Ocular Surface Masquerades

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"Masquerade syndrome" was first described by Theodore and later by Irvine as chronic blepharoconjunctivitis due to an underlying conjunctival malignancy. Conjunctival intraepithelial epithelioma and sebaceous cell carcinoma can mimic chronic blepharoconjunctivitis as they engage in intraepithelial (pagetoid) spread, and 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 who were initially diagnosed with chronic conjunctivitis or blepharoconjunctivitis who had conjunctival malignancies which were undiagnosed and which were masquerading as chronic conjunctival inflammation. One patient had their problem as a consequence of intraepithelial epithelioma, one had the problem as a consequence of invasive squamous cell carcinoma. 2 had conjunctival lymphoma, and 2 had sebaceous carcinoma. The "take home message" of these cases, all of which had been cared for by multiple excellent ophthalmologists, is that one should have a very high index of suspicion for possible underlying malignancy in any patient with unusual, of treatmentresistant conjunctival inflammation. Biopsy, relatively early in the course of the patient's problem, is the most important step in avoiding misdiagnosis, but we would also emphasize that the specimens should be handled and processed very carefully, and, further, should be analyzed by an experienced pathologist who has been alerted to the possibility of malignancy; two of our cases had been misdiagnosed histopathologically by the ophthalmic pathologist who had initially read the specimens, but the same pathologist picked up the malignant features of the biopsy when subsequently appropriately alerted to the distinct possibility a malignant problem. Close follow-up with patients is obviously critical to detect local recurrences and systemic involvement, since these malignancy often recur, require multiple surgeries. and may even be associated with systemic metastases.