An A+ Solution to Triple A Syndrome: Scleral Lenses

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INTRODUCTION

Triple A Syndrome also known as Allgrove Syndrome is an autosomal recessive condition characterized by adrenal insufficiency, achalasia (difficulty swallowing), and alacrima (reduced or absent tear production). Found in fewer than 1000 people in the United States, there is an estimated prevalence of 1 per 1 million individuals. Mutations in the AAAS gene which encodes protein ALADIN are implicated.[1] Although its function is not fully understood, this protein is involved in transporting proteins to the cellular nucleus and maintaining structures within the nucleus. Individuals with Allgrove Syndrome suffer from difficulty swallowing, reduced or absent tear production, fatigue, low blood pressure, peripheral neuropathy, and autonomic dysfunction. From an ocular standpoint, these patients suffer from severe dryness, chronic ocular surface damage, neurotrophic keratopathy, and optic atrophy.[2,,4,5] Typical ocular treatments include frequent artificial tear applications, nighttime ophthalmic gels, and warm compress. This case report strives to investigate the application of scleral contact lenses in patients with this rare condition.

CASE DESCRIPTION

A female patient in her early forties presented to our clinic for management of neurotrophic keratopathy and alacrima secondary to Allgrove Syndrome. Clinical findings revealed a significantly compromised ocular surface and large confluent areas of punctate keratitis of both eyes. Dilated findings revealed optic atrophy of both eyes. Corrected visual acuities were 20/30 OD/OS at our initial visit. She was fit into scleral contact lenses to provide sustained lubrication to her ocular surface. She required significant insertion and removal training due to finger weakness associated with her peripheral neuropathy but learned to become efficient utilizing an insertion stand. She was adjunctively initiated on Cyclosporine 0.05%, sodium chloride hypertonic drops, and nighttime ointment. Her BCVA improved to 20/20 in each eye. Over five years, we have followed her closely to ensure improvement in her overall comfort, quality of vision, and corneal status.

FIGURES

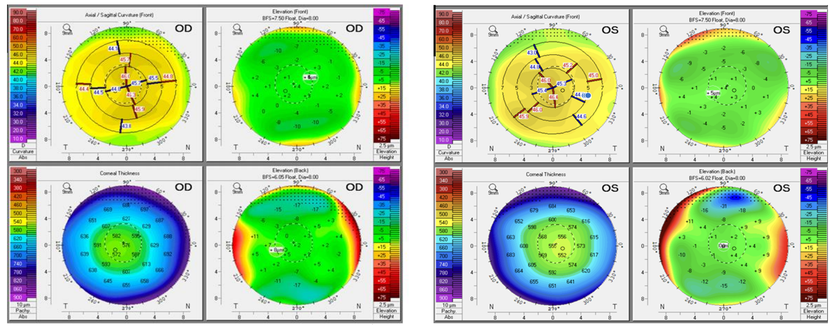

Figure 1. Tomography OD (left) and OS (right)

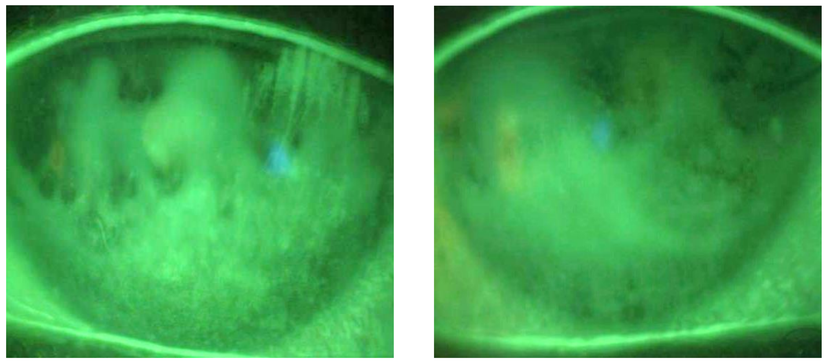

Figure 2. Sodium fluorescein staining OD (left) and OS (right)

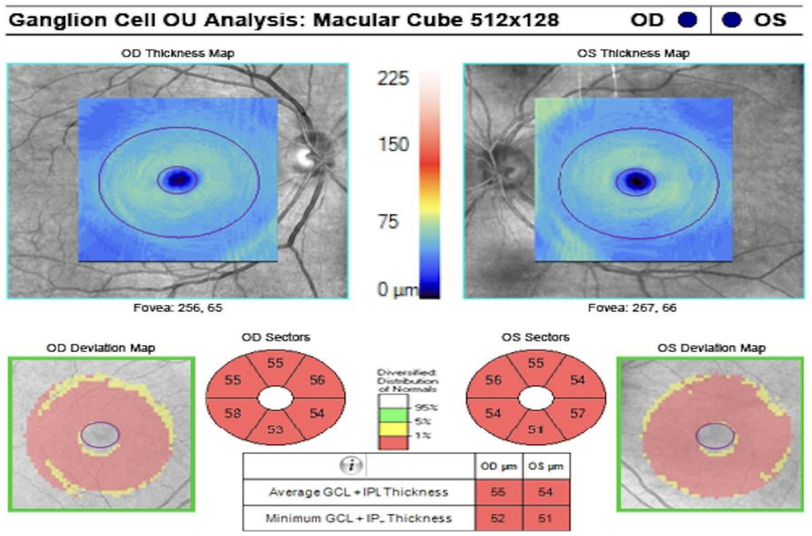

Figure 3. Diffuse ganglion cell loss due to bilateral optic atrophy

FINALIZED SCLERAL LENS RX

	Power	BC	SAG	Diameter	Limbal	Edges	BCVA
OD	-3.50 SPH	7.11 mm	4150 µm	14.80 mm	LCD +75	Flat2/ Steep2	20/20
OS	-1.00 SPH	7.50 mm	4100 µm	14.80 mm	LCD +75	Flat2/ Steep2	20/20

DISCUSSION

Allgrove Syndrome is a rare systemic condition with frequently undermanaged ocular manifestations. Patients with Allgrove Syndrome develop neurotrophic keratitis from two separate processes:

1. **Alacrima related dehydration keratopathy leads to chronic corneal nerve damage**
2. **Autonomic dysfunction (a hallmark of this condition) leads to peripheral neuropathy that extends to the corneal nerves**

The cornea is innervated by the first branch of the trigeminal nerve, which plays a vital role in corneal healing, regeneration, sensory feedback, tear regulation, and maintaining corneal metabolism. Other causes of neurotrophic keratitis include herpetic keratitis, penetrating ocular surgery, chemical burns, Sjogren's syndrome, amyloidosis, uncontrolled diabetes, multiple sclerosis, and vitamin A deficiency. Scleral lenses provide sustained lubrication and protection to the ocular surface to promote corneal healing, improve comfort, and enhance vision.[3]

CONCLUSION

Scleral lenses can play a crucial role in managing the ocular complications of Allgrove Syndrome and other causes of neurotrophic keratopathy or severe dry eye. These lenses vault over the cornea and rest on the sclera, creating a fluid-filled reservoir that keeps the cornea continuously hydrated, prevents corneal damage, improves visual acuity, and reduces discomfort. In patients with Allgrove Syndrome, scleral lenses provide long-lasting relief and protection, eliminating the need for frequent artificial tear application. By improving both visual function and comfort, scleral lenses enhance the quality of life for individuals with this rare disorder.

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