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Ocular marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue masquerading as chalazion: a case report

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Abstract

A case of ocular marginal zone non-Hodgkin B-cell lymphoma of mucosa-associated lymphoid tissue of the conjunctiva masquerading as chalazion is reported in a 57-year-old Chinese man, known to have diabetes mellitus. He presented with painless swelling of the lower lid and redness in the right eye for 3 months duration. A diagnosis of chalazion was made, and incision and curettage were performed by a general ophthalmologist. The swelling worsened and spread to the whole lower lid. Magnetic resonance imaging showed a lesion involving the right periorbital region limited to the anterior orbital septum that was hypointense on T1 and hyperintense on T2. A diagnosis of periorbital cellulitis with possibility of lymphoma was suggested.

When he came to our eye clinic for expert opinion, his visual acuity, anterior segment and fundus were normal except early cataract changes in both eyes. There was a hard, non-tender, immobile mass within the lower eyelid associated with conjunctival injection and chemosis. Histopathology of the conjunctival biopsy showed features of low-grade non-Hodgkin B-cell lymphoma and the immunohistochemistry report was suggestive of marginal zone lymphoma. He was treated with chemotherapy (cyclophosphamide, vincristine, prednisolone) and radiotherapy, following which the swelling resolved.

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When patients do not respond to treatment as expected after incision and curettage of chalazion, a high index of possibility of malignancy should be kept in mind whenever a recurrence of hard swelling is observed at the same site in the eyelid. All suspicious lesions should be biopsied to find out the correct diagnosis.

Keywords: chalazion, conjunctival tumour, marginal zone lymphoma, non-Hodgkin B-cell lymphoma, radiotherapy

Limfoma sel B zon marginal tisu limfoid berkaitan mukosa yang menyamar sebagai kalazion: laporan kes

Abstrak

Kes limfoma sel B non-Hodgkin sel limfoma tisu limfoid yang berkaitan dengan mukosa konjunktiva yang menyamar sebagai kalazion dilaporkan pada seorang lelaki Cina berusia 57 tahun, yang diketahui menghidap diabetes mellitus. Dia mengalami bengkak tanpa rasa sakit pada kelopak mata bawah dan kemerahan di mata kanan selama 3 bulan. Diagnosis chalazion dibuat, dan prosedur insisi dan kuretej dilakukan oleh pakar oftalmologi umum. Pembengkakan menjadi bertambah buruk dan merebak ke seluruh penutup bawah. MRI menunjukkan lesi yang melibatkan kawasan periorbital kanan yang terhad kepada septum orbital anterior yang hypointens pada T1 dan hyperintens pada T2. Diagnosis selulitis periorbital dengan kemungkinan limfoma diusulkan sebagai diagnosis.

Ketika kehadiran beliau ke klinik mata kami untuk konsultasi pakar, ketajaman penglihatan, segmen anterior dan fundusnya normal kecuali perubahan katarak awal pada kedua matanya. Terdapat ketulan yang tidak lembut dan tidak bergerak di kelopak mata bawah yang berkaitan dengan suntikan konjunktiva dan kemosis. Histopatologi biopsi konjunktiva menunjukkan ciri limfoma sel B bukan Hodgkin kelas rendah dan laporan imunohistokimia menunjukkan limfoma zon marginal. Dia dirawat dengan kemoterapi (siklofosfamid, vincristine, prednisolone) dan radioterapi, setelah itu pembengkakannya hilang.

Jika pesakit tidak bertindak balas terhadap rawatan seperti yang diharapkan setelah prosedur insisi dan kuretej kalazion, kecurigaan yang tinggi akan kemungkinan malignan harus diingat setiap kali berlaku pembengkakan yang keras secara berulang diperhatikan di tempat yang sama di kelopak mata. Semua luka yang mencurigakan harus dilakukan biopsi untuk mengetahui diagnosis yang tepat.

Kata kunci: chalazion, limfoma sel B bukan Hodgkin, radioterapi, tumor konjunktiva, limfoma zon marginal

Introduction

Ocular adnexal lymphoma (OAL) or orbital lymphoproliferative disorders include a heterogeneous group of lymphoid cell disorders that can affect orbital soft tissues, conjunctiva, eyelid, or adnexal structures such as the lacrimal drainage system or lacrimal gland.[1] Generally, they are classified as Hodgkin and non-Hodgkin lymphoma (NHL).[2] The most common primary OAL is low-grade malignant extranodal marginal zone B-cell lymphoma (EMZL) of mucosa-associated lymphoid tissue (MALT) type.[1] Ocular and adnexal lymphomas comprise 5–10% of all extranodal lymphomas.[3] However, OAL is the most common orbital tumour, especially in older populations, with an incidence ranging from 11% to 24% of all orbital tumours.[3] Most cases involve adults over 60 years old, with no gender predilection.[4] Bilateral disease is observed in approximately 10–15% of cases.[3] The incidence of NHL in the general population is on the rise, most likely due to immunodeficiency syndromes, organ transplantation, autoimmune diseases, and involvement of several pathogenic viruses. Depending on location, OAL can present with a spectrum of clinical manifestations. Presentations include proptosis, mass at the eyelid, conjunctival mass, ocular motility restriction, or diplopia. However, visual deterioration is uncommon. Conjunctival OAL is classically seen as a salmon pink patch at the bulbar conjunctiva.

A literature search in PubMed, Science Direct, and Google Scholar did not reveal any case reports of ocular marginal zone NHL from Malaysia. Therefore, we report the first case of ocular marginal zone non-Hodgkin B-cell lymphoma of MALT in the eyelid masquerading as a chalazion.

Case report

A 57-year-old Chinese man, known case of type II diabetes mellitus, first presented in August 2016 with a complaint of a painless lower lid swelling and redness in the right eye for 3 months duration. It was gradually increasing in size and associated with foreign body sensation. Initially, he was treated conservatively by a private ophthalmologist as lower eyelid chalazion with oral antibiotics, but the condition did not improve. He sought treatment at another centre, and an incision and curettage of the lesion was performed. However, the lid swelling and conjunctival injection worsened, prompting magnetic resonance imaging of the orbit. The result showed lesions involving the right periorbital region limited to the anterior orbital septum measuring 2.9 cm (W) x 1.3 cm (AP) x 2 cm (CC). The lesions involved the lower lid,

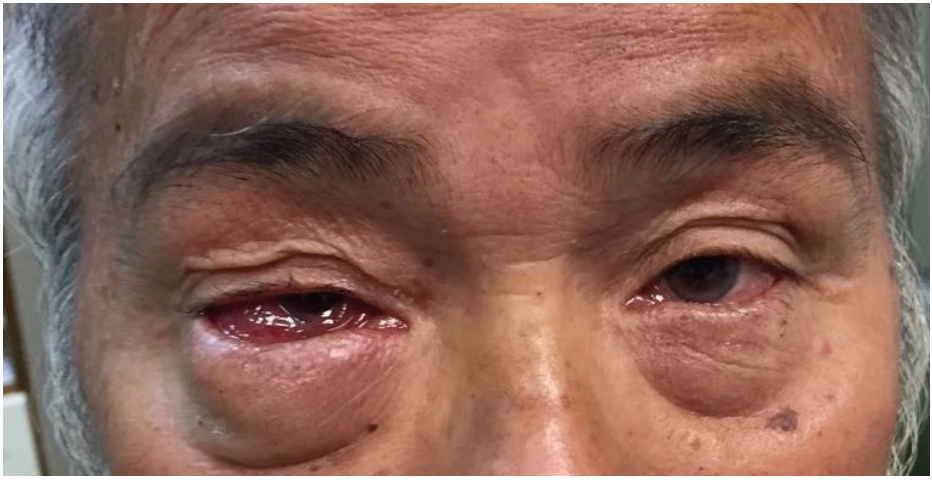

Fig. 1. Mass (4 x2 cm) in the lower eyelid below the lid margin and chemosis of the conjunctiva in the right eye.

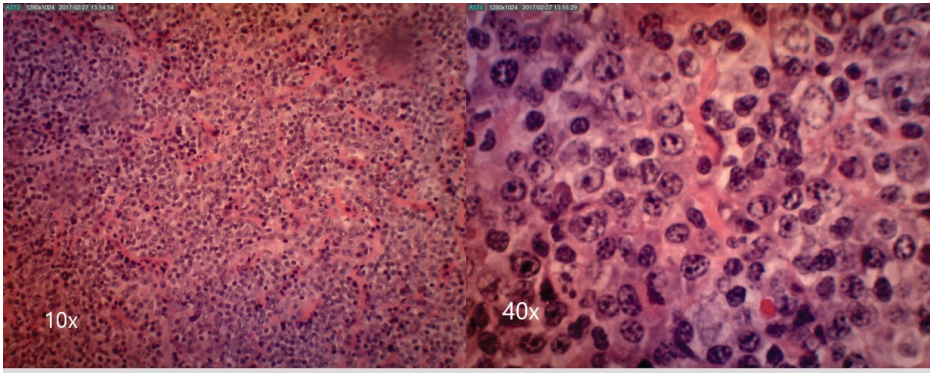

Fig. 2. H&E stain showing the mixed population of lymphoid cells ranging from small, condensed nuclei with scanty cytoplasm to larger vesicular nuclei with prominent nucleoli. Plasma cells are also seen.

which was hypointense on T1 and hyperintense on T2, not suppressed on T2 fat suppression and enhancing on gadolinium. No involvement of other structures was noted. The diagnosis given was suggestive of periorbital cellulitis with a differential diagnosis of lymphoma. He presented to our eye clinic for expert 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, immobile mass measuring 4 x 2 cm within the lower eyelid associated with conjunctival injection and chemosis (Fig. 1). Examination of other ocular structures showed no abnormalities except for early cataract in both eyes.

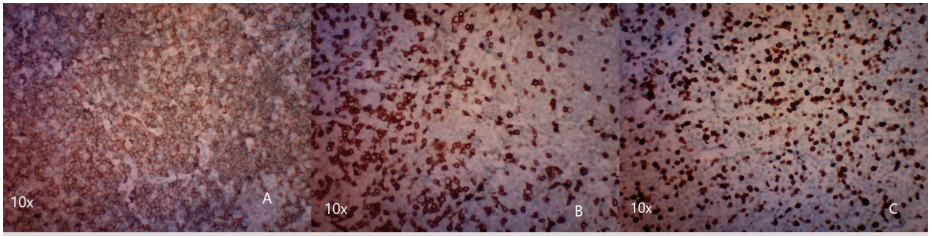

Fig. 3. Immunohistochemistry results. (*A*) Lymphoid cells with larger cytoplasm positive for CD20 (B-cells). (*B*) Lymphoid cells with smaller cytoplasm positive for both CD3 (T-cells) and CD20 (B-cells). (*C*) Ki-67 highlights raised mitotic activity and present in both larger and smaller lymphoid cells.

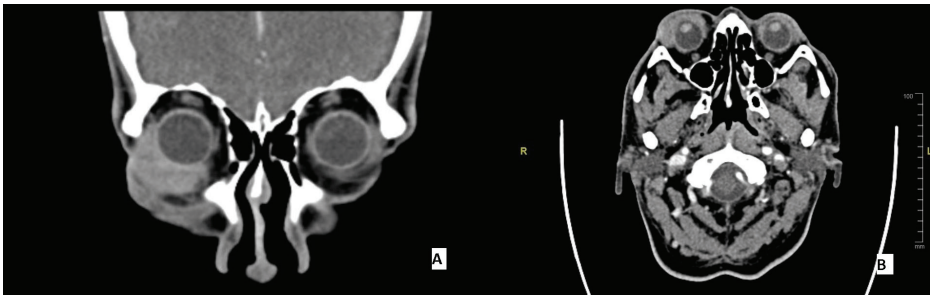

Fig. 4. Coronal computerized tomography cuts of the brain and orbits showing enhancing soft tissue mass in the right lower eyelid (*red arrow*).

Biopsy of the conjunctiva showed features of a low-grade non-Hodgkin B-cell lymphoma (Fig. 2). The histological features and immunohistochemistry results were suggestive of marginal zone lymphoma (Fig. 3). The immunophenotype markers results were positive for CD3 and CD20, negative for CD5, CD10, CD23, and cyclin-D, and 30% Ki-67.

Bone marrow aspiration and trephine examination showed no evidence of systemic NHL. A computerized tomography of the brain, neck, thorax, abdomen, and pelvis done for staging showed an enhanced soft tissue mass in the right lower eyelid measuring 1.0 x 3.0 x 1.4 cm with no evidence of lymphadenopathy (Fig. 4). Thus, the diagnosis was T1cN0M0 based on TNM clinical staging for OALs.[5]

The patient was subsequently referred to the haematology team, where a chemotherapy regime of cyclophosphamide, vincristine, prednisolone (CVP) was started in November 2016. Nonetheless, 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. Ten months post-completion of radiotherapy, the patient showed improvements, with resolved lid swelling and conjunctival injection (Fig. 5). A repeat scan showed stable lymphoma.

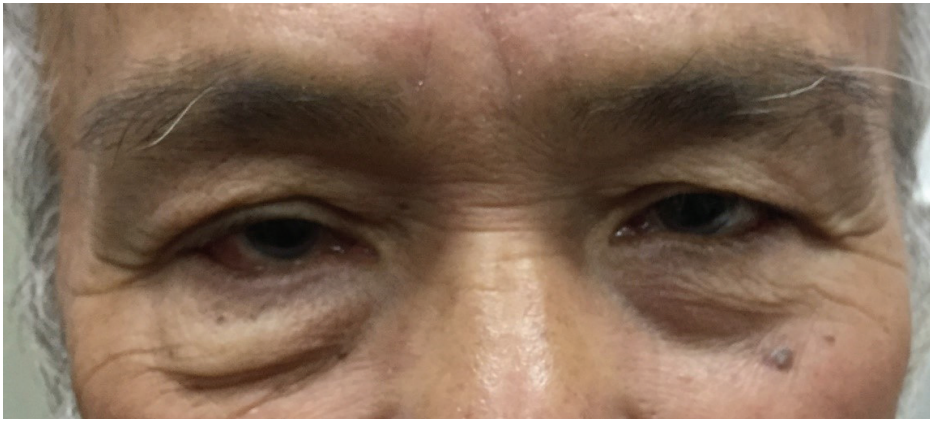

Fig. 5. Right eye showing marked reduction in the size of the conjunctival tumour in the lower eyelid 10 months post-completion of radiotherapy.

Discussion

We have reported the first case of conjunctiva marginal zone-B cell lymphoma of a MALT tissue masquerading as a chalazion in Malaysia. OAL can present with sinister onset, gradually progressive, and non-painful mass that can involve the eyelid, orbital soft tissue, conjunctival tissues, muscle, or lacrimal gland. Age of appearance is generally between 50 and 70 years of age, with no gender predilection.[4] It is the most common orbital tumour (accounting for 24% of cases) in the age group > 60 years. The most common type of OAL is EMZL of MALT lymphoma. It is a feature in more than 50% of cases.[5] EMZL most commonly involve the orbit (60%) followed by conjunctiva (33%), lacrimal gland (4%), and eyelid (3%).[4]

Conjunctival lesions typically present as mobile pink infiltrates ("salmon-pink patch") in the substantia propria either at the palpebral or inferior bulbar conjunctiva, causing conjunctival swelling, redness, and irritation. As the lesion arises from the substantia propria, the covering epithelium is typically normal.

This patient sought treatment at multiple centres for a prolonged period. Initial treatment centred on the diagnosis of localized infection, which did not resolve with antibiotics as well as surgical incision and curettage of the lesion.

There has been a report of conjunctival lymphoma mimicking allergic or chronic conjunctivitis in which presentation involved bilateral, atypical, normal-coloured, papillae-like lesions, and inflammation.[6] Previously, a primary cutaneous EMZL of the eyelid skin has been reported presenting as blepharitis and chalazion.[7]

When patients do not respond to treatment as expected after incision and curettage of chalazion, a high index of possibility of malignancy should be kept in mind whenever a recurrence of hard swelling is observed at the same site in the

eyelid. All suspicious lesions should be biopsied to find out the correct diagnosis. Heightened awareness and quick recognition of malignancy are essential to avoid oversight and misdiagnosis, as well as the subsequent delay in commencing treatment and missing the probable systemic involvement.

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