

Ocular Cicatricial Pemphigoid in a Healthy Elderly Male Filipino Patient

Jose Christopher C. Tesorero III, MD,¹ George Michael N. Sosuan, MD¹ and Ruben Lim Bon Siong, MD^{1,2}

¹Department of Ophthalmology and Visual Sciences, Philippine General Hospital, University of the Philippines Manila, Manila, Philippines

²Eye Institute, St. Luke's Medical Center, Quezon City, Philippines

ABSTRACT

Ocular cicatricial pemphigoid (OCP) is a chronic bilateral, blinding, cicatrizing form of conjunctivitis with relapsing and remitting periods. It has strong evidence for an immune type II hypersensitivity that leads to subconjunctival fibrosis and extensive systemic bullae formation. To the best knowledge of the authors, this is the first reported case of direct immunofluorescence (DIF) assay-proven OCP in an elderly Filipino man.

A 68-year-old male presented with bilateral corneal conjunctivalization, symblepharon, ectropion, conjunctival hyperemia testing positive with conjunctival biopsy for basement membrane antibodies with DIF for the left eye, while turning out negative for the right eye. He was managed as a case of OCP, both eyes, and was given topical steroids and antibiotics. Oral Dapsone was started by Dermatology and Rheumatology Services.

OCP is a rare autoimmune and blinding disease. Early diagnosis and prompt treatment are vital as ocular complications permanently affect the quality of life of patients as seen in our patient. DIF assay remains the gold-standard for diagnosis. Systemic immunosuppression is the mainstay of treatment. Adjunctive supportive topical medication may be given to alleviate ocular discomfort. A multidisciplinary approach is essential to provide holistic care to each patient.

Keywords: ocular cicatricial pemphigoid, Filipino, direct immunofluorescence assay

INTRODUCTION

Ocular cicatricial pemphigoid (OCP) is characterized by a chronic bilateral non-infectious, cicatrizing form of conjunctivitis with relapsing and remitting periods.¹ Strong evidences exist for an autoimmune type II hypersensitivity involving autoantibodies against the integrin and laminin contents located in the hemidesmosome-epithelial membrane complex of the conjunctival squamous epithelium eventually leading to separation of the epithelium from the basement membrane with subsequent bullae formation. The migration of inflammatory cells into the substantia propria typically presents as subconjunctival fibrosis with or without inflammation. In early and acute phases, eosinophils and neutrophils mediate the inflammation; however, lymphocytes have been largely observed in chronic cases.²

OCP is a rare form of potentially blinding cicatrizing conjunctivitis among Filipinos and should not be missed among patients presenting with chronic bilateral conjunctivitis. Written informed consent to publication of information was obtained from the patient. To the best knowledge of the authors, this is the first reported case of ocular cicatricial pemphigoid in a healthy elderly Filipino man confirmed by direct immunofluorescence (DIF) assay. The authors aimed to report this potentially blinding disease and review



eISSN 2094-9278 (Online)
Published: November 28, 2025
<https://doi.org/10.47895/amp.vi0.12049>
Copyright: The Author(s) 2025

Corresponding author: George Michael N. Sosuan, MD
Department of Ophthalmology and Visual Sciences
Sentro Oftalmologico Jose Rizal
Philippine General Hospital
University of the Philippines Manila
Taft Avenue, Ermita, Manila 1000, Philippines
Email: gmsosuan@yahoo.com
ORCID: <https://orcid.org/0000-0002-1306-3727>

its management in the hope of better understanding of the disease process.

CASE PRESENTATION

A non-hypertensive, non-diabetic male patient in his late 60s consulted for progressive blurring of vision on the right eye. The blurring of vision initially presented one year prior to consult (PTC), and was associated with eye redness and progressive development of whitish opacity of the eye. He denied any history of eye trauma or surgery and any adverse reactions to medications. He also reported to have relapsing-remitting blistering lesions on the face, the left foot, and the oral mucosa (palate) after the onset of ocular symptoms. At this time, he consulted an ophthalmologist where epilation of inwardly misdirected eyelashes was done. He was also given unrecalled topical medications which provided no relief.

Six months PTC, he developed the same symptoms on the left eye, but with progression of blurring of vision on the right eye. He consulted an ophthalmologist with an unrecalled diagnosis. He underwent epilation of inwardly misdirected eyelashes bilaterally and was given topical lubricants which provided minimal relief.

A few weeks PTC, the blurring of vision on both eyes progressed now associated with difficulty opening both eyelids. Hence, he opted to consult our institution.

On examination, the visual acuity on the right eye was best corrected to counting fingers at 1 foot and on the left eye was best corrected to 20/20. On further examination, the right eye showed complete obliteration of the inferior fornix with the symblepharon forming between the entire inferior bulbar and palpebral conjunctiva. There was also right lower eyelid entropion, madarosis, and conjunctivalization of the entire cornea with moderate conjunctival hyperemia (Figures 1A and B). The left eye showed a clear cornea with 360 degrees pannus

formation associated with moderate conjunctival hyperemia, lower eyelid entropion, and symblepharon formation on the inferior half of the fornix (Figures 2A and B). The right eye had limited extraocular muscle movement, while the left eye had full range of motion. Intraocular pressure for both eyes were normal. Fundus examination on the left eye was normal while the right eye had no view. Ocular ultrasound of both eyes was normal.

On systemic examination, the patient had associated erythematous blisters on his hard palate (Figure 3A), erythematous raised plaque seen on his upper back (Figure 3B), and excoriated skin lesion on his left foot (Figure 3C).

The patient was assessed to have ocular cicatricial pemphigoid, both eyes (right: stage 4; left: stage 3B). Preservative-free artificial tears every two hours were started on both eyes. Conjunctival biopsy for direct immunofluorescence (DIF) assay was done for both eyes to confirm the diagnosis. He was also co-managed with Dermatology and Rheumatology Services. Post-operatively, the patient was started on tobramycin-dexamethasone ointment three times a day and levofloxacin 0.5% eyedrops every three hours for both eyes.

Table 1 shows the summary of the results of the DIF assay. The right eye revealed a negative result (Figures 4A-

Table 1. Conjunctival Biopsy Results of the Direct Immunofluorescence Assay

Right eye		Left eye
IgA	Negative	(++) Strong linear deposition in the basement membrane zone
Fibrinogen		(+) Weak linear deposition in the basement membrane zone
IgG		
C3		
IgM		Negative

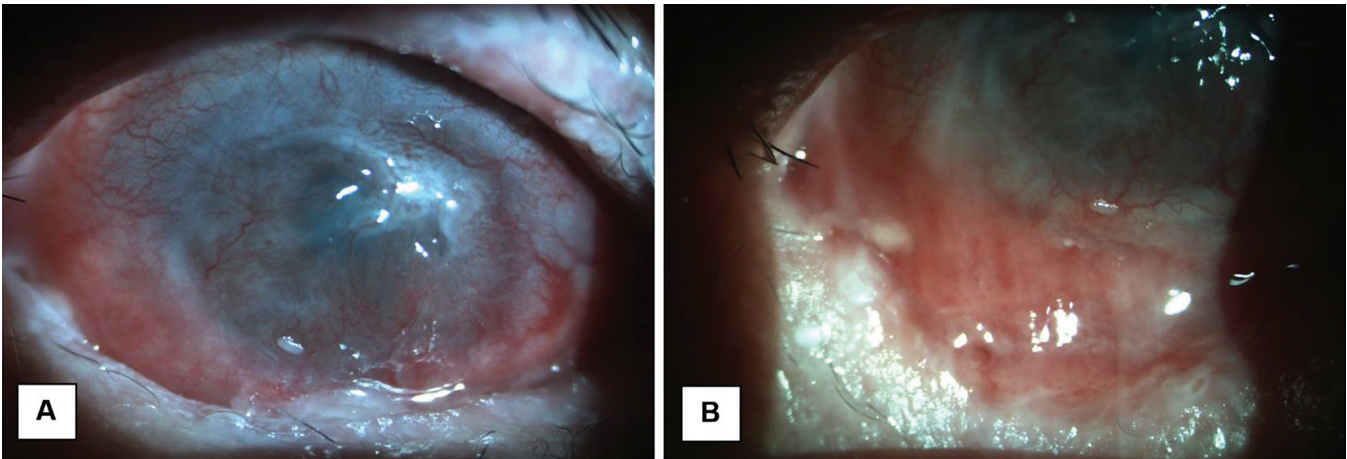


Figure 1. Slit lamp images of the right eye (A) showing conjunctivalization of entire corneal surface with moderate conjunctival hyperemia, and (B) complete obliteration of the inferior fornix with the symblepharon formation between the entire right inferior bulbar and palpebral conjunctiva.

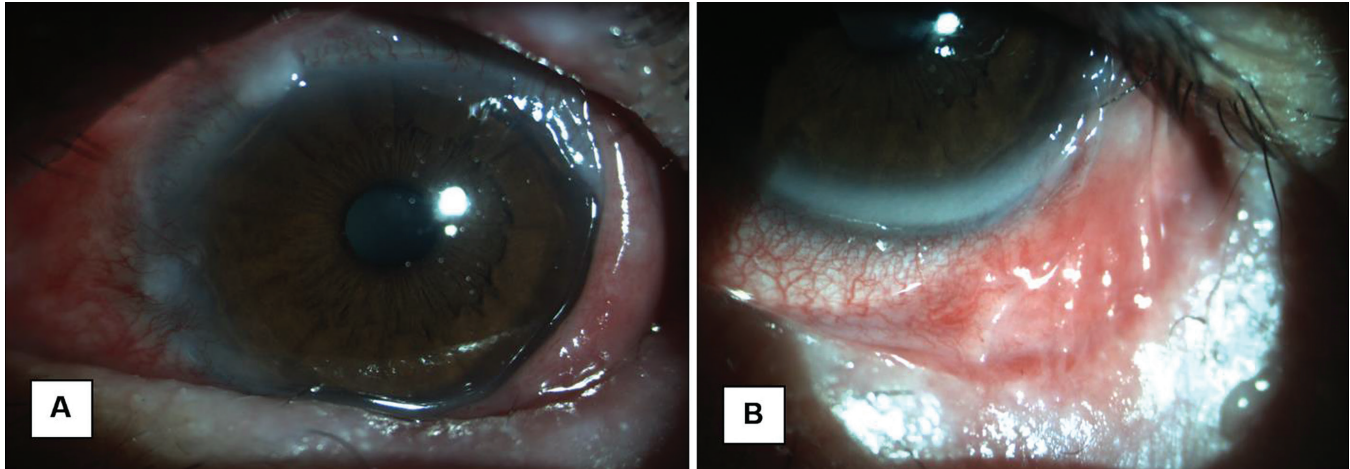


Figure 2. *Slit lamp images of the left eye (A) showing a clear cornea with 360 degrees pannus formation associated with moderate conjunctival hyperemia, and (B) left lower eyelid entropion and symblepharon formation on the inferior half of the fornix.*

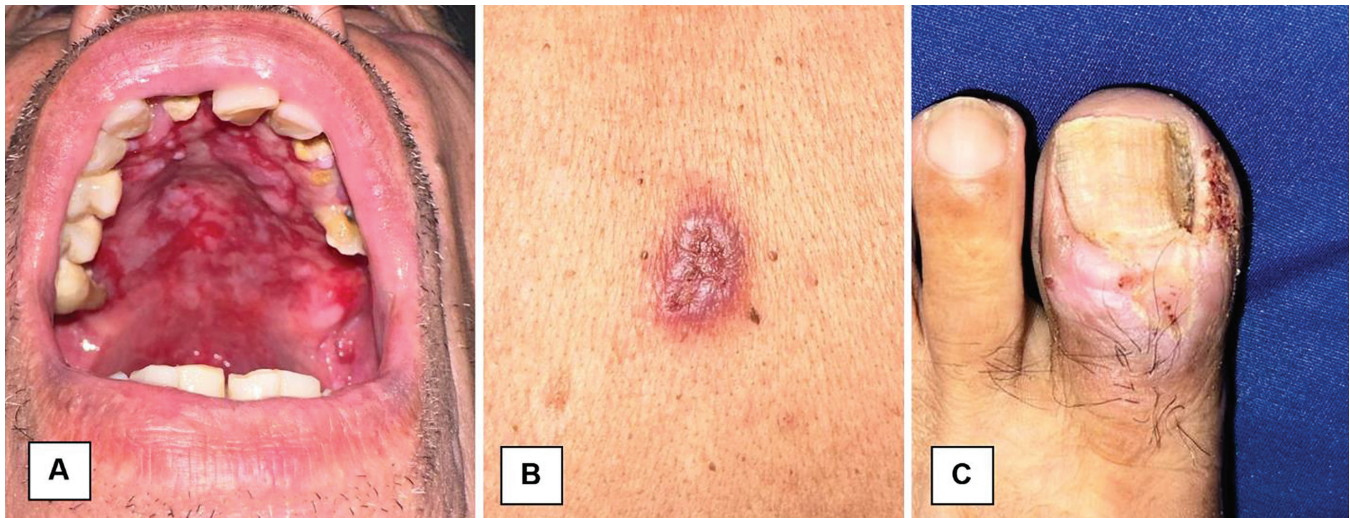


Figure 3. *Extraocular manifestations of OCP existing on the patient. (A) erythematous blisters on the hard palate, (B) erythematous, raised plaque seen on the upper back, and (C) excoriated skin lesions on the left foot.*

E), while the left eye showed positivity for IgA, C3, and fibrinogen (Figures 5A-E).

One important consideration in a patient presenting with chronic cicatrizing conjunctivitis is chronic ocular Steven-Johnson Syndrome (SJS); however, the patient did not present with history of adverse drug reactions to any medication prior to the start of his ocular symptoms. Ocular SJS also has more bulbar involvement of the eye as compared to him with more shortening of the conjunctival fornix. He did not have any history or exposure to trauma (chemical, thermal, radiation) thus making the chronic sequelae of ocular surface burn unlikely. Lastly, he tested positive for conjunctival anti-basement antibodies which is characteristic of OCP.

On post-operative day 3 and after seeing the DIF assay results, the patient was started on Dapsone 100 mg daily by

the Rheumatology service to control the systemic disease. Tobramycin-dexamethasone ointment and levofloxacin 0.5% eyedrops were continued.

On follow up after six months, vision for both eyes remained the same with no progressive worsening of ocular surface cicatrization and shallowing of the inferior fornix. He was maintained on Dapsone 100 mg and topical preservative-free artificial tears every two hours. The blistering lesions on the face, the left foot, and the oral mucosa (palate) have resolved. He was advised to follow-up every month to monitor disease activity. Long term plans include protecting the left eye by preventing progression of cicatrization and possible keratoprosthesis with ocular surface and fornical reconstruction for the right eye once disease activity is controlled.

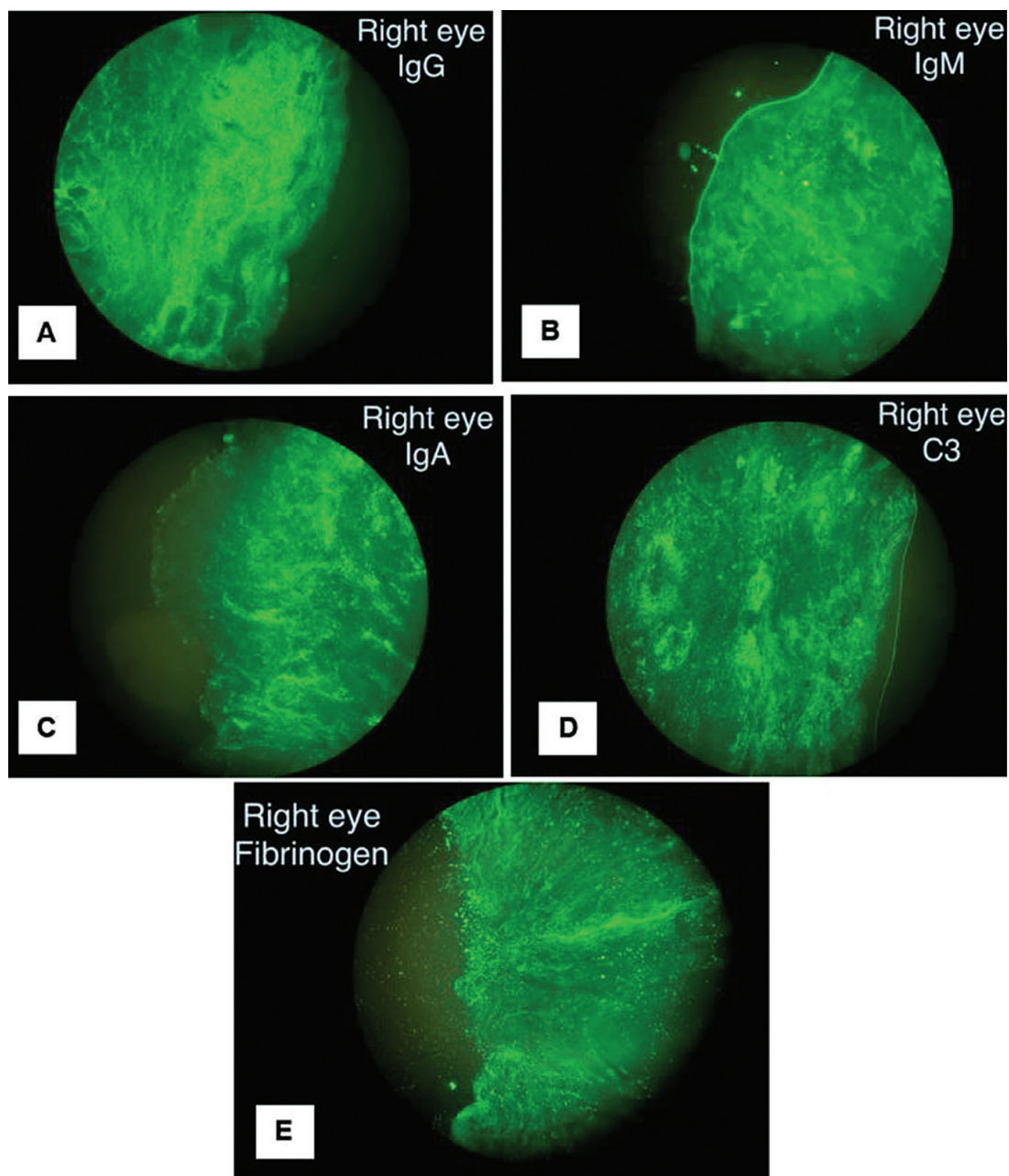


Figure 4. Direct immunofluorescence assay results of the right conjunctiva. Negative for IgG, IgM, IgA, C3, and fibrinogen.

DISCUSSION

Ocular cicatricial pemphigoid is a rare, chronic autoimmune blistering disorder which can affect the skin, the mucous membranes, or both. Ocular cicatricial pemphigoid and mucous membrane pemphigoid may be used interchangeably. For this case, the authors opted to use ocular cicatricial pemphigoid since ocular involvement per se carried a poorer prognosis. Other mucous membrane involvement may include the oral, nasal, genital, or anal mucosae.³

Ocular cicatricial pemphigoid is a type II autoimmune hypersensitivity response which affects 1 out of 10,000 to 50,000 people. It has no racial predilection and affects more females over males with a 2:1 ratio. Due to the rarity of the disease, worldwide prevalence studies are not yet available.⁴ OCP usually presents among the geriatric population with the median age over 65 years old.^{5,6} It is the leading cause of cicatrizing conjunctivitis in developed countries. A study on cicatrizing conjunctivitis in the United Kingdom (UK) estimated a prevalence of 0.8 per 1 million population, with

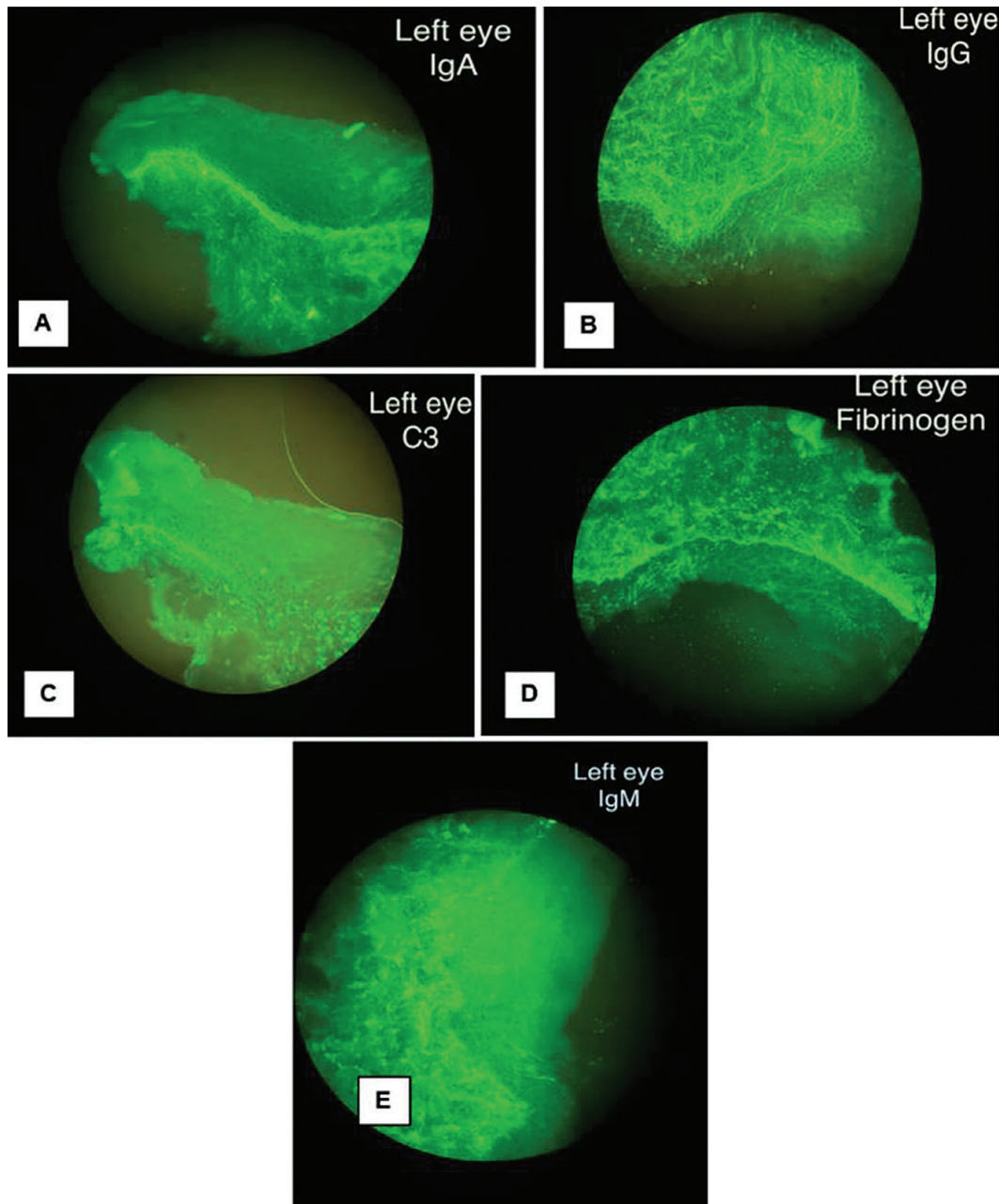


Figure 5. *Direct Immunofluorescence assay results of the left conjunctiva.* (A) IgA is strongly positive, exhibiting distinct deposition in the basement membrane; (B) IgG, (C) C3, and (D) fibrinogen are weakly positive, exhibiting fairly distinct deposition in the basement membrane; (E) IgM showing negative immunofluorescence.

OCP comprising 60% of the cases.⁵ Similar incidences have been found in France (1.13 per million) and in Germany (0.87 per million).⁷ From the census at the External Disease and Cornea Clinic of the Philippine General Hospital from the year 2016 to 2024, this was the only case of OCP diagnosed with DIF assay.

The clinical features of OCP include a history of chronic remitting and relapsing bilateral cicatrizing conjunctivitis

with an insidious onset. In a retrospective study done at Moorfield's Eye Hospital, only 50% with ocular symptoms presented with systemic manifestations. The most common presenting systemic manifestations are blisters in the oral cavity (44%) followed by the pharynx (30%), esophagus, (27%), the nose/sinus (18%), the skin (17%), the anus (5%), and the vagina (5%).⁶ In another study in the UK, only 52% presented with mucocutaneous involvement. The oral

mucosa (40%) is the most commonly involved, followed by skin at 18%.⁸

Ocular involvement in OCP can present as foreign body sensation, tearing, burning sensation, pruritus or redness, similar to other ocular surface inflammatory conditions in the early stages.⁹ However, this patient's presentation pointed towards a more severe and chronic form of this cicatrizing disease that involved difficulty opening the eyelids, entropion, symblepharon formation, and blurring of vision from conjunctivalization and scarring of the cornea associated with systemic findings on the mouth, skin, and left foot.

The Foster System is used for evaluating and grading the severity and disease progression of OCP. It is based on clinical signs and is divided into four stages. The first stage includes subconjunctival scarring and fibrosis, conjunctival inflammation, mucous discharge, and small patches of Rose Bengal staining of the conjunctival epithelium. The second stage is marked by shortening of the inferior conjunctival fornix. Within the second stage are four subcategories that describes the degree of fornix shortening as A, B, C, D corresponding to 0-25%, 25-50%, 50-75% and 75-100%, respectively. The third stage corresponds to symblepharon formation, further divided into percentage of horizontal involvement of symblepharon as A, B, C, D corresponding to 0-25%, 25-50%, 50-75% and greater than 75%, respectively. This stage also includes corneal neovascularization, keratopathy, generalized conjunctival shrinkage, trichiasis, distichiasis, and decreased tear production. The end stage or fourth stage of OCP is defined by keratinization of the ocular surface as well as formation of ankyloblepharon associated with severe Sicca Syndrome.^{10,11}

The right eye was at stage 4 which presented with ocular surface keratinization and neovascularization, symblepharon formation encompassing the entire inferior lid margin and obliteration of the inferior fornix. The left eye was at stage 3B which presented with inferior symblepharon formation encompassing 25-50% of the lateral inferior fornix, with associated limbal conjunctivalization and fibrosis.⁸

For definitive diagnosis of OCP, conjunctival biopsy with DIF assay is the gold standard. Positive results exhibit a linear deposition of IgG, IgA, IgM, complement 3 proteins in the epithelial basement membrane zone. A histologic analysis may show decreased goblet cells and an increased mast cell proliferation in the affected tissue.^{9,10} However, some patients may have clinical signs of ocular cicatricial pemphigoid but still result to a negative biopsy result. A negative biopsy results from concurrent quiescence of the disease, disease burnout, or total loss of conjunctival basement membrane.⁹ Other methods to detect basement membrane antibodies include indirect immunofluorescence and specific autoantibody detection.⁷ The DIF assay of the conjunctiva as listed in Table 1 showed negative staining for all reagents for the right eye. The negative result might be from loss of immune reactions due to the total loss of conjunctival

basement membrane in the later stages of the disease. On the other hand, the left eye was strongly positive for IgA, weakly positive for IgG, fibrinogen, and complement protein C3, and negative for IgM. This result supported that OCP may affect both eyes with marked asymmetry.

Medical therapy with immunosuppressive agents to decrease risk of progression is the mainstay of treatment. The patient's assessment should include systemic manifestations that may cause significant morbidity and mortality. In the end-stage "burned out" disease, treatment is not recommended due to the irreversible scarring and fibrosis at this stage.⁸

Patients with mild to moderate inflammation are usually treated by a stepwise approach with immunosuppressive therapy, initially with diaminodiphenylsulfone (Dapsone), sulphapyridine, or sulfasalazine. Prior to Dapsone treatment, the patients must be checked for sulpha drug allergies and glucose-6-phosphate dehydrogenase deficiency since the most common adverse drug effect of dapsone is hemolytic anemia. A step-up to mycophenolate, azathioprine, or methotrexate is used for moderate inflammation or for diseases not responsive to initial therapy. A trial of cyclophosphamide with a short course of prednisolone or a combination of sulfas and myelosuppressive drugs may be used for severe inflammation. In a retrospective cohort study by Pujari entitled the SITE (Systemic Immunosuppressive Therapy for Eye Disease) Study, the authors discovered that the use cyclophosphamide as monotherapy was effective in controlling inflammation among 80% of all patients resulting to faster tapering of systemic corticosteroids after the first year.¹² For patients unresponsive to conventional therapy, intravenous immunoglobulins and rituximab (anti-CD20 monoclonal antibodies) may be used.^{8,13}

Adjunctive topical medications may be applied depending on the clinical presentation and symptoms unique to each patient. Ocular lubricants, punctal occlusion, and use of contact lenses may counter irritation and associated dry eye symptoms.¹¹ For severe cases, topical steroids, cyclosporine A, tacrolimus and even autologous serum can be used to reduce ocular surface inflammation while also treating ocular surface irritation. Steroids, however, must be used with caution as this may cause cataract and ocular hypertension. The addition of antibiotic ointments may be used to aggressively treat associated blepharitis.²

There are many adjunctive minor and major procedures for OCP. For ocular surface disease, punctal occlusion together with topical lubricants may be provided. For lid problems such as entropion and symblepharon, lower lid retractor plication, lateral tarsorrhaphy, epilation of lashes, release of symblepharon, and cryotherapy may be done. For severe disease, limbal stem cell transplant, mucous membrane autografts, amniotic membrane transplantation may be necessary. For end-stage patients needing treatment of corneal opacity, a Boston keratoprosthesis type 2 or an osteo-odonto-keratoprosthesis (OOKP) is usually done.

CONCLUSION

OCP is a rare autoimmune and blinding disease. Early diagnosis and prompt treatment are vital as ocular complications permanently affect the quality of life of patients as seen in our patient. DIF assay remains the gold-standard for diagnosis. This case documented the first Filipino patient diagnosed by DIF assay with ocular cicatricial pemphigoid who presented with ocular manifestations, skin lesions on the back, the left foot, and the oral mucosa. Systemic immunosuppression is the mainstay of treatment. Adjunctive supportive topical medication may be given to alleviate ocular discomfort. A multidisciplinary approach is essential to provide holistic care to each patient.

Acknowledgments

The authors would like to thank the Department of Ophthalmology and Visual Sciences of the Philippine General Hospital for the support in making this paper.

Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

All authors declared no conflicts of interest.

Funding Source

None.

REFERENCES

1. Branisteanu D, Stoleriu G, Branisteanu D, Boda D, Branisteanu C, Maranduca M, et al. Ocular cicatricial pemphigoid (Review). *Exp Ther Med*. 2020 Jul 7;1(20):3379-82. doi: 10.3892/etm.2020.8972.
2. Tyagi S, Kailash Bhol, Natarajan K, Charalampos Livir-Rallatos, C. Stephen Foster, A. Razzaque Ahmed. Ocular cicatricial pemphigoid antigen: Partial sequence and biochemical characterization. *Proc Natl Acad Sci U S A*. 1996 Dec 10;93(25):14714-9. doi: 10.1073/pnas.93.25.14714.
3. Tolaymat L, Hall MR. Cicatricial Pemphigoid [Internet]. PubMed. Treasure Island (FL): StatPearls Publishing. 2022 [cited 2024 Nov]. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK526120/>. PMID: 30252376.
4. Foster CS. Cicatricial pemphigoid. *Transactions of the American Ophthalmological Society*. 1986;84:527-663. PMID: 3296406. PMCID: PMC1298749.
5. Radford CF, Rauz S, Williams GP, Saw VPJ, Dart JKG. Incidence, presenting features, and diagnosis of cicatrizing conjunctivitis in the United Kingdom. *Eye*. 2012 Jun 22 (9):1199-208.; PMID: 22722485. PMCID: PMC3443826.
6. Elder M, Bernauer W, Leonard JN, Dart JK. Progression of disease in ocular cicatricial pemphigoid. *Br J Ophthalmol*. 1996 Apr;(4):292-6. doi: 10.1136/bjo.80.4.292. PMID: 8703876. PMCID: PMC505451.
7. Dart JK. The 2016 Bowman Lecture Conjunctival curses: scarring conjunctivitis 30 years on. *Eye*. 2017 Jan;31(2):301-332. doi: 10.1038/eye.2016.284. PMID: 28106896. PMCID: PMC5306467
8. Williams GP, Radford C, Nightingale P, Dart JKG, S Rauz. Evaluation of early and late presentation of patients with ocular mucous membrane pemphigoid to two major tertiary referral hospitals in the United Kingdom. *Eye*. 2011 Jul;25(9):1207-18. doi: 10.1038/eye.2011.175. PMID: 21799523. PMCID: PMC3173873.
9. Schonberg S, Stokkermans TJ. Ocular Pemphigoid [Internet]. PubMed. Treasure Island (FL): StatPearls Publishing; 2021 [cited 2024 Nov]. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK526100/>. PMID: 30252356
10. Ahmed M, Zein G, Khawaja F, Foster CS. Ocular cicatricial pemphigoid: pathogenesis, diagnosis and treatment. *Progress in Retinal and Eye Research*. 2004 Nov;23(6):579-92. doi:10.1016/j.preteyeres.2004.05.005.
11. Benitez-del-Castillo JM, Lemp MA. Ocular Surface Disorders [Internet]. Google Books. JP Medical Publishers. 2024 [cited 2024 Nov]. 103-115. Available from: https://books.google.com.ph/books?hl=en&lr=&id=7NCQXp2KA0C&oi=fnd&pg=PP12&ots=AE11FBKQr5&sig=AZyfGdsQYvKadna6fWifzI2w9IU&redir_esc=y#v=onepage&q=ocular%20cicatricial%20pemphigoid&cf=false
12. Pujari SS, Kempen JH, Newcomb CW, Gangaputra S, Daniel E, Suhler EB, et al. Cyclophosphamide for ocular inflammatory diseases. *Ophthalmology*. 2010 Feb;117(2):356 - 65. doi: 10.1016/j.ophtha.2009.06.060.
13. Georgoudis P, Sabatino F, Szentmary N, Palioura S, Fodor E, Hamada S, et al. Ocular mucous membrane pemphigoid: current state of pathophysiology, diagnostics and treatment. *Ophthalmology and Therapy*. 2019 Mar;8(1):5-17. doi: 10.1007/s40123-019-0164-z. PMID: 30694513. PMCID: PMC6393250.