Case Report

Conjunctival Involvement of T-Cell Lymphoma in a Patient with Mycosis Fungoides

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Background. Ocular involvement in mycosis fungoides (MF) cases occurs in one-third of patients with the eyelid being the most frequent site affected; however, conjunctival involvement is rarely reported. Herein, we report a rare case of conjunctival involvement of MF. Case Presentation. A 66-year-old man who was previously diagnosed with MF in 2010 and was treated with total skin electron beam irradiation as well as boost treatments to numerous areas presented to the clinic in 2014 complaining of foreign body sensation and redness in both eyes. Slit lamp examination of both eyes showed erythematous conjunctival growth that extended circumferentially. Physical examination revealed erythematous skin lesions on different body parts. Conjunctival biopsy was performed and revealed a dense, highly polymorphic lymphocytic population. The immunophenotype demonstrated a neoplastic T-cell origin consistent with MF. A diagnosis of conjunctival involvement by MF was made. The conjunctiva was treated with radiotherapy resulting in tumor regression. There were no recurrences at the 6-month follow-up. Conclusion. T-cell lymphoma should be considered in patients with a history of MF presenting with conjunctival and skin lesions.

1. Background

Lymphoproliferative tumors of the eye and ocular adnexa are usually non-Hodgkin’s B-cell lymphomas with the marginal zone type (mucosa-associated lymphoid tissue (MALT)) being the most common one [1, 2]. T-cell lymphoma of the ocular adnexa is rare, and when it occurs it is usually a manifestation of systemic T-cell lymphoma or represents a progression of mycosis fungoides (MF) on the skin [2]. However, only about 2% of people with cutaneous T-cell lymphoma have ophthalmic manifestations with the eyelid being the most common site to be affected [3]. To the best of our knowledge, T-cell lymphoma of the conjunctiva in a patient with MF has only been reported four times in the English literature (Table I) [4–6]. We hereby report a rare case of conjunctival involvement of T-cell lymphoma in a patient with MF. Moreover, this is the first reported case with a comprehensive immunohistochemical analysis.

2. Case Presentation

A 66-year-old man with a known history of hypertension, hypercholesterolemia, and MF (without extracutaneous manifestations) that was diagnosed in 2010 and treated with total skin electron beam irradiation as well as boost treatments to numerous areas presented to the clinic in 2014 with foreign body sensation and redness in both eyes that had persisted for few months. This foreign body sensation was more in the right eye than in the left eye and was not associated with any tearing or decreased vision. Visual acuity at presentation was 20/40 in both eyes. Slit lamp examination of the right eye showed rubbery erythematous growth of the conjunctiva extending circumferentially around the globe; however, it was more prominent in the inferior part of the conjunctiva (Figures I(a) and I(b)). The cornea was not involved. Examination of the left eye showed similar findings but with lesser degree. At this stage, a clinical diagnosis of
Table 1: Cases of T-cell lymphoma of the conjunctiva in patients with mycosis fungoides in the English literature.

<table>
<thead>
<tr>
<th>Author</th>
<th>Publication year</th>
<th>Gender</th>
<th>Age</th>
<th>Ocular structures involved</th>
<th>Method of treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fradkin et al. [7]</td>
<td>1969</td>
<td>F</td>
<td>42</td>
<td>Left caruncle</td>
<td>Radiotherapy with strontium-90</td>
<td>Not reported</td>
</tr>
<tr>
<td>O’Day et al. [5]</td>
<td>1985</td>
<td>F</td>
<td>40</td>
<td>Conjunctiva</td>
<td>Radiotherapy</td>
<td>Not reported</td>
</tr>
<tr>
<td>Rubegni et al. [6]</td>
<td>1997</td>
<td>M</td>
<td>34</td>
<td>Conjunctiva</td>
<td>Surgery with low dose rINF-α2α plus etretinate</td>
<td>No relapse after 1-year follow-up</td>
</tr>
<tr>
<td>Present report</td>
<td>2015</td>
<td>M</td>
<td>66</td>
<td>Conjunctiva</td>
<td>Radiotherapy</td>
<td>No relapse up to 6 months of follow-up</td>
</tr>
</tbody>
</table>

F: female; M: male.

B-cell lymphoma (MALT type) involving the conjunctiva was made; however, the patient gave a history of several similar skin lesions to the ones he had in 2010 that started to appear around the same time of the eye complaint. Physical examination of the body revealed erythematous skin lesions mainly on the back, between the buttocks, and left medial superior thigh. A progression of the disease to involve the conjunctiva was then suspected and a conjunctival biopsy was performed. Microscopically, the conjunctival specimen showed an infiltration of neoplastic lymphocytes; some of them were large cells with large and irregular nuclei. Overall, the neoplastic population was pleomorphic and included cells with small, medium, and large cribiform nuclei as well as blast cells with prominent nuclei and mitotic figures. The immunohistochemical panel (Figures 2(a)–2(f)) was positive for T-cell markers, including CD2, CD3, CD4, and CD5. Conversely, the tumor cells did not express CD7 or CD8 indicating the malignant nature of the cells and favoring the diagnosis of extracutaneous MF. CD20, CD10, and CD23 (B-cell markers), anaplastic lymphoma kinase (ALK), and CD30 were all negative. Ki 67 was positive in 20% of the neoplastic cells. A diagnosis of conjunctival involvement by mycosis fungoides was subsequently made. Magnetic resonance imaging of the brain showed thickening of the anterior periocular tissue in both eyes without orbital extension. The patient received low dose radiotherapy of 4 grays (Gy) in two fractions and showed significant tumor regression with good tolerability. After 6 months of follow-up, no tumor recurrence was noted. All data accumulation was in accordance with Canada and Province of Quebec legislation and the tenets of the Declaration of Helsinki.

3. Discussion

MF is the most common type of primary cutaneous T-cell lymphoma and is usually defined as peripheral epidermotropic cutaneous T-cell lymphoma with small and medium-sized infiltrating T-cells with cerebriform nuclei [8]. The clinical picture of the disease follows a specific stepwise pattern from patches to plaques and mushroom-shaped tumors [9]. Visceral and lymph node involvement is seen at late stages of the disease and usually involves the lungs, spleen, and liver [10, 11]. Ocular involvement can be found in up to one-third of cases with the eyelid being the most common involved site [3, 4, 12, 13]. The most prevalent ocular findings in MF cases are eyelid tumor, cicatrical eyelid ectropion, eyelid thickening, blepharitis, and corneal abnormalities [3]. However, conjunctival involvement is extremely

Figure 1: Slit lamp images of the right eye. (a) Erythematous flesh-like tumor affecting most of the conjunctiva, which is more prominent in the inferior location. (b) After treatment, there was significant lesion regression. Note the scar at the biopsy site (arrow).
Figure 2: Histopathological analysis of the biopsy. (a) The conjunctiva was extensively infiltrated by atypical lymphocytes with epidermotropism (arrowhead; Hematoxylin and Eosin [H and E], 200x). (b) At higher magnification, the tumor showed atypical small to medium sized lymphocytes with irregular nuclei and clumped chromatin (arrow). Macrophages and reactive lymphocytes were also seen (H and E, 400x). (c) The neoplastic cells were positive for CD3 (200x). (d) Decreased coexpression by the neoplastic lymphocytes was noticed (100x). (e) CD20 was only expressed by the scattered reactive B-cells (100x). (f) Proliferative index estimated by nuclear positivity for KI-67 antigen was approximately 20% (400x).
using topical steroids and radiotherapy with a good initial response, but the patient died from complications of the skin tumors [4]. In 1985, O’Day et al. [5] reported treating a patient’s skin lesions with psoralen long-wave ultraviolet (PUVA), but since she was wearing UV blocking glasses during treatment she developed a relapse in an area of the conjunctiva that was not exposed to PUVA, the so-called sanctuary site. This relapse site was then treated by superficial radiotherapy. Rubegni et al. reported surgical excision with low doses of recombinant interferon-α (INF-α) plus etretinate as their treatment of choice in order to avoid cataract formation associated with local radiotherapy [6]. In the present case, our patient was treated by superficial radiation with no documented cataract formation. Moreover, there was no relapse of the tumor after 6 months of follow-up.

4. Conclusion

In conclusion, herein we report a rare case of conjunctival involvement in a patient diagnosed with MF. Full history and physical examination are key factors for the correct differential diagnosis. Although B-cell lymphoma is more common, T-cell lymphoma should be considered in patients presenting with a conjunctival lesion and a history of MF. Immunohistochemistry is a crucial tool for proper diagnosis and classification of lymphoproliferative diseases of the eye and ocular adnexa.

Abbreviations

MF: Mycosis fungoides
MALT: Mucosa-associated lymphoid tissue
PUVA: Psoralen long-wave ultraviolet
Gy: Gray.

Conflict of Interests

The authors declare that there are no competing interests.

Authors’ Contribution

Pablo Zoroquiain, Sarah A. Alghamdi, and Miguel N. Burnier Jr. were the pathologists that performed the histopathological evaluation. Sonia Callejo was the ophthalmologist and the attending physician responsible for providing all the clinical information and clinical pictures. All authors participated in the design of the paper. Sultan S. Aldrees drafted the paper while Sultan S. Aldrees, Pablo Zoroquiain, Patrick T. Logan, Sarah A. Alghamdi, Sonia Callejo, and Miguel N. Burnier Jr. did the final revision of the paper. All authors read and approved the final paper.

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References


