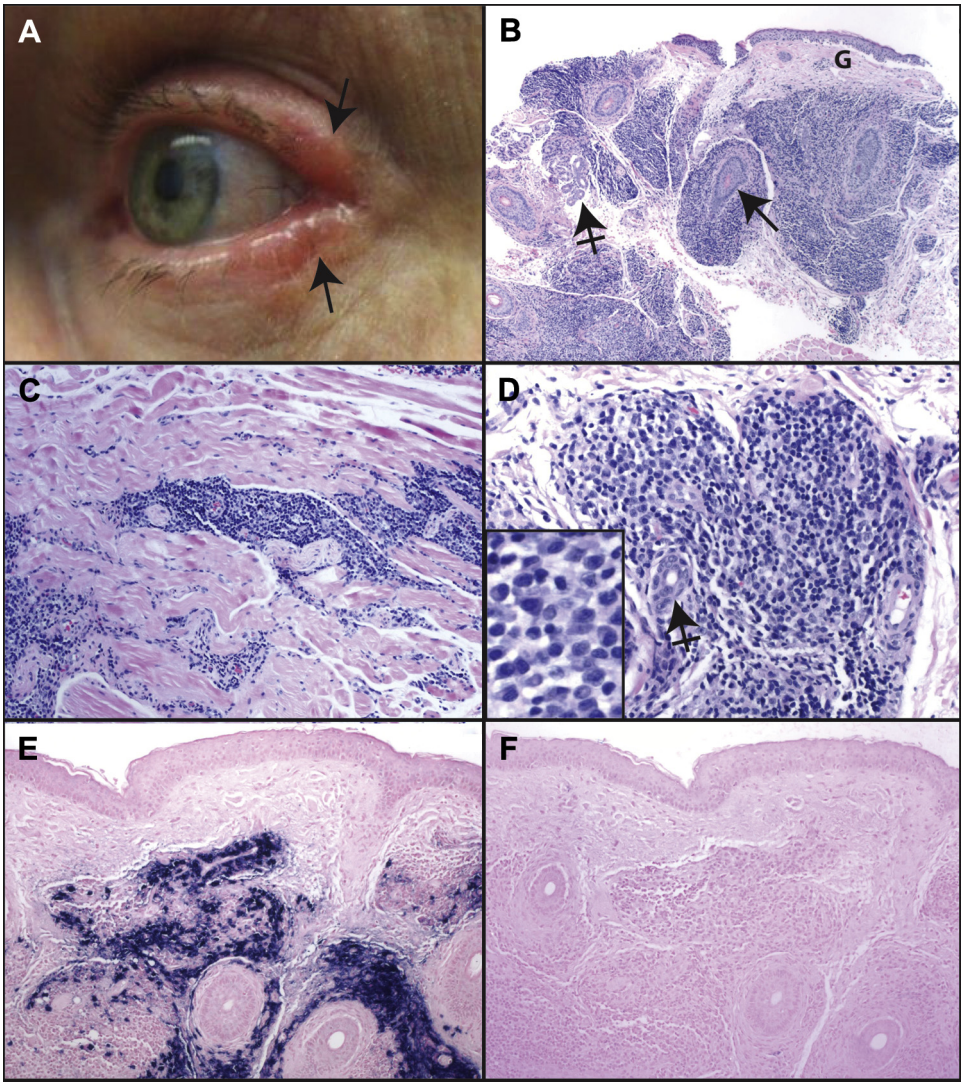

Fig. 1 – Clinical and histopathologic features and *in situ* hybridization studies of primary cutaneous extranodal marginal zone B-cell lymphoma of the eyelid skin. *A:* A 60-year-old man developed lateral loss of eyelashes and eyelid thickening and erythema (*arrows*) of the lateral canthus. *B:* A dense dermal lymphoid infiltrate is seen at low power beneath a normal appearing epidermis. The lymphoid cells surround hairs (*arrow*) and sweat glands (*crossed arrow*). The grenz (border) zone of the upper dermis (G) is free of tumor. *C:* The lymphoid cells infiltrate between orbicularis muscle fibers. *D:* At higher power, the lymphoid cells are noted to be small with slightly irregular nuclei and admixed mature plasma cells, seen to better advantage at higher power in the inset. The *crossed arrow* highlights a sweat duct. *E: In situ* hybridization for kappa immunoglobulin light chain reveals that the plasma cell population is kappa restricted. *F: In situ* hybridization for lambda immunoglobulin light chain is negative. (*B–D*, hematoxylin and eosin, 50 X, 100 X, 200 X, inset 400 X; *E–F*, *in situ* hybridization, 100 X, 100 X).