Anterior segment OCT guided diagnosis of anterior segment ocular malformation.

Ahmad Rehmani[1,2*], Christina Vicidomini[1,2]

[1]Department of Ophthalmology, St John's Episcopal Hospital, Far Rockaway, NY, USA

[2]Department of Ophthalmology, Intefaith Medical Center, Brooklyn, NY, USA

Abstract

Congenital corneal opacities secondary to anterior segment defects is a large category encompassing a wide spectrum of disorders. Depending on circumstances of patient presentation, symptomatology, and ophthalmologic exam, the exact etiology of these defects may be difficult to elucidate or diagnose. We introduce a case of bilateral congenital anterior segment malformation in an asymptomatic 56 year old patient who presented to the clinic for initial routine eye exam. Physical exam was notable for excellent visual acuity, bilateral leukomas overlying areas of inferior iridocorneal touch, normal intraocular pressure and posterior segment. Anterior segment optical coherence tomography revealed intact corneal endothelium throughout the area of iridocorneal adhesions. Anterior segment OCT served as a useful, non-invasive, fast diagnostic tool allowing us to exclude diseases that otherwise may not have been able to be ruled out.

Keywords: Anterior-segment OCT, Iridocorneal adhesions, Corneal opacity, Peter's Anomaly.

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Introduction

Development of the ocular anterior segment occurs during the 7th week of embryogenesis during which 5 corneal layers, trabecular meshwork, iris, pupillary membrane and anterior chamber are formed. The complex interrelationship of these structures in such close proximity in a normal eye is dependent on 3 distinct waves of mesenchymal migration. The first, second and third waves are characterized by development of corneal endothelium, iris/pupillary membrane and corneal stroma/sclera respectively. By the 5th month of gestation, the anterior segment is well defined [1].

Congenital anomalies may develop during any of the many intricate steps of anterior segment formation. When patients present with these malformations, the exact diagnosis may not be so apparent. Anterior segment OCT (AS-OCT) is a powerful tool in the high-resolution cross-sectional imaging of these structures of the eye. It uses low-coherence interferometry with spatial resolution of 10-20 μm [2].

AS-OCT has proven to be a useful diagnostic technique for assessing anterior chamber depth and angle, corneal thickness, as well as abnormalities of the cornea, iris, and lens [2]. We present a unique case of anterior segment malformation presenting as asymptomatic bilateral iridocorneal adhesions diagnosed by anterior segment OCT.

Case Report

A 56-year-old male presented for first time visit to Bishop Walker Eye clinic without complaints for an initial examination stating he has had "white spots" on his eyes since birth. He denied any change in size of the lesions, pain, irritation, or redness throughout his life. He denied any medical, surgical or past ocular history, allergies, medication use, or any family history of ocular issues.

The patient underwent a full ophthalmologic examination including anterior segment OCT, corneal pachymetry, B-scan, A-scan, gonioscopy, as well as laboratory tests for Syphilis, Lyme disease, Herpes Simplex and Zoster.

The patient's vision was 20/25 OD and 20/20 OS with correction. On pupillary exam bilateral iridocorneal adhesions dragged the pupils eccentrically inferior during constriction. On slit lamp examination (Figure 1) the patient was noted to have bilateral inferior iridocorneal adhesions from 4 to 8 o'clock in the same location in both eyes with overlying stromal haze and surrounding pigment.

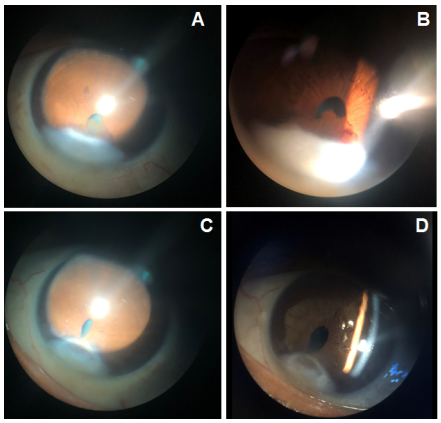

Figure 1. Slit lamp examination of right (A and B) and left (C and D) eyes. Bilateral inferior leukomas are noted in thin slit view (B and D). Eccentrically inferior peaked pupils following pupillary construction due to downward pull by irideocorneal adhesions are observed with full illumination (A and C).

The central cornea was clear. On gonioscopy the patient's iridocorneal angles were Shaffer 4 in both eyes 360 degrees – except in areas of iridocorneal touch where no angle structures were appreciated.

Intraocular pressure was 14 and 16 in the right and left eyes respectively. Dilated fundus exam was unremarkable with no deficits or colobomas noted. Anterior segment OCT revealed the presence of corneal endothelium bilaterally throughout the areas of iridocorneal touch (Figure 2). Corneal pachymetry was >700 in areas of iridocorneal touch. B-scan was negative for any posterior pathology. The workup for Syphilis, Lyme disease, and Herpes was negative.

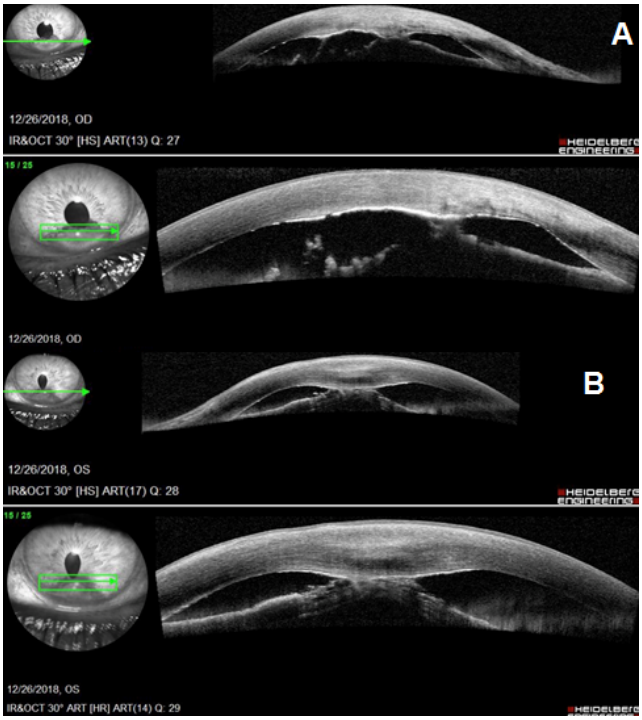

Figure 2. AS-OCT of the right (A) and left (B) eyes show bilateral inferior iridocorneal touch. The presence of intact corneal endothelium throughout the defect can be appreciated in both eyes. Anterior chamber appears normal outside of places of iridocorneal touch.

Discussion

Congenital corneal opacities can be classified as primary or secondary. Primary congenital opacification refers to developmental anomalies of the cornea only and include disorders such as corneal dystrophies, isolated sclerocornea and corneal defects due to dermoids [3].

Secondary congenital corneal opacities refer to defects in other anterior segment structures causing corneal opacification. Under this umbrella, kerato-irido-lenticular dysgenesis encompasses a wide spectrum of disorders including failure of lens to separate from the cornea either congenitally or from posterior pressure and as in our patient, iridocorneal adhesions [3].

Secondary congenital corneal opacities due to iridocorneal touch are often diagnosed as Peter's anomaly; the hallmark of which is characterized by absence of posterior cornea i.e. corneal endothelium/bowman's membrane [1,3-6]. Our patient presented with inferior leukomas in both eyes. However, the bilateral and asymptomatic nature of his condition, excellent vision, no evidence of glaucoma, and late age of presentation suggested another pathologic process.

Using anterior segment OCT (AS-OCT), we were able to visualize the corneal endothelium and thus deduce the patient did not meet the requirement for Peter's anomaly. As the patient had the lesions since birth, which remained unchanged, it is likely instead that the patient underwent a malformation of the anterior segment during embryogenesis that left the eyes largely unaffected other than inferior iridocorneal adhesions.

Other possible causes include childhood illness such as Measles, which has been shown to cause inferiorly located leukomas from iridocorneal adhesions [7]. However, these cases involved Measles infection during early childhood rather than congenitally as in our patient.

Conclusion

AS-OCT is a powerful tool in the high definition imaging of the anterior segment. Studies have shown its applicability particularly for imaging through opacified corneas with a higher resolution than standard ultrasonography [4]. Additionally, the quick, noninvasive, non-contact nature of AS OCT allows for frequent imaging and can be used in patients at every checkup allowing close monitoring of previously poorly understood ocular conditions such as Peter's Anomaly or congenital malformations of the angle [4]. We recommend anterior segment OCT as a valuable tool in evaluating the anterior chamber.

References

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***Correspondence to**

Ahmad Rehmani

Department of Ophthalmology

St. John's Episcopal Hospital

327 Beach 19th St, Far Rockaway, NY 11691, USA

ahmadsrehmani@gmail.com