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Ocular Surface Masquerades

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Ocular surface "Masquerade Syndrome" was first described by Theodore and later by Irvine as chronic blepharoconjunctivitis due to an underlying conjunctival malignancy, such as conjunctival intraepithelial neoplasia and sebaceous cell carcinoma as they engage in intraepithelial (pagetoid) spread. They can even produce chronic cicatrizing conjunctivitis mimicking cicatricial pemphigoid. The lesions have no distinct borders and therefore may be clinically indistinguishable from the uninvolved adjacent tissue. Additionally, conjunctival lymphoma may not present with the classic, salmon patch subepithelial nodular infiltration, but may rather occur diffusely, causing persistent conjunctival inflammation and cicatrizing conjunctivitis, masquerading as chronic conjunctivitis or even scleritis unresponsive to steroid or immunosuppressive therapy.

We have described six patients initially diagnosed with chronic conjunctivitis or blepharoconjunctivitis who were later found to have undiagnosed conjunctival malignancy masquerading as chronic conjunctival inflammation. One patient had intraepithelial epithelioma, one had invasive squamous cell carcinoma, two had conjunctival lymphoma, and two had sebaceous carcinoma.

There are also case reports and case series in the ophthalmology literature describing ocular surface squamous neoplasia (OSSN) misdiagnosed as chronic scleritis until a conjunctival and scleral biopsy confirmed the diagnosis later in the course of disease.

The 'take-home message' of these cases—all of which had been managed by multiple experienced ophthalmologists—is that one should maintain a high index of suspicion for underlying malignancy in any patient with unusual or treatment-resistant conjunctival inflammation.

Early conjunctival biopsy is the most important step in avoiding misdiagnosis; however, careful handling and processing of the specimen and evaluation by an experienced pathologist aware of the possibility of malignancy are essential. Interestingly, two of our cases were initially misdiagnosed histopathologically by the ophthalmic pathologist who first evaluated the specimens; however, the same pathologist subsequently identified malignant features when appropriately alerted to the distinct possibility of malignancy. Additionally, close follow-up of these patients is essential for detecting local recurrence and systemic involvement, given that these malignancies frequently recur, often necessitate multiple surgeries, and may be associated with systemic metastasis.

References

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