# Conjunctival lymphoma

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## DESCRIPTION

A man in his 60s with no history of autoimmune diseases or preceding infections presented with bilateral painless red eyes. For the last few weeks, the patient had noticed painless conjunctival redness in both of his eyes, left worse than right. The redness was associated with some tear production and blurry vision but no purulent discharge. Physical examination revealed diffuse, salmon-coloured, slightly raised lesions involving most of his left conjunctiva and a smaller lesion involving his lateral right conjunctiva (figure 1A). Diffuse lymphadenopathy was noted on nodal survey. Laboratory studies revealed leukocytosis ($14.46\times10^9$/L) with lymphocyte predominance (78%). Serum protein electrophoresis detected the presence of M-component (IgM Lambda, 0.36 g/dL). CT scans of the chest, abdomen and pelvis demonstrated lymphadenopathy involving the axilla, mediastinum, retroperitoneum and pelvis.

The patient underwent bilateral conjunctival biopsies. Pathology revealed infiltrations of small-sized to medium-sized lymphocytes (figure 1B) that were PAX5 (+) (figure 1C), CD3 (−) (figure 1D), CD5 (−) and CD10 (−) consistent with a diagnosis of CD5 (−)/CD10 (−) B lineage non-Hodgkin's lymphoma (NHL). Flow cytometry showed a population of variably sized (predominantly small-sized to medium-sized) cells with the following immunophenotype: CD5 (−), CD10 (−), CD19 (+), CD20 (+), CD38 (partial +), CD45 (+), kappa (−), lambda (+). Bone marrow biopsy demonstrated extensive infiltration with cells of similar morphology and immunophenotype. No chromosomal abnormalities were noted on karyotype. Molecular analysis of the myeloid differentiation primary response 88 (*MYD88*) gene was negative for mutations. Based on the above results and extranodal involvement, this patient was diagnosed with stage IV marginal zone lymphoma with conjunctival involvement (conjunctival lymphoma).

Ocular adnexal lymphoma (OAL) is a rare manifestation of NHL and accounts for 1%–2% of all NHL and 5%–10% of all extranodal lymphomas.[1] About 25%–30% of OALs involve the conjunctiva.[1] Conjunctival lymphoma typically presents in the fifth to seventh decade of life as a painless salmon-pink patch in the bulbar conjunctiva, which can be misdiagnosed as conjunctivitis. Clinically, reactive lymphoid hyperplasia can also appear nearly identical to conjunctival lymphoma, so it is crucial to establish the correct diagnosis via biopsy and further studies.

Treatment of conjunctival lymphoma is determined by the stage and extent of disease. Patients with disease localised to the ocular adnexa are typically treated with external beam radiation therapy at the site of disease, which typically results in excellent disease control.[2 3] Patients with systemic disease are typically treated similarly to those with nodal marginal zone lymphoma. First-line therapies include single-agent rituximab, rituximab and bendamustine, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone) and R-CVP (rituximab, cyclophosphamide, vincristine, prednisone).[2] Recurrence rates after treatment can be significant (30%–80%) and close observation is warranted.[4] This patient was initially treated with single-agent rituximab resulting in a short-lived partial response. Patient was then treated with rituximab and bendamustine with subsequent resolution of adenopathy and conjunctival infiltration.

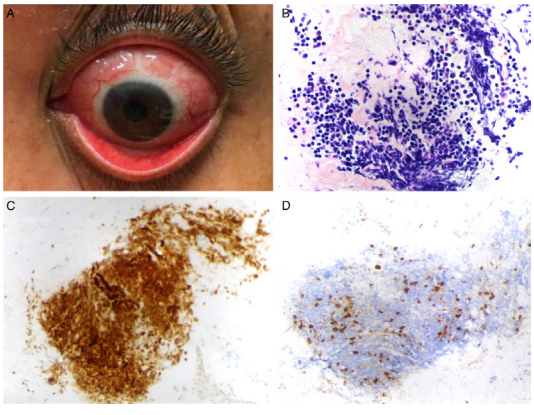


**Figure 1** (A) Salmon-pink patches involving most of the conjunctiva. (B) Infiltrations of small- to medium-sized cells (H&E, 500×) on conjunctival biopsy. (C) Lymphoma cells expressing PAX5 (IHC, 100×). (D) Lymphoma cells lacking CD3 (IHC, 100×).

## Learning points

- Bilateral eye redness is not always conjunctivitis; salmon-coloured patches should raise suspicion for conjunctival lymphoma.
- Clinical features of conjunctival lymphoma are non-specific and the lesions can appear nearly identical to other more benign processes, so it is crucial to obtain biopsies and further studies.
- Disease localised to the ocular adnexa can be treated with external beam radiation therapy, but patients with systemic disease may need chemotherapy.

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