A Case of Paraproteinemic Keratopathy

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Introduction

Ocular paraproteinemia is a constellation of ocular symptoms resulting from monoclonal gammopathies including Multiple Myeloma, Waldenström Macroglobulinemia, and monoclonal gammopathy of undetermined significance (MGUS). Additional systemic diseases, including B-cell lymphoma, plasmacytomas, amyloidosis, chronic lymphocytic leukemia, cryoglobulinemia, polyclonal hypergammaglobinemia, and even systemic IVIG therapy can also present with protein deposition within the eye.

Monoclonal gammopathies can present as a range of ocular diseases, including paraprotein keratopathy (PPK), maculopathy, and retinal vein occlusions, among others.

Differential diagnoses include congenital metabolic disorders, corneal dystrophies (including Macular, Schnyder or congenital stromal corneal dystrophy), and lipid deposition.

Case Report

- 59 year-old male presenting Aug 2021 with 5 months of blurred vision
- Described as ”clouded” vision with glare at night
- Has tried multiple glasses prescriptions with no benefit

Past Medical and Ocular History

- PCIOL OU, 2014
- History of IgG Kappa Multiple Myeloma, diagnosed 2012
  - Initiated Cytoxan, Velcade, Decadron in 2020
  - Progression May 2021 on PET/CT
  - Started Carfilzomib, Cytoxan, Decadron May 2021
  - Switched to Carfilzomib, Revlimid, Decadron
- Recent uncontrolled hyperglycemia in setting of systemic therapies and corticosteroids. Following with endocrinology with improved glycemic control at time of initial consultation

Further Workup and Treatment

- We discussed the nature of his findings and limited options for treatment, especially in the setting of active systemic disease
- Between August 2021 and December 2021, noted to have worsening systemic disease despite treatment
- Underwent autologous stem cell transplant Dec 2021
- He has an upcoming appointment with us in which we will monitor his disease status after stem cell transplantation

Ophthalmic Exam

- BCVA: 20/25 OD, 20/25 OS, J1+ OU
- IOP wnl, EOMs full, no rAPD, CVF full, normal adnexa
- Cornea: Corneal verticillata OU. Subepithelial and stromal crystals throughout cornea
- AC: Deep and Quiet **|** Iris: Flat, Round **|** Lens: PCIOL **|** Vitreous: Clear **|** Fundus: Normal, no retinopathy

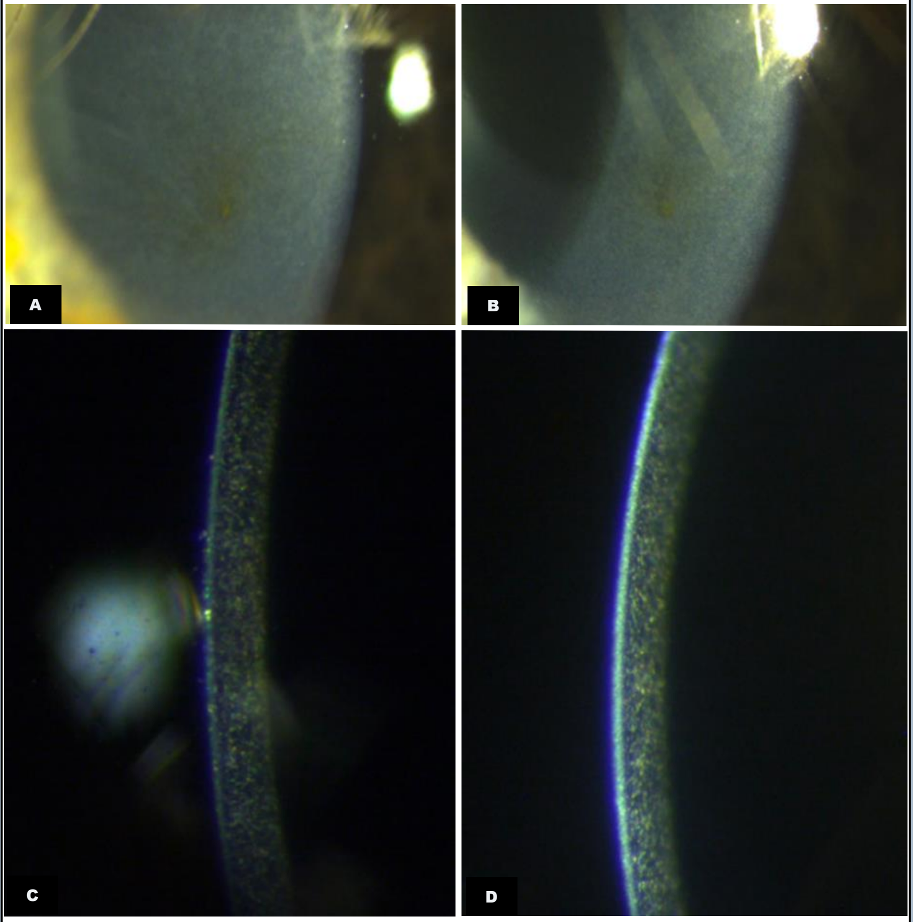

Figure 1: Slit lamp photography. (A) external slit lamp photo revealing corneal verticillata of the right eye. (B) optical cross section of the right cornea revealing crystalline deposition. (C) external slit lamp photo revealing corneal verticillata of the left eye. (D) optical cross section of the left cornea revealing crystalline deposition.

Discussion

Paraproteinemic keratopathy (PPK) is estimated to affect less than 1% of patients with systemic gammopathies and is more common among kappa light chain gammopathies.

They classically appear as bilateral crystalline deposits surrounded by nummular patchy opacities. Additional appearances include crystalline-like, lattice-like, peripheral granular-like, peripheral band-like, peripheral patch-like. These deposits can mimic corneal dystrophies and can pose diagnostic challenges. Genetic testing can be utilized as a method of distinguishing acquired vs inherited conditions.

Corneal verticillata have also been shown to be associated with systemic paraproteinemic states.

Treatment of underlying systemic disease guided by a Hematologist/Oncologist is essential and can result in an improvement in keratopathy.

Paraproteinemic keratopathy can improve, remain stable, progress, or can be relapsing and remitting associated with active systemic disease. There are case reports of resolution following proteasome inhibitor therapies however an associated timeframe has not been identified.

There is currently no widely effective ophthalmic treatment for PPK given the high risk of recurrence. Available treatments include superficial keratectomy, phototherapeutic keratectomy (PTK), penetrating keratoplasty (PK), or deep anterior lamellar keratoplasty (DALK). Recurrence has been documented as early as a few months to a year after transplantation. Topical steroids have not shown to be an effective treatment.

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