

Ocular Cicatricial Pemphigoid in a Young Filipino Male: The First Biopsy-Proven Case Report from the Philippines

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ABSTRACT

Objective: To describe the first biopsy-proven case of ocular cicatricial pemphigoid (OCP) in a young, Filipino male – an atypical presentation for this disease.

Methods: This is a case report.

Case Presentation: A 34-year-old healthy Filipino male presented with a 2-year history of intermittent eye redness, foreign body sensation, and tearing. Slit lamp examination revealed symblepharon formation and fornix shortening of both eyes. Excision of symblepharon with ocular surface reconstruction using amniotic membrane graft was performed, and the conjunctival specimen was sent for immunohistochemical analysis. Direct immunofluorescence was performed which confirmed strong linear deposits of IgG and fibrinogen along the basement membrane zone, confirming diagnosis of OCP. Early diagnosis and prompt treatment is even more important in the younger population as OCP tends to progress more rapidly, and is associated with more severe manifestations. If left untreated, OCP may progress to symblepharon or, in severe cases, ankyloblepharon formation.

Conclusion: OCP is a rare, sight-threatening, chronic, autoimmune disease that involves inflammation and cicatrization of the conjunctiva, which typically affects older females and is rarely seen in younger populations. This case report highlights the need for increased awareness among clinicians in the Philippines to consider OCP as a differential diagnosis for chronic conjunctivitis, even in patients outside the typical demographic. Early recognition and confirmatory biopsy are essential to prevent progressive ocular surface scarring and vision-threatening complications.

Keywords: ocular cicatricial pemphigoid, symblepharon, young, male, Philippines

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INTRODUCTION

Improper mucous membrane pemphigoid (MMP) is a rare, chronic, autoimmune, subepithelial blistering disease where lesions of the mucous membranes heal with scarring. Ocular cicatricial pemphigoid (OCP) is a subtype of MMP which presents clinically as chronic cicatrizing conjunctivitis.¹ Direct immunofluorescence (DIF) staining of the actively involved conjunctival area remains to be the gold standard in diagnosing OCP.² The characteristic pathologic feature of MMP are deposits of IgG, IgM or C3 in the epithelial basement membrane zone.

OCP is a rare disease with an incidence of 1 per 50,000 individuals.¹ It usually affects the elderly population, with a female predominance (2:1 ratio).³ Although uncommon, there are only a few documented cases of OCP in the young population.⁴ In the Philippines, OCP is likely underrecognized and underreported because of its rarity, nonspecific early manifestations, and overlap with more common causes of chronic conjunctivitis. In addition, access to specialized diagnostic modalities such as direct immunofluorescence (DIF) may be limited in some institutions, potentially contributing to delayed diagnosis and underreporting.⁷ To our knowledge, no previously published biopsy-confirmed cases of OCP in a young Filipino male have been reported following a literature search of PubMed, HERDIN, Acta Medica Philippina, and Google Scholar. Available local literature on OCP predominantly describes older patients, including a previously reported biopsy-confirmed case in an elderly Filipino male.⁷

CASE PRESENTATION

A 34-year-old male with no known comorbidities presented with a 2-year history of intermittent eye redness and foreign body sensation associated with tearing of both eyes. During the 2-year period, the patient did not seek any consult and self-medicated with over-the-counter lubricant eye drops. He denied any previous history of chemical injury to the eye, prior ophthalmologic surgical procedures, and other ocular medication use. During the interim, the patient noted on-and-off appearance of a solitary erythematous patch on the

right wrist, and occasional appearance of painful erosions on the oral and nasal mucosa which would spontaneously heal. The patient denied manifestations like formation of cutaneous vesicles, dysphagia, ocular pain, and blurring of vision. Persistence of the ocular symptoms prompted consultation.

Ophthalmologic examination of both eyes showed best corrected visual acuity (BCVA) of 20/32. The slit lamp examination of both eyes showed normal eyelid position with upper eyelid distichiasis with symblepharon formation involving the upper and lower lids from lateral canthus when the right eye was in the adducted position (Figure 1). The conjunctiva was hyperemic with few papillae on the palpebral with subepithelial fibrosis. There is circumferential conjunctival keratinization starting 4 mm from the limbus extending in a centripetal manner, and shortening of the inferior fornix to 4.6 mm in the right eye and 4.8 mm in the left (Modino's Classification Stage 3). There was also noted symblepharon formation involving the lower lid from the lateral canthus of the left eye. The cornea, anterior chamber and lens findings and posterior segment of both eyes were unremarkable.

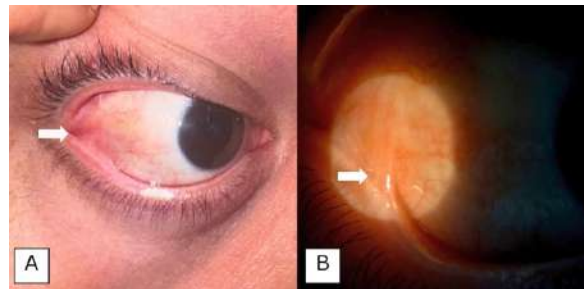


Figure 1. Preoperative photo that shows diffuse, 10x magnification (A) and close up, 16x magnification (B) view of symblepharon formation (white arrow) of the right eye involving the temporal upper and lower lid (white arrow).

The nonspecific nature of the ocular symptoms, coupled with a broad differential diagnosis and the patient's desire for cosmetic improvement, prompted the decision to proceed with ocular surface reconstruction. The patient underwent excision of the temporal symblepharon of the right eye with ocular surface reconstruction using fresh amniotic membrane graft. The excised conjunctival tissue from the symblepharon was sent for hematoxylin and eosin (H&E) and direct DIF evaluation due to it representing clinically affected tissue with ongoing cicatrizing inflammation. Punch

biopsies for H&E and DIF were also done on the lesion of the right upper arm and the left buccal mucosa which revealed nonspecific findings as staining did not match any classic diagnostic immunobullous disease pattern, including pemphigoid. Buccal mucosa biopsy is performed even in purely ocular cases of OCP, as it is an easily accessible mucosal site with a relatively high diagnostic yield for direct immunofluorescence, even in the absence of overt oral lesions.⁹ Cutaneous biopsies, may have a lower diagnostic yield since OCP is a predominantly a mucosal disease, but may be done in addition to the mucosal biopsy to increase overall diagnostic yield.⁸

DIF staining of the conjunctival tissue showed strong linear deposits of IgG and fibrinogen on the basement membrane zone of the conjunctival tissue (Figure 2). H&E staining of the conjunctival mucosa showed non-specific findings. DIF confirmed the diagnosis of ocular cicatricial pemphigoid.

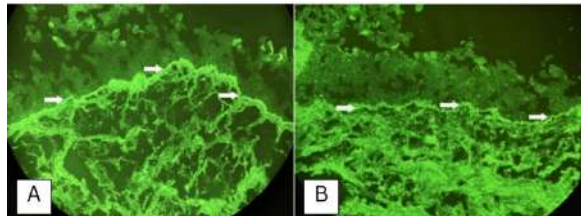


Figure 2. Direct immunofluorescence of conjunctival tissue showing +1 linear deposition (white arrows) of (A) IgG, (B) fibrinogen in the basement membrane zone

One week after the procedure, there was deepening of the inferior fornix and the globe was noted to be more freely mobile with the amnion graft well opposed to the bulbar conjunctiva (Figure 3). Tobramycin plus dexamethasone ophthalmic ointment and preservative-free lubricating eye drops were continued. The left eye was started on topical prednisolone acetate 1% eye drops as well. The patient was referred to internal medicine for clearance prior to oral steroid then subsequent immunosuppressive agent use.

DISCUSSION

OCP is a rare disease involving females in the older age group. Its incidence increases with advancing age, with onset typically at age 50 years old.² In contrast, the patient in our case is a healthy young male in his 3rd decade of life. OCP is even

more uncommon in populations aged younger than 50 years old, with an estimated incidence of about less than 0.5 cases per 1 million individuals in this age group.² The female to male ratio ranges from 2:1 to 4:1.3 To our knowledge, this is the first documented case of a biopsy-confirmed OCP in the younger male population in the Philippines.



Figure 3. Diffuse illumination, 10x magnification, of patient's right eye 1 week after the excision of the symblepharon of the right eye.

OCP is a type II hypersensitivity reaction involving autoantibodies against the surface antigens of the basement membranes. Involvement of other mucosal surfaces may also occur in OCP.¹ If left untreated, OCP progresses to subepithelial fibrosis, shortening of the fornices, symblepharon formation, and eventually ankyloblepharon. Ocular symptoms begin as ocular redness, tearing, and burning or foreign body sensation.⁵ Hence, making the diagnosis of OCP in its initial stage is particularly challenging as it is nonspecific and may mimic other ocular disease such as chronic conjunctivitis and dry eye disease. In this case, the patient initially had nonspecific ocular symptoms, leading to a delay in seeking appropriate medical consultation. Given the nonspecific symptoms, and the physical examination of the patient, other causes of cicatrizing conditions were considered, including Steven-Johnson syndrome, drug-induced pseudopemphigoid, and even a history of a previous chemical burn. However, the lack of prior mucocutaneous illness, history of chemical burn/trauma, and no current use of topical medications made these differentials less likely.

Consequently, by the time the patient decided to seek consult, OCP had progressed to its more advanced stages. Inferior fornix shortening with symblepharon formation was noted, corresponding

to a Foster classification Stage 3 (out of 4). Compared with the few published similar cases of OCP in the young, this case presents with similar signs and symptoms. According to Jinagal et al, most cases presented with a similar physical exam with diffuse conjunctival congestion and subconjunctival fibrosis with or without shortening of the fornices. Similar to our patient, past cases had an initial presentation of Foster Classification Stage 3 or higher.² However, similar to the typical older demographic, the symptomatic presentation of OCP in the young is usually nonspecific.

This makes diagnosis and management of OCP somewhat challenging, especially in the young. As the disease is reported to be more severe and progresses more rapidly in individuals aged younger than 60 years old, it is paramount to identify OCP in its earlier stages.² If left untreated, OCP can progress to ankyloblepharon and corneal scarring, leading to blindness. Management of OCP mainly involves controlling the inflammatory aspect to halt disease progression. Systemic immunosuppression is the mainstay of treatment⁷, as it stops the progression of cicatrization in patients.¹ Oftentimes, induction therapy of steroids followed by steroid-sparing agents is required. However treatment of OCP is relatively more difficult as the presentation of the disease in the young population is usually severe and resistant to medical therapy. There is also a potential for systemic toxicity brought about by long term use of steroids or immunosuppressive agents. Alejab et al details different adverse drug reactions to prolonged steroid use in the young population including weight gain, growth retardation and Cushingoid features.⁶

OCP is a rare, sight-threatening, autoimmune disorder that is often undiagnosed or diagnosed late due to its nonspecific early ocular symptoms. This case highlights the importance of considering OCP in the differential diagnosis of chronic conjunctival inflammation, even in atypical presentations, and contributes valuable local data that may improve early clinical recognition, timely diagnosis and management.

CONCLUSION

OCP may present initially with nonspecific ocular symptoms and should remain a diagnostic consideration in patients with chronic conjunctivitis,

dry eye symptoms, or unexplained conjunctival scarring. This case highlights the importance of maintaining a high index of suspicion for OCP even in the atypical demographic of younger male patients, particularly in the Philippine setting, where underrecognition and underreporting may contribute to delays in diagnosis and treatment.

ETHICS COMPLIANCE STATEMENT

The authors affirm that this case report was prepared in accordance with the ethical standards of the *Philippine Journal of Ophthalmology*, the principles outlined in the Declaration of Helsinki, and applicable institutional and national guidelines on research involving human participants.

Written informed consent for publication of clinical details and accompanying images were obtained from the patient prior to manuscript submission. The authors confirm that all identifying information has been removed or anonymized to protect patient privacy. The authors declare that the case report did not require formal institutional review board (IRB) approval, as per the policies of the authors' institution, because it describes a single clinical case without experimental intervention. Should institutional policies require review, appropriate documentation has been secured and is available upon request.

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