Ocular Marginal Zone B Cell Lymphoma of a Mucosa-associated Lymphoid Tissue(MALT) Masquerading as Chalazion : A Case Report and Review of Literature

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Abstract

Background: To report a case of Ocular Marginal Zone B Cell Lymphoma Of A MALT presenting as chalazion.

Material & Methods: Case report

Result: A 57-year-old Chinese gentleman, known to have Diabetic Mellitus and HBV positive presented with painless right lower lid swelling and conjunctiva injection for 3 months duration. A diagnosis of chalazion was made and incision and curettage (I&C) performed by a general ophthalmologist. The swelling worsened and spread to the whole lower lid. Magnetic resonance imaging showed a lesion involving right periorbital region limited to anterior of the orbital septum that was hypointense on T1, and hyperintense on T2. Histopathology of the conjunctiva biopsy showed features of low-grade Non-Hodgkin B-cell lymphoma and immunohistochemistry report suggestive of marginal zone lymphoma.

Conclusion: This case showed a rare presentation of Ocular Adnexal Lymphoma (OAL) manifested as chalazion

Conflict of interest: None

Keywords: Marginal zone lymphoma, Non-Hodgkin's B cell lymphoma, MALT lymphoma, Chalazion, conjunctiva

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Case report

A 57-year-old gentleman, who is a known Type 2 Diabetes Mellitus (T2DM) first presented in August 2016, complaining of a painless lower lid swelling and conjunctival injection for 3 months duration. It was gradually increasing in size and associated with foreign body sensation. He was initially treated by a private ophthalmologist conservatively as chalazion with oral antibiotics but the condition did not improve. He sought treatment at another center, and an I&C was performed. However, the lid swelling, as well as the conjunctiva injection, worsened, thus MRI orbit was ordered. The result showed lesions involving right periorbital region limited to anterior of orbital septum measuring 2.9cm (W) x 1.3cm (AP) x 2cm (CC). The lesions involved both upper and lower lid which was hypointense on T1, hyperintense on T2, not suppressed on T2 fat suppression and enhancing on gadolinium. No involvement of other structures noted. Impression given was suggestive of periorbital cellulitis with a differential of lymphoma. He presented to our center for an opinion. He denied any constitutional symptoms and family history of malignancies. His visual acuity was 6/6. Examination of the right eye showed a hard, non-tender mass measuring 4x2cm within the lower eyelid associated with conjunctival injection (Figure 1). Examination of other ocular structures showed no abnormalities except for early cataract in both eyes.

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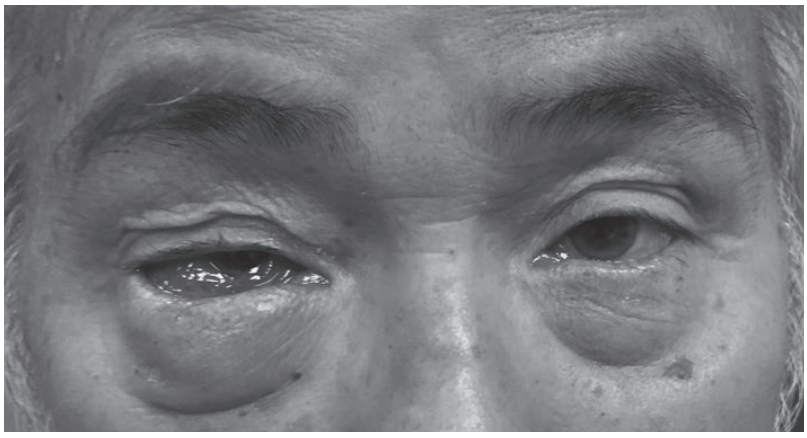

Figure 1 Anterior view on presentation

Biopsy of the conjunctiva showed features of a low-grade Non-Hodgkin B-cell lymphoma (Figure 2).

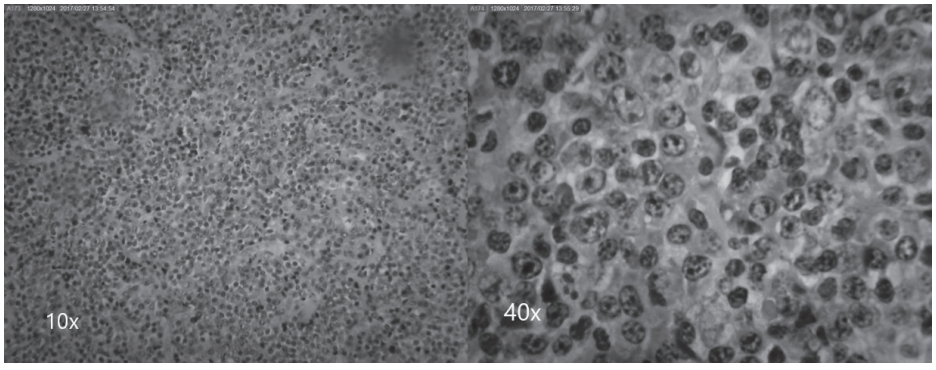

Figure 2 H&E: mixed population of lymphoid cells ranging from small condensed nuclei with scanty cytoplasm to larger vesicular nuclei with prominent nucleoli. plasma cells are seen

Histological features and immunohistochemistry (IHC) are suggestive of marginal zone lymphoma (Figure 3, Table 1).

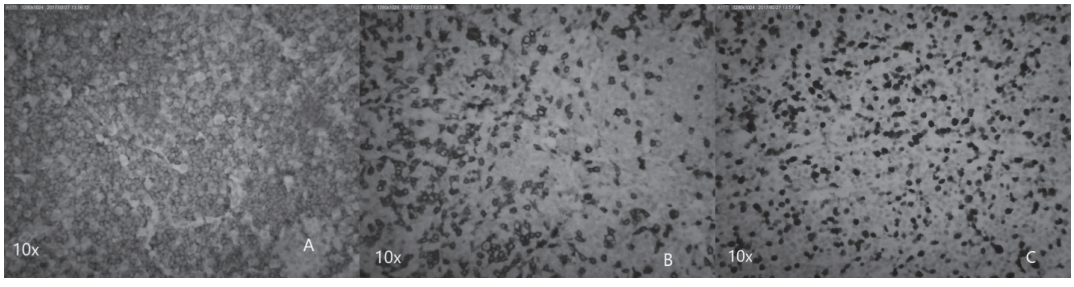

Figure 3- A Lymphoid cells with larger cytoplasm are positive for CD20 (B-cells) **B** Lymphoid cells with smaller cytoplasm are positive for both CD3 (T-cells) and CD20 (B-cells) **C** Ki-67 highlight mitotic activity raised and present in both larger and smaller lymphoid cells

Table 1: Immunophenotypic markers analyzed in the clinical case

CD20	+
CD3	-
CD5	-
CD10	-
CD23	-
Cyclin D1	-
Ki-67	30%
Special stain	-

Bone Marrow Aspiration and Trephine (BMAT) examination were done, showing no evidence of non-Hodgkin's lymphoma. It was noted that during routine baseline blood investigation, he was found to have Hepatitis B positive with chronic hepatitis. A CT Brain, neck, thorax, abdomen and pelvis for staging was performed. The result showed enhanced soft tissue masses in the right upper and lower eyelids measuring 1.0 x 3.0 x 1.4 cm with no evidence of lymphadenopathy (Figure 4), thus the diagnosis was T1cN0M0 based on TNM clinical staging for OALs[18].

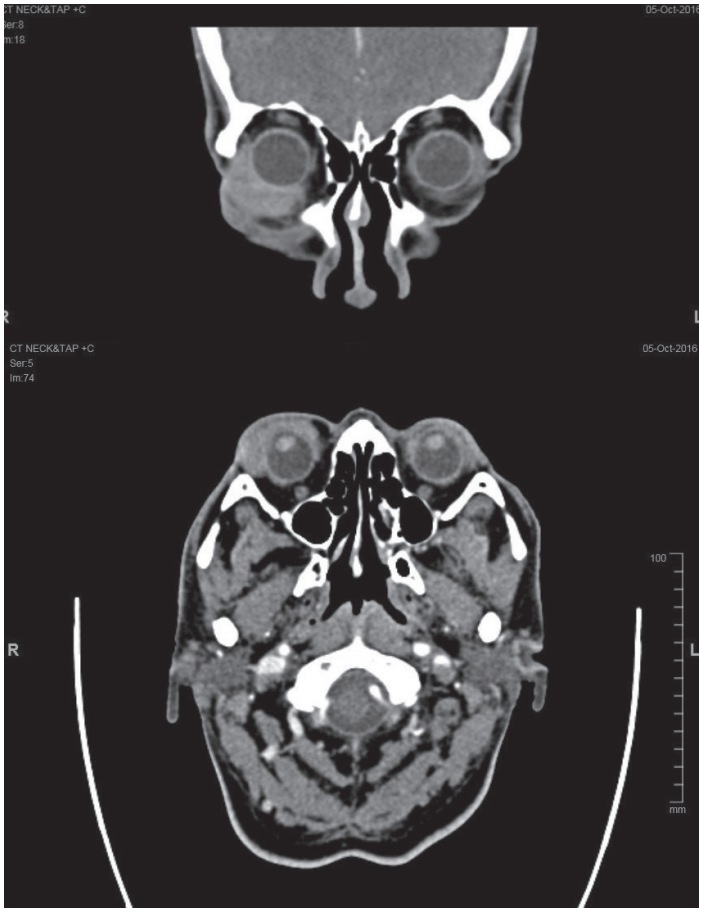

Figure 4 CT brain orbit showing enhancing soft tissue masses in the right upper and lower eyelids

He was subsequently referred to the hematology team where chemotherapy regime cyclophosphamide, vincristine, prednisolone (CVP) was started in November 2016. However, the lesion and swelling worsened. Re-biopsy was consistent with previous findings. He was then started on radiotherapy in March 2017, which he completed for 15 cycles. 10 months post completion of radiotherapy, the patient showed improvement, with reduced lid swelling and conjunctival injection (Figure 5). Repeated scan showed stable lymphoma.

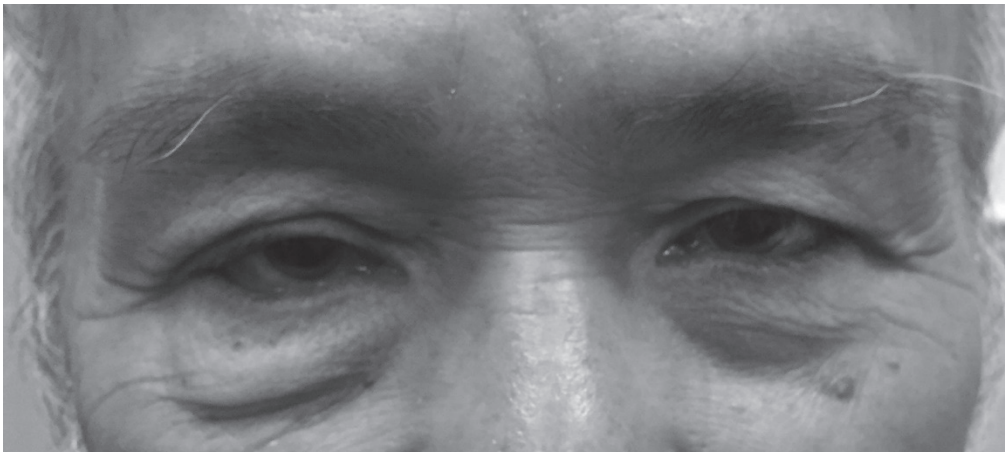

Figure 5 10 months post radiotherapy

Discussion

We have reported the first case of conjunctiva marginal zone B cell lymphoma of a MALT tissue masquerading as a chalazion. OAL can present with sinister onset, gradually progressive and non-painful mass that can involve eyelid, orbital soft tissue, conjunctival tissues, muscle, or lacrimal gland. Age of appearance is generally between 5th to 7th decade of life[2] with no gender predisposition[8] and it is the most common orbital tumor (accounting for 24% cases) in age group >60 years[4]. The most common type of OAL is EMZL or MALT lymphoma. It is a feature in about 50% of cases[9,10]. EMZL most commonly involve the orbit (60%) followed by conjunctiva (33%), lacrimal gland (4%), and eyelid (3%)[10].

Conjunctival lesions typically present as mobile pink infiltrates ("salmon-pink patch") in the substantia propria either at palpebral conjunctiva or inferior bulbar causing conjunctival swelling, redness, and irritation[11]. As the lesion arises from substantia propria, the covering epithelium is typically normal. This patient sought treatment at multiple centers for a prolonged period of time. Initial treatment centered on the diagnosis of localized infection, which did not resolve with antibiotics as well as surgical I&C of the lesion.

There have been reports of conjunctival lymphoma mimicking allergic or chronic conjunctivitis[12-14] in which presentation was bilateral, atypical, normal-colored, papilla-like lesions or inflammation. In another case series, patients initially treated as conjunctivitis and lacrimal duct obstruction, in fact had trivial lesions in the fornices which was later diagnosed as conjunctival lymphoma[15]. Other case reports include conjunctival lymphoma masquerading as scleritis[16]. Previously, a primary cutaneous extranodal marginal zone B-cell lymphoma of the eyelid skin has been reported presenting as blepharitis and chalazion[17].

A high index of suspicion of a chronic lesion should be present when patients do not respond to treatment as expected. Suspicious lesions should be biopsied in order to achieve the diagnosis. Heightened awareness and quick

recognition of these tumors are essential to avoid oversight and misdiagnosis, and the subsequent delays in commencing treatment and probable systemic involvement.

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