

Scleral Lenses for Chronic Graft-versus-Host Disease: A Case Report

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Cornea and Contact Lens

Background

- 49 year old Caucasian female
- Medical History: Chronic Graft-vs-Host Disease (GvHD), history of myelofibrosis x 2012, and allogenic hemopoietic stem cell transplant x 2015.
- Ocular History: LASIK x 2010, Dry eye syndrome OU, Punctal cautery OU x 2014
- Medications: Tacrolimus 5mg BID PO, Autologous serum x 1 q1h
- Social History: Unremarkable

Case Details

- Chief Complaint:** Referred by ophthalmology for scleral lens fitting to treat chronic dry eyes and GvHD. Currently using preservative free artificial tears and autologous serum throughout the day with mild relief.

Visit 1: Pertinent Findings		
Test	OD	OS
Unaided DVA	20/20-2	20/25
Refraction	-0.50-0.50x037	-0.75-0.50x135
Ant Seg	Cauterized punctum, LASIK scar, Diffuse SPK	Cauterized punctum, LASIK scar, Diffuse SPK
Post Seg	Normal findings	Normal findings

Table 1: Visit 1 Pertinent Findings

Visit 1: Diagnosis and Treatment	
Assessment	Treatment
Keratoconjunctivitis Sicca due to Graft-versus-Host Disease	Diagnostically fit with Zenlens Scleral lenses OU.
	Ordered Zenlens with changes made to over-refraction and fit.
	Follow-up for scleral lens dispense and insertion and removal training once lenses arrive.

Table 2: Visit 1 Diagnosis and Treatment

Visit 2: Scleral Lens Dispense Summary	
BCVA	20/20 OD, OS
Background	Trial #1 Dispense and Insertion and Removal training
Subjective findings	Patient reports good comfort and vision. Reports improvement of dryness upon insertion of lenses.
Anterior segment	OU: Slight improvement of SPK, Cauterized punctum, LASIK scar Prescribe Lotemax BID OU.

Table 3: Visit 2 Follow Up Summary

Visit 3: Scleral Lens Progress Evaluation for Trial 2	
BCVA	20/20 OD, OS
Background	OD: ~205um central clearance, good limbal clearance. Nasal edge digs into conjunctiva. (+) inferior conjunctiva prolapse (+) mild blanching Nasal. OS: ~201um central clearance, good limbal clearance. Mild edge lift temporally.
Pertinent Findings	OU: Improvement of SPK, Cauterized punctum, LASIK scar
Treatment	Lotemax BID (while lenses are out). Continue Autologous serum.

Table 4: Visit 3 Follow Up Summary

Visit 4: Scleral Lens Progress Evaluation for Trial 3	
BCVA	20/20 OD, OS
Background	OD: ~175um central clearance, good central clearance. Trace blanching Nasal OS: ~211um central clearance with good limbal clearance. Mild edge lift temporal, (-) blanching 360
Pertinent Findings	OU: No SPK, Cauterized punctum, LASIK scar
Treatment	Discontinue Lotemax BID. Continue Autologous serum. Continue wearing scleral lenses for dry eyes.

Table 5: Visit 4 Follow Up Summary

Pathophysiology

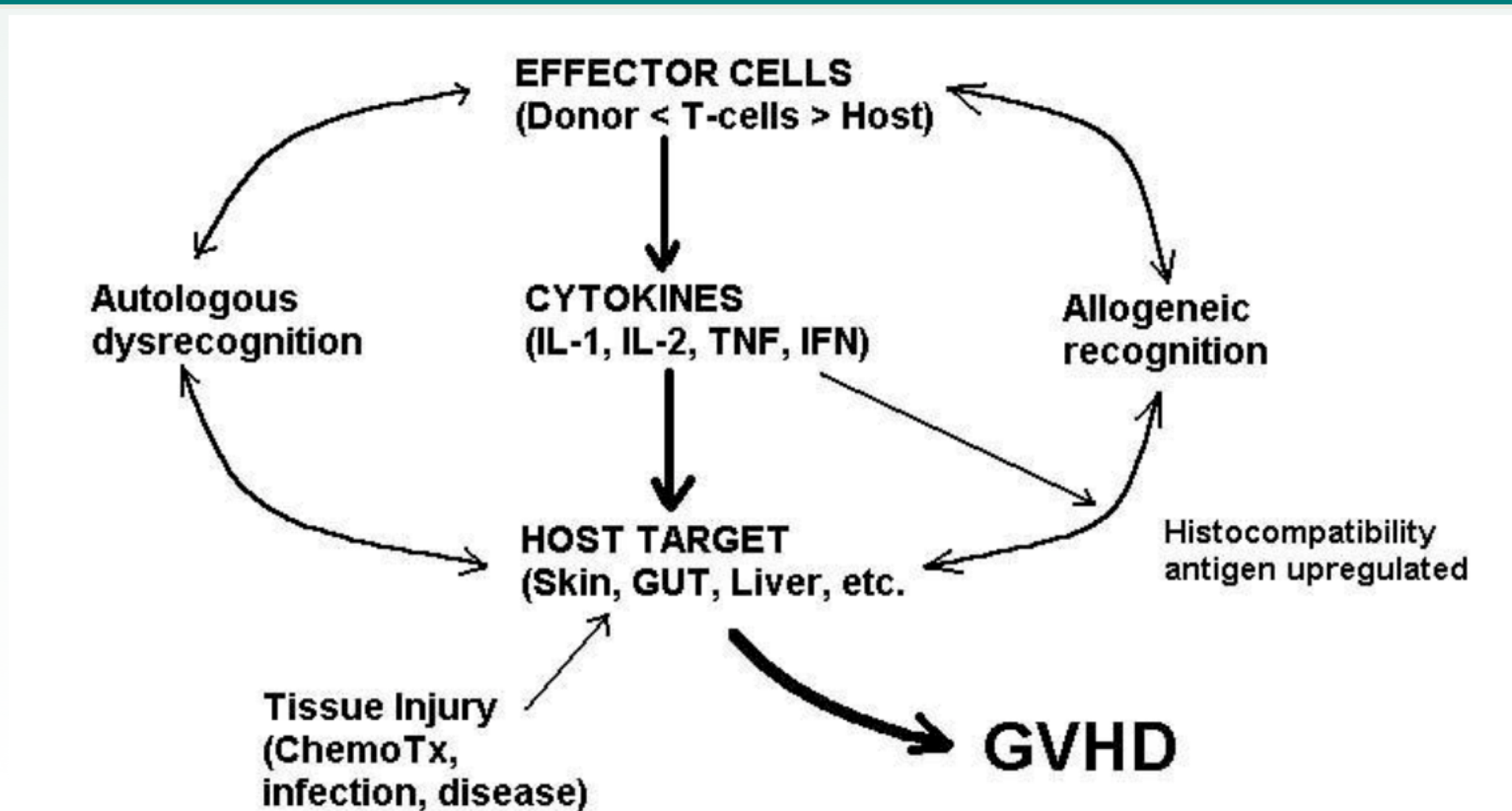


Figure 1. Interaction of factors involved in the mechanism of development of GVHD. Cytokines such as interleukin-1, interleukin-2, tumor necrosis factor alpha, and gamma interferon play an important role in the initiation and propagation of GVHD.

Interactive factors involved in the pathogenesis of graft versus host disease (GVHD.) Courtesy of Romeo A. Mandanas, MD, FACP.

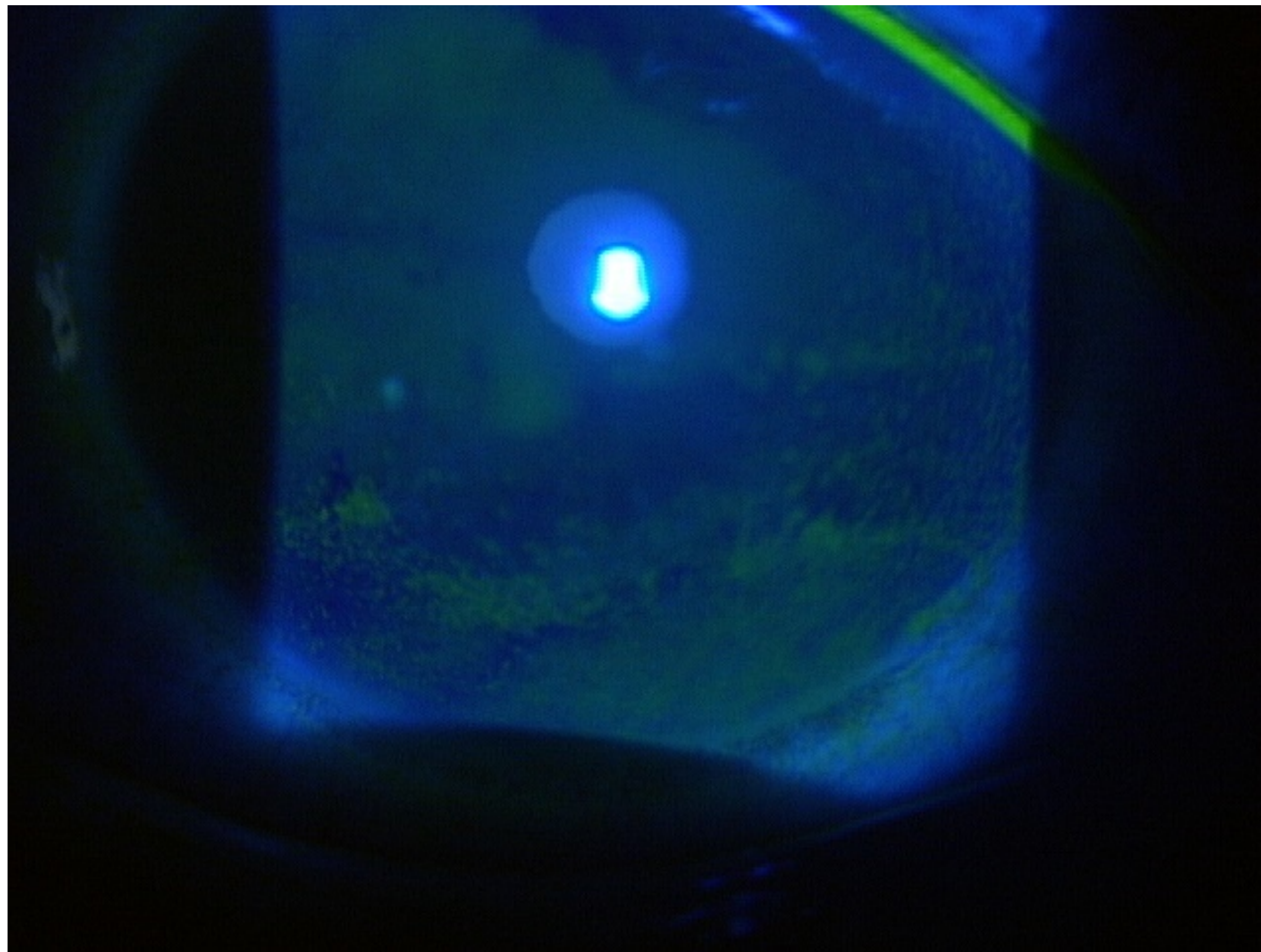


Figure 1 (above): OD captured by anterior segment camera from 08/19/2020

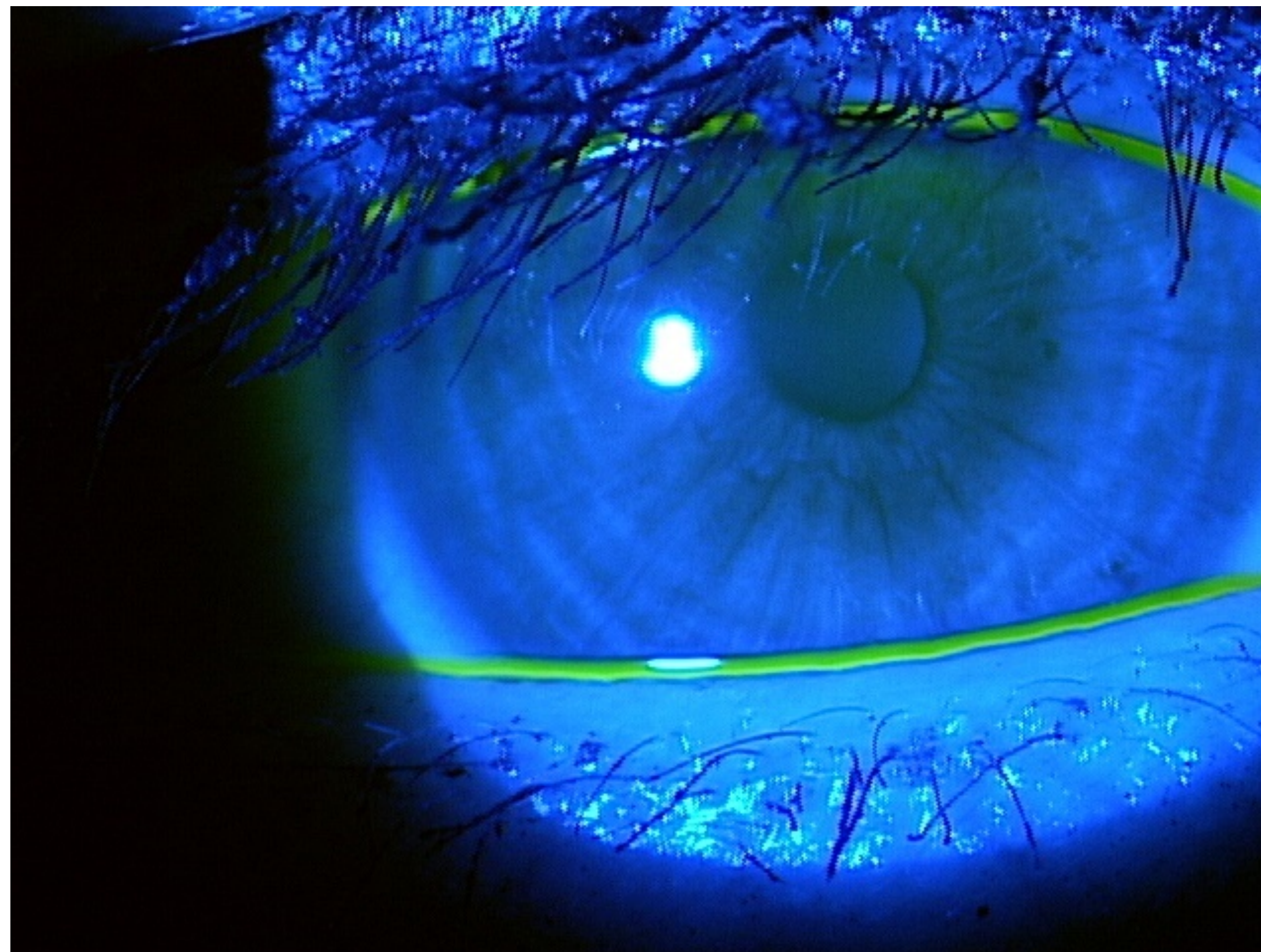


Figure 2 (above): OD captured by anterior segment camera from 11/03/2020

Differential Diagnosis

- Primary Keratoconjunctivitis Sicca
- Sjogren's Syndrome
- Stevens-Johnson's Syndrome
- Ocular Rosacea
- Radiation

Diagnosis and Discussion

Primary Diagnosis: Keratoconjunctivitis Sicca secondary to Chronic Graft versus Host Disease

Graft versus host disease (GvHD) is an immune-mediated disease resulting from a complex interaction between donor cells and a recipient's immune system. It is a disease that mimics autoimmune conditions. GvHD can frequently be triggered by allogenic stem cell transplantations (allo-SCT). GvHD develops in approximately 50% of patients following allo-SCT₄.

There are two types of GvHD, acute and chronic.

Acute:

- Within 100 days of receiving stem cell transplant
- Direct donor T-cells attacking of recipient cells
- Typically affects skin, liver, gastrointestinal system
- Responds well to steroids or immunosuppressants when affecting the eyes

Chronic:

- More than 100 days of receiving stem cell transplant
- Indirect activation of host T-cells attacking other host cells
- Typically affects skin, lungs, eyes, and mucous membranes

In ocular GVHD, patients can experience many dry eye symptoms including dryness, foreign body sensation, burning, stinging, itching, and tearing. Many patients will exhibit keratoconjunctivitis sicca, cicatricial conjunctivitis and a variety of ocular cellular destruction due to an inflammatory response.

A diagnostic evaluation for dry eye is essential to determine the severity of the disease and to monitor for improvement. Clinicians should use Sodium Fluorescein, Rose Bengal or Lissamine Green staining to determine both corneal and conjunctival involvement. Additional diagnostic tests include tear break-up time (TBUT), Schirmer scoring, Meibomian gland evaluation, corneal sensitivity and tear osmolarity.

Treatment and Management

- Preservative free artificial tears and autologous serum tear drops.
- Punctal plugs and punctal cauterization
- Acute treatment of surface inflammation with topical corticosteroid.
- Treat with Cyclosporine or Tacrolimus
- Fit patient in soft bandage contact lenses or scleral lenses to create a desired ocular environment and decrease friction.
- Co-management with primary care physician for systemic GvHD.
- The patient will be re-fit with a larger diameter scleral lens OD in February 2021.

Conclusion

- Diagnosis and treatment of symptoms in ocular GvHD will greatly improve a patients quality of life.
- Scleral lenses may be necessary to provide the desired environment for patient's suffering from ocular GvHD improve the long-term outcome of a patient's condition.
- It's important of eye care providers to work with other health care providers when recognizing the ocular manifestations of systemic disease.

Disclosures

- No financial disclosures to report

References

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