

Ocular Cicatricial Pemphigoid: Understanding Diagnosis and Management in Ophthalmological Practice

Shadi Gociman, MD¹ | Abram Beshay, MD² | Morgan M. Miller, PhD³ | Rachel Coddan, MPH³ | Thomas Oberg, MD⁴ | Christopher M. Hull, MD¹ | John J. Zone, MD¹ | Jamie L.W. Rhoads, MD, MS¹

¹Department of Dermatology, University of Utah School of Medicine, Salt Lake City, UT | ²Kaiser Permanente, Southern California Medical Group | ³Division of Epidemiology, Department of Internal Medicine, University of Utah School of Medicine, Salt Lake City, UT | ⁴Still Canyon Eyelid and Facial Plastic Surgery, Salt Lake City, UT

Background

- Ocular cicatricial pemphigoid (OCP) is a rare, chronic, inflammatory, form of mucous membrane pemphigoid that affects the ocular mucosa and clinically presents as a chronic cicatrizing conjunctivitis¹
- Early detection and treatment of OCP are necessary to prevent long term complications such as permanent blindness¹
- There are limited data available on the preferred treatment options by clinicians for this condition
- The objective of this survey study was to understand the awareness, diagnosis, and management of OCP in local ophthalmological practices

Methods

- A 23-question survey was distributed in person to board certified/eligible practicing ophthalmologists present at the 2023 Utah Ophthalmological Society Annual Meeting via a survey link
- Survey data were collected and managed using REDCap electronic data capture tool hosted at the University of Utah^{2,3}
- REDCap (Research Electronic Data Capture) is a secure, web-based software platform designed to support data capture for research studies^{2,3}

Results

- 44 participants completed the survey (response rate 89.8%)
- Only 46% were comfortable in diagnosing patients with OCP
- Most (62%) had not diagnosed a patient with OCP in the past year
- Most (63%) had not performed a conjunctival biopsy in the past year to evaluate for OCP
- When disease was clinically suspected, 80% opted to refer to a provider specialized in the care of patients with OCP as next step rather than performing a biopsy or indirect immunofluorescence
- When asked the stage of majority of their patients with OCP at first presentation, 37% responded that none of their patients have OCP, 19% responded stage 1, 19% responded stage 2, 26% responded stage 3, 0% responded stage 4
- Most (59%) were uncomfortable managing the disease
- Regarding next steps in management once OCP diagnosed, 18% chose to administer topical or systemic therapy, 59% would refer to an ophthalmologist specialized in the care of OCP, 9% opted to refer to a dermatologist specialized in this area, and 7% would refer to a rheumatologist.
- Most (92%) had not administered systemic therapy for OCP in the past year
- Most respondents (75%) did not prefer a particular systemic therapy
- Free text comments emphasized the rarity of the condition, lack of clarity on biopsy protocols and who to refer to, as well as degree of difficulty in findings specialists to manage systemic immunosuppression

Conclusions

- In general, respondents were not comfortable diagnosing or managing OCP and prefer to refer patient out when disease is suspected
- The results highlight the importance of multidisciplinary coordination of care across specialties as well as the need to streamline the referral process for further medical management once diagnosis has been made
- Practice variation by region and respondent subspecialty limits generalizability of study data

Table 1. Respondent demographics and practice characteristics

Characteristics	n (%) unless otherwise specified
Age	
Average (min, max)	46 (32, 85)
Gender	
Female	7 (17%)
Male	35 (83%)
Year residency completed	
Average (min, max)	2008 (1961, 2022)
Practice setting	
Academic	8 (19%)
Solo private practice	8 (19%)
Single Specialty Group with 2-3 providers	14 (33%)
Single Specialty Group with >4 providers	7 (17%)
Multispecialty Group	5 (12%)
Primary specialty	
General	25 (61%)
Cornea	8 (20%)
Retina	4 (10%)
Glaucoma	1 (2%)
Oculoplastics	2 (5%)
Neuro-ophthalmology	1 (2%)
Practice volume	
Average (min, max)	101 (10, 200)

References

- Kirzhner et al. Ocular cicatricial pemphigoid a review of clinical features, immunopathology, differential diagnosis, and current management. *Semin Ophthalmol.* 2011 Jul-Sep;26(4-5):270-7.
- Harris et al. A metadata-driven methodologic and workflow process for providing translational research informatics support. *J Biomed Inform.* 2009 Apr;42(2): 377-81.
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