Complete detachment of the Schwalbe's line in a case of Axenfeld-Reiger anomaly - A rare presentation

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Key words: Axenfeld–Reiger anomaly, Axenfeld–Reiger syndrome, developmental glaucoma, Schwalbe's line

A 21-year-old female presented with a dimness of vision in both eyes. Her best-corrected visual acuity (BCVA) was 20/80 and 20/400 in her right and left eyes, respectively. There was posterior embryotoxon (PE) with areas of peripheral anterior synechiae (PAS) in both eyes. The anterior segment of the right eye showed a completely detached rolled-up white cord-like structure with adhesions to the iris tissue [Fig. 1a and b]. The left eye showed a similar white cord-like structure traversing the anterior chamber with adhesions to the iris tissue and point attachment to the angle inferiorly [Fig. 1c and d]. The IOP was 42 and 44 mmHg in the right and left eyes, respectively. Gonioscopy showed anterior insertion of the iris into the trabecular meshwork, prominent iris processes, and broad-based synechiae. The cord-like structure was seen taking origin at the level of the Schwalbe's line. Both optic disks showed advanced cupping with 0.9 cup-disk-ratio (CDR) [Fig. 2a and b]. She had no dental, facial, umbilical, or pituitary abnormalities. Automated perimetry, done using the Humphrey's Field Analyzer, HFA 720 II (Carl Zeiss-Humphrey Systems, Dublin, CA, USA), of the right eye showed advanced visual field defect, while in the left eye, the test could not be done [Fig. 3a and b]. The optical coherence tomography (OCT) of the retinal nerve fiber layer (RNFL) using the Spectralis spectral domain-OCT (Heidelberg Engineering, Heidelberg, Germany) of both eyes showed gross RNFL thinning [Fig. 3c]. A diagnosis of the Axenfeld–Reiger anomaly (ARA) was made. The medical management was started with dorzolamide (2% w/v) and timolol maleate (0.5% w/v) fixed-dose combination (FDC) eye drops twice daily and travoprost (0.004% w/v) eye drop once daily in both eyes. Two weeks post-treatment, her IOP was 24 mmHg in both eyes. Surgical intervention was planned. She was asked to undergo a trabeculectomy with mitomycin-C in her right eye under a very guarded visual prognosis followed by a palliative trabeculectomy with mitomycin-C in her left eye.

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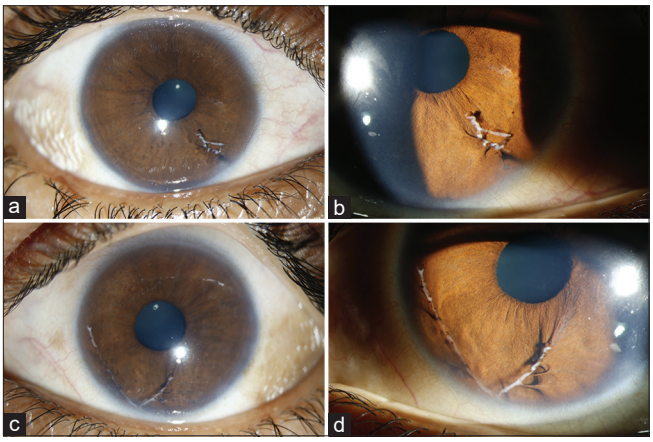

Figure 1: (a) Diffuse and (b) slit-beam illumination photographs of the right eye of the patient showing a completely detached rolled-up white cord-like structure adherent to the iris. This is the completely detached Schwalbe's line adherent to the iris tissue in the anterior chamber. (c) Diffuse and (d) slit-beam illumination photographs of the left eye showing a similar white cord-like structure traversing the anterior chamber with point attachment to the angle inferiorly. This is the partially detached Schwalbe's line with adherent iris tissue strands. PAS can be seen inferiorly and infero-temporally

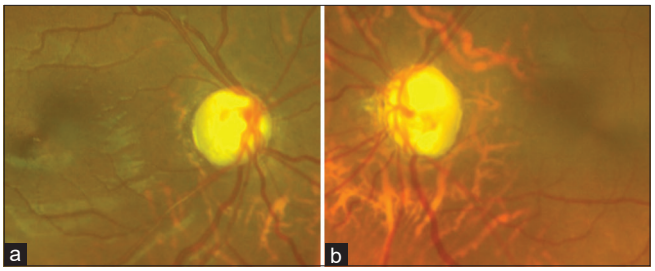

Figure 2: Disk photographs showing advanced, near-total cupping of the optic disks in the right eye (a) and left eye (b)

Discussion

ARS is a bilateral, heterogeneous condition and may include developmental abnormalities of the angle, trabecular meshwork, and iris.[1] Correctopia, polycoria, ectropion uveae, PE, and increased IOP are common ophthalmological findings. This spectrum of anomalies was initially described by Axenfeld and Rieger in the early twentieth century. ARS is subcategorized into three groups.[2] Axenfeld anomaly (AA) patients present with angle anomalies only. Reiger anomaly (RA) patients

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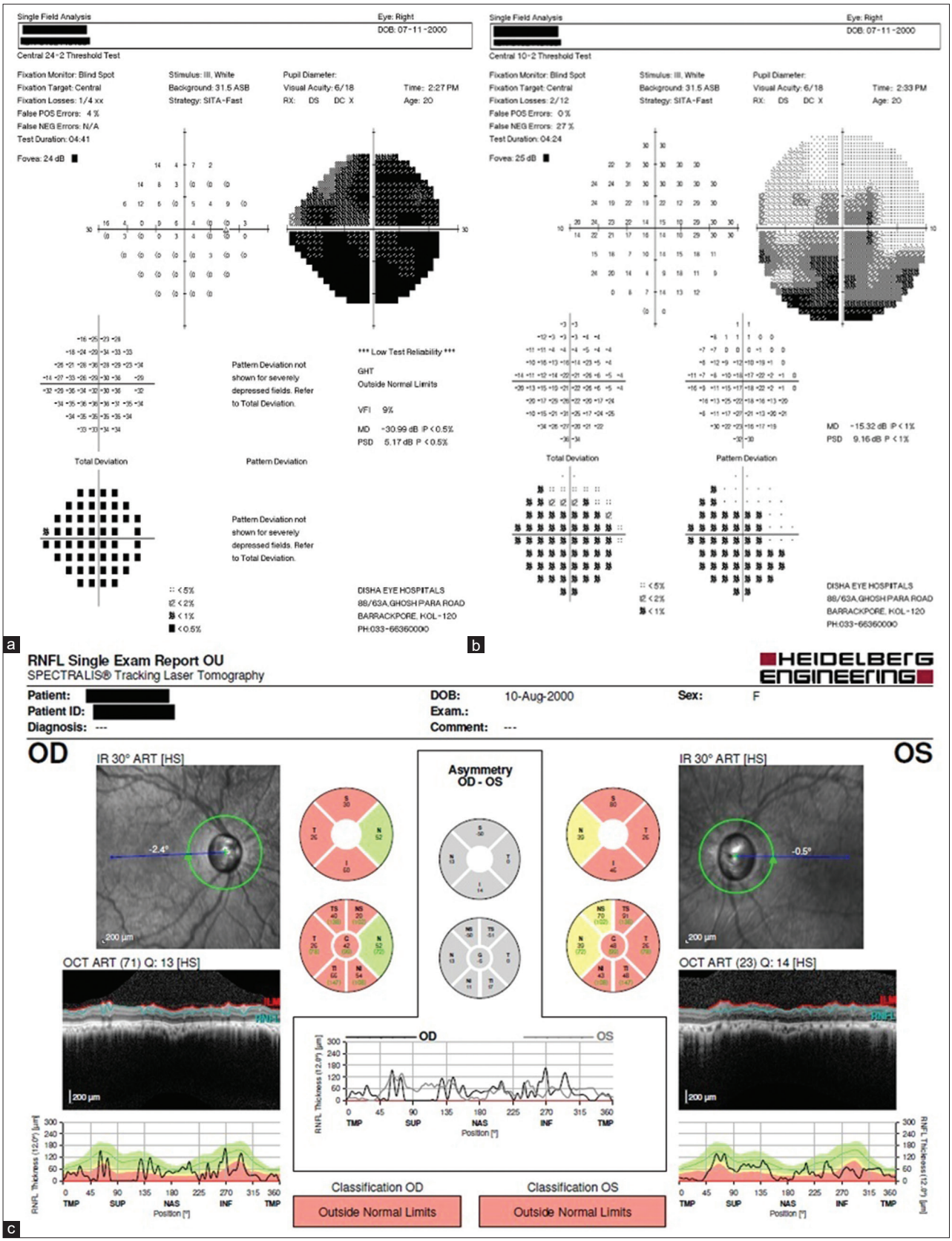

Figure 3: (a) Humphrey 24-2 single-field analysis of the right eye of the patient showing advanced visual field damage. (b) Humphrey 10-2 single-field analysis of the right eye of the patient showing dense scotoma in the inferior hemifield. (c) Spectralis SD-OCT RNFL single exam report of the patient showing gross RNFL thinning in all the quadrants in both the eyes

characteristically present with iris defects and the Reiger syndrome (RS) describes patients with Reiger anomaly in addition to systemic features.[3] Impaired neural crest cell migration and differentiation during embryonic development are considered important in the pathogenesis of ARS. Schwalbe's line and iris stroma have a common origin from the neural crest cells which could possibly explain the presence of detached Schwalbe's line with attached iris tissue strands in the anterior chamber.[4]

This thickened, prominent Schwalbe's line being completely detached in one eye and partially detached in the other eye in a case of ARA is an extremely rare presentation and has been reported by very few authors.[5] This presentation should be kept in mind as a rare clinical sign in ARA or ARS patients with developmental glaucoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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